Bedside Clinical Guidelines Partnership

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Paediatric Guidelines

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N: NEUROLOGY

R: RENAL

R: RHEUMATOLOGY

S: SAFEGUARDING

This book has been compiled as an aide-memoire for all staff concerned with the management of general medical paediatric patients, especially those who present as emergencies.

Guidelines on the management of common medical conditions

No guideline will apply to every patient, even where the diagnosis is clear-cut; there will always be exceptions. These guidelines are not intended as a substitute for logical thought and must be tempered by clinical judgement in the individual patient.

The guidelines are advisory, NOT mandatory

Prescribing regimens and nomograms

The administration of certain drugs, especially those given intravenously, requires great care if hazardous errors are to be avoided. These guidelines do not include all guidance on the indications, contraindications, dosage and administration for all drugs. Please refer to the British National Formulary for Children (BNFc).

Practical procedures

DO NOT attempt to carry out any of these Practical procedures unless you have been trained to do so and have demonstrated your competence.

National guidelines

Where there are different recommendations the following order of prioritisation is followed: NICE > NPSA > SIGN > RCPCH > National specialist society > BNFC > Cochrane > Meta-analysis > systematic review > RCT > other peer review research > review > local practice.

Evidence base

These have been written with reference to published medical literature and amended after extensive consultation. Wherever possible, the recommendations made are evidence based. Where no clear evidence has been identified from published literature the advice given represents a consensus of the expert authors and their peers and is based on their practical experience.

Supporting information

Where possible the guidelines are based on evidence from published literature. It is intended that the evidence relating to statements made in the guidelines and its quality will be made explicit.

Where supporting evidence has been identified it is graded I to V according to standard criteria of validity and methodological quality as detailed in the table below. A summary of the evidence supporting each statement is available, with the original sources referenced (ward-based copies only). The evidence summaries are being developed on a rolling programme which will be updated as each guideline is reviewed.

Level of evidence	Strength of evidence
I	Strong evidence from at least one systematic review of multiple well-designed randomized controlled trials
II	Strong evidence from at least one properly designed randomized controlled trial of appropriate size
III	Evidence from well-designed trials without randomization, single group pre-post, cohort, time series or matched case-control studies
IV	Evidence from well-designed non-experimental studies from more than one centre or research group
V	Opinions of respected authorities, based on clinical evidence, descriptive studies or reports of expert committees

JA Muir-Gray from Evidence Based Healthcare, Churchill Livingstone London 1997

Feedback

Evaluating the evidence-base of these guidelines involves continuous review of both new and existing literature. The editors encourage you to challenge the evidence provided in this document. If you know of evidence that contradicts, or additional evidence in support of the advice given in these guidelines please contact us.

The accuracy of the detailed advice given has been subject to exhaustive checks. However, if any errors or omissions become apparent contact us so these can be amended in the next review, or, if necessary, be brought to the urgent attention of users. Constructive comments or suggestions would also be welcome.

Contact

Partners in Paediatrics, via www.networks.nhs.uk/nhs-networks/partners-in-paediatrics or Bedside clinical guidelines partnership via e-mail: bedsideclinicalguidelines@uhns.nhs.uk

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MANAGEMENT

- Stimulate patient to assess for signs of life and shout for help
- Establish basic life support: Airway – Breathing – Circulation
- Connect ECG monitor: identify rhythm and follow **Algorithm**
- Control airway and ventilation: preferably intubate
- Obtain vascular access, peripheral or intraosseous (IO)
- Change person performing chest compressions every few minutes

Airway (A)

- Inspect mouth: apply suction if necessary
- Use either head tilt and chin lift or jaw thrust
- Oro- or nasopharyngeal airway
- Intubation:
  - endotracheal tube sizes
  - term newborns 3–3.5 mm
  - aged 1 yr 4.5 mm
  - aged >1 yr: use formula [(age/4) + 4] mm for uncuffed tubes; 0.5 smaller for cuffed
- If airway cannot be achieved, consider laryngeal mask or, failing that, cricothyrotomy

Breathing (B)

- Self-inflating bag and mask with 100% oxygen
- Ventilation rate
  - unintubated: 2 inflations for every 15 compressions
  - intubated:10–12/min, with continuous compressions
- Consider foreign body or pneumothorax

Circulation (C)

- Cardiac compression rate: 100–120/min depressing lower half of sternum by at least one third: push hard, push fast
- Peripheral venous access: 1–2 attempts (<30 sec)
- Intraosseous access: 2–3 cm below tibial tuberosity (see **Intraosseous infusion** guideline)
- Use ECG monitor to decide between:
  - a non-shockable rhythm: asystole or pulseless electrical activity (PEA) i.e. electromechanical dissociation
    OR
  - a shockable rhythm: ventricular fibrillation or pulseless ventricular tachycardia

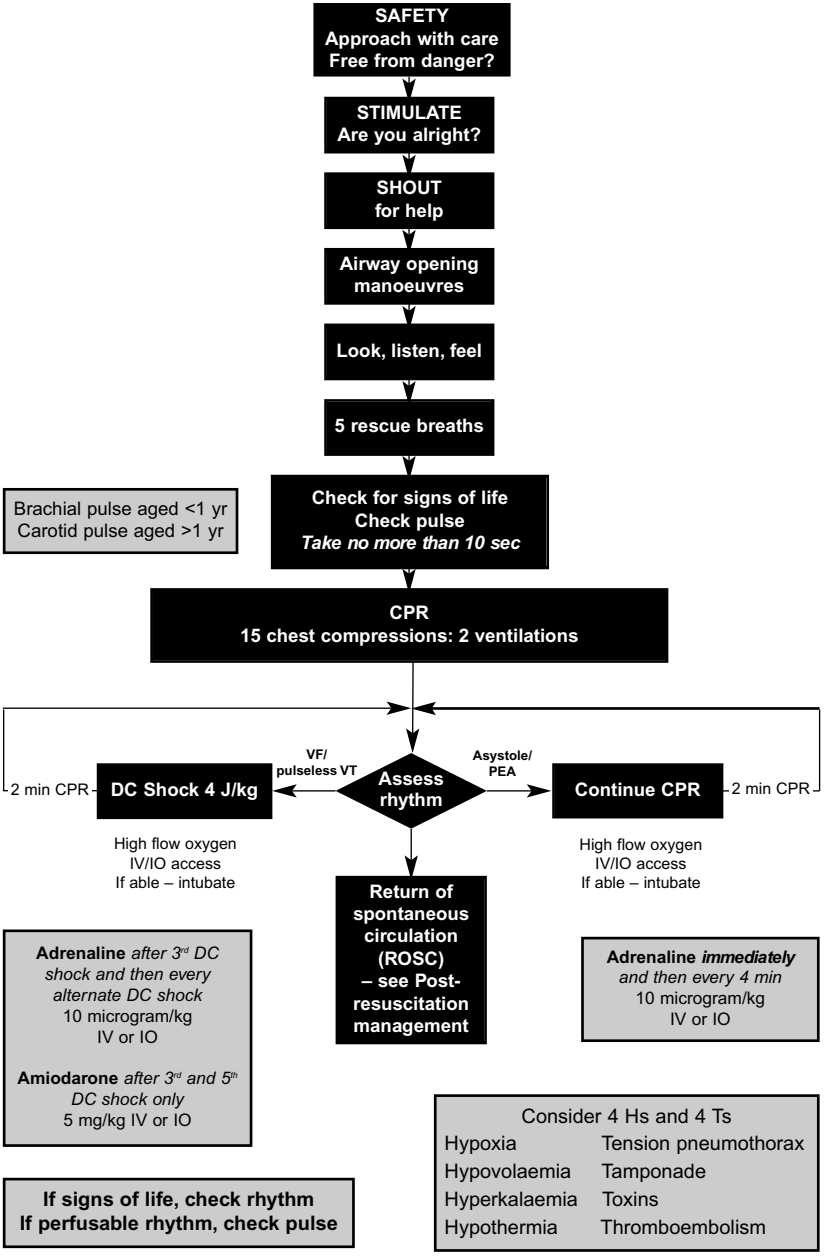
Algorithm for managing these rhythms follows:

- If arrest rhythm changes, restart **Algorithm**
- If organised electrical activity seen, check pulse and for signs of circulation

Adrenaline doses for asystole

Route	Aged <12 yr	Aged 12 yr – adult	Notes	
IV rapid bolus/ intraosseous	10 microgram/kg (0.1 mL/kg of 1:10,000)	1 mg (10 mL of 1:10,000)	Initial and usual subsequent dose	If given by intraosseous route flush with sodium chloride 0.9%
	100 microgram/kg (0.1 mL/kg of 1:1000 or 1 mL/kg of 1:10,000)	5 mg (5 mL of 1:10,000)	Exceptional circumstances (e.g. beta-blocker overdose)	Maximum dose 5 mL of 1:1000
Endotracheal tube (ETT)	100 microgram/kg (0.1 mL/kg of 1:1000 or 1 mL/kg of 1:10,000)	5 mg (5 mL of 1:1000)	-	

APLS - CARDIORESPIRATORY ARREST • 2/3



Modified from ALSG 2010, reproduced with permission

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Defibrillation

- Use hands-free paediatric pads in children, may be used anteriorly and posteriorly
- Resume 2 min of cardiac compressions immediately after giving DC shock, without checking monitor or feeling for pulse
- Briefly check monitor for rhythm before next shock: if rhythm changed, check pulse
- Adrenaline and amiodarone are given after the 3rd and 5th DC shock, and then adrenaline only every other DC shock
- Automatic external defibrillators (AEDs) do not easily detect tachyarrythmias in infants but may be used at all ages, ideally with paediatric pads, which attenuate the dose to 50–80 J

PARENTAL PRESENCE

- Evidence suggests that presence at their child's side during resuscitation enables parents to gain a realistic understanding of efforts made to save their child. They may subsequently show less anxiety and depression
- Designate one staff member to support parents and explain all actions
- Team leader, not parents, must decide when it is appropriate to stop resuscitation

WHEN TO STOP RESUSCITATION

- Unless exceptions exist, it is reasonable to stop after 30 min of CPR if you find:
  - no detectable signs of cardiac output **and**
  - no evidence of signs of life (even if ECG complexes)
- Exceptions include:
  - hypothermia (<32ºC)
  - overdoses of cerebral depressant drugs (successful resuscitation has occurred with 24 hr CPR)
- Discuss difficult cases with consultant before abandoning resuscitation

POST-RESUSCITATION MANAGEMENT

Identify and treat underlying cause

Monitor

- Heart rate and rhythm
- Oxygen saturation
- CO_2 monitoring
- Core and skin temperatures
- BP
- Urine output
- Arterial blood gases
- Central venous pressure

Request

- Chest X-ray
- Arterial and central venous gases
- Haemoglobin and platelets
- Group and save serum for crossmatch
- Sodium, potassium, urea and creatinine
- Clotting screen
- Blood glucose
- LFTs
- 12-lead ECG

- Transfer to PICU
- Hold a team debriefing session to reflect on practice

APLS - RECOGNITION AND ASSESSMENT OF THE SICK CHILD • 1/4

SUMMARY OF RAPID CLINICAL ASSESSMENT

Assessment

Airway (A) and Breathing (B)

- Effort of breathing
  - respiratory rate
  - recession
  - use of accessory muscles
  - additional sounds: stridor, wheeze, grunting
  - flaring of nostrils
- Efficacy of breathing
  - chest movement and symmetry
  - breath sounds
  - SpO_2 in air

Circulation (C)

- Heart rate
- Pulse volume
  - peripheral
  - central (carotid/femoral)
- Blood pressure
- Capillary refill time
- Skin colour and temperature

Disability (D)

- Conscious level
- Posture
- Pupils

Exposure (E)

- Fever
- Skin rashes, bruising

Don't Ever Forget Glucose (DEFG)

- Glucose stix

Actions

- Complete assessment should take <1 min
- Treat as problems are found
- Once airway (A), breathing (B) and circulation (C) are clearly recognised as being stable or have been stabilised, definitive management of underlying condition can proceed
- Reassessment of ABCDE at frequent intervals necessary to assess progress and detect deterioration
- Hypoglycaemia: glucose 10% 2 mL/kg followed by IV glucose infusion

CHILD AND PARENTS

- Give clear explanations to parents and child
- Allow and encourage parents to remain with child at all times

RECOGNITION AND ASSESSMENT OF THE SICK CHILD

Weight

Anticipated weight in relation to age

Age	Weight (kg)
Birth	3.5
5 months	7
1 yr	10

Weight can be estimated using following formulae:

- 0–12 months: wt (kg) = [age (m) / 2] + 4
- 1–6 years: wt (kg) = [age (y) + 4] x 2
- 7–14 years: wt (kg) = [age (y) x 3] + 7

Airway

Primary assessment of airway

- Vocalisations (e.g. crying or talking) indicate ventilation and some degree of airway patency
- Assess patency by:
  - **looking** for chest and/or abdominal movement
  - **listening** for breath sounds
  - **feeling** for expired air

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Re-assess after any airway opening manoeuvres

- Infants: a neutral head position; other children: 'sniffing the morning air'
- Other signs that may suggest upper airway obstruction:
  - stridor
  - intercostal/subcostal/sternal recession

Breathing

Primary assessment of breathing

- Assess
  - effort of breathing
  - efficacy of breathing
  - effects of respiratory failure

Effort of breathing

- Respiratory rates 'at rest' at different ages

Age (yr)	Resp rate (breaths/min)
<1	30–40
1–2	25–35
3–5	25–30
6–12	20–25
>12	15–20

- Respiratory rate:
  - tachypnoea: from either lung or airway disease or metabolic acidosis
  - bradypnoea: due to fatigue, raised intracranial pressure, or pre-terminal
- Recession:
  - intercostal, subcostal or sternal recession shows increased effort of breathing
  - degree of recession indicates severity of respiratory difficulty
  - in child with exhaustion, chest movement and recession will decrease
- Inspiratory or expiratory noises:
  - stridor, usually inspiratory, indicates laryngeal or tracheal obstruction
  - wheeze, predominantly expiratory, indicates lower airway obstruction
  - volume of noise is not an indicator of severity
- Grunting:
  - a sign of severe respiratory distress
  - can also occur in intracranial and intra-abdominal emergencies
- Accessory muscle use
- Gasping (a sign of severe hypoxaemia and can be pre-terminal)
- Flaring of nostrils

Exceptions

- **Increased effort of breathing DOES NOT occur in three circumstances:**
  - **exhaustion**
  - **central respiratory depression** (e.g. from raised intracranial depression, poisoning or encephalopathy)
  - **neuromuscular disease** (e.g. spinal muscular atrophy, muscular dystrophy or poliomyelitis)

Efficacy of breathing

- Breath sounds on auscultation:
  - reduced or absent
  - bronchial
  - symmetrical or asymmetric
- Chest expansion and, more importantly in infants, abdominal 'see-sawing'
- Pulse oximetry

Effects of respiratory failure on other physiology

- Heart rate:
  - increased by hypoxia, fever or stress
  - bradycardia a pre-terminal sign

- Skin colour:
  - hypoxia first causes vasoconstriction and pallor (via catecholamine release)
  - cyanosis is a late and pre-terminal sign
  - some children with congenital heart disease may be permanently cyanosed and oxygen may have little effect
- Mental status:
  - hypoxic child will be agitated first, then drowsy and unconscious
  - pulse oximetry can be difficult to achieve in agitated child owing to movement artefact

Circulation

- Heart rates 'at rest' at different ages

Age (yr)	Heart rate (beats/min)
<1	110–160
1–2	100–150
3–5	95–140
6–12	80–120
>12	60–100

Pulse volume

- Absent peripheral pulses or reduced central pulses indicate shock

Capillary refill

- Pressure on centre of sternum or a digit for 5 sec should be followed by return of circulation in skin within 2 sec
  - can be prolonged by shock or cold environmental temperatures
  - not a specific or sensitive sign of shock
  - should not be used alone as a guide to response to treatment

BP

- Cuff should cover >80% of length of upper arm
  - expected systolic BP = 85 + (age in yrs x 2)
- Hypotension is a late and pre-terminal sign of circulatory failure

Effects of circulatory inadequacy on other organs/physiology

- Respiratory system:
  - tachypnoea and hyperventilation occurs with acidosis
- Skin:
  - pale or mottled skin colour indicates poor perfusion
- Mental status:
  - agitation, then drowsiness leading to unconsciousness
- Urinary output:
  - <1 mL/kg/hr (<2 mL/kg/hr in infants) indicates inadequate renal perfusion

Features suggesting cardiac cause of respiratory inadequacy

- Cyanosis, not relieved by oxygen therapy
- Tachycardia out of proportion to respiratory difficulty
- Raised JVP
- Gallop rhythm/murmur
- Enlarged liver
- Absent femoral pulses

Disability

Primary assessment of disability

- Always assess and treat airway, breathing and circulatory problems before undertaking neurological assessment:
  - respiratory and circulatory failure will have central neurological effects
  - central neurological conditions (e.g. meningitis, raised intracranial pressure, status epilepticus) will have both respiratory and circulatory consequences

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Neurological function

- Conscious level: **AVPU** (**Figure 1**); a painful central stimulus may be applied by sternal pressure or by pulling frontal hair
- Posture:
  - hypotonia
  - decorticate or decerebrate postures may only be elicited by a painful stimulus
- Pupils – look for:
  - pupil size, reactivity and symmetry
  - dilated, unreactive or unequal pupils indicate serious brain disorders

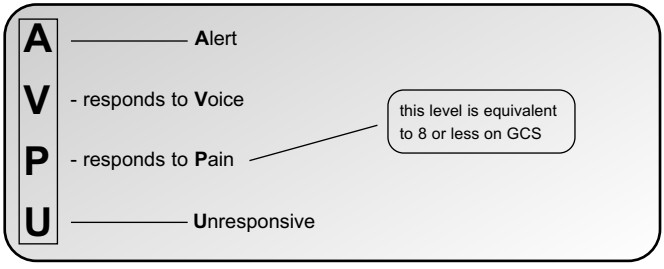
Respiratory effects

- Raised intracranial pressure may induce:
  - hyperventilation
  - Cheyne-Stokes breathing
  - slow, sighing respiration
  - apnoea

Circulatory effects

- Raised intracranial pressure may induce:
  - systemic hypertension
  - sinus bradycardia

Figure 1: Rapid assessment of level of consciousness



INTRAOSSEOUS INFUSION • 1/2

INDICATIONS

- Profound shock or cardiac arrest, when immediate vascular access needed and peripheral access not possible (maximum 2 attempts)
- allows rapid expansion of circulating volume
- gives time to obtain IV access and facilitates procedure by increasing venous filling

EQUIPMENT

- Intraosseous infusion needles for manual insertion or EZ-IO drill and needles (<40 kg: 15 mm pink; >40 kg: 25 mm blue) on resuscitation trolley
- Alcohol swabs to clean skin
- 5 mL syringe to aspirate and confirm correct position
- 10 mL sodium chloride 0.9% flush
- 20 or 50 mL syringe to administer fluid boluses
- Infusion fluid

Manual insertion is painful, use local anaesthetic unless patient unresponsive to pain. Infiltrate with lidocaine 1% 1–2 mL (maximum dose 0.3 mL/kg) and wait 90 sec

PROCEDURE

Preferred sites

Avoid fractured bones and limbs with fractures proximal to possible sites

Proximal tibia

- Identify anteromedial surface of tibia 1–3 cm below tibial tuberosity
- Direct needle away from knee at approx 90º to long axis of tibia
- Needle entry into marrow cavity accompanied by loss of resistance, sustained erect posture of needle without support and free fluid infusion
- Connect 5 mL syringe and confirm correct position by aspirating bone marrow contents or flushing with sodium chloride 0.9% 5 mL without encountering resistance
- Secure needle with tape
- Use 20 or 50 mL syringe to deliver bolus of resuscitation fluid

Figure 1: Access site on proximal tibia – lateral view

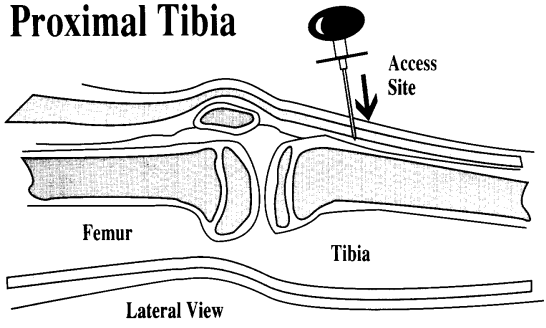


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Figure 2: Access site on proximal tibia – oblique view

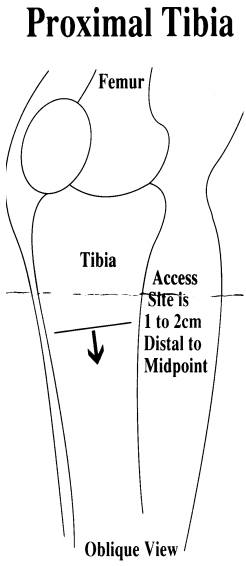
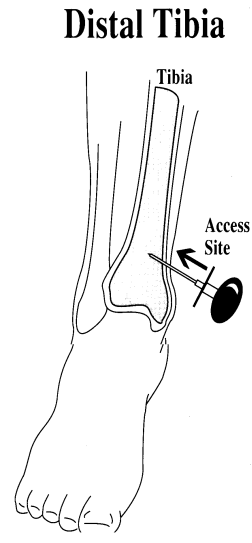


Figure 3: Access site on distal tibia



Distal tibia

- Access site on medial surface of tibia proximal to medial malleolus

Distal femur

- If tibia fractured, use lower end of femur on anterolateral surface, 3 cm above lateral condyle, directing needle away from epiphysis

COMPLICATIONS

- Bleeding
- Infection
  - revert to central or peripheral venous access as soon as possible
- Compartment syndrome
  - observe and measure limb circumference regularly
  - palpate distal pulses and assess perfusion distal to IO access site
- Pain from rapid infusion: give lidocaine 2% 0.5 mg/kg slow infusion

APPARENT LIFE THREATENING EVENT (ALTE) • 1/2

DEFINITION

A sudden, unexpected change in an infant's behaviour that is frightening to the observer and includes changes in two or more of the following:

- Breathing: noisy, apnoea
- Colour: blue, pale
- Consciousness, responsiveness
- Movement, including eyes
- Muscle tone: stiff, floppy

INVESTIGATION OF FIRST ALTE

Clinical history

- Feeding
- Sleeping
- Infant and family illness and medicines
- Gestation at delivery

Examination

- Full examination including signs of non-accidental injury

Assessment

- SpO_2
- Fundoscopy by paediatric ophthalmologist if:
  - recurrent
  - severe events (e.g. received CPR)
  - history or examination raises child safeguarding concerns (e.g. inconsistent history, blood in nose/mouth, bruising or petechiae, history of possible trauma)
  - anaemic

Investigations

Indicated if:

- Aged <1 month old
- <32 weeks gestation
- Previous illness/ALTE
- Examination abnormal
- Severe ALTE

Immediate

- FBC
- U&E, blood glucose
- Plasma lactate
- Blood gases
- Blood culture

Urgent

- Nasopharyngeal aspirate for virology
- Per-nasal swab for pertussis
- Urine microscopy and culture (microbiology)
- Urine biochemistry: store for possible further tests (see below)
- Chest X-ray
- ECG

If events recur during admission, discuss with senior role of further investigations (see below)

MANAGEMENT

Admit for observation

- SpO_2, ECG monitoring
- Liaise with health visitor (direct or via liaison HV on wards)
- Check if child known to local authority children's social care or is the subject of a child protection plan

After 24 hr observation

- If event brief and child completely well:
  - reassure parents and offer resuscitation training
  - discharge (no follow-up appointment)

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APPARENT LIFE THREATENING EVENT (ALTE) • 2/2

- All patients in following categories should have consultant review and be offered Care of Next Infant (CONI) Plus programme and/or home SpO_2 monitoring:
  - parents remain concerned despite reassurance
  - recurrent ALTE
  - severe ALTE (e.g. needing cardiopulmonary resuscitation/PICU)
  - <32 weeks gestation at birth
  - a sibling was either a sudden unexplained death (SUD) or had ALTEs
  - family history of sudden death

If events severe (e.g. CPR given) or repeated events

- Multi-channel physiological recording

Further investigations

Exclude following disorders:	
Gastro-oesophageal reflux	pH study +/- contrast swallow
Seizures	EEG
Intracranial abnormalities	CT or MRI brain
Cardiac arrhythmias	ECG and 24 hr ECG
Upper airway disorder	Sleep study
Hypocalcaemia	Ca and bone screen
Metabolic assessment	Urinary amino and organic acids Plasma amino acids and acylcarnitine
Abuse	Skeletal survey (including CT brain) Blood and urine toxicology (from admission) Continuous physiological or video recordings

DEFINITION

Sudden onset systemic life-threatening allergic reaction to food, medication, contrast material, anaesthetic agents, insect sting or latex, involving either:

- Circulatory failure (shock)
- Difficulty breathing from one or more of following:
  - stridor
  - bronchospasm
  - rapid swelling of tongue, causing difficulty in swallowing or speaking (hoarse cry)
  - associated with GI or neurological disturbance and/or skin reaction

Document

- Acute clinical features
- Time of onset of reaction
- Circumstances immediately before onset of symptoms

IMMEDIATE TREATMENT

Widespread facial or peripheral oedema with a rash in absence of above symptoms do not justify adrenaline or hydrocortisone. Give chlorphenamine orally

- See **Management of anaphylaxis** algorithm
- Remove allergen if possible
- Call for help
- **IM** adrenaline: dose by age (see **Algorithm**) or 10 microgram/kg:
  - 0.1 mL/kg of 1:10,000 in infants (up to 10 kg = 1 mL)
  - 0.01 mL/kg of 1:1000 (max 0.5 mL = 0.5 mg)
  - give in anterolateral thigh
- ABC approach: provide BLS as needed
  - if airway oedema, call anaesthetist for potential difficult airway intubation
  - if not responding to IM adrenaline, give nebulised adrenaline 1:1000 (1 mg/mL) 400 microgram/kg (max 5 mg)
  - treat shock with sodium chloride 0.9% 20 mL/kg bolus
  - monitor SpO_2, non-invasive blood pressure and ECG (see **Algorithm**)
- Repeat IM adrenaline after 5 min if no response

Do not give adrenaline intravenously except in cardiorespiratory arrest or in resistant shock (no response to 2 IM doses)

SUBSEQUENT MANAGEMENT

- Observe for a minimum of 6 hr to detect potential biphasic reactions and usually for 24 hr, especially in following situations:
  - severe reactions with slow onset caused by idiopathic anaphylaxis
  - reactions in individuals with severe asthma or with a severe asthmatic component
  - reactions with possibility of continuing absorption of allergen
  - patients with a previous history of biphasic reactions
  - patients presenting in evening or at night, or those who may not be able to respond to any deterioration
  - patients in areas where access to emergency care is difficult
- Monitor SpO_2, ECG and non-invasive BP, as a minimum

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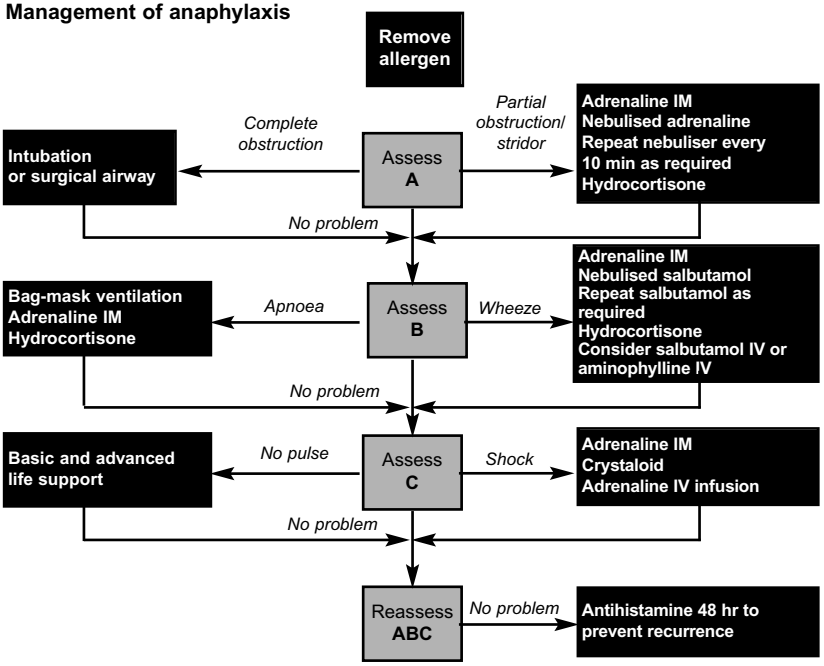
- Sample serum (clotted blood – must get to lab immediately) for mast cell tryptase if clinical diagnosis of anaphylaxis uncertain and reaction thought to be secondary to venom, drug or idiopathic at following times and send to immunology:
  - immediately after reaction
  - 1–2 hr after symptoms started when levels peak
  - >24 hr after exposure or in convalescence for baseline
- If patient presenting late, take as many of these samples as time since presentation allows
- Write mast cell tryptase on immunology lab request form with time and date of onset and sample to allow interpretation of results

DISCHARGE AND FOLLOW-UP

- Discuss all children with anaphylaxis with a consultant paediatrician before discharge
- Give following to patient, or as appropriate their parent and/or carer:
  - information about anaphylaxis, including signs and symptoms of an anaphylactic reaction
  - information about risk of a biphasic reaction
  - information on what to do if an anaphylactic reaction occurs (use adrenaline injector and call emergency services)
  - a demonstration of correct use of the adrenaline injector and when to use it
  - advice about how to avoid suspected trigger (if known)
  - information about need for referral to a specialist allergy service and the referral process
  - information about patient support groups
- Discharge with an emergency plan, including 2 adrenaline pen auto-injectors after appropriate training
- If still symptomatic give oral antihistamines and steroids for up to 3 days
- Refer as out-patient to a consultant paediatrician with an interest in allergy

Management of anaphylaxis



Drugs in anaphylaxis	Dosage by age			
	<6 months	6 months–6 yr	6–12 yr	>12 yr
Adrenaline IM: pre-hospital practitioners	150 microgram (0.15 mL of 1:1000)		300 microgram (0.3 mL of 1:1000)	500 microgram (0.5 mL of 1:1000)
Adrenaline IM: in-hospital practitioners	10 microgram/kg 0.1 mL/kg of 1:10,000 (infants and young children) OR 0.01 mL/kg of 1:1000 (older children)[1]			
Adrenaline IV	1 microgram/kg = 0.01 mL/kg of 1:10,000 over 1 min			
Crystalloid	20 mL/kg			
Hydrocortisone (IM or slow IV)	25 mg	50 mg	100 mg	200 mg

[1] Strength of IM adrenaline not intended to be prescriptive, 1:1000 or 1:10,000 is used depending on what is practicable: e.g. use of 1:1000 involves drawing up too small volumes when used in infants

ALSG: APLS Anaphylaxis Algorithm: Updated January 2010 reproduced with permission

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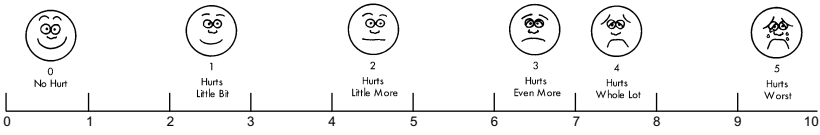
PAIN ASSESSMENT • 1/1

FLACC Behavioural	**SUGGESTED AGE GROUP:** 2 months to 7 years		
	SCORING		
CATEGORIES	**0**	**1**	**2**
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant quivering chin, clenched jaw
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking, or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid or jerking
Cry	No cry (awake or asleep)	Moans or whimpers, occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed	Reassured by occasional touching, hugging or being talked to, distractible	Difficult to console or comfort
Each of the five categories: **(F)** Face; **(L)** Legs; **(A)** Activity; **(C)** Cry; **(C)** Consolability; is scored from 0 - 2 which results in a total score between 0 and 10 *(Merkel et al, 1997)*			

WONG AND BAKER PAIN ASSESSMENT – SELF REPORT

- Suggested age group ≥4 yr
- Point to each face using the words to describe the pain intensity
- Ask child to choose a face that best describes their own pain and record the appropriate number



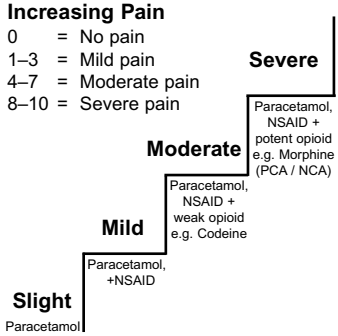
From Wong D.L., Hockenberry-Eaton M., Wilson D., Winkelstein M.L., Schwartz P.: Wong's Essentials of Pediatric Nursing, ed. 6, St. Louis, 2001, p. 1301. Copyrighted by Mosby, Inc. Reprinted by permission

Analgesic interventions

Analgesic ladder

Increasing Pain

0 = No pain
1–3 = Mild pain
4–7 = Moderate pain
8–10 = Severe pain



Play Specialist

Intervention by play staff

Preparation aid used: doll, verbal

Explanation, photos

Distraction: toys, bubbles, music, multi sensory, books

Refer all in need of analgesia and with behavioural concerns

NB: Check BNFc for contraindications/interactions/precautions etc

- For combination of analgesics to use, see **Analgesic ladder** in **Pain assessment** guideline

TOPICAL

Age group	Preparation	Time to onset	Comments
<1 month	Glucose syrup on pacifier (available as a tootsweet)	During procedure	For venepuncture or cannulation
>1 month	Ametop	30 min	Causes itch, lasts 4 hr
	LMX4	30 min	Wait 5 min after removing cream before cannulation
	EMLA	1 hr	Remove after 1 hr
>5 yr	Ethyl chloride	Immediately	If cannot wait for cream

MILD PAIN (pain score 1–3)

Drug and preparation	Dose	Maximum dose	Comments
Paracetamol [oral/nasogastric (NG)] ● Suspensions: ○ 120 mg/5 mL ○ 250 mg/5 mL ● Tablets/soluble 500 mg	● First dose 20 mg/kg **THEN** ○ aged 1–3 months: 30–60 mg 8-hrly ○ aged 3–6 months: 60 mg ○ aged 6–24 months: 120 mg ○ aged 2–4 yr: 180 mg ○ aged 4–6 yr: 240 mg ○ aged 6–8 yr: 250 mg ○ aged 8–10 yr: 375 mg ○ aged 10–12 yr: 500 mg ○ aged 12–16 yr: 750 mg ○ aged >16 yr: 500 mg –1 g	Max total dose in 24 hr ● Aged <1 month: 60 mg/kg/day ● Aged ≥1 month–18 yr: 90 mg/kg (max 4 g)	● For mild pain ● Increase dose interval in renal impairment ● Avoid large doses in dehydration, malnutrition, hepatic impairment
Paracetamol (rectal) ● Suppositories ○ 60 mg ○ 125 mg ○ 250 mg ○ 500 mg ○ 1 g	● First dose 30 mg/kg **THEN** ○ birth–3 months: 20 mg/kg 8-hrly ○ aged 3 months–12 yr: 20 mg/kg 4-hrly ○ aged >12 yr: 500 mg–1 g 4-hrly	● Max total dose in 24 hr: ○ aged <3 months: 60 mg/kg ○ aged ≥3 months: 90 mg/kg for 48 hr then 60 mg/kg ○ aged >12 yr: 4 g	● As for oral paracetamol ● For mild pain when oral/NG route not possible ● Suspension can be given rectally
Paracetamol (IV) 10 mg/mL (<33 kg use 50 mL vial via burette or in syringe) Prescribe in mg (not mL)	● <10 kg: 10 mg/kg 6-hrly ● 10–50 kg: 15 mg/kg 6-hrly ● >50 kg: 1 g 6-hrly	● Aged <1 month: 30 mg/kg/day ● <50 kg: 60 mg/kg/day ● >50 kg: 60 mg/kg/day ● Up to 4 g daily	● As for oral paracetamol ● For mild pain when oral/NG/PR route not possible ● Give over 15 min

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MODERATE PAIN (pain score 4–7)

Drug and preparation	Dose	Maximum dose	Comments
Ibuprofen ● Liquid 100 mg/5 mL ● Tablets 200 mg and 400 mg	● Aged 3 months–12 yr: 5 mg/kg 6–8 hrly ● Aged ≥12 yr: 200–600 mg 6–8 hrly	● Aged <12 yr: max 30 mg/kg/day ● Aged ≥12 yr: max 2.4 g/day	● If aged <3 months or <5 kg use only if recommended by consultant ● Avoid in renal dysfunction ● Contraindications: ○ shock ○ bleeding disorders ○ hypersensitive to aspirin or other NSAID ● Can be given to asthmatics if no history of NSAID-induced wheeze and chest clear on auscultation ● Caution in hypertension, heart failure
Diclofenac ● Tablets: ○ dispersible 50 mg (can be used to give smaller doses) ○ enteric coated 25 mg and 50 mg ● Suppositories 12.5 mg, 25 mg, 50 mg and 100 mg	● Aged >6 months: 300 microgram–1 mg/kg 8-hrly	● Max 150 mg/day	● As ibuprofen
Codeine ● Liquid 25 mg/5 mL ● Tablets 15 mg, 30 mg and 60 mg	● Aged <12 yr: 500 microgram–1mg/kg 4–6 hrly ● Aged ≥12 yr: 30–60 mg 4–6 hrly	● Max 240 mg/day	● For moderate pain ● Caution in hepatic impairment ● If aged <1 yr, use only if recommended by consultant ● Repeated doses increase risk of respiratory depression ● Caution if renal impairment, obstructive or inflammatory bowel disease, raised ICP, compulsive disorders ● Contraindications: ○ acute respiratory depression ○ paralytic ileus ● Not to be given with other opioids ● Prescribe laxatives if given for >24 hr

SEVERE PAIN IN CHILDREN AGED >1 YR (pain score 8–10)

In head injuries/respiratory difficulties/upper airway obstruction, use opioids only with consultant advice. Monitor children needing oxygen and parenteral opioids with SpO_2 +/- $TcCO_2$ in an HDU setting

Analgesic method and technique	Dose	Monitoring
Oral morphine ● Single dose before painful procedure may be useful ● Use if no IV access or for weaning from IV opioid ● If to be taken regularly consider use of prophylactic laxative	● Aged >1–12 yr: ○ 200–300 microgram/kg 4-hrly ● Aged >12 yr: ○ 5–10 mg 4-hrly (max 10 mg)	● **Respiratory rate**, maintain: ○ aged 1–2 yr, >16 breaths/min ○ aged 2–9 yr, >14 breaths/min ○ aged 10–16 yr, >12 breaths/min ○ if rate reduced, contact medical staff
Morphine patient/nurse-controlled analgesia (PCA/NCA) ● PCA suitable for children aged >5 yr (understand and will press button); NCA otherwise ● Nurses must be certified competent in use of PCA/NCA ● Use anti-reflux valve unless dedicated cannula ● Use morphine 1 mg/kg made up to 50 mL with sodium chloride 0.9% ○ maximum of 50 mg/50 mL	● If loading dose required: ○ experienced staff only ○ 50–100 microgram/kg ● Background infusion if used ○ 4–10 microgram/kg/hr ● Bolus dose ○ 10–20 microgram/kg ● Lockout time ○ 5–30 min ● Maximum dose in 4 hr of 400 microgram/kg	Hourly observations ● Pain score ● Sedation score ● Pump displays ● Syringe movement ● Respiratory rate ● SpO_2 if needed ● $TcCO_2$ if needed 4 hourly observations ● Vomiting/itching ● Urinary retention ● Inspection of IV site
Morphine infusion ● Use for severe pain when unable to use PCA/NCA ● Use anti-reflux valve unless dedicated cannula ● Use anti-siphon valve on line ● Use morphine 1 mg/kg made up to 50 mL with sodium chloride 0.9% ○ max of 50 mg/50 mL	● Loading dose of 100 microgram/kg given over 5–20 min (max 5 mg) ● Continuous infusion of 10–30 microgram/kg/hr ● Start at 20 microgram/kg/hr except after major surgery when start at 30 microgram/kg/hr and adjust according to pain and sedation scores	Hourly observations ● Pain score ● Sedation score ● Respiratory rate (as above) ● SpO_2 monitoring ● Syringe movement ● IV site for infection ● Urinary retention
IV intermittent morphine ● Infusion preferable	● Give slowly over 5 min ● Aged 1–12 yr: ○ 100 microgram/kg 4-hrly ● Aged >12 yr: ○ 2.5–5 mg 4-hrly	Hourly observations ● Pain score ● Sedation score ● Respiratory rate (as above) ● SpO_2 monitoring
SC intermittent morphine ● IV preferable ● Site 22/24 G SC cannula at time of surgery or using EMLA cream ○ suitable sites: uppermost arm, abdominal skin	● Flush with sodium chloride 0.9% 0.3 mL ● Prime cannula with morphine solution ● Morphine: ○ 100–200 microgram/kg 4-hrly ○ max 6 times in 24 hr	● Pain score ● Sedation score ● Respiratory rate (as above)

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SEVERE PAIN IN CHILDREN AGED <1 YR (pain score 8–10)

In head injuries/respiratory difficulties/upper airway obstruction, only use opioids with consultant advice. Monitor children requiring oxygen and parenteral opioids with SpO_2 +/- $TcCO_2$ in an HDU setting

Analgesic method and technique	Dose	Monitoring
Oral morphine ● Use if no IV access or for weaning from IV opiate	● Aged 1–6 months: 50–100 microgram/kg 4-hrly ● Aged 6–12 months: 100–200 microgram/kg 4-hrly	● Pain score ● Sedation score ● **Respiratory rate**, maintain: ○ if aged <6 months, >20 breaths/min ○ if aged ≥6 months, >16 breaths/min ○ if rate reduced, contact medical staff ● SpO_2 as appropriate
Morphine infusion ● Use anti-reflux valve unless dedicated cannula ● Use anti-siphon valve on line ● Use morphine 1 mg/kg made up to 50 mL with sodium chloride 0.9% ○ thus 1 mL/hr = 20 microgram/kg/hr	● Aged <1 month: 50 microgram/kg over 5 min then 5–20 microgram/kg/hr ● Aged 1–12 months: ○ 100 microgram/kg over 5 min then 10–30 microgram/kg/hr (0.5–1.5 mL/hr) ● Adjust in increments of 5 microgram/kg/hr according to response	Hourly observations ● Pain score ● Sedation score ● Respiratory rate (as above) ● SpO_2 monitoring ● Syringe movement ● Site for infection ● Urinary retention
IV intermittent morphine ● Infusion preferable	● Aged <1 month: 50 microgram/kg 6-hrly ● Aged 1–12 months: 100 microgram/kg 4-hrly	● Hourly observations for 24 hr then 4-hrly if stable ● Pain score ● Sedation score ● Respiratory rate (as above) ● SpO_2 monitoring
SC intermittent opiate ● IV preferable ● Site 24 G SC cannula at time of surgery or using EMLA cream ○ suitable sites: uppermost arm, abdominal skin	● Flush with sodium chloride 0.9% 0.3 mL ● Morphine: ○ aged <1 month: 100 microgram/kg 6-hrly ○ aged 1–6 months 100–200 microgram/kg 6-hrly (aged ≥6 months 4–6 hrly)	● Pain score ● Sedation score ● Respiratory rate (as above) ● SpO_2 as appropriate

SEDATION • 1/3

ASSESSMENT

Sedation and anaesthesia belong to the same spectrum of impaired consciousness

- In sedation, patient maintains the following vital functions without assistance:
  - protection of airway, swallowing, cough reflex
  - respiration
  - cardiovascular stability

Cautions

Discuss with anaesthetist before sedation if any of following present:

- Abnormal airway (including large tonsils)
- Sleep apnoea
- Respiratory failure
- Respiratory disease with significant functional compromise
- Active respiratory tract infection
- Cardiac failure
- Raised intracranial pressure
- Decreased conscious level
- Neuromuscular disease
- Bowel obstruction
- Significant gastro-oesophageal reflux
- Renal impairment
- Liver impairment
- Previous adverse reaction to sedation
- Very distressed child

Potential difficulties

Sedation can be difficult in children:

- Taking anti-epileptics (can result in increased or reduced effect of sedating drug)
- Already taking sedating drugs
- With behavioural difficulties

PREPARATION FOR SEDATION

Information required

- Age
- Weight
- Procedure for which sedation required
- Previous sedation history
- Other drugs being taken
- Other major diagnoses and implications in terms of respiratory function and upper airway competence
- Current health, including coughs, colds, pyrexia
- Oral intake status

Consent for sedation (all cases)

Discuss with parent(s):

- Unpredictable response to medication
- Paradoxical excitation
- Failure of sedation (may need repeat dose or general anaesthetic at future date)
- Over-sedation (maintaining airway, aspiration)

Fasting for moderate–heavy sedation

- There should be the following interval before procedure:
  - after a full meal: 6 hr
  - after milk: 4 hr
  - after clear fluids: 2 hr

For short, painless procedures (e.g. CT or X-ray), give infants aged <4 months normal milk feed only and allow them to sleep naturally

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EQUIPMENT

- Portable oxygen
- Portable suction
- Appropriately sized face mask and self-inflating resuscitation bag
- Two healthcare professionals trained in airway management with patient during sedation

DRUG CHOICE

Sedation drugs

Drug	Route	Onset	Duration	Dose	Comments
Chloral hydrate	● Oral ● Rectal	30 min–1 hr	1–2 hr	● Night sedation 30 mg/kg ● Pre-anaesthesia 50 mg/kg ● Scans 70 mg/kg ● max dose 2 g	● More efficacious in infants <15 kg or aged <18 months
Melatonin	● Oral	15–30 min	2–5 hr	● Aged ≤5 yr: 5 mg ● Aged >5 yr: 10 mg	● Use for sedation before EEG ● Use 5 mg initially, if no response, give further 5 mg
Temazepam	● Oral	45–90 min	up to 4 hr	● 0.5 mg/kg ● Up to 1 mg/kg for scans ● Max 30 mg	● Only if aged ≥2 yr ● CT, MAG3 scan
Midazolam	● Oral	15–30 min	1–2 hr	● Aged 1 month–18 yr: 500 microgram/kg (max 20 mg)	● Have flumazenil ready to give
	● Rectal			● Aged 6 months–12 yr: 300–500 microgram/kg (max 20 mg)	
	● Buccal			● Aged 6 months–10 yr: 200–300 microgram/kg (max 5 mg) ● Aged >10 yr: 6–7 mg	
	● IV	2–3 min		● 25–50 microgram/kg over 2–3 min, 5–10 min before procedure (aged 1–6 yr max 2 mg; aged 6–12 yr max 6 mg; aged 12–18 yr max 7.5 mg)	● IV cannulation (+ EMLA or local anaesthetic) ● More suitable for older children (not suitable for infants) ● Not for CT scan
Morphine sulphate	● Oral	15 min	2–3 hr	● Aged >1 yr: 200–300 microgram/kg (max 20 mg)	● May be combined with midazolam 500 microgram/kg oral for painful procedures

MONITORING

- Keep under direct observation
- Once asleep or if <1 yr, monitor saturation continuously
- Record saturation, heart rate and colour every 15 min
- Discontinue once conscious level returned to normal

SUBSEQUENT MANAGEMENT

Failed sedation

- Repeat maximum dose of initial drug used after expected period of onset
- If repeat dose fails:
- call anaesthetist who may give IV sedation (apply EMLA), or
- reschedule procedure for later date under general anaesthetic

Paradoxical excitement

- Do not attempt further drug dose
- Discuss with anaesthetist. If unavailable that day, reschedule procedure for later date under general anaesthetic

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IV FLUID THERAPY • 1/1

For previously well children aged 1 month–16 yr (excluding renal, cardiac, endocrinology, diabetic ketoacidosis and acute burns patients)

Hyponatraemia may develop as a complication of any fluid regime

If shock present, administer sodium chloride 0.9% 20 mL/kg or compound sodium lactate (Hartmann's) (10 mL/kg in the setting of trauma)
Repeat if necessary and call for senior help
Consider blood or colloid if relevant

Symptomatic hyponatraemia is a medical emergency

Estimate any fluid deficit and replace as sodium chloride 0.9% (with or without glucose 5%) OR compound sodium lactate (Hartmann's) over a minimum of 24 hr
Check plasma electrolytes

Calculate volume of maintenance and replacement fluids and select fluid type

VOLUME OF INTRAVENOUS MAINTENANCE FLUID

<10 kg: 100 mL/kg/day
10–20 kg: 1000 mL + 50 mL/kg/day for each kg >10 kg
>20 kg: 1500 mL + 20 mL/kg/day for each kg >20 kg

- **Up to a maximum of 2500 mL/day (males) or 2000 mL/day (females)**

Use bags with potassium chloride premixed
Check serum potassium

VOLUME OF INTRAVENOUS REPLACEMENT FLUID
(to replace losses, reassess every 4 hr)

- Fluids used to replace ongoing fluid losses should reflect the composition of fluid being lost. Sodium chloride 0.9% or sodium chloride 0.9% with potassium 0.15% will be appropriate in most cases

TYPE OF INTRAVENOUS MAINTENANCE FLUID

In following circumstances, administer isotonic fluids such as sodium chloride 0.9% with potassium 0.15%

- Plasma sodium <135 mmol/L
- Intravascular volume depletion
- CNS infection
- Peri- and post-operative patients
- Hypotension
- Head injury
- Bronchiolitis
- Sepsis
- Excessive gastric or diarrhoeal losses
- Salt wasting conditions e.g. diabetes, CF
- Hypernatraemic dehydration (Na >160 mmol/L)

Otherwise, children may be safely administered sodium chloride 0.45% with glucose 5% and potassium 0.15%

Patients requiring both maintenance fluids and replacement of ongoing losses should receive a single isotonic fluid such as sodium chloride 0.9% with potassium 0.15% or sodium chloride 0.9% with glucose 5%

Monitoring

- Weigh patient before starting fluid therapy, then daily if possible
- Check plasma electrolytes before commencing infusion, except before majority of elective surgery
- Check plasma electrolytes every 24 hr whilst intravenous fluids are being administered
  - if abnormal or if plasma sodium <130 mmol/L measure every 4–6 hr
- Check plasma electrolytes immediately if clinical features suggestive of hyponatraemia; features include nausea, vomiting, headache, irritability, altered level of consciousness, seizure or apnoea
- Maintain fluid balance chart to record input and output. Oliguria may be due to inadequate fluid, renal failure, obstruction or effect of ADH
- Some acutely ill children with increased ADH secretion may benefit from restriction of maintenance fluids to ⅔ of normal recommended volume
- Contact senior paediatrician, PICU or paediatric anaesthetist if uncertain or ongoing fluid losses

When using volumetric pump to administer IV fluids

- Do not leave bag of fluid connected (blood components excepted)
- Nurse to check following hourly:
  - infusion rate
  - infusion equipment
  - site of infusion
- Close all clamps and switch off pump before removing giving set

LONG LINE INSERTION • 1/3

INDICATIONS

- 'Short' long lines in patients requiring 5–14 days IV therapy either in hospital or at home
- Peripherally inserted central catheter (PICC) for drugs that have to be given centrally (e.g. if they cause phlebitis), if risk of infection high (e.g. parenteral nutrition) or for access >14 days

EQUIPMENT

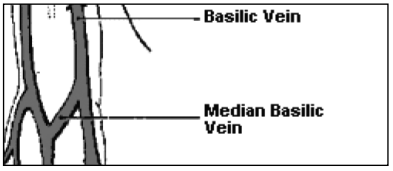
- Assistant
- Long line
- 'Short' long line:
- Leaderflex 22 G (2.5 F) line 8 or 20 cm
- PICC:
- Vygon PICC 3, 4 or 4.5 F 60 cm Lifecath (expert silver coated)
- Vygon Nutriline 2, 3 or 4 F 30 cm
- Vygon Neocath or Epicutaneo-cave catheter 2 F (23 G) 15, 30 or 50 cm has different insertion technique, not recommended except neonates

DO NOT ATTEMPT INSERTION UNLESS YOU ARE FULLY TRAINED
Use whichever line you have been trained to use

- Flush solution: sodium chloride 0.9% 5 mL
- Single dressing pack
- Sterile gloves
- Sterile scissors
- 2 extra sterile towels
- 5 mL syringe/green needle
- Tape measure
- Sterile clear dressing (e.g. Opsite®/Tegaderm®)
- Incontinence pad
- 2 extra packs gauze swabs
- Alcoholic chlorhexidine (or other skin antiseptic)
- 1 injectable bung
- 3 wide Steri-strips® (optional to secure line)
- Sterile untoothed forceps (to feed line up butterfly)

PROCEDURE

PICC line preparation



- Assess whether patient will need sedation. Rarely, children with needle phobia will need the line inserted under general anaesthetic. Arrange appropriate person to administer sedation
- If necessary, shave arm to avoid hair plucking when dressing removed
- Specify exactly where you would like topical local anaesthetic cream sited. Basilic vein (medial) is usually best. Apply anaesthetic cream to chosen veins (3 sites) at least 1 hr before starting procedure
- A BP cuff inflated to 80 mmHg is a more reliable tourniquet than either an elastic strip or a nurse's squeeze
- Check patient's notes for comments about previous line insertions. Some veins can be particularly difficult and patient can often provide guidance
- Check whether blood samples are required
- Gather all necessary equipment including a spare line (unopened)

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Consent

- Explain procedure and reassure patient
- Obtain and record consent

Premedication and position of patient

- Position patient seated in chair or lying with his/her arm stretched out and supported by table or bed (on a utility drape)
- ensure patient in position and comfortable, and lighting optimal
- Measure distance from site of insertion to sternal notch (if inserting in arm) or xiphisternum (if inserting in leg) so catheter tip is placed outside heart

Aseptic non touch technique (ANTT)

- Wash hands, and put on apron/gown and sterile gloves
- Clean patient's skin thoroughly with alcoholic chlorhexidine and allow to dry in area of planned insertion
- Drape sterile sheet to expose only chosen vein, and cover surrounding areas to provide working room and a flat surface on which to rest your line, forceps and flush

Nutriline PICC line

- Assemble line fully and flush with sodium chloride 0.9%1 mL to ensure patency
- Place everything you will need onto sterile sheet within reach
- Ask assistant to apply tourniquet (or squeeze patient's arm), but remain ready to release
- Check patient is ready for you to start
- Be careful: introducer for the PICC line is **much stiffer** than a standard cannula and more likely to perforate the entire vein
- Insert peelable cannula until blood flowing freely (it is not necessary to thread needle into vein) in some patients this will come quite quickly so have catheter ready
- Ask assistant to release tourniquet to reduce blood flow
- Taking the PICC line in forceps, pass it up through cannula. At about 5 cm, you will reach tip of the cannula. If line passes easily beyond 6 cm, you have probably succeeded. Resistance at any point usually indicates failure to thread vein, or curling of line. Rotating butterfly needle so that the bevel faces downwards may help to introduce line into vein if it will not thread more than 5 cm
- Insert line to previously measured distance from site of insertion
- When tip of line is judged to be in correct position, carefully withdraw sheath and remove from around line by pulling apart the two blue wings
- Pressing firmly on insertion site with a piece of gauze, remove cannula
- Without releasing pressure on entry site (it may bleed for a few minutes), reassemble line and flush with sodium chloride 0.9% 2 mL
- With sterile scissors, cut rectangle of gauze (1 x 2 cm) to prevent hub of line rubbing skin
- Check all connections are firmly tightened. Coil any unused line next to insertion site and secure with Steri-strips®
- Cover entry site, connections and all exposed line with one piece of clear dressing (e.g. Opsite®)
- X-ray line with 0.5 mL of contrast (e.g. Omnapaque 240) in the line to check tip position if near heart or if no blood flushes back up line. Do not draw blood back up line (this increases risk of line blockage)
- Flush once more and line is then ready to use

Leaderflex lines

- These are inserted using Seldinger technique
- Cannulate target vein with either needle provided or a blue cannula
- Feed guidewire into vein through cannula sheath and remove sheath leaving wire *in situ*
- Feed line over guidewire and into vein with a gentle twisting action. It is important that, at any time, operator is able to grasp directly either free end of wire or wire itself as it passes through skin, to ensure that it does not pass entirely into vein
- Remove guidewire and secure line in place
- It is not necessary to verify position of 8 cm lines radiologically

Use an aseptic technique when accessing the system or for dressing changes

AFTERCARE

- Aim to insert to 20 cm and tape remaining silastic length to skin with an adhesive dressing e.g. Steri-strip®
- Place a folded half gauze swab under the blue hub before taping down with adhesive, then cover with transparent dressing, minimising contact between gauze and transparent dressing in case removal is required for troubleshooting
- Flush after each use with sodium chloride 0.9% 2 mL

LONG LINE CARE

- Keep dressing clean and intact
- Maintain aseptic technique for accessing system and dressing changes. Before accessing system, disinfect hub and ports with disinfectant compatible with catheter (e.g. alcohol or povidone-iodine)
- Assess site at least daily for any signs of infection and remove if signs of infection are present (only short-term CVCs)
- Replace administration sets every 24 hr and after administration of blood, blood products and lipids. Routine catheter replacement is unnecessary
- Assess need for device daily and remove as soon as possible
- Document insertion and all interventions in patient notes

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PRE-OP FASTING • 1/1

PRINCIPLES

- Do not fast patients for longer than necessary for their safety under general anaesthesia
- Do not deny fluids for excessively long periods; allow patients to drink within these guidelines
- Use theatre time efficiently

Ideally give all children (especially those aged <2 yr) clear fluids up to 2 hr pre-operatively. Liaise closely with theatre to discover approximate time of patient's operation

POLICY

- Solid food and milk (including formula) up to 6 hr before elective surgery
- Breast milk up to 4 hr before elective surgery
- Encourage patients to take clear oral fluids up to 2 hr before elective surgery. Thereafter, sips of water may be taken to enable tablets to be swallowed
- clear fluids do **not** include fizzy drinks

PROCEDURE

All children aged ≥1 yr

Morning operating lists

- No solid food after midnight
- Water or diluted squash to finish before 0630 hr

Afternoon operating lists

- Light breakfast (including toast, or small bowl of cereal), to finish before 0700 hr
- Water or diluted squash to finish before 1100 hr

Infants/children aged <1 yr

Morning operating lists

- Last formula milk feed before 0230 hr
- Last breast milk feed before 0430 hr
- Water or diluted squash to finish before 0630 hr

Afternoon operating lists

- Last formula milk feed before 0700 hr
- Last breast milk feed before 0900 hr
- Water or diluted squash to finish before 1100 hr

Nursing and medical staff should ensure that all children are encouraged to drink clear fluids (e.g. water or diluted squash) until 2 hr before anaesthesia/surgery

POST GA MONITORING EX-PREMATURE INFANTS • 1/1

- Risk of apnoea after general anaesthetic (GA)
  - increased if anaemic
  - with chronic lung disease who have required oxygen treatment within last 6 months

MANAGEMENT

Pre-operative

- Check haemoglobin
  - if Hb <90 g/L, arrange transfusion
- Arrange overnight stay for post-operative monitoring if age (weeks) <[3 x (38 – gestational age in weeks)] e.g. baby born at 30 weeks gestation would be kept overnight after GA if <24 weeks old. A 36 week baby would be allowed home after GA if >6 weeks old

Immediate post-GA period

- Transfer patient with oxygen supply, continuous SpO_2 monitoring and full resuscitative equipment
- Admit patient to a designated HDU ward area

Subsequent post-GA management

- High dependency nursing care
- Monitoring to include:
  - continuous pulse oximetry
  - continuous ECG
  - continuous respiratory rate
  - transcutaneous CO_2
- If apnoea >15 sec:
  - immediate respiratory support by nurse (airway manoeuvres, bag and mask ventilation)
  - contact on-call SpR
  - liaise with anaesthetist responsible for patient
  - review period of HDU care

DISCHARGE AND FOLLOW-UP

- Discharge patient home same day or next day as calculated by above formula providing there have been no apnoeic episodes

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RECOGNITION AND ASSESSMENT

Definition

Asthma is a chronic inflammatory disorder of the airways with reversible obstruction

Symptoms and signs

- Breathlessness
- Wheeze
- Cough
- Nocturnal cough
- Tight chest
- Bilateral wheeze

- Symptoms and signs tend to be:
  - variable
  - intermittent
  - worse at night
  - provoked by triggers, including exercise

Mild/moderate

- Normal vital signs
- Mild wheeze
- Speaks in complete sentences or feeding
- SpO_2 >92% in air
- PEF >50% in patient aged ≥7 yr

Severe

- Too breathless to talk/feed
- Tachypnoea (>40 breaths/min if aged <5 yr; >25 breaths/min if aged >5 yr)
- Tachycardia (>140 beats/min if aged <5 yr; >125 beats/min if aged >5 yr)
- Use of accessory muscles, recession subcostal and intercostal, flaring of alae nasi
- SpO_2 <92% in air
- Peak expiratory flow (PEF) ≤50% predicted/best

Life-threatening

- Cyanosis/pallor
- Decreased air entry/silent chest
- Poor respiratory effort
- Altered conscious level
- Irritable/exhausted
- SpO_2 <92% in air
- PEF ≤33% in those aged ≥7 yr

Patients with severe or life-threatening attacks may not be distressed and may not have all these abnormalities. Presence of any one of these should alert doctor

Differential diagnosis

- Foreign body
- Pneumonia
- Pneumothorax
- Aspiration
- Cystic fibrosis
- Tracheobronchomalacia
- Gastro-oesophageal reflux

Assessment

- Record:
  - respiratory rate and effort
  - recession
  - heart rate
  - air entry
  - oxygen saturation in air
  - if aged ≥7 yr, peak expiratory flow (PEF)
  - conscious level

Do not take any samples for routine blood tests or routine blood gases. Routine chest X-ray is unnecessary in a child with asthma

IMMEDIATE TREATMENT

- Follow algorithm **Management of acute wheezing in children**

Senior assessment

If you are worried about child's conscious level or there is no response to nebulised salbutamol or poor respiratory effort:

- Call senior doctor for further assessment
- Site an IV line
- Initial dose of salbutamol IV over 5 min (max 250 microgram)
  - aged <2 yr: 5 microgram/kg
  - aged >2 yr: 15 microgram/kg
- Using 500 microgram/mL injection preparation dilute to a concentrate of 50 microgram/mL with sodium chloride 0.9%
  - e.g. withdraw 250 microgram = 0.5 mL and make up to a total volume of 5 mL using sodium chloride 0.9% = 250 microgram in 5 mL

Not responding within 15 min

- Salbutamol 1–2 microgram/kg/min continuous infusion
  - use 1 mg/mL solution for IV infusion dilute 10 mg (10 mL) to concentration of 200 microgram/mL made up to 50 mL with sodium chloride 0.9%
- If not responding increase up to 5 microgram/kg/min for 1 hr then reduce back to 2 microgram/kg/min
- If requiring >2 microgram/kg/min admit to PICU
- Use $TcCO_2$ monitor
- Continue with high flow oxygen and continuous salbutamol nebuliser while waiting

Drug doses

- Salbutamol nebulised, driven by 6–8 L/min oxygen:
  - aged <5 yr, 2.5 mg
  - aged >5 yr, 2.5–5 mg
- Ipratropium bromide (Atrovent®) nebulised:
  - aged <12 yr, 250 microgram
  - aged >12 yr, 500 microgram
- Prednisolone 0.5 mg/kg oral:
  - aged <2 yr = max 10 mg once daily
  - aged 2–5 yr = max 20 mg once daily
  - aged >5 yr = max 30 mg once daily
- Hydrocortisone slow IV injection:
  - aged <2 yr, 4 mg/kg (max 25 mg) 6-hrly
  - aged 2–5 yr, 50 mg 6-hrly
  - aged 5–18 yr, 100 mg 6-hrly

Monitoring

- Record heart rate and respiratory rate every 10 min
- Continuous SpO_2
- Cardiac monitoring
- Baseline U&Es (capillary blood gas for potassium)

SUBSEQUENT MANAGEMENT

Follow the algorithm **Management of acute wheezing in children**

Previous history

- When recovering, ask about:
  - previous episodes of wheeze, similar episodes
  - triggering factors, seasonal variation
  - nocturnal cough
  - family history of asthma, hay fever, eczema, other atopy
  - smokers in the family (including child)

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- days off school because of asthma
- number of courses of prednisolone used in last year
- pets
- drug history (device and dose) especially any bronchodilators/inhaled corticosteroids and their effect, particularly need to use beta-agonists

DISCHARGE AND FOLLOW-UP

Discharge criteria

- SpO_2 in air >94%
- Respiratory rate: <40 breaths/min aged <5 yr; <30 breaths/min aged >5 yr
- Heart rate: <140 beats/min aged <5 yr; <125 beats/min aged >5 yr
- Peak flow: ≥75% predicted/best in those aged >7 yr
- Stable on 4-hrly treatment

Discharge home same day if:

- Child has made a significant improvement and has remained stable for 4 hr
- Parents:
- understand use of inhalers
- have a written personal asthma action plan
- have a written discharge/weaning salbutamol information leaflet
- know how to recognise signs of deterioration and the actions to take

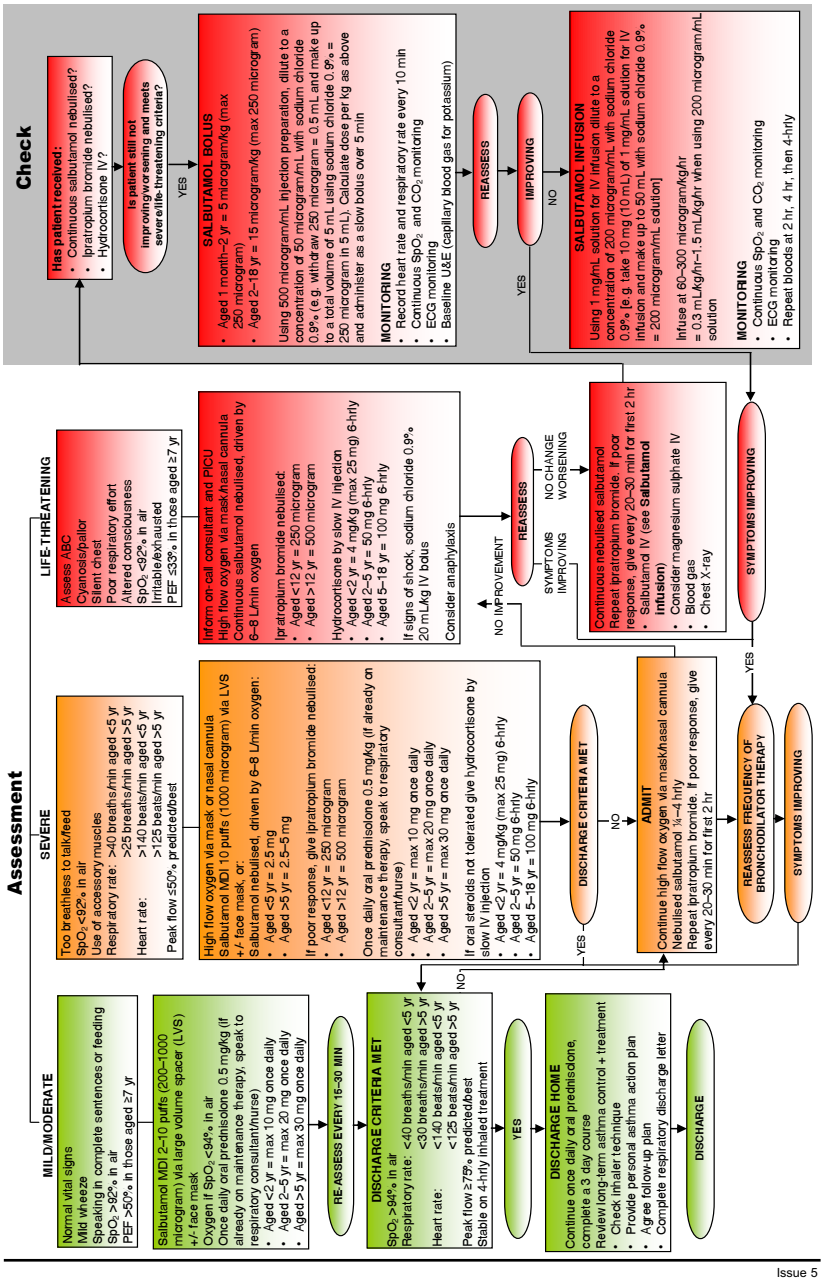
Discharge treatment

- Prescribe beta-agonist with spacer
- Give prednisolone 0.5 mg/kg daily for 3–5 days (if already on oral prednisolone maintenance therapy speak to respiratory consultant/nurse)
- Educate on use of PEF meter if aged >6 yr (not if child has never used one before)
- Discuss follow-up in either nurse-led asthma clinic or consultant clinic

Chronic management

- Give inhaled corticosteroid if any of following:
- frequent episodes
- bronchodilators used most days
- nocturnal and/or exercise-induced symptoms
- other atopic symptoms and strong family history of atopy
- If recurrent upper respiratory tract problems or allergic rhinitis triggering attacks, give oral antihistamines +/- steroid nasal spray

Algorithm: Management of acute wheezing in children



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RECOGNITION AND ASSESSMENT

Definition

- Acute viral inflammatory illness of small airways that occurs in winter epidemics and affects children aged <2 yr, with peak incidence at around 6 months

Symptoms and signs

- Coryzal symptoms for 2–5 days before presentation
- Cough (sometimes paroxysmal)
- Intermittent wheeze
- Irritability and poor feeding
- Mild pyrexia – rarely higher than 38.5ºC
- Respiratory distress with progressive tachypnoea, flaring of alae nasi and intercostal recession
- Apnoea or hypoventilation
- Hyperinflated chest on examination
- Widespread fine crackles and wheeze over both lung fields

Differential diagnosis

- Recurrent viral-induced wheeze
- Early asthma
- Cystic fibrosis
- Pertussis
- Recurrent aspiration
- Foreign body in trachea
- Congenital lung anomaly

Investigations

- SpO_2 while breathing air
- Capillary blood gas if:
  - respiratory rate >80 breaths/min
  - transcutaneous PCO_2 >6 kPa
  - SpO_2 <92% in >50% inspired oxygen
  - severe respiratory distress
- Avoid tests that do not contribute to immediate management. Perform following only for specific indications:
  - viral nose swab for influenza for oseltamivir if admission required when flu prevalence high
  - nasopharyngeal aspirate for respiratory virus immunofluorescence in severely immunocompromised patient to plan antiviral treatment
  - chest X-ray if there are localising signs, cardiac murmur or atypical presentation (e.g. aged >18 months)
  - U&E if there is a plan for IV fluids
  - blood cultures if signs of sepsis or temperature >38.5ºC

IMMEDIATE TREATMENT

- Nurse in cubicle, or in bay with children with same diagnosis
- Strict hand washing to support infection control and use apron for patient contact
- Nurse head up to reduce splinting of diaphragm
- Clear airway by suction of nares and mouth
- Use sodium chloride 0.9% nose drops before suction
- Nebulised sodium chloride 3% 4 mL 6-hrly

Respiratory

- If oxygen saturation ≤92% in air, give oxygen via face mask with a reservoir bag
- humidify oxygen
- if mask not tolerated, use nasal prongs for oxygen flow up to 1 L/min in children ≤5 kg body weight or up to 2 L/min in children >5 kg
- use humidifier if available to warm oxygen

- In patients with impending respiratory failure, review hourly. Consider additional respiratory support with CPAP or humidified high flow cannulae (e.g. Vapotherm) if two or more of following are present:
- respiratory rate >60 breaths/min or bradypnoea
- severe intercostal recession and indrawing
- need for >2 L/min oxygen via nasal prongs: SpO_2 <90% in >50% oxygen or cyanotic episodes despite supplemental oxygen (except cyanotic congenital heart disease)
- rising $PaCO_2$ (>3 kPa from baseline)
- respiratory acidosis (pH <7.20)

Circulation and hydration

- Assess circulation and treat shock if present
- Correct dehydration if present
- Use IV fluids if oral fluids not tolerated or significantly increased work of breathing
- restrict intake to 80% of estimated maintenance requirements (see I**V fluid therapy** guideline) using sodium chloride 0.9% with 10 mmol potassium chloride per 500 mL
- check U&E at least once every 12 hr while giving intravenous fluids (more frequently if abnormal), and adjust volume and potassium content accordingly

Feeds

- Normal feeds (breast, bottle, solids) if tolerated
- NG tube feeds if:
- oral intake by normal route insufficient **and**
- airway protective reflexes test normal on suctioning **and**
- patient well enough to tolerate NG feeds
- IV fluids (as above) if:
- persistent respiratory rate >80 breaths/min
- persistent vomiting
- oxygen saturation <92% despite supplemental oxygen
- deterioration of respiratory status during nasogastric feeding
- marked increase in work of breathing with poor coordination of sucking, swallowing and breathing

Drug treatment

- In immunocompetent patients, drug treatment and physiotherapy (in acute phase) are ineffective. **Do not routinely prescribe** salbutamol, ipratropium bromide (Atrovent®), adrenaline, antibiotics or corticosteroids
- For babies aged <6 weeks or patients with temperature >39ºC, discuss antibiotics with consultant
- If symptoms <48 hr and influenza test positive or high prevalence influenza (see www.hpa.org.uk) and risk factors (chronic respiratory, renal, liver, neurological or cardiovascular disease, diabetic or immunocompromised) give oseltamivir

Criteria for admission

Absolute

- Apnoea
- Underlying cardiac defects, especially large left to right shunt
- SpO_2 <92% in air in a child in the early phase of the illness
- Inadequate feeding
- Dehydration
- Diagnostic uncertainty

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Relative

- Re-attends A&E in <48 hr
- Aged <6 weeks (corrected gestational age)
- Unsatisfactory family circumstances and impaired ability to care for unwell child
- Younger children (i.e. aged <6 months), presenting earlier in illness (<3 days symptoms)
- Pre-existing lung disease, including chronic lung disease, ex-preterm, cystic fibrosis: inform speciality consultant
- Other pre-existing chronic disease (e.g. neurodegenerative)

MONITORING TREATMENT

- Standard nursing observations
- Continuous oxygen saturation monitoring if patient requires supplemental oxygen
- Transcutaneous CO_2 monitoring if patient using oxygen via nasal prongs at ≥2 L/min (approximately ≥60% oxygen) or has history of apnoea or colour changes
- Continuous heart and respiratory rate monitoring if patient requires additional respiratory support

SUBSEQUENT MANAGEMENT

- Fluid balance
- Oxygen support:
  - test the need for support 6-hrly
  - keep oxygen saturation ≥92% in recovery phase
  - wean from nasal prongs to air as tolerated

DISCHARGE AND FOLLOW-UP

- Discharge home when:
  - fully fed orally
  - SpO_2 >92% in air
- Hospital follow-up if:
  - ventilated on PICU
  - consolidation on chest X-ray (first reassess clinically, do not request 'routine' follow-up X-ray)
  - ex-preterm with chronic lung disease
- GP follow-up in all other cases

DEFINITION

- Acute viral inflammation of upper airway causing oedema of larynx and trachea and presenting with stridor
- Causative agent: parainfluenza virus (sometimes influenza, respiratory syncytial virus, rhinovirus)

Aetiology

- Aged 6 months–6 yr (peak age 2 yr)
- Seasonal peak: Spring and Autumn
- Transmission: usually by droplet spread
- Incubation period 2–6 days

Differential diagnosis of stridor

Acute

- Croup
- Epiglottitis (rare since immunisation against *Haemophilus influenzae* type B)
- Bacterial tracheitis
- Foreign body

Chronic

- Allergic airways disease (recurrent croup)
- Congenital abnormality e.g. laryngeal haemangioma
- Laryngomalacia
- Foreign body
- Laryngeal papilloma

CROUP

Symptoms and signs

- Preceding coryzal illness
- Fever
- Harsh bark/seal-like cough
- Hoarse voice
- Inspiratory stridor
- Symptoms worse at night
- Child does not look toxic

Assessment

- Record croup severity:
  - C – Cyanosis
  - R – Recession of chest
  - O – Oxygen saturations (keep >92%)
  - UP – Upper airway obstruction e.g. stridor
  - respiratory rate
  - heart rate
  - level of consciousness
- **Do not examine throat as it may cause acute severe/total obstruction**
- Do not distress child
- Any clinical concerns call consultant paediatrician immediately

Severity

Mild croup

- Barking cough
- Stridor
- No recession
- No cyanosis

Moderate croup

- Intermittent stridor at rest
- Mild recession
- Alert and responsive

Severe croup

- Stridor at rest
- Cyanosis
- Oxygen saturation <92% in air
- Moderate to severe recession
- Apathetic/restless

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Investigations

- No investigations necessary, do not attempt to take blood or put in cannula
- If diagnosis unclear, or child severely unwell, call consultant as an emergency measure

IMMEDIATE MANAGEMENT

Mild to moderate croup

- Antipyretics
- Adequate fluid intake
- Leaflet on croup and reassurance
- Oral dexamethasone 150 microgram/kg, can be repeated 12 hr later if symptoms persist
- Admit/observe for 4 hr and reassess
- If better, discharge with 1 dose of dexamethasone 150 microgram/kg oral, telling parents to use if symptoms persist 12–24 hr later. If dexamethasone not available as TTO, discharge with prednisolone 1 mg/kg as a single dose 12–24 hr after dexamethasone if symptoms persist

If parents do not clearly understand what to do, do not discharge

Severe croup

- Keep child and parents calm: do not upset child e.g. by forcing oxygen mask onto face or examining throat; nurse on parents lap and in position they find comfortable
- Nebulised adrenaline 400 microgram/kg to max 5 mg (0.4 mL/kg to max 5 mL 1:1000) relieves symptoms, but short duration of action
- Dexamethasone 150 microgram/kg oral (or if child refuses to swallow oral medication, nebulised budesonide 2 mg)
- High flow oxygen 15 L/min via mask with reservoir bag
- Contact on-call consultant paediatrician urgently to assess clinical situation
  - discuss whether to involve on-call paediatric anaesthetist and ENT surgeon
- If no sustained improvement with adrenaline and dexamethasone:
  - secure airway in theatre by experienced anaesthetist
  - transfer to PICU

DISCHARGE AND FOLLOW-UP

- Leaflet on croup
- Antibiotics, antitussives and humidified air do not help
- Advise paracetamol to control fever and encourage oral fluid intake
- Advise parents to seek help urgently if any of the following are present:
  - drooling
  - laboured breathing
  - persistent fever
  - biphasic/worsening stridor
  - cyanosis
  - reduced level of consciousness/confusion
- No need for follow-up of croup

CYSTIC FIBROSIS – ADMISSION • 1/2

ARRANGING ADMISSION

- Via CF nurse specialist with ward sister
- Refer to admission plan in notes or clinic letter
- Always admit to a cubicle

ADMISSION PROCEDURE

- Plot baseline weight, height
- Perform flow volume loop spirometry on admission day (aged ≥6 yr)
- Write up drug chart before parents leave
- Check whether annual bloods could conveniently be taken now (see **Annual bloods**)
- Ask nursing staff to inform physiotherapist and dietitian on day of admission
- Check specific aspects of management or investigations, as described by CF team
  - for IV antibiotics, see **Cystic fibrosis – Exacerbation** guideline
  - for bowel blockage, see **Cystic fibrosis – Distal intestinal obstructive syndrome (DIOS)** guideline

INVESTIGATIONS

Annual bloods

- All children attending CF clinics have annual blood screening
- Perform annual bloods if admission within a month of annual screening (usually at time of birthday):
  - during insertion of a long line or Port-a-cath needle, or when checking tobramycin level

All ages

- FBC and film
- Vitamins A, D, E
- Parathyroid hormone
- U&E, creatinine, chloride, calcium, magnesium, phosphate, albumin, total protein, alkaline phosphatase, bilirubin, AST/ALT, GGT, CRP
- Glucose

If aged >5 yr

All of the above **plus:**

- If symptoms could be caused by allergic bronchopulmonary aspergillosis, specific IgE to aspergillus and aspergillus precipitins. Also total IgE
- If diabetic, HbA_{1c}

If aged ≥10 yr

- Add glucose tolerance test (at 0, 60 and 120 min)
- Baseline DEXA scan (repeated every 2–3 yr)

Chest X-ray

- Most children have a chest X-ray every 6–12 months so another may not be necessary
- Check when latest was taken and, if in doubt, discuss with CF consultant

Lung function and oxygen saturation

- Measure FVC and FEV_1 using ward spirometer (physiotherapist/trained nurse can take these measurements if requested):
  - in all children who can blow reliably (usually from aged 6 yr)
  - on admission and at least weekly, preferably before ward rounds
  - towards the end or after completion of a course of IV antibiotics, take measurements before and after inhalation of salbutamol MDI 4–8 puffs via a spacer
- Monitor oxygen saturation overnight for first 2 nights after admission
  - if saturations <91%, give oxygen via nasal cannulae or face mask

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Microbiology

- In hospital, request twice weekly sputum/cough swab
- usually performed by physiotherapist but check this has been done
- If new pathogen found, see **Cystic fibrosis – Microbiology** guideline and cross-infection

Screening for hyperglycaemia

About 8% of children with CF develop diabetes after age 10 yr, usually manifests as weight loss; ketoacidosis is rare

- If taking regular oral corticosteroids, screen for glucose intolerance at admission
- During first 24 hr after admission, request glucose stick profile before breakfast, 1–2 hr after every meal, and at 0200 hr if on overnight feeds
- If prednisolone started or dosage increased during admission, repeat glucose stick profile
- If blood glucose elevated, discuss with CF team

NUTRITION

- Always involve dietitians
- Weigh twice weekly, **in nightwear** and **before breakfast** (weigh babies naked if possible)
- Continue normal supplements

Pancreatic enzyme supplements

- Continue same type and dose of pancreatic supplement as already prescribed

Starting dosage for newly diagnosed child

- **Infants**
  - Creon Micro for children ½ scoop (2500 units lipase) to 1 scoop (5000 units lipase) per 120 mL milk or breast feed

OR

  - Creon 10,000 one-quarter (2500 units lipase) to one-half capsule (5000 units lipase) per 120 mL milk or breast feed

- **Children**
  - starting dose Creon 10,000 2 capsules per meal, 1 capsule per snack
- Dose titrated with fat content of meals and snacks to control symptoms of malabsorption
  - maximum 10,000 units lipase/kg/day, higher doses can result in colonic strictures

Signs of malabsorption

- Fatty pale stools, frequent, smelly, orange oil, excess flatulence, abdominal pains
  - discuss with CF team

H_2-receptor antagonists

- If taking large doses of pancreatic enzymes (e.g. >10,000 units lipase), discuss with CF team need for concurrent ranitidine to reduce deactivation of pancreatin

Vitamins A, D and E

Starting dosage for newly-diagnosed

- **Infants**
  - 0.6 mL Dalivit® or/and 0.5 mL (50 mg) Vitamin E

- **Children**
  - 1.2 mL Dalivit® or 3 BPC multivitamins capsules and 100 mg Vitamin E (2 x 50 mg capsule)

OR

  - continue dose as prescribed in CF clinic
- Vitamin levels are checked annually and dosage adjusted accordingly

Oral sodium chloride

- Only if prescribed by CF team
- Often needed in first year of life after diagnosis has been made

CYSTIC FIBROSIS – EXACERBATION • 1/2

RESPIRATORY INFECTION/EXACERBATION

If unusual symptoms, such as haemoptysis, abdominal pain (distal intestinal obstruction syndrome), or bleeding varices, discuss urgently with CF consultant

Symptoms and signs

- Increasing cough and sputum
- Increasing dyspnoea
- Weight loss with loss of appetite
- Thick, tenacious sputum
- Coarse crepitations
- Haemoptysis
- Signs of right heart failure

Investigations

- See investigations in **Cystic fibrosis – Admission** guideline

Differential diagnosis

- Non-CF bronchiectasis
- Chronic obliterative bronchiolitis

ADDITIONAL ADMISSION PROCEDURE

- If IV antibiotics required, discuss with CF team re procedure:
  - agree a clear individualised procedure for every patient
  - discuss with CF team whether anaesthetic team needed for needle-phobic patients
- Trained nursing staff needed to needle Port-a-cath

IMMEDIATE TREATMENT

- Use IV antibiotic regimen suggested following discussion with CF team
- If no discussion possible, stop oral antibiotics and give first-line regimen (see below)
- Take into account any past allergic reactions

First-line regimen

- Sputum culture
  - *Pseudomonas aeruginosa:* ceftazidime 50 mg/kg 8-hrly (max 3 g/dose) and tobramycin 10 mg/kg once daily (max 660 mg) given over 30 min
  - no *Pseudomonas aeruginosa:* cefuroxime 50 mg/kg 8-hrly (max 1.5 g/dose)
- Courses usually last 2 weeks
- For cephalosporins (but not tobramycin), aim to use whole vials by rounding doses +10% considering vial size
- After satisfactory tobramycin blood levels established, CF team will teach parents to give antibiotics at home. Discuss with pharmacy as well

Nebulised antibiotics

- Give children colonised with *Pseudomonas*, colomycin 1 million units made up to 4 mL with sodium chloride 0.9%, nebulised 12-hrly

Oral antibiotics

- Children are rarely given oral antibiotics during admission but may resume an oral agent on discharge

Bronchodilators

- Prescribe salbutamol by MDI and spacer before chest physiotherapy in hospital

Inhaled corticosteroids

- There is no evidence these are of benefit. Discuss with CF team re stopping

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TOBRAMYCIN MONITORING

Once daily regimen:

- Trough level immediately before 2nd and 8th dose
- Should be <1 mmol/L
- **High levels need to be discussed with CF consultant**
- No need to determine peak
- **Always discuss dose changes with CF team beforehand**
- Do not check tobramycin dose via Port-a-cath or long line

SUBSEQUENT MANAGEMENT

- Do not change antibiotics before discussing with CF team

Oral corticosteroids

- If no chest improvement after a week of IV antibiotics, consider starting 7 day course of prednisolone 1 mg/kg/day
- If already taking alternate-day prednisolone at lower dosage, review dosage needed at discharge
- For children with allergic broncho pulmonary aspergillosis (ABPA), continue prednisolone for longer (e.g. at least one month)

Dornase alfa (DNAse)

- Discuss need for dornase alfa with CF team
- Indications for use are:
- cough productive of sputum or sputum difficult to expectorate
- Give dornase alfa (2.5 mg/daily) via nebuliser after morning physiotherapy
- Patients should bring their own nebuliser (usually a modified Sidestream®) and compressor into hospital

DISCHARGE AND FOLLOW-UP

- On advice of CF team

Self-administration of IV antibiotics – home IV therapy

- Service managed by CF nurse in conjunction with hospital pharmacy
- Discuss fully with CF nurse before making any changes or arrangements

Criteria for home administration of IV antibiotics

Ensure that:

- CF team and ward staff happy for patient to be discharged
- Patient and parents **entirely happy, confident and competent** to administer IV antibiotics at home
- Patient/parent has been assessed before discharge by CF team
- Parents have written guidelines and **24 hr** contact numbers
- If patient considered responsible enough to self-administer IV antibiotics, important that parent/carer also has adequate instruction and guidance
- Anaphylaxis kit at home and family know how to use
- Notify CF liaison nurses of any patient discharged on home antibiotic therapy so they can arrange support at home or at school if necessary
- CF liaison nurse will visit patient at home during his/her course of IV therapy, to monitor progress
- Feedback any concerns to CF team

CYSTIC FIBROSIS – MICROBIOLOGY • 1/2

In addition to standard precautions and hand hygiene, the following precautions are required for patients infected/colonised with transmissible pathogens

- Do not share equipment between patients
- Nurse children with CF in a cubicle
- Prevent contact between CF patients

PATIENT NEWLY DIAGNOSED WITH CF

- Prophylaxis with flucloxacillin 125 mg oral 12-hrly until aged 2 yr
- If newly diagnosed CF patient has chest infection requiring IV antibiotics:
  - commence cefuroxime IV for 2 weeks, then co-amoxiclav oral for 3–4 weeks
- Subsequent treatment depends on microbiology

PSEUDOMONAS AERUGINOSA

First isolations in sputum or cough/throat swabs

- If asymptomatic with first isolation from sputum/cough swab:
  - ciprofloxacin: aged 1 month–18 yr 20 mg/kg oral 12-hrly (max 750 mg) and colomycin aged <2 yr 1 million units 12-hrly, aged ≥2 yr 2 million units 12-hrly via nebuliser for 3 months
- If recurrent isolation from sputum/cough swab:
  - ciprofloxacin and colomycin or intravenous antibiotic – discuss with CF team
- If symptomatic:
  - tobramycin and ceftazidime IV for 2 weeks, followed by: ciprofloxacin and colomycin as directed by CF team

Pseudomonas chronic infection

- Defined as **3 or more** isolations in 6 months from sputum/cough swab samples taken at least 1 month apart

Nebulised antibiotics

- If chronically infected with *Pseudomonas*, give colomycin 1 million units made up to 4 mL with sodium chloride 0.9%, nebulised 12-hrly
- Nebulised tobramycin to be decided by CF team

BURKHOLDERIA CEPACIA COLONISATION

- **Report any new cases to CF team immediately**
- Nurse children with *B. cepacia* colonisation in a cubicle on a separate ward from other CF children
- Use **separate spirometer with disposable filters**

MRSA COLONISATION

- **Report any new cases to CF team immediately**
- Use normal spirometer with a **disposable filter**

CHICKENPOX AND CF

- Varicella infection can have serious consequences in immunosuppressed children
- CF patients taking oral corticosteroids are at high risk
- If no history of chickenpox and no antibodies, vaccinate

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Exposure

- Ask about exposure to a known case:
  - being in the same room (e.g. in the house, classroom or hall in school) for ≥15 min
  - face-to-face contact, for example whilst having a conversation
- If exposure significant, check notes to determine immune status (history of chickenpox or antibody status before corticosteroids)
- If non-immune and taking a high dose of oral corticosteroid (prednisolone 1 mg/kg/day for one month or 2 mg/kg/day for 1 week), and exposure occurred <1 week earlier, give varicella-zoster immunoglobulin (VZIG) aged <6 yr 250 mg; aged 6–10 yr 500 mg; aged 11–14 yr 750 mg; aged >15 yr 1 g, or IV immunoglobulin 0.2 g/kg
- If non-immune and taking a modest dose of oral corticosteroid (prednisolone <1 mg/kg/day), give aciclovir prophylaxis 6-hrly: aged <2 yr 200 mg; aged >2 yr 400 mg 7–21 days after exposure

Infected

- If chickenpox appears in a child not taking oral corticosteroid, give aciclovir 10 mg/kg oral 6-hrly for 7 days and a course of oral antibiotics (e.g. amoxicillin and flucloxacillin)

INFLUENZA AND PNEUMOCOCCAL VACCINE

- Influenza vaccine every October
- Conjugate pneumococcal vaccine (Prevenar13®)
- Usually prescribed by patient's own GP but obtainable from pharmacy

PORT-A-CATH

- Use in children requiring frequent IV antibiotics
- Manufacturer's instructions found on ward
- Observe sterile precautions whenever Vascuport accessed
- Accessed only by trained nursing staff

Routine flushing of Port-a-cath (usually by nursing staff)

- Every 4 weeks (coincide with clinic appointment where possible)
- Use a straight Port-a-cath needle and 4 mL heparinised sodium chloride 0.9% 100 units/mL (e.g. Canusal®, not Hepsal®), withdrawing needle while injecting last mL

CYSTIC FIBROSIS – DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS) • 1/1

RECOGNITION AND ASSESSMENT

- Faeces can accumulate in distal ileum and caecum causing varying degrees of intestinal obstruction
- Patients present with intermittent abdominal pain, constipation and faecal masses, usually in right or left iliac fossa

MANAGEMENT

- If symptoms mild, prescribe daily macrogol laxative (e.g. Movicol) see BNFc and encourage fluids
- Consider adjusting pancreatic enzymes – but discuss with CF team
- If unresponsive, or symptoms more severe:
  - single dose of sodium amidotrizoate (Gastrografin): see dose and treatment practice in BNFc
  - repeat dose after 12–18 hr, encourage drinks, monitor fluid balance and allow food
- If no effect after 24–48 hr or if patient deteriorates, give balanced electrolyte solution (discuss with CF team and gastro team)
- Bowel lavage with Klean-Prep® (usually requires a nasogastric tube)
- 1 sachet Klean-Prep® in 1 L give:

  10 mL/kg/hr for 30 min

  then 20 mL/kg/hr for 30 min

  then 25 mL/kg/hr up to max total dose of 100 mL/kg or 4 L
- Start early in the morning and continue until stools are yellow, watery and free of solid matter
- 2 L in first instance, increasing to 3 or 4 L depending on response, age and size of child (most children with DIOS will be teenagers)
- Withhold food but, if success not achieved after 12 hr, stop, give an evening meal and resume following morning
- Monitor effectiveness with pre- or post-plain abdominal X-ray before and after lavage
- If signs of complete intestinal obstruction, stop lavage, give IV fluids and discuss contrast enema with CF team

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If aged <1 month-old, refer to Neonatal guidelines

RECOGNITION AND ASSESSMENT

Definition

- Inflammation and consolidation of the lung caused by a bacterial, viral or mycoplasma infection
- Absence of clinical signs **AND** negative CXR makes pneumonia unlikely
- Up to 35% of lower respiratory tract infections have single virus as causative organism
- Can be presenting illness in cystic fibrosis and immunodeficiency states

Symptoms and signs

- Cough
- Fever
- Irritability
- Poor feeding
- Vomiting
- Tachypnoea at rest (most useful sign)

Beware: awake or unsettled infants can have high respiratory rate on a single measurement; measure at rest and repeat

Table 1: WHO definition of tachypnoea

Age	Counted breath rate
<2 months	≥60/min
2–11 months	≥50/min
1–5 yr	≥40/min

- Bronchial breathing, inspiratory crackles
- Recession
- Abdominal pain (referred pleural pain)
- Aged >5 yr, headache, arthralgia, sore throat (suggests mycoplasma)

Investigations

- Pulse oximetry
- CXR
- Full blood count, blood culture
- Serum electrolytes (may have hyponatraemia owing to SIADH), CRP
- If mycoplasma pneumonia suspected, mycoplasma titre (indicate date of onset on request form)
- Sputum if able to provide good quality specimen
- Nasopharyngeal aspirate or nasal swab in viral transport medium for respiratory viruses
- If pertussis suspected, pernasal swab in charcoal transport medium
- Pleural fluid culture and PCR if aspirated
- If severe pneumonia, pneumococcal antigen in urine

Differential diagnosis

- Bronchiolitis with atelectasis (usually aged <1 yr)
- Foreign body aspiration
- Tumour ('round' pneumonia)
- Empyema/lung abscess
- Tracheobronchitis
- Whooping cough

IMMEDIATE TREATMENT

See **Flowchart**

Pleural effusion

- See **Pleural effusion** guideline

SUBSEQUENT MANAGEMENT

- Change from IV to oral within 24–48 hr
- Total antibiotic course 5–7 days
- If atypical or staphylococcal pneumonia, treat for 14 days uncomplicated CAP and 14–21 days for severe CAP
- Physiotherapy once cough productive
  - important if neuromuscular impairment results in poor clearance
- Maintain hydration
  - oral fluids if tolerated
  - if unable to take oral fluids and Na >135 mmol give sodium chloride 0.45% with glucose 5% and potassium chloride 10 mmol/500 mL via IV infusion. If Na <135 mmol use sodium chloride 0.9% with glucose 5% with potassium
  - restrict IV fluid replacement to 80% maintenance
  - monitor electrolytes

MONITORING TREATMENT

- Continuous SpO_2 monitoring if needing oxygen
- 1–4 hrly observation depending on severity of illness
- If no improvement in 24–48 hr, review diagnosis (repeat chest X-ray) or treatment

DISCHARGE AND FOLLOW-UP

- Follow-up within 6–8 weeks with CXR if:
  - lobar collapse
  - significant pleural effusion
  - 'round' pneumonia on CXR
  - previous lower respiratory tract infections
  - failure to thrive
- GP follow-up for all others within 6–8 weeks
- Convalescent mycoplasma titre can be obtained at this visit (indicate date of onset on request form)

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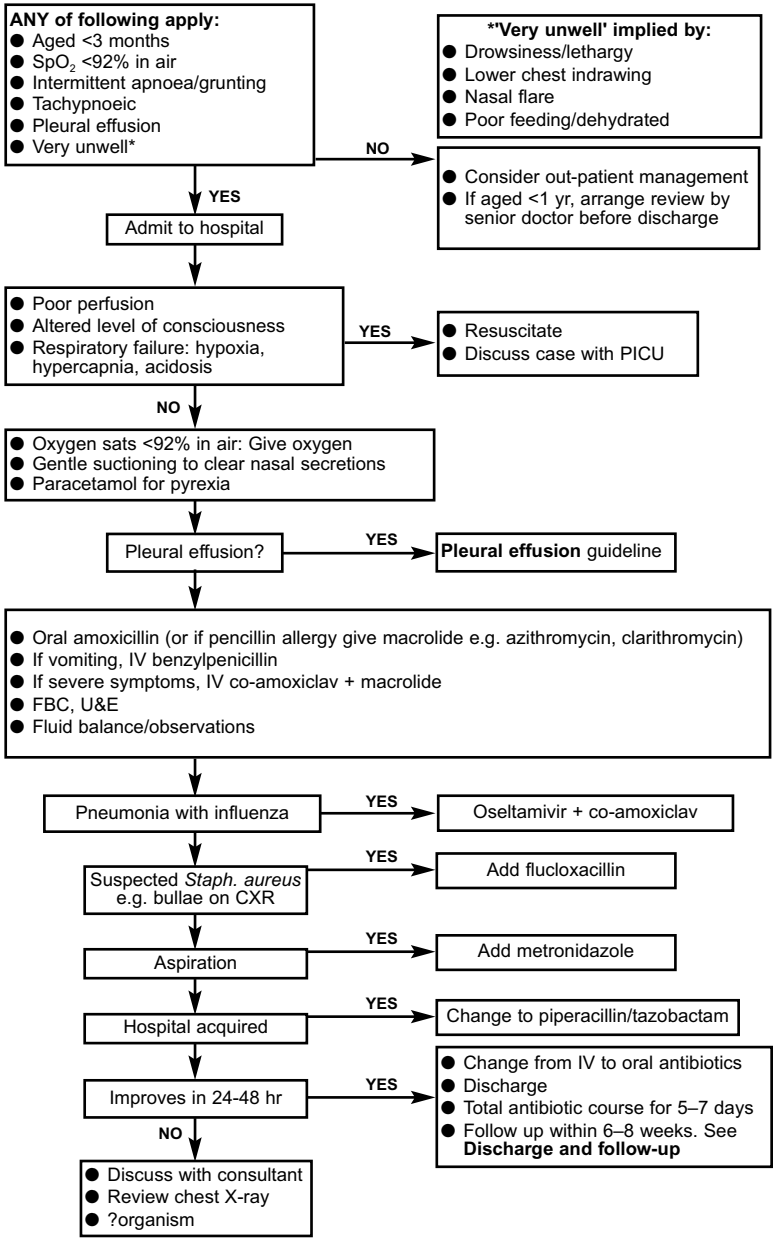
Flowchart: Management of community acquired pneumonia in a previously well patient aged >1 month-old



RECOGNITION AND ASSESSMENT

Symptoms and signs

- Investigate for effusion if persistent pyrexia or unwell 48 hr after treatment started for pneumonia

Differential diagnosis

- Uncomplicated pneumonia
- Malignancy
- Heart failure
- Pancreatitis
- Pulmonary embolism

Investigations

- Chest X-ray PA or AP (no need for lateral)
- Ultrasound (US) scan to:
  - confirm presence of effusion
  - ascertain volume
  - ask radiologist to mark optimal position for chest drain
  - differentiate between simple and complicated effusion (e.g. loculations, heterogeneous material)
- If history, chest X-ray or US suggestive of malignancy, request CT chest
- If risk factors for coagulopathy or thrombocytopenia check and correct before drain insertion
- Pleural fluid analysis for:
  - Gram stain and bacterial culture
  - differential cell count
  - AAFB and TB PCR and culture
- LDH, protein, glucose and pH (via blood gas analyser)
  - **at same time,** blood samples for FBC, clotting screen, U&E, LDH, protein, albumin, glucose
- CRP
- Blood cultures
- Sputum culture, if possible
- If recurrent infections, investigate for immune deficiency (first line: FBC, IgG, A, M, functional antibodies and HIV antibody)

It is not necessary to obtain sample for pleural fluid culture routinely before chest drain insertion if cause likely to be infective. If alternative cause suspected, try to avoid unnecessary chest drain insertion by obtaining diagnostic aspirate of pleural fluid for cytology

Table: Fluid/serum protein and LDH ratios are best discriminators between transudate and exudate

	Transudate	Exudate
Appearance	Serous	Cloudy, bloody
Leucocyte count	<10,000/mm^3	>50,000/mm^3
Protein	≤30 g/L	>30 g/L
Fluid/serum protein ratio	≤0.5	>0.5
LDH	≤200 IU	>200 IU or >$^2/_3$ local upper limit of serum LDH
Fluid/serum LDH ratio	≤0.6	>0.6
Glucose	≥3.3 mmol/L	<3.3 mmol/L
pH	≥7.4	≤7.3
Gram stain/culture	No organisms	Organisms on stain or culture

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IMMEDIATE TREATMENT

Supportive

- ABC
- Oxygen and fluid resuscitation as indicated
- Analgesia

Refer to respiratory paediatrician

- Remember that underlying cavitating disease may lead to bronchopleural fistulae. Assess likelihood of this problem before inserting any chest drain
- Small effusions (<2 cm deep) which are not enlarging or compromising respiratory function do not need to be drained
- Early active treatment reduces length of illness

Antibiotic therapy

Type of effusion suspected	Choice of antibiotics
Effusion following community-acquired pneumonia	Co-amoxiclav IV + clindamycin IV or oral
Effusion following hospital-acquired pneumonia or in immune-compromised child	Piperacillin/tazobactam
Effusion possibly tuberculous	Discuss with TB team

- Narrow antibiotic spectrum with culture results

Chest drain insertion

- Drain inserted by experienced team
- Discuss with respiratory team, consultant paediatrician, paediatric anaesthetic team (usually GA used)
- support may also be required from cardiothoracic team +/- interventional radiologist
- Consider simultaneous insertion of long line during general anaesthetic, if possible
- Ensure vascular access before starting procedure
- CXR after drain insertion

Chest drain management

- Ensure nursing staff trained in care of children with chest drains
- Attach chest drain to low level suction (5–10 cm H_2O) via underwater seal
- If altitude chest drainage system used, set wall suction to 160 mmHg/22 kPa and set dial on drainage system to 20
- Keep underwater seal below level of chest at all times
- After 10 mL/kg has been drained, clamp chest drain for 1 hr
- **Never clamp a bubbling chest drain** – this indicates presence of pneumothorax
- If clamped and chest pain or breathlessness, unclamp immediately
- Ensure adequate analgesia (see **Analgesia** guideline) and encourage patient to move freely when well enough

Intrapleural fibrinolytics

- Indicated if thick fluid with loculations or pus
- Instill urokinase in all patients, as follows:
  - ≥10 kg, urokinase 40,000 units in 40 mL sodium chloride 0.9%
  - <10 kg, urokinase 10,000 units in 10 mL sodium chloride 0.9%
  - administer via chest drain 12-hrly for 3 days (total 6 doses)
  - clamp chest drain for 4 hr after instillation of urokinase, then drain for 8 hr
- Record fluid volumes into and out of pleural space carefully and accurately

SUBSEQUENT MANAGEMENT

Act on response to treatment and clinical assessment of patient

- Monitor symptoms and re-examine patient to assess progress
- Repeat CRP as needed
  - if falling rapidly, continue with current regimen
  - if not falling after 72 hr, treat as non-resolution (see below)
- Chase pleural fluid aspirate results
  - if unexpected organisms grown, adjust antibiotic therapy with antibiotic sensitivities
  - if differential cell count shows lymphocytosis, discuss with TB team, send aspirate for cytology and consider CT scan of chest
- Chase blood and sputum culture results – if no growth, continue empirical treatment until patient improves
- Remove chest drain when drainage minimal and in agreement with respiratory paediatrician: appose skin with Steristrips® rather than sutures
- Continue IV antibiotics at least until afebrile. Change to oral co-amoxiclav when clinical improvement obvious. Complete minimum 14 days antibiotics
- Continue antibiotics until CRP <10
- Encourage early mobilisation and exercise

Non-resolution

- Non-resolution of effusion after 3 days or further complications occur, consider CT scan of chest
- If no fluid draining, check for obstruction by flushing
- If drain can not be unblocked, remove and replace if significant effusion remains
- Discuss referral for thoracotomy with respiratory paediatrician

Surgery

- Discuss with paediatric thoracic surgeon if:
  - effusion has not resolved
  - child is still septic

DISCHARGE AND FOLLOW-UP

- Arrange review by respiratory paediatrician, initial appointment 6 weeks after discharge (CXR on arrival)
  - if symptoms persist or recur, early referral to respiratory paediatrician

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PNEUMOTHORAX • 1/2

RECOGNITION AND ASSESSMENT

Symptoms and signs

Tension pneumothorax (very rare)

- Severe dyspnoea
- Circulatory compromise
- Trachea +/- apex beat displaced

Treat immediately

- Give oxygen 15 L/min with mask with reservoir bag
- Insert a large bore cannula of at least 4.5 cm in length into 2nd anterior intercostal space, midclavicular line
- Insert intercostal tube
- Remove emergency cannula when bubbling in underwater seal system confirms intercostal tube system functioning

Spontaneous pneumothorax

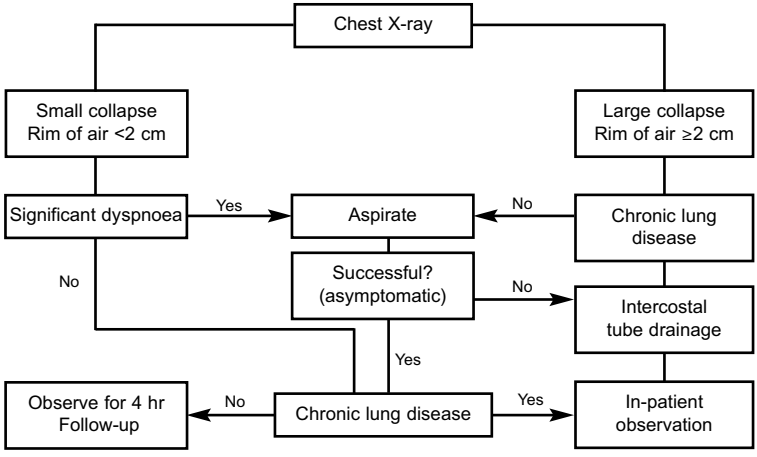
- Sudden onset, occasionally at rest
- Chest pain (unilateral)
- Dyspnoea
- Resonance on percussion, with reduced vocal fremitus and breath sounds (if moderate-large)

Investigations

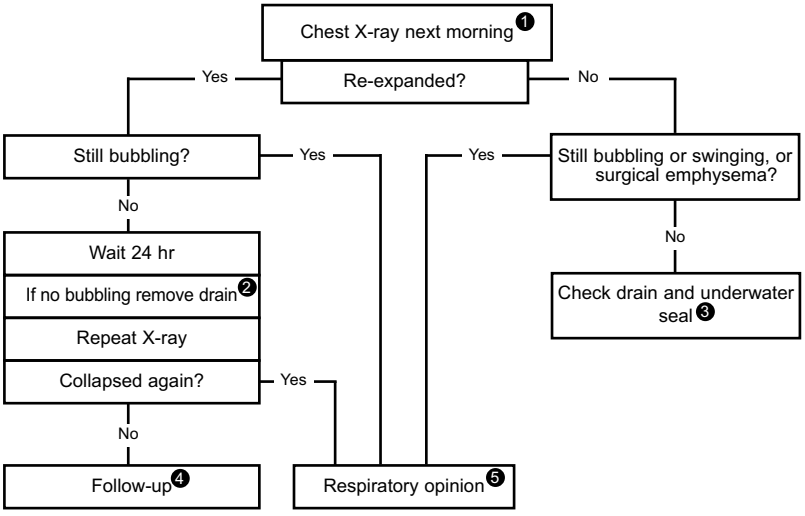
- PA chest X-ray
- If findings are unclear on PA, lateral (if possible, decubitus) film may help
- If findings obscured by surgical emphysema or complex bulla disease, CT scan may help

BEWARE: suspected basal pneumothorax usually implies a bulla. CT scan will differentiate bullae from pneumothorax

IMMEDIATE TREATMENT



Management of intercostal drains



Do not clamp chest tube unless advised by respiratory paediatrician or thoracic surgeon. If clamped and chest pain or breathless unclamp immediately

- **1: Chest X-ray**
  - keep underwater seal below level of chest at all times

- **2: Removal of chest drain:**
  - bubbling stopped for at least 24 hr
  - cut drain-securing suture
  - withdraw tube while patient holds breath in expiration
  - close wound with remaining sutures

- **3: Check drain:**
  - if lung not re-inflated and no bubbling in underwater bottle: Try to remove **block** or **kink**
  - if unsuccessful, remove drain. Insert new drain through clean incision

- **4: Follow-up:**
  - at clinic in 7–10 days
  - patient given discharge letter and written advice to return immediately if deteriorates
  - no air travel until chest X-ray resolved

- **5: Respiratory paediatrician's opinion:**
  - if no re-expansion consider air leak, displaced/blocked tube, bronchopleural fistula, underlying pulmonary disease
  - use of high volume/low pressure suction, 1–2 kPa/Barr, (8–16 mmHg; 8–20 cm H_2O)
  - if altitude chest drainage system used, set wall suction to 160 mmHg/22 kPa and set dial on drainage system to 20
  - early thoracic surgery. Refer when pneumothorax fails to resolve after 5 days of above management or after 3 days if patient has chronic lung disease

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RECOGNITION AND ASSESSMENT

Symptoms and signs

- Central cyanosis may be respiratory or cardiac in origin
- Respiratory illness producing cyanosis will usually have signs of respiratory distress (e.g. cough, tachypnoea, recession and added respiratory sounds)
- Cardiac decompensation may occur with a respiratory infection: they may co-exist
- Cyanosis more likely due to cardiac disease if:
- SpO_2 responds poorly to high flow oxygen (15 L/min) via face mask and reservoir bag
- marked tachycardia
- enlarged heart (clinically or on CXR)
- gallop rhythm/murmur
- enlarged liver/raised JVP
- basal crackles
- absent femoral pulses
- finger clubbing occurs after a few months (also consider endocarditis)

Causes of cardiac cyanosis

Significant right-to-left shunt

- Transposition with inadequate mixing, pulmonary or tricuspid atresia
- Fallot's tetralogy: hypercyanotic episodes follow emotional or painful upset

Duct-dependent pulmonary circulation

- Commonly presents in first 10–14 days of life
- severely blue, breathless or shocked
- pulmonary atresia
- critical pulmonary valve stenosis
- tricuspid atresia
- severe Fallot's tetralogy
- transposition of the great arteries without septal defect
- single ventricle anatomy

Acute pulmonary outflow obstruction (cyanotic episodes)

- Fallot's tetralogy or other complex congenital cyanotic heart disease
- severe pallor
- loss of consciousness
- convulsions

Physical examination

- Remember to check femoral pulses
- If coarctation of the aorta suspected: check BP in upper and lower limbs – normal difference <15 mmHg

Investigations

If infant cyanosed or in heart failure, discuss urgency of investigations with consultant

SpO_2

- Check pre- (right arm) and post-ductal (lower limbs)
- when breathing air before oxygen given
- after giving 15 L/min oxygen by mask with a reservoir bag for 10 min

Chest X-ray

- For cardiac conditions, specifically record:
- cardiac situs (normal or right side of chest)
- aortic arch left or right-sided
- bronchial situs (is right main bronchus on the right?)
- cardiac size and configuration
- size of pulmonary vessels and pulmonary vascular markings

Electrocardiogram

See **ECG interpretation** guideline.

Nitrogen washout in cyanosed babies

- Monitor SpO_2 in air then in headbox after breathing 100% oxygen for 10 min
- in cyanotic congenital heart disease, PaO_2 will remain below 20 kPa with SpO_2 unchanged
- not as reliable as echocardiogram

Echocardiogram

- Locally, if available, or refer to regional paediatric cardiac centre

IMMEDIATE TREATMENT

If infant cyanosed or in heart failure, discuss urgency of referral to local paediatric cardiac surgical centre with consultant

Duct-dependent congenital heart disease

- Immediate treatment before transfer to a paediatric cardiac centre:
- open duct with prostaglandin E1 (alprostadil) or E2 (dinoprostone) same dose:
- 5–10 nanogram/kg/min IV infusion to start
- increasing in steps of 5–10 nanogram/kg up to max 100 nanogram/kg/min
- then reducing to lowest dose needed
- May cause apnoea and patients may need ventilation
- Beware of giving high concentrations of oxygen as this encourages duct closure

Acute pulmonary outflow obstruction (cyanotic episodes)

- Immediate treatment before transfer to a paediatric cardiac centre:
- do not upset child
- give morphine 50–100 microgram/kg IV over 5 min or IM
- provide high concentration face mask oxygen (15 L/min with reservoir bag)
- if Fallot's tetralogy has been diagnosed by echocardiography, discuss with cardiologist use of IV beta-blocker

SUBSEQUENT MANAGEMENT

- On advice of consultant and paediatric cardiac centre

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CAUSES

- Congenital heart malformations
  - aortic stenosis
  - coarctation of the aorta
  - hypoplastic left heart
- Cardiomyopathies
- Pericardial effusion
- Myocarditis
- Arrhythmias
- Hypoxia
- Hypovolaemia
- Acidosis
- Toxins

RECOGNITION AND ASSESSMENT

Presentation

- Usually during first few weeks of life
- Later triggered by an intercurrent infection, with associated myocarditis or prolonged arrhythmia

Symptoms and signs

- Failure to thrive
- Rapid weight gain
- Sweating
- Breathlessness, particularly during feeding
- Tachypnoea
- Tachycardia
- Absent or low volume peripheral or central pulses
- Enlarged heart
- Prominent cardiac impulses
- Quiet heart sounds in pericardial effusion
- Thrill
- Gallop rhythm
- Enlarged liver

Recognition of cardiogenic shock

- For definition of shock see **Septicaemia** guideline
- Following cardiopulmonary resuscitation with adequate fluid replacement in patients with:
  - septic shock that fails to improve after adequate fluid replacement (e.g. ≥40 mL/kg)
  - a known heart condition and shock
  - a large heart on chest X-ray but previously well
  - shock, who have a history of poisoning
  - a murmur or pulmonary oedema, or both

INVESTIGATIONS

- Check BP in upper and lower limbs (normal <15 mmHg difference)

SpO_2

- Check pre- (right arm) and post-ductal (lower limbs)
- In air and after giving oxygen

Chest X-ray

- For cardiac conditions, specifically record:
  - cardiac situs (normal or right side of chest)
  - aortic arch left- or right-sided
  - bronchial situs (is right main bronchus on the right?)
  - cardiac size and configuration
  - size of pulmonary vessels and pulmonary vascular markings

Electrocardiogram

- See **ECG interpretation** guideline

Echocardiogram

- Locally, if available, or refer to local paediatric cardiac centre

MONITORING

- ECG monitor
- Non-invasive BP
- Pulse oximetry
- Core-skin temperature difference
- Daily weights
- Urine output (≥1 mL/kg/hr)
- If shocked or ≥40 mL/kg fluid resuscitation:
  - intra-arterial BP monitoring
  - CVP

THERAPEUTIC MEASURES

In all children with heart failure

1. If breathless, elevate head and trunk
2. If infant not feeding well, give nasogastric feeds
3. In moderate-to-severe failure or if patient hypoxic or distressed, give oxygen therapy via nasal cannulae (up to 2 L/min) or via a face mask with reservoir bag (up to 15 L/min)
4. Diuretics: furosemide 1 mg/kg oral or by slow IV injection (max 4 mg/min) and amiloride 100 microgram/kg (max 10 mg) oral 12-hrly (doses can be repeated if not responding to initial dose)
5. If serum potassium <4.5 mmol/L, give additional potassium chloride 1 mmol/kg 12-hrly enterally
6. Correct acidosis, hypoglycaemia and electrolyte imbalance
7. Relieve pain with morphine: loading dose 100 microgram/kg IV over 5 min, followed by 50 microgram/kg IV 4–6 hrly over 5 min or 10 microgram/kg/hr via IV infusion (doses can be doubled if necessary)
8. If anaemic (Hb <100 g/L), correct with infusion of packed cells over 4 hr to bring Hb to 120–140 g/L

If cardiogenic shock present

1. Monitor CVP and ensure adequate pre-load: give Human Albumin Solution (HAS) 4.5% 10 mL/kg as IV bolus or, if HAS not available, sodium chloride 0.9% 10 mL/kg as IV bolus
2. If shock severe (see **Septicaemia** guideline), start mechanical ventilation with positive end-expiratory pressure early; if pulmonary oedema present, start urgently
3. If shock severe, give early inotropic drug support: dopamine, dobutamine, adrenaline or noradrenaline as per NNU/PICU protocols

DUCT-DEPENDENT CONGENITAL HEART DISEASE

- May present in first two weeks of life

Duct-dependent systemic circulation

- Breathless, grey, collapsed, poor pulses
  - severe coarctation of the aorta
  - critical aortic stenosis
  - hypoplastic left heart syndrome

Duct-dependent pulmonary circulation

- Blue, breathless or shocked
  - pulmonary atresia
  - critical pulmonary valve stenosis
  - tricuspid atresia
  - severe Fallot's tetralogy
  - transposition of the great arteries

Treatment

- See **Cyanotic congenital heart disease** guideline

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ECG INTERPRETATION • 1/4

- All ECGs, check:
  - P-wave size and axis
  - axis of QRS complex
  - R-S pattern in chest leads
  - P-R, QRS and Q-T intervals
  - P- and T-wave configuration
  - size of QRS in chest leads

PAPER SPEED

- ECG normally recorded at 25 cm/sec
  - 1 mm (1 small square) = 0.04 sec
  - 5 mm (1 large square) = 0.2 sec

P WAVE

- Reflects atrial activity
- Duration shorter than in adults
  - infants: 0.04–0.07 sec
  - adolescents: 0.06–0.1 sec
- Height ≤2.5 mm
- Varying P wave morphology may indicate wandering atrial pacemaker

Right atrial hypertrophy (RAH)

- Increased P wave amplitude in leads II, V1, and V4R

Causes

- Pulmonary hypertension
- Pulmonary stenosis
- Pulmonary atresia
- Tricuspid atresia

Left atrial hypertrophy (LAH)

- Biphasic P wave (later depolarization of LA)

Causes

- Mitral valve disease
- LV obstruction and disease

P-R INTERVAL

- Atrial depolarization varies with age and rate

Normal range of P-R interval (time in sec)

Heart rate	P-R interval (sec)			
	0–1 month	0–12 months	1–12 yr	12–16 yr
<60	-	-	-	0.1–0.19
60–99	-	-	0.1–0.16	0.1–0.17
100–139	0.08–0.11	0.08–0.12	0.1–0.14	-
140–180	0.08–0.11	0.08–0.12	0.1–0.14	-
>180	0.08–0.09	0.08–0.11	-	-

Prolonged interval

- Normal
- Myocarditis
- Ischaemia
- Drugs
- Hyperkalaemia

Short interval

- Wolff-Parkinson-White syndrome
- Lown-Ganong-Levine syndrome
- Glycogen storage disease

Variable interval

- Wandering atrial pacemaker
- Wenckebach phenomenon

QRS COMPLEX

- Ventricular activity
- Duration: 0.06–0.08 sec

Prolonged

- Ventricular hypertrophy
- Bundle branch block
- Electrolyte disturbance
- Metabolic disease
- Drugs (e.g. digoxin)

Normal range of R and S waves (height in mm)

Age	R and S waves (height in mm)					
	V4-R	V1-R	V1-S	V5-R	V6-R	V6-S
Birth	4–12	5–20	0–20	2–20	1–13	0–15
6–12 months	2–7	3–17	1–25	10–28	5–25	0–10
1–10 yr	0–7	2–16	1–12	5–30	5–25	0–7
>10 yr	0–6	1–12	1–25	5–40	5–30	0–5

Q WAVE

- Normal in II; III; aVF; V5-6
- Depth 2–3 mm
- pathological if >4 mm (i.e. septal hypertrophy)
- May be found in other leads in:
- anomalous coronary arteries
- hypertrophic obstructive cardiomyopathy
- transposition of great arteries (with opposite polarity)

Q-T INTERVAL

Inversely proportional to rate

- Calculate ratio of Q-T interval to R-R interval
- $QTc = \frac{Q\text{-}T}{\sqrt{R\text{-}R^{1}}}$
- QTc is usually less than 0.44 s
- prolonged QTc is associated with sudden death: alert consultant immediately

Prolonged interval

- Hypocalcaemia
- Myocarditis
- Jervell-Lange-Nielsen syndrome
- Romano-Ward syndrome
- Head injuries or cerebrovascular episodes
- Diffuse myocardial disease
- Antiarrhythmics

Short interval

- Hypercalcaemia
- Digitalis effect

T WAVE

- Ventricular repolarization

Normal

- T inversion V4R/V1 (from third day of life until 10 yr)
- Amplitude is 25–30% of R-wave
- Aged <1 yr: V5 ≤11 mm; V6 ≤7 mm
- Aged >1 yr: V5 ≤14 mm; V6 ≤9 mm
- Adolescence reduces amplitude

Peaked T wave

- Hyperkalaemia
- LVH
- Cerebrovascular episode
- Post-MI

Flat T wave

- Normal newborn
- Hypothyroidism
- Hypokalaemia
- Hyper/hypoglycaemia
- Hypocalcaemia
- Peri/myocarditis
- Ischaemia
- Digoxin effect

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MEAN QRS AXIS

Vertical plane (limb leads)

Normal axis in vertical plane

- Birth +60° to +180° (av +135°)
- Aged 1 yr +10° to +100° (av +60°)
- Aged 10 yr +30° to +90° (av +65°)

Right axis deviation

- Right ventricular hypertrophy (RVH)
- Left posterior hemiblock
- Ostium secundum atrial septal defect (ASD)/right bundle branch block (RBBB)

Left axis deviation

- Left ventricular hypertrophy (LVH)
- Ostium primum ASD (+ RBBB)
- Often in conduction defects

Horizontal plane (anterior chest leads)

Normal

- Transition at around V3

Clockwise rotation

- S>R in V4 = RA/RV hypertrophy

Anticlockwise rotation

- R>S in V2 = cardiac shift (e.g. pneumothorax)

LEFT VENTRICULAR HYPERTROPHY

Diagnosis

- SV1 + RV5 ≥40 mm (30 mm aged <1 yr)
- +/- prolonged QRS
- Flat T wave
- T wave inversion V5-V6 (LV strain)
- Left bundle branch block

Causes include

- Aortic stenosis
- Aortic regurgitation
- Hypertension
- Moderate VSD
- Hypertrophic obstructive cardiomyopathy
- Patent ductus arteriosus
- Mitral regurgitation

RIGHT VENTRICULAR HYPERTROPHY

Diagnosis

- RAD and RV1 > SV1 (aged >1 yr)
- SV6 above maximum for age:
  - 0–6 months 15 mm
  - >6 months 10 mm
  - >12 months 7 mm
  - 10 yr 5 mm
- R waves in V4R/V1 >normal
- T wave changes
  - upright in V1/V4R (aged from 3 days to 10 yr)

Causes include

- Pulmonary stenosis/atresia
- Transposition of great arteries
- Pulmonary regurgitation
- Total anomalous pulmonary drainage
- Tricuspid regurgitation
- Fallot's tetralogy
- Pulmonary hypertension

BIVENTRICULAR HYPERTROPHY

Diagnosis

- R + S >50 mm in V3-V4
- LVH + bifid R <8 mm in V1
- RVH + LV strain
- Q waves V3-V6 imply septal hypertrophy

TYPICAL ECG ABNORMALITIES

Heart lesion	ECG abnormalities
PDA	LVH > RVH; LAH
VSD	LVH > RVH; +/- RBBB; T inv LV. leads
ASD	Secundum RAD; RBBB; +/- increased P-R; AF Primum LAD; RBBB; BVH; RAH
Eisenmenger's	RVH; P pulmonale
Aortic stenosis	LVH + strain
Aortic regurgitation	LVH
Coarctation	Newborn: RVH Older: Normal or LVH +/- strain; RBBB
Mitral regurgitation	LVH
Pulmonary stenosis	RVH; RAH
Ebstein's anomaly	Prolonged P-R interval; gross RAH; RBBB
Fallot's tetralogy	Newborn: Normal or T +ve V1 Older: RVH; RAH
Pulmonary atresia	RAH
Tricuspid atresia	LAD; RAH; LVH

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SUPRAVENTRICULAR TACHYCARDIA

Early diagnosis and effective management of supraventricular tachycardia (SVT) are vital as there is a small risk of mortality

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Recurrent condition
  - family may identify as 'another attack'
- Infants
  - gradual onset of increasing tachypnoea
  - poor feeding
  - pallor
  - occasionally more dramatic presentation with a rapid onset of severe cardiac failure
- Toddlers
  - recurrent episodes of breathlessness, cold sweats and pallor
- Older children
  - recurrent palpitations, episodes of dizziness and pallor

Investigations

- Confirm diagnosis with 12-lead ECG
- Continuous ECG monitoring is essential
- Assess for cardiac failure

Differential diagnosis

- Sinus tachycardia, particularly in infants, can be >200/min. However, rates of 220–300/min are more likely to be SVT
- If first presentation, check for any other cause of cardiac failure
- Failure to respond to adenosine can be used to distinguish origin of a tachycardia in a stable patient

Causes of tachyarrhythmias

- Re-entrant congenital conduction pathway abnormality (common)
- Poisoning
- Metabolic disturbance
- After cardiac surgery
- Cardiomyopathy
- Long QT syndrome

ECG DIAGNOSIS

Infants

- Majority have a P wave before every QRS complex, usually by >70 msec (2 mm at 25 mm/sec)
- QRS complexes are generally normal but may be wide
- Accessory pathway frequently capable of anterograde as well as retrograde conduction
  - this will be revealed during normal sinus rhythm by short P-R interval and presence of a delta wave (classic Wolff-Parkinson-White syndrome)

Older children

- Nodal tachycardias become more common with increasing age
  - characterised by fast, regular, narrow QRS complexes without visible P waves
- Wide QRS complex or bundle branch block in childhood is rare
  - changes also present in sinus rhythm
  - review previous ECGs

If in doubt, seek more experienced help

IMMEDIATE TREATMENT

- Resuscitate (ABC) first
- If first presentation, refer to consultant
- See following **Algorithms**

Vagal manoeuvres

These may include:

- Diving reflex
- wrap infants in a towel and immerse their whole face into iced water for about 5–10 sec
- in children, place a bag or rubber glove containing iced water over face
- One side carotid massage
- Valsalva manoeuvre
- Where possible, maintain ECG monitoring and recording during all procedures

Do NOT use eyeball pressure because of risk of ocular damage

Adenosine

- Drug of choice as it has a rapid onset of action and not negatively inotropic
- Very short half-life (10–15 sec) giving short-lived side-effects (flushing, nausea, dyspnoea, chest tightness)
- Effective in >80% of junctional tachycardias and will not precipitate ventricular tachycardias into ventricular fibrillation
- Can be used in broad-complex tachycardia of uncertain origin
- Must be given as a rapid bolus IV via a large peripheral or central vein and followed by sodium chloride 0.9% flush
- In patients with sinus tachycardia, heart rate will slow to bradycardia but will rapidly increase again

Other drugs

- If adenosine ineffective, seek advice from a paediatric cardiologist
- In refractory Wolff-Parkinson-White tachycardia, flecainide is particularly useful
- In refractory atrial tachycardia, amiodarone is useful

Do not use verapamil and propranolol in same patient, as both have negative inotropic effects. Do not use verapamil in children aged <1 yr

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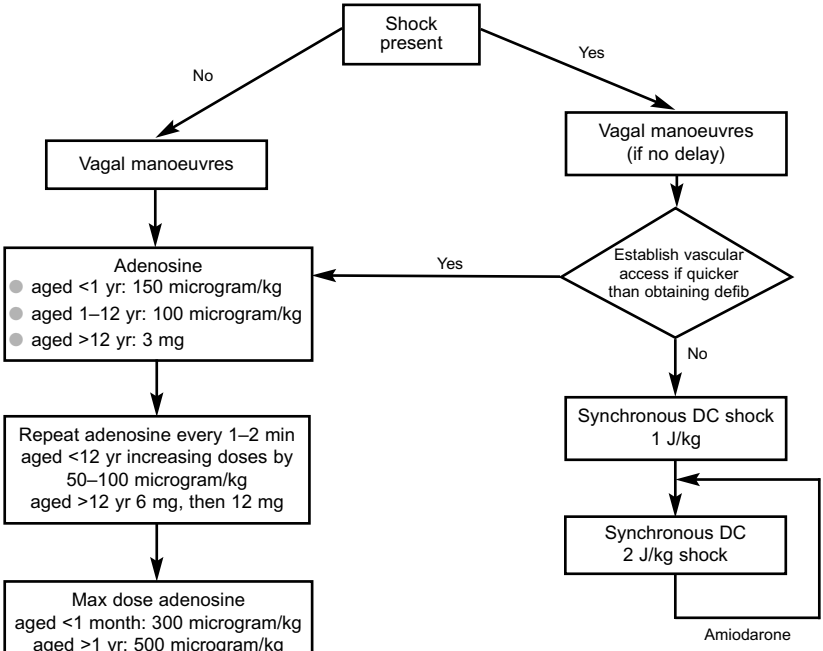
Supraventricular tachycardia



- Discuss with cardiologist
- Consider:
  - synchronous DC shock
  - other anti-arrhythmics (seek advice)

- Adenosine may be used in preference to electrical shock
- An anaesthetic must be given for DC shock if patient responsive to pain

WIDE COMPLEX TACHYCARDIA

RECOGNITION AND ASSESSMENT

Definition

- Ventricular tachycardia
  - ≥3 successive ectopic ventricular beats
  - sustained if it continues >30 sec

Causes

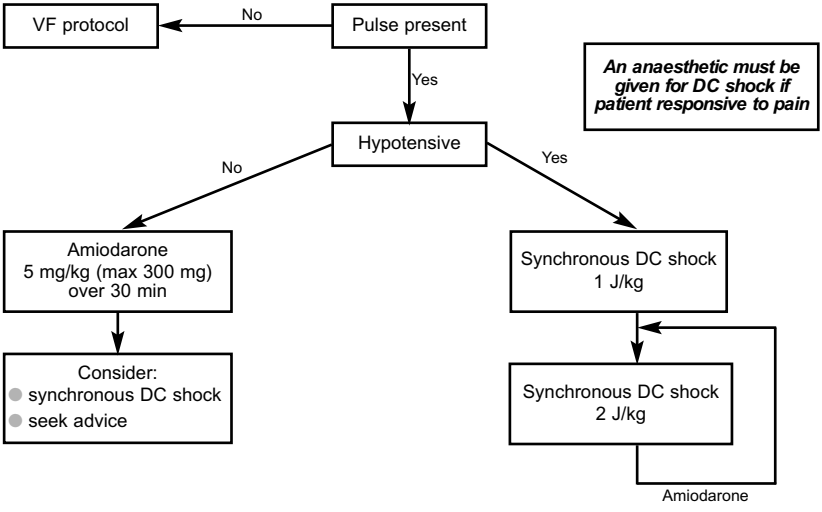
- Underlying cause (e.g. myocarditis, cardiomyopathy, or patient with congenital heart disease)
- Poisoning (e.g. phenothiazines, tricyclic antidepressants, quinidine and procainamide)
- Electrolyte disturbance (e.g. hypokalaemia, hypomagnesaemia)
- Ventricular tachycardia can degenerate into ventricular fibrillation

Diagnosis

- Wide-QRS SVT (SVT with aberrant conduction) is uncommon in infants and children. Correct diagnosis and differentiation from VT depends on careful analysis of at least a 12-lead ECG +/- an oesophageal lead
- Assess patient and obtain family history to identify presence of an underlying condition predisposing to stable ventricular tachycardia
- SVT or VT can cause haemodynamic instability: response to adenosine can help identify underlying aetiology of the arrhythmia, but adenosine should be used with extreme caution in haemodynamically stable children with wide-complex tachycardia because of the risk of acceleration of tachycardia and significant hypotension. This should not delay definitive treatment in children with shock
- Seek advice
- Ventricular tachycardia not always obvious on ECG, clues are:
  - rate varies between 120 and 250 beats/min (rarely 300 beats/min)
  - QRS complexes are almost regular though wide
  - QRS axis abnormal for age (normal for aged >6 months is <+90º)
  - no preceding P wave, or A-V dissociation
  - fusion beats (normally conducted QRS complex merges with an abnormal discharge)

IMMEDIATE TREATMENT

Ventricular tachycardia



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- Treatment of haemodynamically stable child with ventricular tachycardia should always include early consultation with a paediatric cardiologist. They may suggest amiodarone: can cause hypotension, which should be treated with volume expansion
- Use synchronous shocks initially, as these are less likely than an asynchronous shock to produce ventricular fibrillation. If synchronous shocks are ineffectual, and child is profoundly hypotensive, subsequent attempts will have to be asynchronous
- Treatment of torsade de pointes ventricular tachycardia is magnesium sulphate 25–50 mg/kg (up to 2 g) diluted to 100 mg/mL in sodium chloride 0.9% over 10–15 min. Can be repeated once if necessary
- Amiodarone 5 mg/kg (max 300 mg) may be given over 3 min in ventricular tachycardia if child in severe shock

BRADYARRHYTHMIAS

- Urgently manage:
  - pre-terminal event in hypoxia or shock
  - raised intracranial pressure
  - vagal stimulation

Investigations

- ECG to look for:
  - conduction pathway damage after cardiac surgery
  - congenital heart block (rare)
  - long QT syndrome

Management

- ABC approach: ensure adequate oxygenation and ventilation
- If vagal stimulation is cause, give atropine 20 microgram/kg (min 100 microgram; max 600 microgram)
- Consider IV isoprenaline infusion
- Contact paediatric cardiologist for advice
  - fax ECG to cardiologist

ENDOCARDITIS PROPHYLAXIS • 1/1

Endocarditis prophylaxis is not recommended unless undergoing gastro-intestinal or genito-urinary tract procedure at a site where infection is suspected

INDICATIONS

Cardiac risk factors

- Acquired valvular heart disease with stenosis or regurgitation
- Valve replacement
- Structural congenital heart disease, including surgically corrected or palliated structural conditions, but excluding isolated atrial septal defect, fully repaired ventricular septal defect or fully repaired patent ductus arteriosus, and closure devices judged to be endothelialised
- Previous infective endocarditis
- Hypertrophic cardiomyopathy

If there is uncertainty, seek advice from cardiology team at local paediatric cardiac surgical centre

MANAGEMENT

- Patients at risk of endocarditis should be:
  - advised to maintain good oral hygiene
  - told how to recognise signs of infective endocarditis, and advised when to seek expert advice
- Investigate promptly any infection in patients at risk of endocarditis and treat appropriately to reduce the risk of endocarditis
- If patients at risk of endocarditis are undergoing a gastro-intestinal or genito-urinary tract procedure **at a site where infection suspected** seek senior advice

POISONING AND DRUG OVERDOSE • 1/3

The poisoned

- Toddlers (accidental poisoning)
- Older children, particularly girls (intentional self-poisoning)

The poisoners

- Most childhood poisonings are accidental
- Intentional poisoning may be by the child or an adult
- Inadvertent poisoning may occur in a medical setting

The poison

- Children will eat and drink almost anything

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Depressed respiration suggests centrally-acting drug
- Skin blisters (between knees/toes) common after barbiturates and tricyclics
- Hypothermia after exposure or barbiturates
- Venepuncture marks and pinpoint pupils suggest opioid overdose
- Burns around mouth

Life-threatening features

- Coma
- Cyanosis
- Hypotension
- Paralytic ileus

Poison(s)/drug(s) information

- Ask patient, relatives, GP, ambulance crew. Retain any containers found
- if identification doubtful, send parents home for poison
- Ask about visitors to the house/visits to other houses (e.g. grandparents)
- Quantity ingested: Difficult to quantify but parents may know how full a bottle should have been
- assume child has ingested something even if found with a few tablets and an empty bottle
- Time of ingestion, including multiple doses
- Other possible poisons/drugs taken

Investigations

- U&E
- Blood gases and acid-base
- Save blood and urine for toxicological analysis. Urgent measurement of plasma/serum concentrations essential in diagnosis and management of poisoning with ethylene glycol, iron, lithium, methanol, paracetamol, theophylline and salicylate. With exception of paracetamol, no need to measure concentrations of these substances unless clear history of ingestion. Specify which drugs suspected or urine for 'drugs of abuse'

Request plasma paracetamol concentration in all unconscious patients in whom drug overdose considered

Seek advice

- Use Toxbase: www.toxbase.org access and password available in A&E
- if further information required, contact National Poisons Information Service (0844 892 0111)

Always admit a child who is symptomatic or who has ingested iron, digoxin, aspirin or a tricyclic antidepressant. If child not admitted always consult on-call paediatric SpR before sending home

IMMEDIATE TREATMENT

Separate guidelines give more detailed advice on management of overdose with alcohol, iron, paracetamol, phenothiazines, salicylates and tricyclic antidepressants

Assess airway, breathing and circulation

- Maintain airway
- if airway not protected, may need intubation and ventilation
- if cyanosed or rate and depth of respiration obviously low, arterial blood gases indicated
- if $PaCO_2$ high or rising, mechanical ventilation indicated
- Correct hypotension
- raise foot of bed
- if in haemodynamic shock, give IV bolus of sodium chloride 0.9% (20 mL/kg over 10 min). Assess and repeat if still in shock
- consider need for central venous pressure (CVP) monitoring

Neurological

- Control convulsions
- if unconscious, treat as head injury until proved otherwise

Drug absorption

- Give antidote if appropriate
- Activated charcoal in patients who have ingested life-threatening amounts of a toxic agent up to 1 hr previously, provided patient conscious or airway can be protected. Give 1 g/kg (max 50 g) oral (disguised with soft drink/fruit juice) or via nasogastric tube. Activated charcoal does not affect absorption of acids, alkalis, alcohols, cyanide, ethylene glycol, iron or lithium
- Do not give ipecacuanha, it does not empty the stomach reliably and can be dangerous
- Stop any regular medication that might enhance effect of substance taken in overdose

SUBSEQUENT MANAGEMENT

- If unconscious, admit to a high-dependency nursing area and attach an ECG monitor
- Supportive care alone required for majority of acutely poisoned patients
- If deliberate self harm, refer to CAMHS – see **Self harm** guideline

MONITORING TREATMENT

- Monitor conscious level, temperature, respiration, pulse and BP until these return to normal
- No need to monitor drug concentrations other than to guide use of measures to enhance drug elimination
- If unconscious, make full head injury observations
- Record pulse, respiratory rate, BP, pupil size and reaction, and level of consciousness hourly for at least 4 hr then increase interval if stable

PSYCHIATRIC REVIEW

- Offer all patients admitted after deliberate acute self-poisoning or drug overdose an interview with the psychiatric priority referral team within 24 hr of admission or regaining consciousness

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DISCHARGE AND FOLLOW-UP

- **When discharged from hospital patients should have:**
  - been conscious and alert with normal vital signs for at least 6 hr
  - no evidence of significant organ dysfunction as a result of poisoning/drug toxicity
  - been interviewed by a member of the psychiatric priority referral team where indicated
  - follow-up appointment in psychiatric clinic (if recommended by psychiatrist)
  - follow-up appointment in paediatric clinic (if persistent sequelae of poisoning require review)

ALCOHOL POISONING • 1/2

Refer all children with features of alcohol intoxication to hospital

RECOGNITION AND ASSESSMENT

Symptoms and signs

Table 1: Assessment of alcohol poisoning

Mild toxicity	● Impaired visual acuity and co-ordination ● Emotional lability
Moderate toxicity	● Slurred speech, diplopia, blurred vision, ataxia, lack of co-ordination, blackouts, sweating, tachycardia, nausea, vomiting, incontinence ● Acidosis, hypoglycaemia, hypokalaemia
Severe toxicity	● Cold clammy skin, hypothermia, hypotension, stupor, coma, dilated pupils, depressed or absent tendon reflexes ● Severe hypoglycaemia, convulsions, respiratory depression, metabolic acidosis ● Cardiac arrhythmias (e.g. atrial fibrillation, atrio-ventricular block)
Potentially fatal	● Deep coma, respiratory depression or arrest, circulatory failure

Alcoholic drinks/preparations

- Spirits are particularly dangerous
- Alcopops
- Perfumes, colognes and aftershaves
- Mouth washes (some)
- Methylated spirit
- Hand gels
- Detergents
- Fake vodka may contain propranolol and methanol
- Other drugs often taken too but not disclosed

IMMEDIATE TREATMENT

- Ensure clear airway and adequate ventilation
- Gut decontamination is unlikely to be of benefit
- activated charcoal does not significantly reduce rate of absorption
- Correct hypoglycaemia as quickly as possible
- if awake, give oral glucose
- if drowsy or unconscious, give glucose 10% 2 mL/kg IV
- check blood glucose hourly until consciousness regained
- Correct hypotension (see **Poisoning and drug overdose** guideline)
- Correct acid-base and metabolic disturbance
- Correct hypothermia using conventional means (e.g. Bair Hugger, blankets)
- Control convulsions with IV lorazepam
- If blood ethanol >5 g/L (108.5 mmol/L) or if arterial pH <7.0, consider haemodialysis
- discuss with National Poisons Information Service (0844 892 0111)

Investigations

- Blood glucose
- In moderate to severe toxicity
- U&E
- arterial blood gases
- blood ethanol concentration
- toxicology blood and urine for drugs of abuse
- 12-lead ECG

Assessment of severity

- Blood ethanol is a guide to severity of poisoning
- 0.2–1.0 g/L (4–22 mmol/L) mild toxicity
- 1–2 g/L (22–43 mmol/L) moderate toxicity
- >2–4 g/L (>43 mmol/L) severe toxicity and potentially fatal
- Observe for at least 4 hr if >0.4 mL/kg body weight of absolute ethanol had been ingested (i.e. 1 mL/kg 40% spirit, 4 mL/kg 10% wine or 8 mL/kg 5% beer)
- Monitor pulse, blood pressure and body temperature

SUBSEQUENT MANAGEMENT

- See **Poisoning and drug overdose** guideline
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from National Poisons Information Service (0844 892 0111)

IRON POISONING • 1/2

RECOGNITION AND ASSESSMENT

Symptoms and signs

Time	Symptoms and signs
<6 hr after ingestion	● Nausea, vomiting, abdominal pain and diarrhoea ● Vomit and stools often grey or black ● Polymorph leucocytosis and hyperglycaemia suggest toxicity but their absence does not exclude it
6–12 hr after ingestion	● Early features improve in mild cases ● Possibly persistent hyperglycaemia/metabolic acidosis in more serious cases
>12 hr after ingestion	● In serious cases, evidence of hepatocellular necrosis appears with jaundice, bleeding, hypoglycaemia, encephalopathy and metabolic acidosis. Hypotension may occur
2–5 weeks after ingestion	● Gastric stricture or pyloric stenosis may start to cause obstructive symptoms

Investigations

- If ingested dose >20 mg/kg elemental iron, measure serum iron 4 hr after ingestion
- U&E, creatinine
- INR
- Blood glucose
- If presenting within 2 hr of ingestion, request plain abdominal X-ray
  - tablets are sometimes visible in the stomach or small bowel
  - if patient could be pregnant, do **NOT** X-ray

Assessment of severity

Review both clinical and laboratory features

- Estimate ingested dose of elemental iron, BNFc lists quantity of elemental iron in various preparations
  - <20 mg/kg mild or no toxicity
  - ≥20 mg/kg toxicity likely
  - >200 mg/kg severe toxicity, possibly fatal
- Coma and shock indicate severe poisoning: urgent treatment required
- Serum iron taken at 4 hr after ingestion is best laboratory measure
  - <3 mg/L (55 micromol/L) mild toxicity
  - 3–5 mg/L (55–90 micromol/L) moderate toxicity
  - >5 mg/L (90 micromol/L) severe toxicity
- Absence of visible tablets on X-ray does not eliminate possibility of ingestion

Action

- Discuss **all** cases with National Poisons Information Service (NPIS) (0844 892 0111) for advice

IMMEDIATE TREATMENT

Unconscious or in shock

- Assess airway, breathing, circulation
- Secure airway, treat shock, control seizures
- IV fluids to replace losses
- Commence desferrioxamine IV (see **Desferrioxamine**)
  - if this is before time when serum iron should be taken (4 hr), take sample for serum iron immediately before commencing desferrioxamine
  - do **not** delay starting desferrioxamine IV

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- Monitor cardiac rhythm, BP and urine output
- Check U&E, FBC, blood glucose, LFTs, INR and arterial blood gases
- Discuss whole bowel irrigation with NPIS if ingested dose >60 mg/kg and/or tablets seen on X-ray and child presents **within 1 hr**
- do not use activated charcoal as it does not adsorb iron

Conscious and not in shock

- Check serum iron concentration at 4 hr
- Interpret serum iron concentration in view of child's clinical condition and history

Moderate poisoning: serum iron 3–5 mg/L

- Repeat serum iron measurement after further 2 hr, even if asymptomatic
- If concentration falling, no further treatment required
- If concentration rising and child symptomatic, give desferrioxamine IV (see **Desferrioxamine**)

Severe poisoning: serum iron >5 mg/L

- If asymptomatic, repeat serum iron after 2 hr: if concentration falling treatment unlikely to be required
- If symptomatic, give desferrioxamine IV (see **Desferrioxamine**)

Desferrioxamine

- Before starting treatment, contact NPIS (0844 892 0111) for advice
- Starting dose is 15 mg/kg/hr
- Reduce after 4–6 hr
- maximum 80 mg/kg in 24 hr
- desferrioxamine commonly causes hypotension if infused more rapidly than recommended rate and turns urine red/orange but rarely causes rashes or anaphylactic reactions

SUBSEQUENT MANAGEMENT

- See **Poisoning and drug overdose** guideline
- If slow release preparations ingested, repeat serum iron after further 6–8 hr
- In patients with severe toxicity:
- arterial blood gases and correct acidosis
- If metabolic acidosis persists despite correction of hypoxia and adequate fluid resuscitation, give IV sodium bicarbonate 1 mL/kg 8.4% bicarbonate diluted in glucose 5% or sodium chloride 0.9% 500 mL at 2–3 mL/kg/hr
- monitor renal and liver function
- be alert for evidence of gut perforation or infarction
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from NPIS (0844 892 0111)

PARACETAMOL POISONING • 1/5

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Common: nausea and vomiting
- Rare: coma and metabolic acidosis
- Late: abdominal pain

Management for

- Paracetamol dose >6 g or >75 mg/kg
- Staggered overdose [including chronic therapeutic excess >75 mg/kg/d (>60 mg/kg in neonate)]
- Symptomatic

OR

- INR >1.3 or ALT >upper limit of normal, or abnormal acid/base or bicarbonate
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from National Poisons Information Service (NPIS) 0844 892 0111
- If there is **absolute certainty** that a single dose of paracetamol of <6 g and <75 mg/kg has been ingested, plasma paracetamol need not be measured and child requires no antidote

Investigations

- Plasma paracetamol 4–16 hr (but not outside this interval) is a reliable guide to the need for treatment after single overdose ingested of <60 min
- If patient presents >8 hr after single overdose; or after staggered overdose; request baseline:
  - FBC, INR
  - U&E, liver function, phosphate
  - acid-base (venous sample)

IMMEDIATE TREATMENT

- Compare plasma paracetamol with treatment graph (**Figure 1**)
  - if above, or on, the 'treatment line', give IV acetylcysteine in glucose 5%
- Time interval is critical in assessing need for treatment. Detailed questioning essential

If there is doubt about timing or need for treatment, treat

Time from overdose (hr)	Guidance on use of acetylcysteine
<1	Give activated charcoal 1 g/kg (max 50 g) oral or via nasogastric tube (gastric lavage is not indicated)
4–7	Await paracetamol level if available <8 hr from ingestion. Treat if level ≥ 'treatment line' OR if biochemical tests (INR, ALT) suggest acute liver injury
8–14	Give at once while awaiting paracetamol concentration result. Cease if concentration well below appropriate 'treatment line' and ALT within normal limit, INR ≤1.3
15–24	Give at once. Cease at 24 hr after ingestion if patient asymptomatic, and INR ≤1.3, and ALT <upper limit of normal. Otherwise complete antidote course
Multiple/ staggered overdose	Plasma paracetamol will confirm ingestion but cannot be related to nomogram. Start acetylcysteine and discuss with NPIS
>24	Give if paracetamol still detectable in the blood (>5 mg/L), or INR >1.3 or ALT >twice upper limit of normal, or symptomatic. If patient has, or is at risk of developing, fulminant hepatic failure (see **life-threatening features** below), continue to give 50 mg/kg in 500 mL every 8 hr Discuss with NPIS and follow Toxbase guidance

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Acetylcysteine dosage (see BNFC*)

Weight <20 kg (including neonates)	● **First phase:** 150 mg/kg IV in 3 mL/kg glucose 5% over 60 min, then ● **Second phase:** 50 mg/kg IV in 7 mL/kg glucose 5% over 4 hr, then ● **Third phase:** 100 mg/kg IV in 14 mL/kg glucose 5% over 16 hr ● Then careful review at 16 hr, in case of need to continue third infusion phase
Weight 20–40 kg	● **First phase:** 150 mg/kg IV in 100 mL glucose 5% over 60 min, then ● **Second phase:** 50 mg/kg IV in 250 mL glucose 5% over 4 hr, then ● **Third phase:** 100 mg/kg IV in 500 mL glucose 5% over 16 hr ● Then careful review at 16 hr, in case of need to continue third infusion phase
Weight >40 kg (dose capped at 110 kg body weight)	● **First phase:** 150 mg/kg IV (max 16.5 g) in 200 mL glucose 5% over 1 hr, then ● **Second phase:** 50 mg/kg IV (max 5.5 g) in 500 mL glucose 5% over 4 hr, then ● **Third phase:** 100 mg/kg IV (max 11.0 g) in 1000 mL glucose 5% over 16 hr ● Then careful review at 16 hr, in case of need to continue third infusion phase

* BNFC dose calculation and prescription method is simple to prescribe and give. Alternatively, the dosage calculation described in Toxbase yields the same drug dose but is prepared differently and prescribed by volumes

Alternative dosage chart for body weight >40 kg

● See **Adult acetylcysteine dose and administration** in the **BNF**

Adult acetylcysteine prescription (each ampoule = 200 mg/mL acetylcysteine)						
Regimen	**First infusion**		**Second infusion**		**Third infusion**	
Infusion fluid	200 mL glucose 5% or sodium chloride 0.9%		500 mL glucose 5% or sodium chloride 0.9%		1000 mL glucose 5% or sodium chloride 0.9%	
Duration of Infusion	1 hr		4 hr		16 hr	
Drug dose	150 mg/kg acetylcysteine		50 mg/kg acetylcysteine		100 mg/kg acetylcysteine	
Patient weight	**Ampoule volume**	**Infusion rate**	**Ampoule volume**	**Infusion rate**	**Ampoule volume**	**Infusion rate**
kg	**mL**	**mL/hr**	**mL**	**mL/hr**	**mL**	**mL/hr**
40–49	34	234	12	128	23	64
50–59	42	242	14	129	28	64
60–69	49	249	17	129	33	65
70–79	57	257	19	130	38	65
80–89	64	264	22	131	43	65
90–99	72	272	24	131	48	66
100–109	79	279	27	132	53	66
≥110	83	283	28	132	55	66

● If for any reason glucose 5% unsuitable, substitute sodium chloride 0.9%

Prepare and check infusion bags carefully.
Administration errors are common

Acetylcysteine can cause a pseudo-allergic reaction (wheezing, flushing, hypotension) that is usually relieved by stopping infusion but occasionally chlorphenamine and hydrocortisone are required. Once reaction has subsided, recommence infusion at lower rate of 50 mg/kg/hr to complete the 150 mg/kg (max 16.5 g), then start the second phase infusion of 50 mg/kg (max 5.5 g) over 4 hr

MONITORING TREATMENT

- Severe liver damage in the context of paracetamol poisoning has been defined as a peak plasma ALT activity exceeding 1000 iu/L

Time of presentation after overdose (hr)	Monitoring/continued treatment	Discharge policy
<8	● INR, AST/ALT, creatinine, bicarbonate 24 hr after overdose or when antidote treatment complete ○ if INR >1.3 or creatinine raised, or patient acidotic, repeat third infusion phase until INR <1.3 ○ recheck INR and U&E 12-hrly until clearly falling ● Do NOT correct INR with vitamin K without prior discussion with tertiary liver unit, see below for management of life-threatening conditions including use of FFP	● Discharge if INR ≤1.3, AST/ALT and plasma creatinine normal at 24 hr after overdose, or after antidote treatment complete, with warning to return if vomiting or abdominal pain occur
8–15	● INR, AST/ALT, creatinine, bicarbonate and phosphate 24 hr after overdose or when antidote treatment complete ○ if INR >1.3 or creatinine raised, or patient acidotic, repeat third infusion phase until INR <1.3 ○ recheck INR and U&E 12-hrly until clearly falling ○ discuss with NPIS	● If INR ≤1.3, AST/ALT and plasma creatinine normal: ○ discharge asymptomatic patients 12 hr after antidote treatment with warning to return if vomiting or abdominal pain occur ○ if INR >1.3 and rises, and/or plasma creatinine raised after antidote treatment, monitor and observe until these indices are falling and INR <2.0

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Time of presentation after overdose (hr)	Monitoring/continued treatment	Discharge policy
≥16	● Observe for signs of encephalopathy (mental confusion, drowsiness, spatial disorientation, asterixis) ● Urine output (maintain good flow†) ● Capillary blood glucose 4-hrly ● Blood gases and acid-base daily ● INR, AST/ALT, creatinine, bicarbonate and phosphate 24 hr after overdose or when antidote treatment complete ○ if INR >1.3 and rises, or creatinine raised, or patient acidotic, repeat third infusion phase until INR <2.0 ○ recheck INR and U&E 12-hrly until INR clearly falling and creatinine <10% higher than start value	● If INR ≤1.3, AST/ALT and plasma creatinine normal: ○ discharge asymptomatic patients 12 hr after end of antidote treatment with warning to return if vomiting or abdominal pain occur ○ if INR >1.3 and rises, and/or plasma creatinine raised after antidote treatment, monitor and observe until these indices are falling and INR <2.0 ● Patients presenting 24–36 hr after overdose can develop hepatic dysfunction after this time, even if INR, ALT and creatinine normal at time of presentation: repeat these indices 12 hr later

Life-threatening features

- A poor prognosis indicated by:
  - INR >3.0
  - serum creatinine >200 µmol/L
  - blood pH <7.3
  - signs of encephalopathy
- If any of these features are present after overdose, **seek advice from local tertiary liver unit**
  - †insert urinary catheter to monitor urine flow and rehydrate to maintain urine output >2 mL/kg/hr or 100 mL/hr whichever is smaller
  - if unresponsive to IV fluids, give furosemide and consider low-dose dopamine
  - insert CVP line to monitor response to IV fluids only if INR normal
- Patients with incipient or established hepatic failure may be candidates for liver transplantation
- Treat haemorrhage with fresh frozen plasma
- Hypophosphataemia usually occurs after paracetamol poisoning and correlates well with degree of hepatic damage

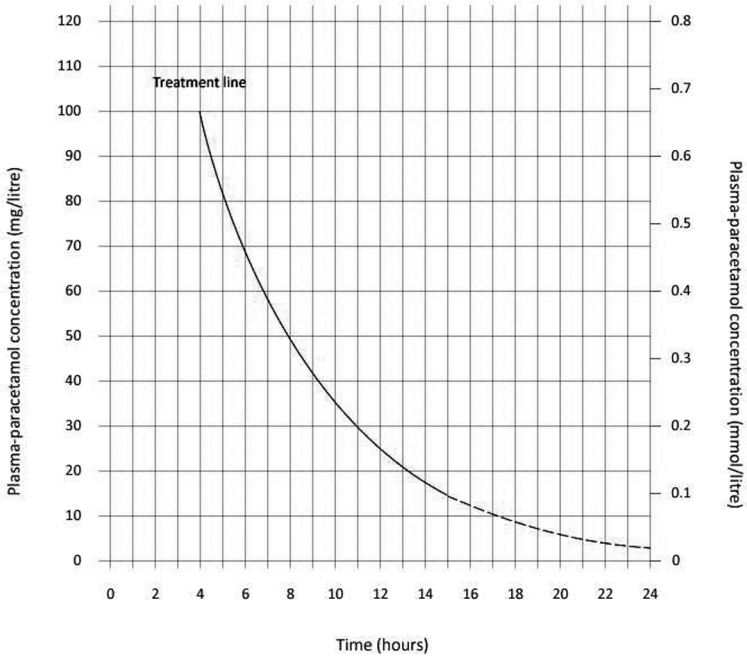
Psychiatric review

- Offer all patients admitted after acute self-poisoning or deliberate drug overdose an interview with a member of the psychiatric priority referral team within 24 hr of admission or regaining consciousness

DISCHARGE AND FOLLOW-UP

- See **Poisoning and drug overdose** guideline
- Advise all patients to return to hospital if vomiting or abdominal pains develop or recur

Figure 1: Treatment graph for paracetamol overdose



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RECOGNITION AND ASSESSMENT

Symptoms and Signs

- Drowsiness
- Confusion

Common preparations

- Chlorpromazine
- Levomepromazine
- Perphenazine
- Prochlorperazine
- Promazine
- Trifluoperazine
- Metoclopramide

Extrapyramidal side effects

Not dose-related

- Dystonia (e.g. oculogyric crises, spasmodic torticollis)
- Dyskinesia
- Appear after only a few doses

Complications

- Convulsions
- Hypothermia
- Hypotension
- Arrhythmias (e.g. sinus tachycardia, QT and QRS prolongation, VT/VF, bundle branch/atrio-ventricular block)
- Respiratory depression
- Rhabdomyolysis
- Renal failure

IMMEDIATE TREATMENT

- If patient presents within 1 hr of ingesting a potentially toxic dose, give activated charcoal 1 g/kg (max 50 g)
- Maintain clear airway and adequate ventilation
- Correct hypotension (see **Poisoning and drug overdose** guideline)
- Correct hypothermia using conventional means (e.g. Bair Hugger, blankets)
- Correct acid-base and metabolic disturbance with bicarbonate infusion (see **Salicylate poisoning** guideline)
- Control convulsions with IV lorazepam

Treatment of extrapyramidal side effects

- Procyclidine orally
- in severe reactions give IV or IM
- subsequent oral doses may be needed for 2–3 days
- if procyclidine not available, give trihexyphenidyl hydrochloride or diazepam

MONITORING TREATMENT/SUBSEQUENT MANAGEMENT

- See **Poisoning and drug overdose** guideline
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from National Poisons Information Service (0844 892 0111)

SALICYLATE POISONING • 1/2

RECOGNITION AND ASSESSMENT

Symptoms and signs

Common features

- Vomiting
- Dehydration
- Tinnitus
- Vertigo
- Deafness
- Sweating
- Warm extremities with bounding pulse
- Increased respiratory rate
- Hyperventilation
- Acid-base disturbance:
  - aged >4 yr usually mixed respiratory alkalosis and metabolic acidosis with normal or high arterial pH
  - aged <4 yr usually a dominant metabolic acidosis with low arterial pH

Uncommon features

- Haematemesis
- Hyperpyrexia
- Hypoglycaemia
- Hypokalaemia
- Thrombocytopenia
- Increased INR/PTR
- Intravascular coagulation
- Renal failure
- Non-cardiac pulmonary oedema
- Confusion
- Disorientation
- Coma
- Convulsions

Preparations

- Aspirin tablets
- Methyl salicylate (Oil of Wintergreen), very toxic
- Choline salicylate (dental gels)
- Numerous over-the-counter analgesics/antipyretics contain aspirin

Investigations

- U&E, creatinine
- INR
- Arterial blood gases
- Blood glucose (capillary)
- In asymptomatic patients with a reliable history of ingestion of <125 mg/kg of aspirin, plasma salicylate not required
- In those who have ingested >125 mg/kg, measure plasma salicylate level
  - repeat after 2 hr: if rising, repeat levels every 3 hr until falling
- If coincident paracetamol overdose, check salicylate level before administration of N-acetylcysteine
- Urine pH

Assessment of severity

- Severity cannot be assessed from plasma salicylate concentrations alone
- Neurological features (e.g. confusion and impaired consciousness), metabolic acidosis, and high salicylate concentrations indicate severe poisoning
- Risk factors for death include:
  - aged <10 yr
  - CNS features
  - acidosis
  - hyperpyrexia
  - late presentation
  - pulmonary oedema
  - salicylate concentration >5.1 mmol/L

IMMEDIATE TREATMENT

- If ingested >125 mg/kg salicylate within previous hour, give oral activated charcoal 1 g/kg (maximum 50 g), mixed with soft drink/fruit juice if necessary to disguise taste
- Rehydrate orally (IV if vomiting)

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Interpretation of plasma salicylate concentrations

- Clinical presentation is most important factor
- Late presenting patient may have a subtoxic salicylate concentration, but serious acid-base or CNS disturbances
- **If levels still rising, repeat oral activated charcoal**
- Plasma salicylate <350 mg/L (2.5 mmol/L) and mild clinical effects:
  - continue maintenance management
- Plasma salicylate 350–700 mg/L (2.5–5.0 mmol/L) and moderate clinical effects:
  - continue maintenance management and start alkaline diuresis in children aged <5 yr
  - if plasma salicylate >500 mg/L (3.6 mmol/L) in children aged ≥5 yr start alkaline diuresis
- Plasma salicylate >700 mg/L (5.0 mmol/L) and severe clinical effects
  - use **haemodialysis**

Children aged <10 yr have an increased risk of salicylate toxicity and may require haemodialysis at an earlier stage

Alkaline diuresis

- If serum potassium low, give potassium chloride 1 mmol/kg oral
  - or if not tolerated sodium chloride 0.45%/glucose 5% with 20 mmol potassium in 500 mL at 100% maintenance
- If serum potassium within normal range, alkalinise urine to enhance salicylate excretion (optimum urine pH 7.5–8.5)
  - give sodium bicarbonate 8.4% 1 mL/kg (1 mmol/kg) in 500 mL sodium chloride 0.9% or glucose 5% at 2–3 mL/kg/hr, and repeat if necessary to maintain urine pH 7.5–8.5
  - repeat salicylate levels and potassium level every 1–2 hr

Do not use volumes of IV fluids above maintenance requirements (forced diuresis) – they do not increase salicylate elimination and can cause pulmonary oedema

Haemodialysis

Use in patients with severe poisoning

- Plasma concentrations >700 mg/L (5.0 mmol/L)
- Renal failure
- Congestive cardiac failure
- Non-cardiogenic pulmonary oedema
- Convulsions
- CNS effects not resolved by correction of acidosis
- Persistently high salicylate concentrations unresponsive to urinary alkalinisation
- Severe metabolic acidosis
- Children aged <10 yr who have an increased risk of salicylate toxicity

MONITORING TREATMENT/SUBSEQUENT MANAGEMENT

- See **Poisoning and drug overdose** guideline
- During alkaline diuresis, check U&E, blood glucose, acid-base hourly
- Repeat plasma salicylate 2-hrly until falling
  - if plasma salicylate continues to rise, consider a second dose of activated charcoal
- Continue therapy until patient improving and plasma salicylate falling
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from National Poisons Information Service (0844 892 0111)

TRICYCLIC POISONING • 1/2

RECOGNITION AND ASSESSMENT

Symptoms and signs

Early in poisoning

- Anticholinergic effects (tachycardia, hot, dry skin, dry mouth and tongue, dilated pupils, urinary retention)
- Ataxia, nystagmus
- Drowsiness
- Metabolic acidosis
- Hypokalaemia

Severe cases

- Hypotension
- Increased tone, hyperreflexia
- Coma
- Seizures
- Respiratory depression
- Cardiac arrhythmias

Preparations

- Amitriptyline
- Amoxapine
- Clomipramine
- Dosulepin
- Doxepin
- Imipramine
- Lofepramine
- Nortriptyline
- Trimipramine

Investigations

- U&E
- Arterial blood gas
- 12-lead ECG, large doses cause prolongation of P-R and QRS intervals

IMMEDIATE TREATMENT

If a benzodiazepine has also been taken, do NOT give flumazenil

- Correct any hypoxia
- if $PaCO_2$ >6 kPa in respiratory failure arrange assisted ventilation
- If high dose (e.g. amitriptyline >4 mg/kg) within previous hour, give activated charcoal 1 g/kg (max 50 g) either oral (mixed with soft drink/fruit juice if necessary to disguise taste) or, if drowsy or unconscious, by nasogastric tube (provided airway can be protected)
- Admit to HDU
- Treat arrhythmias by correction of hypoxia and acidosis
- sodium bicarbonate 8.4% diluted in an equal volume of glucose 5% and give a 'calculated' dose: dose (in mmol) = desired change in base deficit (current-target) x 0.3 x weight (kg) to a maximum of 50 mmol (ideally via central vein). For rapid correction administer over 20 min, otherwise administer at a rate of 1 mmol/min. **Caution required if solution to be given by peripheral venous line, as irritant to veins and can cause local necrosis in cases of extravasation**
- if unresponsive discuss using lipid emulsion with National Poisons Information Service (NPIS) (0844 892 0111)
- do not use arrythmics
- consult local paediatric cardiac team
- Hyperpyrexia (>39ºC): treat with ice bags and sedation: if persists give dantrolene, discuss with NPIS

Prolonged resuscitation (up to 1 hr) may be successful after cardiac arrest

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MONITORING TREATMENT

- See **Poisoning and drug overdose** guideline
- Cardiac monitor for at least 6 hr
- asymptomatic patients with normal ECG after 6 hr are unlikely to develop late complications

SUBSEQUENT MANAGEMENT

- See **Poisoning and drug overdose** guideline
- Follow guidance on www.toxbase.org (password from A&E)
- Further advice from NPIS (0844 892 0111)
- Consider second dose of charcoal after 2 hr if:
- sustained-release formulation taken
- CNS/respiratory depression
- In severe cases, correct hypotension by raising foot of bed or, if necessary, expanding intravascular volume
- Control convulsions with lorazepam IV
- If patient hypothermic, rewarm slowly using conventional means (e.g. Bair Hugger, blankets)
- Treat skin blisters as burns
- monitor for rhabdomyolysis (look for coca-cola coloured urine testing positive for blood, measure creatine kinase)
- Forced diuresis, haemodialysis or haemoperfusion **are of no value**
- Agitation and visual and auditory hallucinations are common during recovery and may require treatment with high doses of diazepam

Management of children with diabetes undergoing surgery and other procedures that require fasting

PRINCIPLES

Reasons to control diabetes well in peri-operative period

- To prevent hyperglycaemia and keto-acidosis, resulting from:
  - omission of insulin
  - stress hormone response to surgery
  - catabolic state
- To minimise risk of infection, enhanced by hyperglycaemia
- To prevent hypoglycaemia, resulting from:
  - starvation pre- and post-operatively
  - anorexia post-operatively

Careful regular monitoring of blood glucose is required throughout peri-operative period

- Diabetic patients should preferably be first on morning list
- Give usual insulin evening before procedure
- Recommend normal age-dependent fasting – see **Pre-op fasting** guideline

If any concerns, contact diabetes team

MINOR SURGERY (able to eat within 4 hr of procedure)

Pre-operative care

First on the morning list

- Advise usual doses of insulin on night before procedure
- On day of procedure omit insulin and breakfast
- Allow clear fluids, including sweet drinks, up to 0600 hr
- Measure and record capillary blood glucose pre-operatively and half-hourly during operation

First on the afternoon list

- Advise usual doses of insulin evening before procedure
- Advise child to have normal breakfast no later than 0700 hr
- Breakfast insulin dose
  - if using multiple daily injection (MDI) regimen, give usual breakfast insulin
  - if using twice daily insulin regimen give one-third of total morning dose as rapid-acting insulin (e.g. Novorapid®)
- Allow clear fluids until 3 hr before operation
- Measure and record capillary blood glucose on arrival in theatre
- Measure and record capillary blood glucose hourly once nil-by-mouth and half-hourly during operation
- If any concerns (e.g. vomiting, prolonged operation), start IV fluids and insulin as for **Major surgery** (see below)

Post-operative care

- Monitor capillary blood glucose in recovery and then hourly for 4 hr
- If well on return to ward and using multiple daily injection (MDI) regimen
  - give dose of rapid-acting insulin appropriate for carbohydrate content of next meal (if advice needed, contact diabetes team) **and**
  - give next dose of long-acting insulin at usual time
- If using twice daily premixed insulin regimen and able to eat by lunch on same day of procedure, give one-third of total morning dose as rapid-acting insulin (e.g. Novorapid®)
  - with meal
  - teatime on same day of procedure, it may be appropriate to give child's usual insulin dose or a reduced dose: contact diabetes team for advice

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- If any concerns (e.g. vomiting or prolonged operation), start IV fluids and insulin as for **Major surgery** (see below)
- when child ready to eat, see **Post-operative care**

MAJOR SURGERY (unable to eat within 4 hr of start of procedure

Pre-operative care

- Admit on day before surgery
- Check pre-meal and bedtime capillary blood glucose measurements on ward

First on the morning list

- If using multiple daily injections (MDI), give usual mealtime short-acting insulin **but half usual dose of long-acting insulin evening before procedure**
- If using twice-daily insulin regimen, give usual doses of insulin with meal evening before procedure
- On day of procedure, omit insulin and breakfast
- Allow clear fluids, including sweet drinks, up to 0600 hr
- Insert 2 IV cannulae if possible. These can be inserted in theatre, if necessary
- Start a glucose and sliding scale insulin infusion in theatre – see **below**
- Measure and record capillary blood glucose pre-operatively and half-hourly during operation

First on the afternoon list

- Advise usual doses of insulin evening before procedure
- Advise child to have a normal breakfast no later than 0700 hr
- Breakfast insulin dose
- if using multiple daily injection (MDI) regimen, give usual breakfast insulin
- if using twice daily insulin regimen, give one-third of total morning dose as rapid-acting insulin (e.g. Novorapid®)
- Allow clear fluids until 3 hr before operation
- Measure and record capillary blood glucose on arrival in theatre
- Insert 2 IV cannulae if possible. These can be inserted in theatre, if necessary
- Start a glucose and sliding scale insulin infusion in theatre – see **below**
- Measure and record capillary blood glucose hourly once nil-by-mouth and half-hourly during operation

EMERGENCY SURGERY

Emergency procedures differ from elective ones as children run the risk of developing ketoacidosis if they are ill. Prolonged starvation associated with delayed surgery poses additional complications

Pre-operative care

- **Inform diabetes team immediately**
- Do not give any SC insulin while child starved
- Check venous U&E, glucose, blood gas when child cannulated
- Commence glucose and sliding scale insulin infusion – see **below**
- Measure and record capillary blood glucose hourly once nil-by-mouth and half-hourly during operation

If patient ill or diabetes not well controlled, follow Diabetic ketoacidosis guideline and postpone operation until patient stabilised.
Operate once patient rehydrated, with stable blood pressure, serum sodium and potassium within normal range and blood glucose <17 mmol/L

INTRAVENOUS INSULIN AND FLUIDS

Maintenance fluid infusion

- Use premixed 500 mL bags of sodium chloride 0.9% and glucose 5% with 20 mmol/L of potassium chloride
- If blood glucose >14 mmol/L give sodium chloride 0.9% with 20 mmol/L potassium chloride
- If blood glucose <5 mmol/L stop insulin, recheck after 10–15 min and if still <5 mmol/L change fluid to glucose 10% with sodium chloride 0.9% and 20 mmol/L of potassium chloride

Insulin infusion

- Add 50 units soluble insulin (e.g. Actrapid®) to 50 mL of sodium chloride 0.9% to make a 1 unit/mL solution
- Administer via syringe pump, do not add directly to fluid bag
- Administer via a Y connector with one-way valve (e.g. Vygon-Protect-a-Line 2 extension)
- Determine infusion rate from hourly capillary blood glucose results, according to sliding scale – see **Table 1**
- Insulin sliding scale should **always to be given in conjunction with a glucose infusion**
- If blood glucose >15 mmol/L, check for ketones, if positive, contact doctor

Do not switch off insulin and/or maintenance fluids in transit to and from theatre
Do not give ANY SC insulin until child ready to come off sliding scale

Monitoring

- Adjust insulin infusion rate according to blood glucose – see **Table 1.** Aim to keep blood glucose between 5 and 10 mmol/L
- Check capillary blood glucose half-hourly during surgery and adjust insulin infusion according to **Table 1**

Table 1

Capillary blood glucose (mmol/L)	Insulin infusion rate (mL/kg/hr)
≥28.1	**Call Doctor**
18.1–28	0.1
12.1–18	0.075
8.1–12	0.05
4.1–8	0.025
≤4	**Stop insulin, treat hypo**. Recheck blood glucose after 30 min and follow sliding scale

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Post-operative care

- Check capillary blood glucose half-hourly for the first 2 hr and then hourly
- Continue glucose and sliding scale insulin infusion until taking adequate oral fluids and snacks. While on insulin sliding scale, child may safely eat and drink
- If taking adequate oral fluids and snacks and using multiple daily injection (MDI) regimen
  - give dose of rapid-acting insulin appropriate for carbohydrate content of next meal (if advice needed, contact diabetes team) **and**
  - give next long-acting insulin dose at usual time. If child was treated using a sliding scale overnight, and usual dose of long-acting insulin was omitted previous night, give half usual dose of long-acting insulin with breakfast if ready to eat
- If using twice daily premixed insulin regimen and taking adequate oral fluids and snacks by:
  - lunch on same day of procedure, give one-third of total morning dose as rapid-acting insulin (e.g. Novorapid®)
  - teatime on same day of procedure or breakfast on next day, it may be appropriate to give child's usual insulin dose or a reduced dose – contact diabetes team for advice

Give non-analogue insulin 30 min before meal. Give analogue insulin 5 min before meal. After 30 min stop infusion

Patients unlikely to resume eating and drinking

- If after 48 hr, patient still unable to eat or drink enough post-operatively:
  - assess for enteral or parenteral feeding – contact nutrition support team
  - contact diabetic nurse specialist for advice on prescribing regular SC insulin

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Thirst
- Weight loss
- Polyuria
- Abdominal pain/vomiting
- Tachypnoea
- Sighing respiration (Kussmaul breathing)
- Odour of ketones
- Dehydration
- Drowsiness
- Coma
- Biochemical signs:
  - ketones in urine or blood
  - elevated blood glucose (>11 mmol/L)
  - acidaemia (pH <7.3)

Assessment

- Airway, breathing, circulation
  - record respiratory rate, heart rate, BP, peripheral pulse volume
- Conscious level: look for signs of cerebral oedema (see **Glasgow coma score**)
- Headache, confusion, irritability, abnormal movements, slow pulse, high BP, papilloedema, small and irregular pupils
- Presence of infection
- Height, weight

Degree of dehydration

- Assessment degree of dehydration as 3%, 5% and 8% (for most children use 5–8% dehydration to calculate fluids)

Investigations

- Insert IV cannula (as large as appropriate for child)

All cases

- Capillary blood glucose
- FBC
- Blood glucose
- Blood gas
- Haemoglobin A_{1c}
- Blood osmolality, sodium, potassium, urea, bicarbonate, creatinine, pH
- Urine ketones on urinalysis
- Blood ketones
- Infection screen: blood and urine culture; if meningism consider lumbar puncture

Moderate and severe cases

- Liver function tests and amylase
- Group and save

Newly diagnosed case

- Thyroid and coeliac disease antibody screen
- Islet cell antibodies
- GAD antibodies
- Thyroid function tests, TSH, Free T4
- Immunoglobulin A

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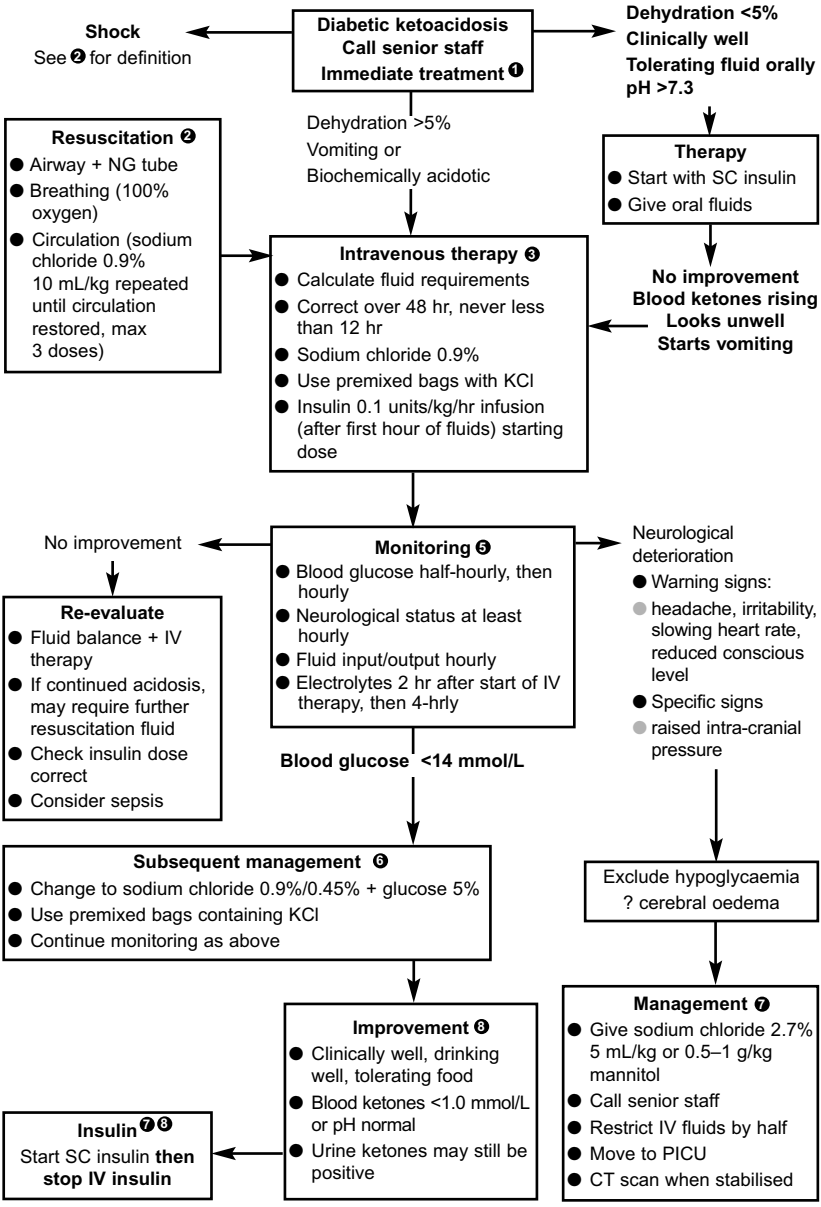
ALGORITHM (cross-referenced to text)

- Remember paediatric type 2 patients can present in DKA



IMMEDIATE TREATMENT ❶

Inform senior staff

Admission

- If alert and not shocked, admit to ward/HDU
- If shock or GCS <8, admit to PICU
- Discuss with PICU if:
  - pH <7.1 and marked hyperventilation
  - aged <2 yr

General

- Nil-by-mouth for first 8–12 hr
  - if vomiting, abdominal pain, no bowel sounds or decreased GCS, insert nasogastric tube
- Place on weigh-bed (if available)
- Strict fluid balance: catheterise children requiring HDU or PICU
- Start flow-sheet to record biochemistry and blood gases
- Monitor ECG for T wave changes
- Initiate IV fluids and insulin (see below)

Shock and resuscitation ❷

- Patient is shocked (very rare in DKA):
  - tachycardia
  - reduced peripheral pulse volume
  - mottled cool peripheries
  - prolonged capillary refill time (poor sign)
  - altered state of consciousness
  - AND acidosis
  - with or without hypotension
- Give sodium chloride 0.9% 10 mL/kg rapidly and reassess; repeated until circulation restored, max 3 doses

If still shocked despite giving 2 boluses of sodium chloride 0.9% 10 mL/kg, discuss with consultant

- When no longer shocked and circulated blood volume has been restored, calculate volume of fluid required (see below)

INTRAVENOUS FLUIDS ❸

Volume of fluid

- Total fluid requirement is the addition of four categories:
  - fluid to re-expand circulating volume if shocked
  - maintenance fluids
  - deficit
  - continuing losses, do not include continuing urinary losses at this stage

Maintenance fluids

- Patient will be nil-by-mouth and will need normal fluid requirement IV

Weight (kg)	Rate (mL/kg/24 hr)	Rate (mL/kg/48 hr)
0–12.9	80	160
13–19.9	65	130
20–34.9	55	110
35–59.9	45	90
>60	35	70

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Fluid deficit

- Estimated amount of fluid patient has lost, (i.e. how dehydrated – see **Assessment**)
- Calculate from weight loss
- Most accurate method
- weigh child and compare with recent weight
- gives good estimate of fluid loss (1 kg weight loss = 1 L fluid deficit)
- Clinical assessment
- deficit in mL = % dehydration x body weight (kg) x 10 (e.g. for a 10 kg child with 5% dehydration, the deficit is 5x10x10 = 500 mL)
- do not use more than 8% dehydration in calculations

Total Amount

- Hourly rate of fluid replacement = (48 hr maintenance requirements + deficit – resuscitation fluid already given)/48
- Weight should rise gradually with rehydration
- If available use weigh-bed to record weight hourly to obtain accurate assessment

Type of fluid

- Initially use sodium chloride 0.9% with potassium chloride and continue this concentration for at least 12 hr
- Use **commercially premixed bag**
- Maximum rate potassium 0.2 mmol/kg/hr (ward)
- If femoral line used give dalteparin 100 units/kg/day (max 5000 units)

Table 1

K^+ <3.5	K^+ 3.5–5.5	K^+ >5.5
500 mL sodium chloride 0.9% with potassium chloride 40 mmol via **central line**	Sodium chloride 0.9% with potassium chloride 40 mmol/L	Sodium chloride 0.9%

If serum potassium <2.5 mmol/L, transfer to PICU. Discuss with consultant whether to give potassium chloride 0.2 mmol/kg in sodium chloride 0.9% by separate infusion over 1 hr. Before infusing bag containing potassium, connect patient to cardiac monitor. If possible, use commercially premixed bag. Only in exceptional circumstances (with consultant agreement and 2 doctors checking procedure) should potassium chloride be added on the ward to a bag of sodium chloride 0.9% (mix well)

- Further fluid and K^+ as dictated by the patient's condition and serum K^+ (**Table 1**), repeated until glucose fallen to 14 mmol/L, then move to **Subsequent management**

YOU MUST obtain consultant authorisation before using bicarbonate infusion (not recommended)

Insulin infusion ❹

- **Start 1 hr after IV fluids**
- Soluble insulin (e.g. Actrapid®) infusion 1 unit/mL in sodium chloride 0.9% via IV syringe pump at 0.1 units/kg/hr (or 0.05 units/kg/hr if local policy)
- If no fall in glucose after 2 hr (very unusual, check pump and patency of IV cannula), increase by 20%. If no fall after 4 hr, consult senior medical staff and re-evaluate (e.g. sepsis, insulin errors)
- If blood glucose falls exceeds 5 mmol/L/hr, reduce insulin infusion rate by 20%
- Do **not** stop insulin infusion. Check capillary glucose in 1 hr
- If IV fluids and insulin given through same cannula use anti-reflux valve

Do not give insulin bolus. Do not add insulin directly to fluid bags

MONITORING TREATMENT ❺

- Hourly capillary blood gas and glucose
- Check U&E, glucose, osmolality pH and capillary ketones 2-hrly until improving, then 4-hrly
- Neurological status, heart rate and blood pressure hourly
- Complete DKA summary sheets

SUBSEQUENT MANAGEMENT ❻

After a minimum 12 hr of initial intravenous therapy, if plasma sodium level stable or increasing, change sodium concentration to sodium chloride 0.45%
When blood glucose falls below 14 mmol/L add glucose to fluid

- Maintenance fluid dependent on sodium, glucose and potassium:

Table 2: Sodium

Blood sodium	**Fluid:** with glucose (see **Table 3**) and potassium chloride (see **Table 4**)
First 12 hr and if falling after 12 hr	Sodium chloride 0.9%
After 12 hr if stable or increasing	Sodium chloride 0.45%

Table 3: Glucose

Blood glucose	**Fluid:** sodium chloride (see **Table 2**) with potassium chloride (see **Table 4**) and
0–8.0	Glucose 10%
8.1–14.0	Glucose 5%
>14	No glucose

Table 4: Potassium

Blood potassium	**Fluid:** sodium chloride (see **Table 2**) and glucose (see **Table 3**) and
K^+ <3.5	**Discuss with consultant**
K^+ 3.5–5.5	Potassium chloride 20 mmol in 500 mL
K^+ >5.5	No potassium

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- If pH >7.3 reduce insulin infusion rate to 0.05 units/kg/hr
- Blood glucose may rise as a result, **but do not revert to sodium chloride 0.9% unless plasma pH falls**
- if pH falls, reassess fluid deficit and regimen
- If glucose falls below 4 mmol/L, give 2 mL/kg glucose 10% IV. Reduce insulin infusion rate by 20%. Check capillary glucose in 1 hr
- **To make glucose 10% with sodium chloride 0.45% (with or without potassium): remove 50 mL from 500 mL bag of glucose 5%, sodium chloride 0.45% (with or without potassium) and add 50 mL of glucose 50%**
- Continue with IV fluids and insulin infusion until urine is negative for ketones and child tolerating oral fluids and food
- Continue IV insulin pump after first SC dose of insulin for 1 hr if SC dose was soluble (e.g. Humulin S®) or 10 min if SC dose of insulin was aspart or lispro (e.g. Novorapid®)

If acidosis not improving, consider:

- Insufficient insulin to switch off ketones
- Inadequate resuscitation
- Sepsis
- Hyperchloraemic acidosis
- Salicylate or other prescription or recreational drugs

Cerebral oedema ❼

- Observe for headache, any change in symptoms, pH <7.2, or persistently low serum sodium as glucose corrects
- Exclude hypoglycaemia
- If cerebral oedema suspected, inform consultant immediately
- Give 5 mL/kg of sodium chloride 2.7% over 5–10 min
- if not available give mannitol 0.5 g/kg (2.5 mL/kg of 20%) over 20 min, repeat mannitol once or twice after 2 hr if required
- restrict IV fluid intake to half maintenance and replace deficit over 72 hr
- if patient unconscious, insert urethral catheter
- admit to PICU
- consider CT scan/MR scan

Converting to SC insulin ❽

- Inform diabetes team (consultant, diabetic nurse and dietitian)
- Children usually require insulin 0.25–1.0 units/kg/day (pre-pubertal usually 0.6–0.8 units/kg/day; higher in puberty)
- **If converting to multiple daily dose regimen:**
- give 40% as long-acting insulin at night
- 20% short-acting insulin with each of the 3 main meals
- Adjust ratio if necessary, depending on child's eating patterns
- **If converting to biphasic insulin regimen divide total daily dose as follows:**
- $^2/_3$ dose 20 min before breakfast (give insulin analogues 5 min before meal)
- $^1/_3$ dose 20 min before evening meal (give insulin analogues 5 min before meal)
- Choose insulin preparation most suitable for child: discuss with diabetes team
- Continue IV insulin pump after first SC dose of insulin for 60 min if SC dose was soluble (e.g. Actrapid®) or 10 min if SC dose of insulin was aspart or lispro (e.g. Novorapid®/Humalog®)

DISCHARGE AND FOLLOW-UP

- Prescribe following as TTO for all new patients:
  - brand and strength of regular insulin, specify if pre-filled pen or cartridges
  - brand of soluble insulin, specify if pre-filled pen or cartridges
  - needles 5 mm
  - 1 pack hypostop triple pack
  - 1 packet glucose tablets
  - 1 box lancets (e.g. Microfine plus)
  - GlucaGen HypoKit (glucagon) 1 kit 500 microgram <25 kg, 1 mg ≥25 kg (if local policy)
  - 1 box blood glucose strips appropriate to blood glucose monitor
  - 1 box Ketostix (ketones in urine)
  - 1 box blood ketone testing strips (particular to local policy)
- Organise out-patient follow-up

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Any child or young person presenting to GP or A&E with symptoms suggestive of diabetes should be referred (by phone) immediately to paediatric diabetes team

RECOGNITION AND ASSESSMENT

Definition

Elevated blood glucose with no ketonuria/blood ketones

- Symptoms + random plasma glucose ≥11 mmol/L
- Or symptoms + fasting plasma glucose ≥7 mmol/L
- No symptoms but random plasma glucose ≥11 mmol/L
- No symptoms but fasting plasma glucose ≥7 mmol/L on 2 tests on 2 separate days

Symptoms and signs

- Change in school performance
- Thirst
- Weight loss
- Thrush
- Polyuria
- Nocturia
- May be absent
- if obese, no ketonuria or evidence of insulin resistance (e.g. acanthosis nigricans), consider type 2 diabetes

Investigations

- Height and weight
- Blood:
- glucose
- electrolytes
- pH
- ketones
- haemoglobin A_{1c}
- FBC
- cholesterol and triglycerides
- TSH and FT4
- immunoglobins A, G and M
- autoantibody screen for thyroid, coeliac GAD and islet cell antibodies
- Urine
- ketones
- glucose
- C-peptide if considering type 2 diabetes

Do not arrange a fasting blood glucose or glucose tolerance test

IMMEDIATE TREATMENT

- Admit under admitting consultant of day/week
- Inform diabetes team, consultant or diabetes nurse specialist
- Start on SC insulin, total daily dose of 0.4 units/kg
- **If starting on multiple daily dose regimen:**
- give 40% as long-acting insulin at night
- 20% short-acting insulin with each of the 3 main meals
- Adjust ratio if necessary, depending on child's eating patterns
- **If starting on biphasic insulin regimen divide total daily dose as follows:**
- $^2/_3$ of total dose 10–20 min before breakfast (give insulin analogues 5 min before meal)
- $^1/_3$ of total dose 10–20 min before evening meal (give insulin analogues 5 min before meal)
- For advice on which insulin to use, discuss with consultant with special interest in diabetes

SUBSEQUENT MANAGEMENT

- If tolerating food, allow patient to eat according to appetite for first 24–48 hr
- Adjust insulin according to child's eating habits
- Refer to dietitians

MONITORING TREATMENT

- Glucose stick monitoring pre-meals and at 0000 and 0400 hr

DISCHARGE AND FOLLOW-UP

- Out-patient appointment to see consultant 1–2 weeks after discharge
- Prescribe as TTO:
  - brand and strength of regular insulin, specify if pre-filled pen or cartridges
  - brand of soluble insulin, specify if pre-filled pen or cartridges
  - needles 5 mm
  - 1 pack glucogel triple pack
  - 1 packet glucose tablets
  - 1 box lancets (e.g. Microfine plus)
  - GlucaGen hypoKit (glucagon) 1 kit 500 microgram <25 kg, 1 mg >25 kg (dependent on local policy)
  - 1 box blood glucose sticks appropriate to blood glucose monitor
  - 1 box ketostix (ketones in urine)
  - if appropriate, 1 box blood ketone sticks (particular to local policy)

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Issued: May 2013
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Unexplained and prolonged

RECOGNITION AND ASSESSMENT

Definition

- For the purposes of this guideline, hypoglycaemia defined as a blood glucose <2.6 mmol/L in child aged >1 month-old

Symptoms and signs

- Neuroglycopenia:
  - lethargy
  - lassitude
  - tremulousness
  - loss of consciousness
  - seizure
- Autonomic effects:
  - sweating
  - shaking
  - trembling
  - tachycardia
  - anxiety
  - hunger

Previous history

- Ask about:
  - antenatal history (e.g. small-for-dates)
  - prematurity
  - history of hypoglycaemia on the neonatal unit
  - early or prolonged jaundice
  - family history of sudden death (MCAD, LCAD)
  - history of neuroglycopenia/autonomic symptoms when glucose intake decreased, (e.g. during minor illnesses)
  - development, especially developmental regression
  - medication
  - access to glycopenic agents (e.g. metformin)
  - oral hypoglycaemics
  - nutritional intake

Investigations

Certain pointers to cause of unexplained hypoglycaemia are detectable only during episode. Take blood samples BEFORE correcting blood glucose

Immediate samples

- Before treating hypoglycaemia, take venous blood for assay using correct blood bottles (**Table 1**)
  - once samples have been obtained, correct hypoglycaemia. See **Immediate treatment**
  - inform laboratory immediately so samples arrive as quickly as possible (within 20 min)
- **Ensure first voided urine specimen after hypoglycaemia episode is obtained to test for ketone bodies, organic/amino acid metabolites and reducing substances. Check with laboratory**

Table 1: Total blood requirement (5 mL minimum)

Fluoride	1.3 mL
Lithium heparin	1.3 mL
Clotted	2.6 mL

Investigations

- In all prolonged unexplained hypoglycaemia:
  - glucose sticks
  - capillary blood gas
  - true glucose
  - lactate
  - ACTH
  - growth hormone
  - insulin
  - C-peptide
  - cortisol
  - urea and electrolytes
  - urinary ketones
  - 17 O HP in infant if hyponatraemia present

- Store blood and urine for these investigations depending on above results:
  - IGF1
  - beta-hydroxybutyrate
  - free fatty acids
  - carnitine
  - urinary-reducing substances
  - organic and amino acids

Physcial examination

- Height and weight
- Midline defects, micropenis, optic nerve hypoplasia (pituitary disorder)
- Dysmorphic features: macroglossia, macrosomia, ear lobe crease (Beckwith-Wiedemann)
- Skin hyperpigmentation (adrenal insufficiency)
- Hepatomegaly (glycogen storage disorder)

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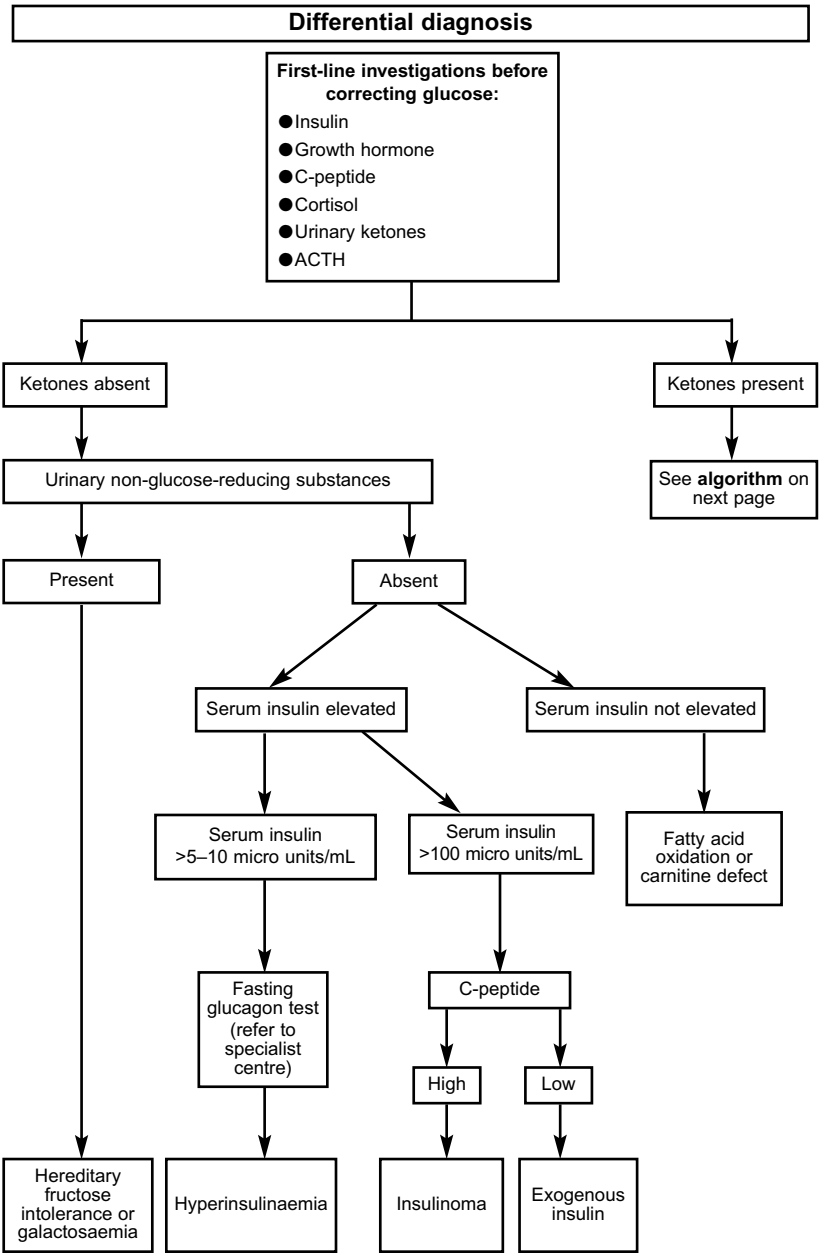
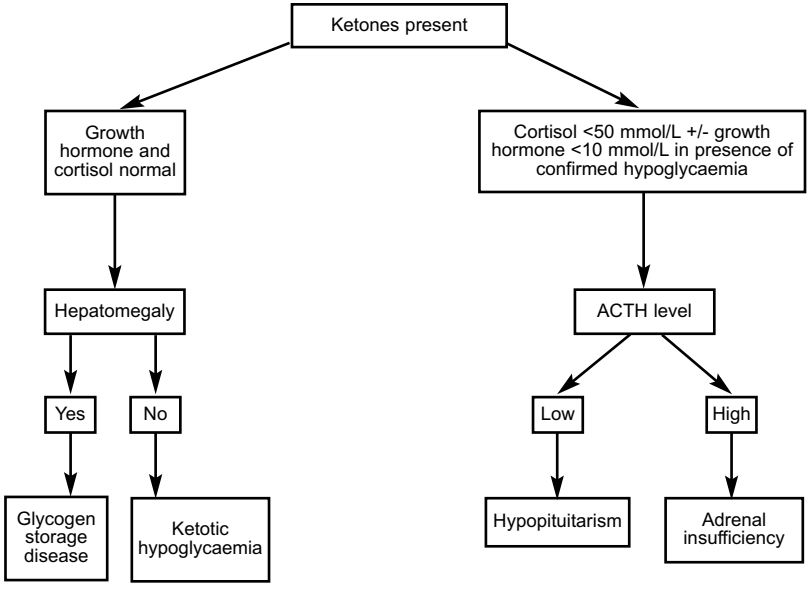

Differential diagnosis
First-line investigations before correcting glucose:
Insulin
Growth hormone
C-peptide
Cortisol
Urinary ketones
ACTH
Ketones absent
Ketones present
Urinary non-glucose-reducing substances
See algorithm on next page
Present
Absent
Serum insulin elevated
Serum insulin not elevated
Serum insulin >5–10 micro units/mL
Serum insulin >100 micro units/mL
Fatty acid oxidation or carnitine defect
Fasting glucagon test (refer to specialist centre)
C-peptide
High
Low
Hereditary fructose intolerance or galactosaemia
Hyperinsulinaemia
Insulinoma
Exogenous insulin

Algorithm: Ketones present



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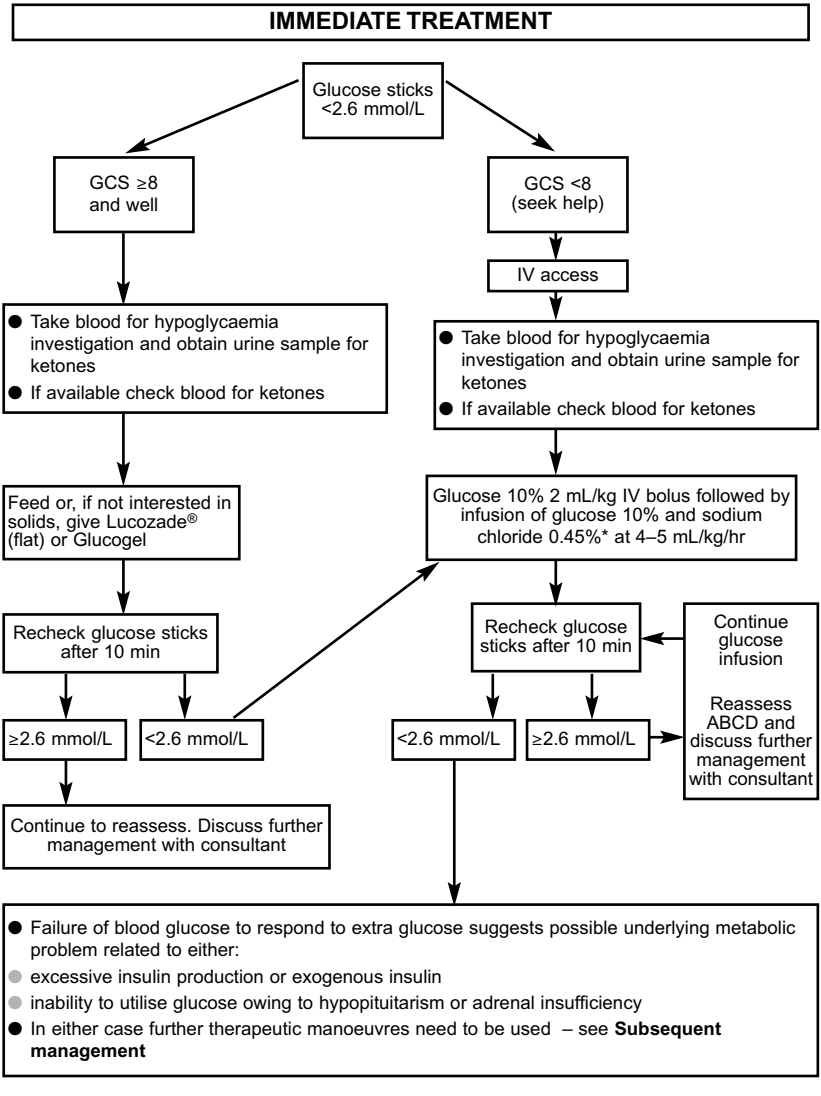
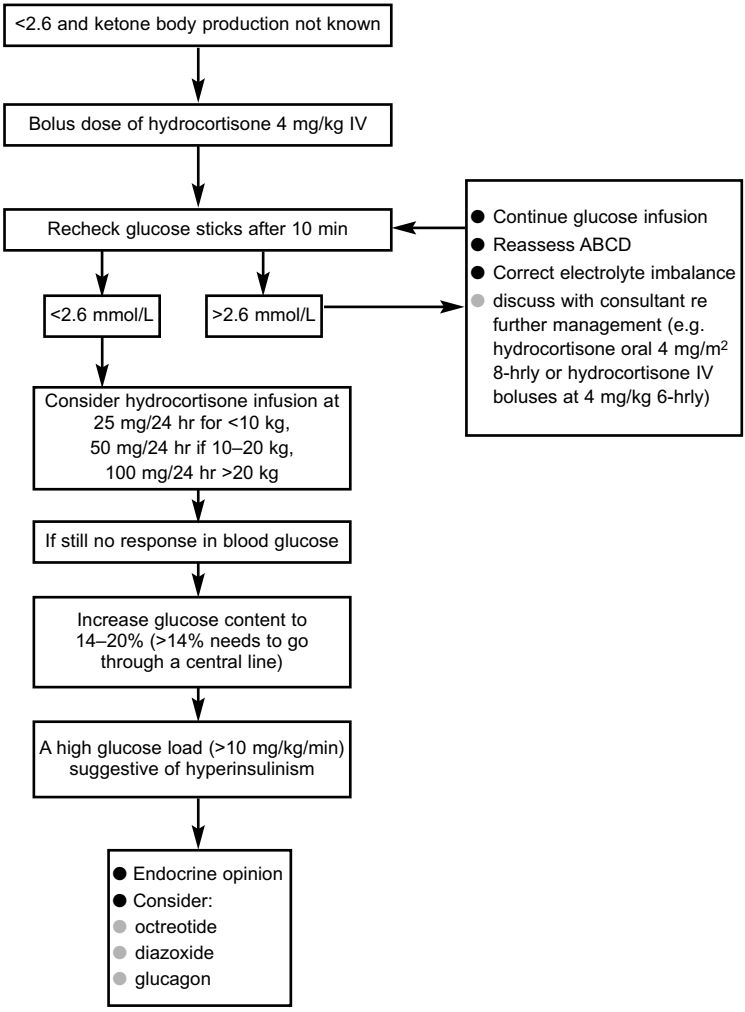


* Remove 50 mL from 500 mL sodium chloride 0.45% and glucose 5%, add 50 mL glucose 50%

SUBSEQUENT MANAGEMENT



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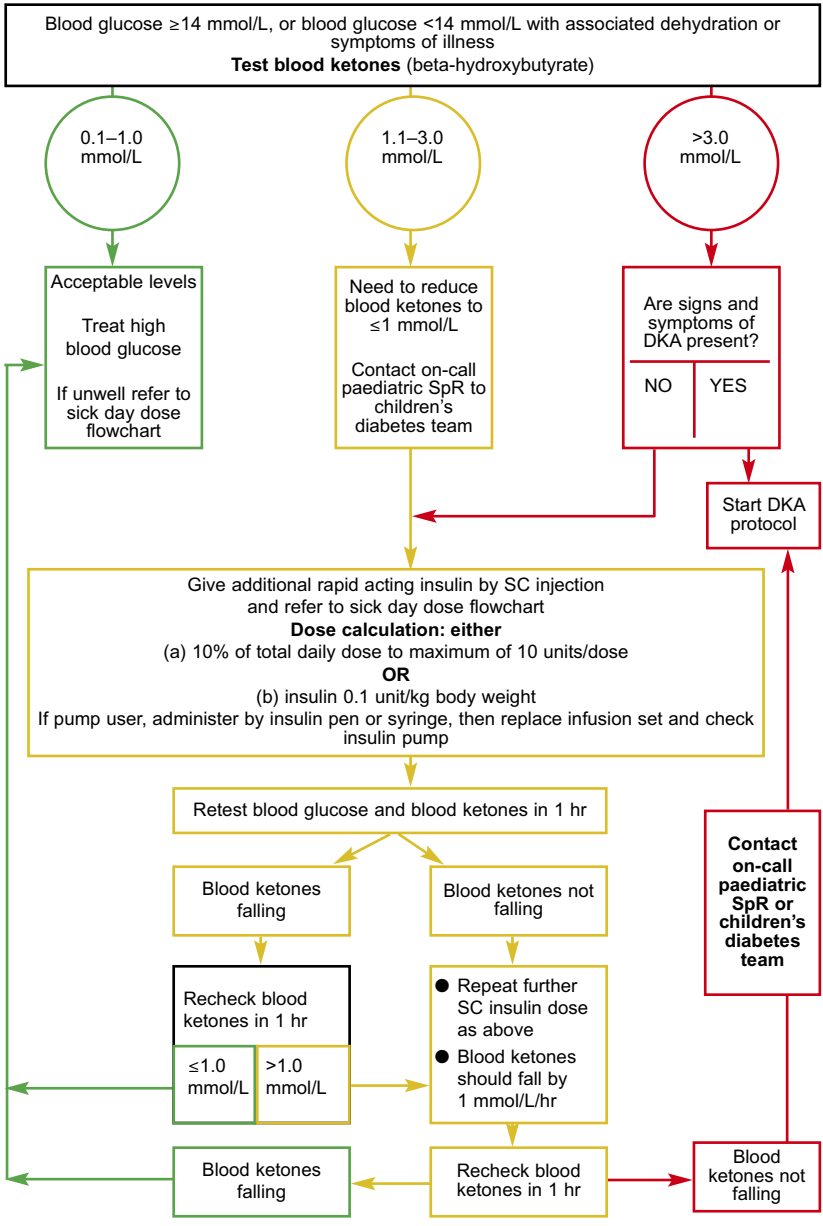
KETONE MONITORING • 1/1

Applicable for all subcutaneous insulin regimes and insulin pump therapy



If unsure, especially if child aged <5 yr, contact children's diabetes team

STEROID DEPENDENCE • 1/2

Pituitary-adrenal axis impairment

RECOGNITION AND ASSESSMENT

Definition

- Children with the following conditions are corticosteroid-dependent with a depressed or absent pituitary-adrenal axis:
  - hypopituitarism
  - adrenal insufficiency
  - congenital adrenal hyperplasia
  - growth hormone insufficiency
  - prolonged corticosteroid use for immunosuppression
  - severe asthma requiring oral corticosteroids or high-dose inhaled corticosteroids

When shocked or stressed corticosteroid-dependent children cannot mount an appropriate adrenal response

- Corticosteroid-dependent children are encountered in a number of ways:
  - at presentation and first diagnosis
  - for elective surgical and investigative procedures
  - for emergency surgery or when acutely unwell
  - with hyponatraemia, hyperkalaemia +/- hypoglycaemia and hypotension

MANAGEMENT

Elective surgical and investigative procedures

- Check whether pre-operative discussion of endocrine management has taken place
  - if no plan for corticosteroid manipulation, prescribe hydrocortisone 2–4 mg/kg IV at induction then 6-hrly until child capable of taking oral medication, then give double usual daily maintenance dosage of hydrocortisone for subsequent 48 hr
- Continue usual medication with:
  - fludrocortisone
  - growth hormone
  - levothyroxine
  - desmopressin

Acute illness

- During illness, corticosteroid-dependent children can usually be managed at home
- Moderate illness with temperature ≤38ºC give double hydrocortisone dose, if temperature >38ºC give treble hydrocortisone dose
  - if unable to take oral corticosteroids (e.g. vomiting or acute collapse), parents to administer IM hydrocortisone 2 mg/kg or aged <1 yr 25 mg, 1–6 yr 50 mg, 100 mg thereafter
- If IM hydrocortisone required, hospital assessment necessary with training of parents to administer IM
- Continue usual dose of other medication
  - failure to do so may lead to hypoglycaemia
- Some units do not prescribe IM hydrocortisone. Check locally
- Patients must carry a steroid card

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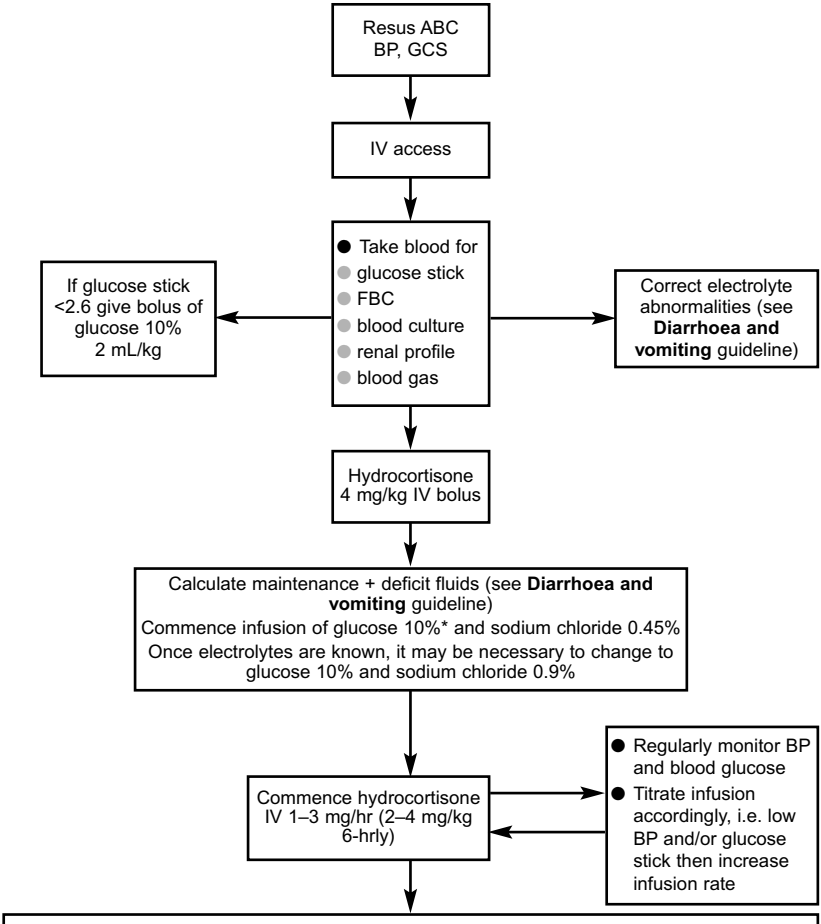
Algorithm for the management of unwell corticosteroid-dependent children



* Glucose 10% can be made by adding 50 mL glucose 50% to a 500 mL bag of glucose 5% with sodium chloride 0.9% or 0.45%

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Pain may be localised or generalised
- Vomiting
- Anorexia
- Fever
- Crying and irritability

Typical features of some important causes of acute abdominal pain in children

Appendicitis

- History of localised pain with increased severity on RIF
- On examination:
  - fever
  - mid-abdominal pain migrating to RIF
  - guarding and rebound tenderness
  - pain on percussion
  - young children may not have typical features

Intussusception

- Typical age of presentation: 2 months–2 yr
- History of intermittent colicky abdominal pain 2–3 times/hr initially with increasing frequency
- Looks pale with pain
- Lethargic between episodes of pain
- Vomiting prominent feature
- Diarrhoea common
- Passage of blood and/or mucus *per rectum* (redcurrant jelly stools) late sign
- Follows respiratory or diarrhoeal illness
- Clinical features of intestinal obstruction
- On examination:
  - a sausage-shaped mass crossing midline in the epigastrium or behind umbilicus
  - may be associated with Henoch-Schönlein purpura
  - abdominal distension and hypovolaemic shock late signs

Mid-gut volvulus

- Presents mainly in neonatal period
- History of:
  - bowel obstruction
  - abdominal pain
  - distension
  - bilious vomiting
- On examination
  - abdominal distension, tenderness

Pneumonia and empyema

- History of fever and cough
- On examination:
  - tachypnoea
  - recession +/- focal signs at one base
  - decreased breath sounds and dullness to percussion

Differential diagnosis

Surgical problems

- Acute appendicitis
- Intussusception
- Intestinal obstruction
- Torsion of ovary or testis
- Meckel's diverticulitis
- Hydronephrosis
- Renal or biliary calculus
- Enterocolitis secondary to Hirschprung's disease

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Medical problems – relatively common

- Mesenteric adenitis
- Constipation
- Gastroenteritis
- Inflammatory bowel disease
- Lower lobe pneumonia
- Acute pyelonephritis
- Henoch-Schönlein purpura
- Hepatitis
- Acute cholecystitis
- Gastritis/peptic ulcer

Medical problems – rare but important

- Lead poisoning
- Diabetes
- Sickle cell crisis
- Acute porphyria
- Pancreatitis
- Primary peritonitis
- Non-accidental injury

Gynaecological problems

- Ectopic pregnancy
- Torsion of ovarian cyst
- Miscarriage
- Pelvic inflammatory disease (PID)
- Mittelschmerz pain

INVESTIGATIONS

- Urine testing and analysis
- FBC, ESR
- Blood and stool culture
- CRP, U&E, amylase, glucose, LFT
- Consider group and save
- Consider pregnancy test in adolescent females (inform patient)

Imaging

- Only if bowel obstruction or perforation suspected: **abdominal X-ray**
- If child stable and suspect appendicitis, intussusception, torsion of ovary or testis, renal problems, pancreatitis or cholecystitis: **ultrasound scan of abdomen**
- If respiratory symptoms: **chest X-ray**
- Do not delay surgical review awaiting scans if acute surgical problem suspected (e.g. torsion of testis, intussusception)

MANAGEMENT

- If present, treat hypotension and shock
- Stop feeding if you suspect surgical problem
- IV access if surgical cause likely
- Nasogastric tube free drainage if bowel obstruction
- IV antibiotics if perforation (e.g. cefuroxime and metronidazole)

Indications for surgical review

- Localised right iliac fossa pain
- Rebound tenderness/pain on percussion
- Migration of pain
- Redcurrant jelly stools and bleeding *per rectum*
- Bile-stained vomiting
- Marked abdominal distension
- Inguino-scrotal pain or swelling
- Increasing abdominal pain with progressive signs of deterioration

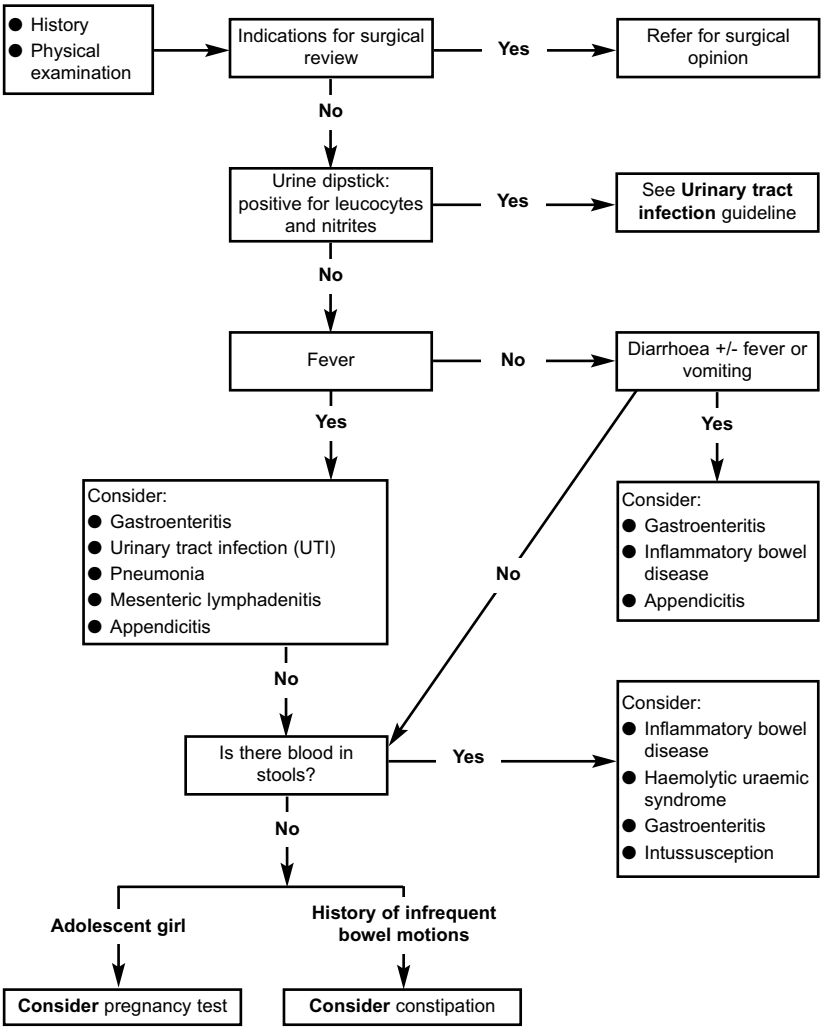
Observation

- If stable, period of observation may be useful to make diagnosis

Analgesia

- Do not withhold analgesia pending surgical review: opioids may be necessary (see **Analgesia** guideline)

Management of acute abdominal pain



DISCHARGE AND FOLLOW-UP

- Discharge usually within 24 hr of symptoms settling (e.g. fever, abdominal pain)
- Follow-up usually appropriate in primary care/GP

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RECOGNITION AND ASSESSMENT

Definition

- **Constipation:** infrequent bowel evacuation of hard faeces or difficult/painful defecation for ≥1 month
- **Faecal soiling** (overflow as a result of faecal impaction): passage of loose and offensive stools in child's underwear over which child has no control
- **Encopresis** (functional non-retentive soiling): inappropriate passage of normal stools in inappropriate places. Often associated with behavioural problems
- **Faecal incontinence:** soiling in the presence of an anatomical or organic lesion
- **Faecal impaction:** hard faecal mass in lower abdomen, a dilated rectum impacted with stool or excessive stool in the colon identified radiologically

KEY POINTS IN HISTORY

- Frequency, volume and type of stool using Bristol stool chart
- Overflow soiling in older children
- Distress and/or straining on opening bowels
- Holding behaviour (crossing legs, back arching or tiptoeing)
- Time of passing meconium after birth
- Bleeding *per rectum*
- Any trigger factors i.e. diet change, infection, potty training or starting nursery/school

KEY POINTS IN PHYSICAL EXAMINATION

- Weight and height
- Abdominal examination to look for abdominal distension, faecal loading
- Lower limb neuromuscular examination in long standing cases
- Spinal examination
- Inspection of perianal area for appearance, position of anus or evidence of streptococcal infections

Symptoms and signs suggestive of organic constipation ('red flags')

- Early onset of constipation (first few weeks of life)
- Failure to thrive/growth failure
- Neuropathic bowel:
  - lack of lumbosacral curve
  - pilonidal dimple or tuft of hair
  - sacral agenesis
  - flat buttocks
  - patulous anus
  - absent cremasteric reflex/absent anal wink
  - decreased lower extremity tone and/or strength
  - absence or delay in relaxation phase of lower extremity deep tendon reflex
  - urinary symptoms
- Hirschsprung's disease
  - delayed passage of meconium for more than 24 hr after birth in a term baby
  - abdominal distension
  - tight empty rectum in presence of palpable faecal mass
  - gush of liquid stool and air from rectum on withdrawal of finger
  - rarely causes soiling

- Anteriorly displaced anus
- Anal stenosis:
- tightness or stricture felt when *per rectum* digital examination done using lubricated fifth finger in newborn and infants up to 6 months
- Dairy protein intolerance in first 3 yr of life

DIFFERENTIAL DIAGNOSIS

- Idiopathic functional constipation (90–95%). Most common cause of constipation beyond neonatal period

Organic constipation (suspected in presence of red flags)

- Constipation secondary to anal anatomic malformation (ano-rectal examination required)
- Neurogenic constipation due to spinal cord anomalies or trauma, neurofibromatosis and tethered cord (lower limb neurological examination required)
- Constipation secondary to endocrine/metabolic disorders (hypothyroidism, hypercalcaemia, hypokalemia, CF)
- Constipation induced by drugs (opioids)
- Coeliac disease

INVESTIGATIONS

- Most children with chronic constipation require minimal investigation:
- careful history and physical examination will help determine appropriate investigation
- Consider testing for coeliac disease and thyroid function in intractable cases

Abdominal X-ray

- Has little or no value in the diagnosis of idiopathic constipation
- lower spine X-ray may be useful in an encopretic child with no faecal masses on abdominal and rectal examination

When to consider referral for rectal biopsy

- History of delayed passage of meconium
- Constipation since neonatal period
- History of abdominal distension and vomiting
- Failure to thrive or faltering growth
- Family history of Hirschprung's

MANAGEMENT OF FUNCTIONAL CONSTIPATION

- See **Constipation management flowchart**
- Refer children with organic cause to gastroenterologist

Principles of treatment

- Education
- Diet and lifestyle
- Behavioural management
- Medication
- Supporting child and family

Education

- Give parents clear explanation of pathophysiology of constipation and soiling

Diet and lifestyle

- Use in combination with laxatives
- Ensure adequate fluid intake
- High fibre diet is recommended
- Encourage physical activities

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Behavioural management

- Use of behavioural management in combination with medications decreases time to remission
  - regular toileting: unhurried time on the toilet after meals
  - correct toilet position
  - maintain diaries of stool frequency combined with reward system
  - regular review and positive reinforcement
  - discourage negative responses to soiling from family
  - encourage older children to take responsibility
- May need counselling or a psychology referral in case of motivational or behavioural problems

Medication

- Disimpaction in the presence of impacted stools

DISIMPACTION

Aged <1 yr, refer to paediatric gastroenterologist

1. A macrogol laxative [polyethylene glycol (e.g. Movicol paediatric plain)]; faecal impaction dose, see below up to a maximum of 7 days
2. Use stimulant laxative, senna or sodium picosulphate (Picolax) if no result with macrogol or if not tolerated
3. Review all children within/after one week of disimpaction (in hospital or by GP)

Disimpaction dosage

Age (yr)	Day 1	Day 2	Day 3	Day 4	Day 5	Day 6	Day 7
	Number of Movicol paediatric plain sachets daily divided into 2–3 doses						
1–5	2	4	4	6	6	8	8
5–12	4	6	8	10	12	12	12
	Number of adult Movicol preparation for children aged >12 yr						
>12–18	4	6	8	8	8	8	8

Rectal disimpaction (only if oral disimpaction fails)

- Sodium citrate micro-enemas (Relaxit)
- Small volume sodium citrate enemas (Microlax) is preferable to large volume phosphate enemas
- Phosphate enemas (only if oral medications and Microlax enemas failed). Use only under specialist supervision. Consider sedation if child is distressed

Manual evacuation

- If all above have failed, consider manual evacuation under general anaesthetic. Consult with paediatric gastroenterologist or paediatric surgeon

MAINTENANCE THERAPY

- After disimpaction, or if child had no impaction, focus treatment on prevention of recurrence and establishment of a regular bowel habit to allow bowel to regain normal tone and sensation

- Continue maintenance therapy for 4–6 months then reduce dosage gradually
  - half the dismpaction dose of Movicol is a useful guide for initial maintenance dose

Laxatives

- Use macrogols as first line maintenance treatment ($^1/_2$–1 sachet daily in children aged <1 yr)
- If not improved within a month or to prevent recurrence of impaction, add a stimulant laxative such as senna, bisacodyl or sodium picosulphate syrup. Using stimulants is recommended only for short periods of time and intermittently. Use with faecal softener e.g. sodium docusate and/or fibre
- Aim for soft/loose stools initially daily
- High doses (up to 4–6 sachets daily of macrogols) may be required and doses may need frequent adjustment by child and parent to maintain a regular bowel action. Advise parents to reduce doses gradually and to increase again if no bowel action in 3 days
- If macrogols not tolerated, use sodium docusate or lactulose
- Aged <6 months:
  - give infant glycerol suppository once/day
  - change milk to hydrolysed formula if dairy intolerance

Supporting child and family

- Organise review within a week then regular and frequent local contact and by telephone to prevent re-impaction
- Provide a contact telephone number for parents if available
  - discuss timing of doses for convenience with bowel action
  - emphasise need for good compliance
- Use outreach nursing support if available
- Liaise with the child's health visitor, community paediatric nurse and/or school nurse. Send copies of consultations with parental agreement to help provide a unified approach
- Child psychology support when available is invaluable

Withdrawal of laxatives

- Once regular bowel habit has been established for a few months, and child has good sensation to pass stools, gradually withdraw laxatives over a period of months

INDICATIONS FOR SEEKING ADVICE OF PAEDIATRIC GASTROENTEROLOGIST

- Organic cause of constipation suspected
- Disimpaction orally/rectally unsuccessful
- Soiling/abdominal pain continues despite treatment
- Children aged <1 yr with faecal impaction or not responding to maintenance therapy

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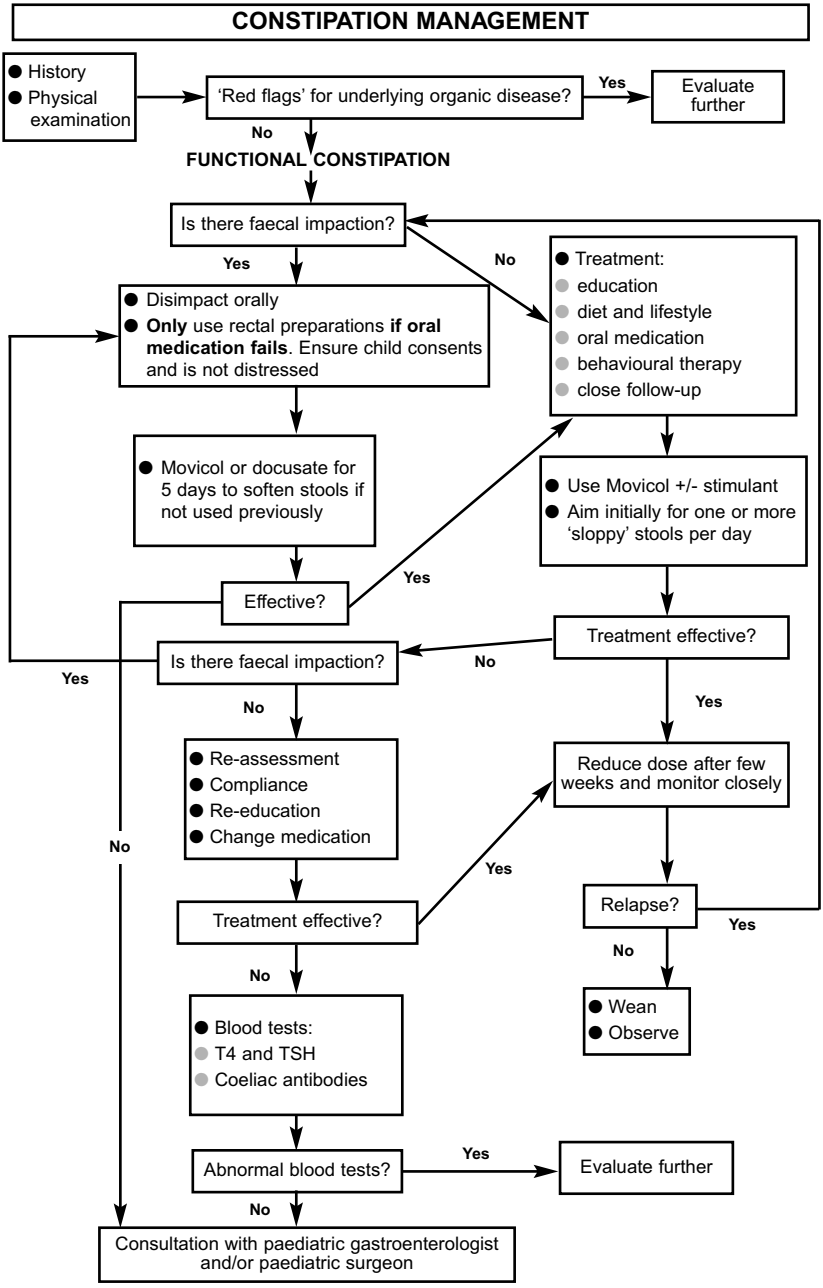
CONSTIPATION MANAGEMENT
History
Physical examination
'Red flags' for underlying organic disease?
Yes
Evaluate further
No
FUNCTIONAL CONSTIPATION
Is there faecal impaction?
Yes
No
Disimpact orally
Only use rectal preparations if oral medication fails. Ensure child consents and is not distressed
Treatment:
education
diet and lifestyle
oral medication
behavioural therapy
close follow-up
Movicol or docusate for 5 days to soften stools if not used previously
Use Movicol +/- stimulant
Aim initially for one or more 'sloppy' stools per day
Effective?
Yes
Treatment effective?
Is there faecal impaction?
Yes
No
No
Yes
Re-assessment
Compliance
Re-education
Change medication
Reduce dose after few weeks and monitor closely
No
Yes
Treatment effective?
Relapse?
Yes
No
No
Wean
Observe
Blood tests:
T4 and TSH
Coeliac antibodies
Abnormal blood tests?
Yes
Evaluate further
No
Consultation with paediatric gastroenterologist and/or paediatric surgeon

RECOGNITION AND ASSESSMENT

Definition of diarrhoea

- Passage of loose watery stools at least three-times in 24 hr
- Most common cause is acute infective gastroenteritis

Diarrhoea and vomiting in infants may be a sign of sepsis

Symptoms and signs

- Sudden onset of diarrhoea (D) or vomiting (V), or both (D&V)
- Fever, malaise, lethargy
- Abdominal cramps
- Loss of appetite

Patient history

- Ask about:
  - duration of illness
  - frequency of stools and associated vomiting (>6 stools more likely to become dehydrated)
  - colour of vomit (if green bilious vomit, consider obstruction)
  - nature of stools, including presence of blood in stool
  - feeds (fluid and food intake)
  - urine output (number of wet nappies)
  - contacts/exposure to infection
  - recent travel abroad
  - recent antibiotic use
  - symptoms of other causes of D&V (e.g. high pyrexia, shortness of breath, severe/localised abdominal pain or tenderness, symptoms of meningitis/septicaemia)
  - weight loss
  - underlying problems e.g. low birth-weight, malnutrition, neuro-disability

Inform public health if outbreak of gastroenteritis suspected

Assessment

- Weight, including any previous recent weight
- Temperature, pulse, respiratory rate
- Degree of dehydration (see **Table 1**) and/or calculate from weight deficit
- Complete systemic examination to rule out other causes of D&V
- Children aged <1 yr are at increased risk of dehydration

Calculating fluid deficit

- Deficit in mL = % dehydration x weight (kg) x 10
- e.g. for a 10 kg child with 5% dehydration deficit is 5 x 10 x 10 = 500 mL

Calculating maintenance fluids

Weight (kg)	Fluid volume
<10	100 mL/kg/day
10–20	1000 mL + 50 mL/kg/day for each kg >10 kg
>20	1500 mL + 20 mL/kg/day for each kg >20 kg

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Table 1: Assessment of degree of dehydration

Increasing severity of dehydration			
	No clinically detectable dehydration (<5%)	**Clinical dehydration 5–10% dehydrated**	**Clinical shock >10% dehydration**
Symptoms (remote and face-to-face assessment)	Appears well	Appears to be unwell or deteriorating	–
	Alert and responsive	Altered responsiveness (e.g. irritable, lethargic)	Decreased level of consciousness
	Normal urine output	Decreased urine output	–
	Skin colour unchanged	Skin colour unchanged	Pale or mottled skin
	Warm extremities	Warm extremities	Cold extremities
Signs (face-to-face assessment)	Alert and responsive	Altered responsiveness (e.g. irritable, lethargic)	Decreased level of consciousness
	Skin colour unchanged	Skin colour unchanged	Pale or mottled skin
	Warm extremities	Warm extremities	Cold extremities
	Eyes not sunken	Sunken eyes	–
	Moist mucous membranes (except for 'mouth breather')	Dry mucous membranes (except after a drink)	–
	Normal heart rate	Tachycardia	Tachycardia
	Normal breathing pattern	Tachypnoea	Tachypnoea
	Normal peripheral pulses	Normal peripheral pulses	Weak peripheral pulses
	Normal capillary refill time	Normal capillary refill time	Prolonged capillary refill time
	Normal skin turgor	Reduced skin turgor	–
	Normal blood pressure	Normal blood pressure	Hypotension (decompensated shock)

Investigations

- If vomiting a major feature or vomiting alone, or if baby aged <3 months: urine for MC&S
- If septicaemia suspected, child immunocompromised, or if stools bloody, mucous or chronic diarrhoea present, send stools for MC&S and virology
- If recent antibiotics, send stool for *Clostridium difficile* toxin
- If severe dehydration, possible hypernatraemic dehydration (see **Hypernatraemic dehydration** below) or diagnosis in doubt:
  - FBC, U&E, chloride, glucose, blood and urine cultures. Blood gas or venous bicarbonate
  - if decreased level of consciousness consider lumbar puncture, especially in babies

IMMEDIATE TREATMENT

See **Flowchart – Management of acute gastroenteritis in young children (aged <4 yr)**

General advice to parents

- Adequate hydration important
- Encourage use of oral rehydration solution (ORS)
  - 'clear fluids' (water alone/homemade solutions of sugar and fruit) lack adequate sodium content and are inappropriate
  - sugar, fruit juices and cola have a high osmolar load and little sodium, and can worsen diarrhoea
- Recommend early re-feeding with resumption of normal diet (without restriction of lactose intake) after 4 hr rehydration
- Do not use anti-diarrhoeal agents
- Anti-emetics (e.g. ondansetron melts) can be given for vomiting

Continue breastfeeding throughout episode of illness, ORS can be given in addition

Treatment of dehydration

- Admit if:
  - patient ≥10% dehydrated
  - failure of treatment (e.g. worsening diarrhoea and/or dehydration)
  - other concerns (e.g. diagnosis uncertain, child aged <3 months, irritable, drowsy, potential for surgical cause)

Step 1: Mild dehydration (<5%)

- Can be managed at home
- Emphasise to parents importance of adequate hydration
- Rehydrate orally using ORS (prescribe sachets and give clear instructions: if genuinely not tolerated, parents may substitute with diluted sugar containing juice)
  - calculate fluid deficit and replace over 4 hr with frequent small volumes (5 mL every 1–2 min)
  - continue to supplement with ORS for each watery stool/vomit (10 mL/kg per watery stool)
- Do not withhold food unless vomiting
  - full feeding appropriate for age well tolerated with no adverse effects

Step 2: Moderate dehydration (6–10%)

- If improving after 4 hr observation, can be managed at home provided social circumstances are appropriate/parents are happy. Otherwise, admit
- Calculate deficit and aim to replace with ORS 50 mL/kg oral over 4 hr
- Give small frequent feeds (5 mL every 1–2 min)
- If not tolerating oral rehydration (refuses, vomits, takes insufficient volume), use NG tube
- Review after 4 hr
  - when rehydrated start a normal diet, and continue maintenance fluids and supplementary ORS for each watery stool or vomit (10 mL/kg per watery stool)
  - if dehydration persists, continue the same regimen but replace fluid deficit with ORS over the next 4 hr
  - if this fails, e.g. vomiting ORS, consider IV rehydration (see below)
- If improving move to **Step 1**

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Step 3: Severe dehydration (>10%) – see flowchart

Beware hypernatraemic dehydration. See* Hypernatraemic dehydration *section

- If child in shock, first resuscitate with sodium chloride 0.9% (20 mL/kg) and reassess
- If >10% dehydration, obtain IV access, especially if child drowsy
- Calculate deficit using recent normal weight if available
- If alert, rehydrate orally with ORS, replacing deficit (plus maintenance requirement) over 4 hr
- Use NG tube if necessary
- If oral/NG rehydration not possible, replace deficit with sodium chloride 0.9% with glucose 5% over 24 hr
- give isotonic fluid e.g. sodium chloride 0.9% or sodium chloride with glucose 5%
- if hypoglycaemic or at risk of hypoglycaemia use sodium chloride 0.9% with glucose 5% and potassium chloride
- start normal diet as soon as tolerated
- continue to replace ongoing losses with ORS for each watery stool or vomit (5 mL/kg per watery stool)
- when improves move to **Step 2**

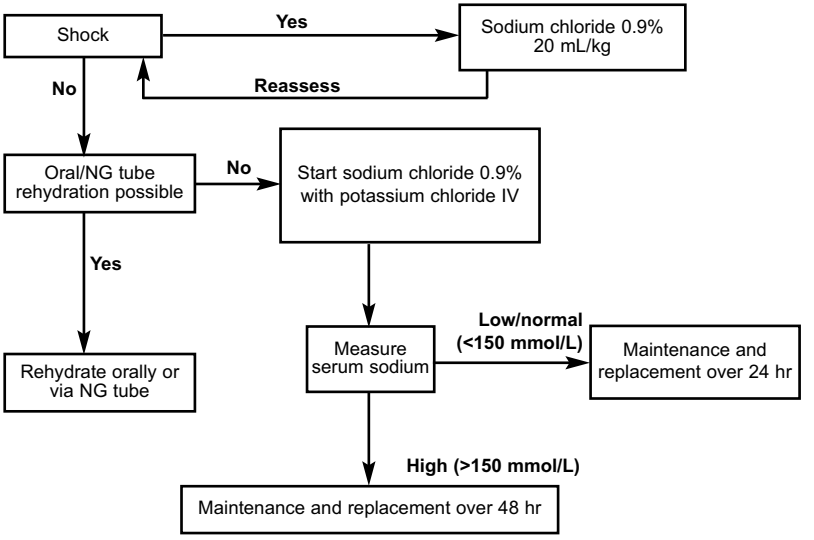
Hypernatraemic dehydration (Na >150 mmol/L)

- In hypernatraemic dehydration, there are fewer signs of dehydration
- skin feels warm and doughy, child lethargic and irritable/jittery with hypertonia and hyperreflexic. They may have seizures
- if in shock, resuscitate with sodium chloride 0.9% 20 mL/kg bolus
- if Na >170 mmol/L, contact PICU
- if child has passed urine, add potassium to IV fluid – initially at 10 mmol/500 mL, adjust according to blood results when available

In hypernatraemic dehydration, aim to reduce sodium by no more than 10 mmol/L in 24 hr

- After initial resuscitation, give ORS: replace deficit (+ maintenance) over 48 hr – via NG if necessary
- Check U&E after 1 hr
- If ORS not tolerated or sodium drops >0.5 mmol/L/hr, start IV rehydration with sodium chloride 0.9%, replacing deficit (+ daily maintenance) over 48 hr
- Recheck U&E after 1–4 hr (depending on rate of drop of serum sodium and starting value)
- If sodium dropping by >0.5 mmol/L/hr, reduce rate by 20%
- Once rehydrated, start normal diet including maintenance fluids orally

MANAGEMENT OF SEVERE DEHYDRATION



DISCHARGE AND FOLLOW-UP

- If dehydration was >5%, ensure child has taken and tolerated two breast or bottle feeds, or at least one beaker of fluid
- Check child has passed urine
- Tell parents diagnosis and advise on management and diet
- Explain nature of illness, signs of dehydration, and how to assess and deal with continuing D&V (explain flagged symptoms in table of dehydration)
- Emphasise importance of adequate hydration. If dehydration recurs will need further rehydration
- If symptoms persisting, aged <1 yr or low birth weight, continue to supplement with ORS at 5 mL/kg per watery stool or vomit
- Do not withhold food, (especially breast milk), full feeding appropriate for age if well tolerated after initial rehydration
- Advise parents how to prevent transmission to other family members and contacts
- patient should not share towels with others
- hand-washing with soap and warm water after using toilet or changing nappy. Dry hands properly
- Exclude from school/nursery until 48 hr from last episode of diarrhoea or vomiting
- Exclude from swimming for 2 weeks following last episode of diarrhoea
- Give open access if appropriate, ensure parents aware of how to seek help if needed
- If diarrhoea persists for >10 days, advise to return for medical reassessment

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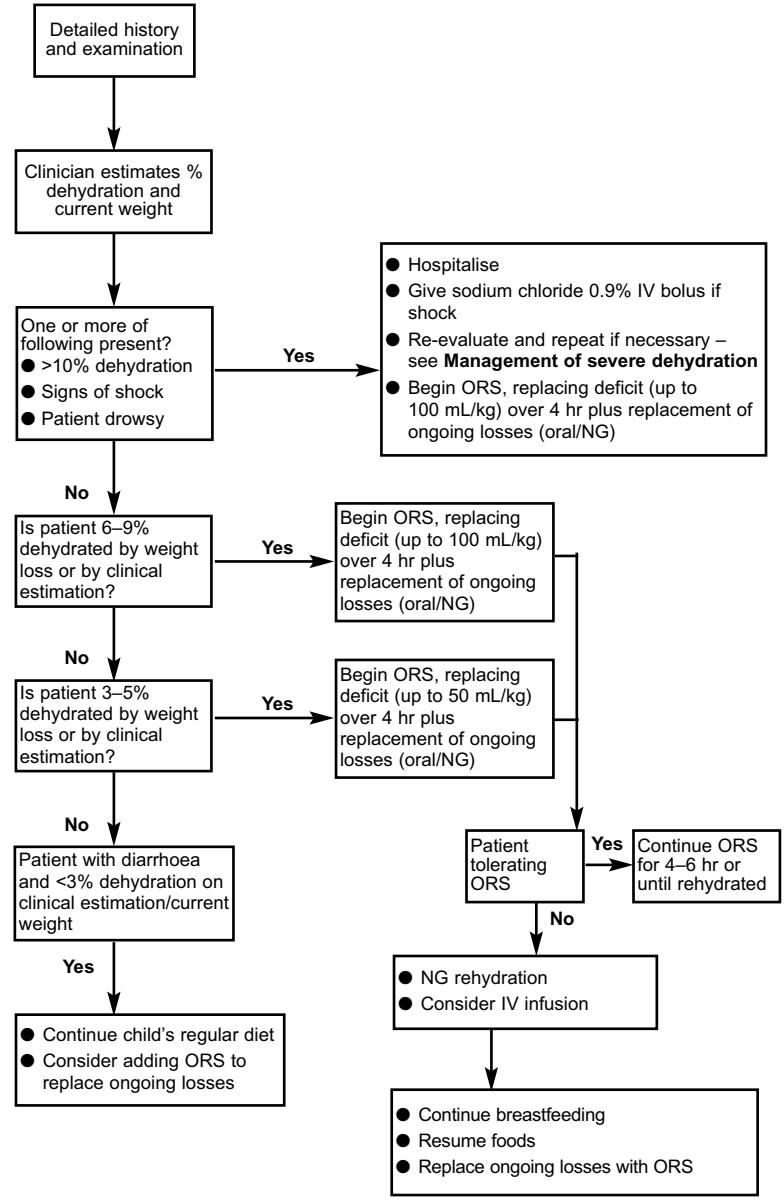
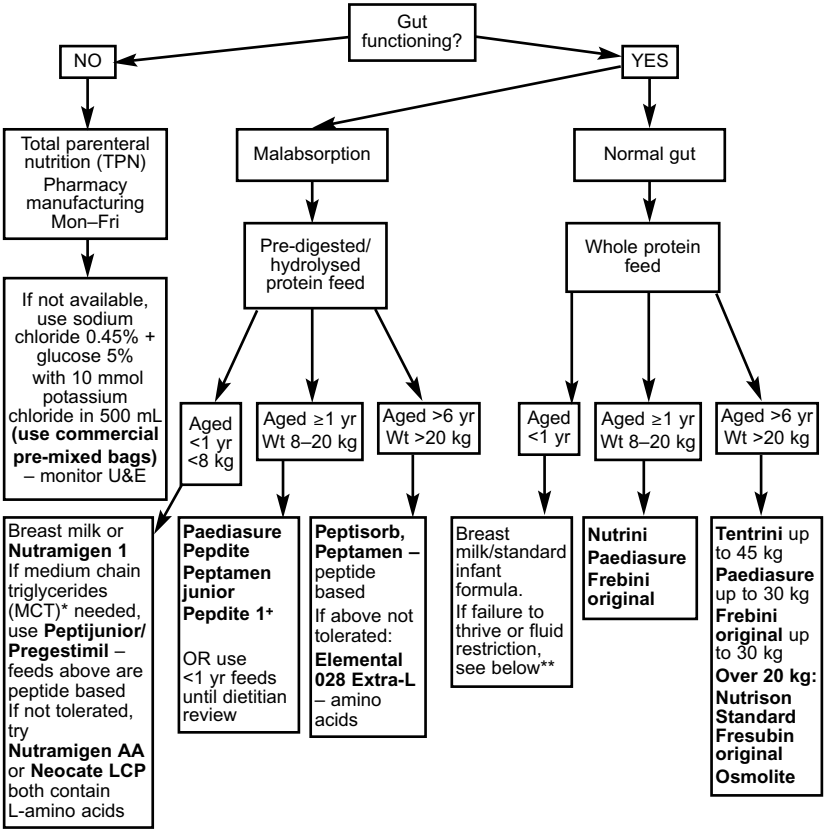
MANAGEMENT OF ACUTE GASTROENTERITIS IN YOUNG CHILDREN (AGED <4 YR)



NUTRITIONAL FIRST LINE ADVICE • 1/3

Initial guide to feeding when child not able to eat normally and dietitian **not available**

If patient nil-by-mouth see* IV fluid therapy guideline *before starting total parenteral nutrition (TPN) to ensure hypotonic fluids are not used if contraindicated



- Contact dietitian to assess individual requirements and appropriate feed at the first available opportunity Monday–Friday
- Feeds in **bold** must be prescribed
- Hospital pharmacy will advise which feed is used locally (all similar composition for ages but different manufacturer)
- See **Table 1 for daily fluid and nutritional requirements**

* Indications for MCT: malabsorption, or problems with digestion, absorption or transport of long chain fats e.g. cholestasis, short gut, pancreatic insufficiency

** If failure to thrive or fluid restricted:

- If using breast milk, dietitian to advise on fortification of breast milk
- **nutriprem** breast milk fortifier 1 sachet (2.1 g) per 50 mL expressed breast milk (EBM) can be added until dietitian advice given
- If using standard infant formula, change to **Similac High Energy or Infatrini**

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Table 1: Nutritional and fluid requirements

Age	Average weight (kg)	Fluid mL/kg per day	Energy *EAR/day	Energy Kcal/kg per day	Protein g/kg per day	Sodium mmol/kg per day	Potassium mmol/kg per day
0–3 months	6	150		100–115	2.1	1.5	3.4
4–6 months	7.5	130		95	1.6	1.6	2.8
7–9 months	9	120		95	1.5	1.6	2.0
10–12 months	10	110		95	1.5	1.5	1.8
1–3 yr female male	 12 12.5	95	 1165 1230	 95 95	1.1	1.7	1.6
4 yr female male	 16 16.5	95	 1460 1520	 87 94	1.1	1.9	1.6
5 yr female male	 18 18.5	95	 1550 1720	 82 88	1.1	1.9	1.6
6 yr female male	 20.5 21	85	 1620 1810	 76 84	1.1	1.9	1.6
7–10 yr female male	 27 27	75	 1740 1970	 70 70	1.0	1.8	1.8
11–14 yr female male	 43 40	55	 1845 2220	 47 55	1.0	1.6	1.8
15–18 yr female male	 56 62.5	50	 2110 2755	 40 45	1.0	 1.3 1.0	 1.6 1.4

*EAR - estimated average requirements = BMR (basal metabolic rate) x 1.4–1.5

Nutritional composition of milks – for further information use BNFc

Per 100 mL	Kcal	Protein g	Fat g	CHO g	Na mmol	K mmol	Osmolality
Breast milk (mature)	70	1.3	4.2	7.2	0.6	1.5	276
Cow & Gate Premium	67	1.4	3.5	7.5	0.8	1.7	330
Infatrini	100	2.6	5.4	10.4	1.0	2.6	310
Similac High Energy	101	2.6	5.4	10.3	1.1	2.3	333
SMA High Energy	91	2.0	4.9	9.8	1.0	2.3	415
Nutramigen 1	68	1.9	3.4	7.4	1.4	1.9	290
Peptijunior	67	1.8	3.6/50% MCT	6.9	1.4	1.9	190
Neocate LCP	71	2.0	3.5	6.6	0.9	1.6	360
Nutramigen AA	68	1.9	3.6	7.0	1.4	1.9	348
Nutrini	100	2.8	4.5	12.2	2.3	2.6	250
Paediasure	100	2.8	5.0	11.2	2.6	2.8	320
Nutrison Standard	100	4.0	3.9	12.3	3.5	3.5	310
Osmolite	100	4.0	3.4	13.6	3.83	3.8	288
Paediasure Pepdite	100	3.0	4.0/50% MCT	13.0	3.0	3.9	320
Peptamen junior	100	3.0	3.9/60%MCT	13.8	2.9	3.5	260
Pepdite 1+	100	3.1	3.9/35% MCT	13.0	2.1	3.0	465
Peptisorb	100	4.0	1.7/47% MCT	17.6	4.3	3.8	520
Peptamen	100	4.0	3.7/70% MCT	12.7	2.6	2.8	240
Aged >6 yr Elemental Extra 028	85	2.5	3.5/35% MCT	11	2.7	2.4	700

How to calculate energy requirements for tube feeds

- Choose appropriate feed for age. **If very underweight for age,** use appropriate feed for **actual bodyweight**
- Calculate amount of feed to use in 24 hr based on:
  - Kcal/kg in children aged <1 yr
  - estimated average requirements (EAR) for age/weight for aged >1 yr
- Calculate fluid requirement; if restricted, continue to use feeds above until reviewed by dietitian
  - if extra fluid required, give water
- Feeding method depends on clinical condition of child:
  - if child at risk of re-feeding syndrome (e.g. anorexia nervosa, Crohn's), introduce feed slowly over 3–4 days starting at 25% of Kcal intake day 1. Increase daily by 25% until full feeds at day 4
- Bolus feed can be given 1, 2, 3, 4 hourly intervals depending on tolerance
- If on continuous feeds (i.e. over 24 hr), start feed at a quarter of final hourly requirement. Increase to half requirement, three-quarters, and full every 4–6 hr as tolerated. When full feeds tolerated, aim to give full requirement over 20 hr

Monitoring

- Check plasma electrolytes daily with particular reference to phosphate, potassium and magnesium: correct accordingly. Stop once clinical condition stable
- Re-feeding syndrome may occur in the first few days of re-feeding but can occur up to 2 weeks after. Continue biochemical monitoring for 2 weeks or until electrolyte parameters are stable

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RECOGNITION AND ASSESSMENT

Definition

- An infant or older child who fails to gain weight as expected
- Growth below the 2nd percentile or a change in growth that has crossed downwards 2 major growth percentiles in a short time (approximately 4 months, or longer period in older child)
- Associated features include:
  - developmental delay
  - apathy
  - misery

Symptoms and signs

- Gastrointestinal problems
  - vomiting
  - voracious appetite
  - anorexia
  - diarrhoea
- Physical examination
  - dysmorphic features
  - heart murmurs
  - abdominal distension
  - wasting
  - bruising

Patient and family history

Child

- Take a full feeding history
  - type of milk given (breast milk, baby milk, cow's milk)
  - volume given at each feed
  - frequency of feeding
  - method of making up feeds (correct strength)
  - introduction of solids: age and type of solid
  - any difficulty with feeding process (e.g. breathless, uncomfortable)
- Perform direct observation of child at mealtimes:
  - oral, motor, co-ordination, behaviour (e.g. crying, tantrums), appetite, family interaction

Family

Ask about socio-emotional factors

- Family composition (other children, age, FTT?)
- Ask parental ages, health, educational status
  - were either parents in care during childhood?
  - do parents have a history of psychiatric illness or depression (including post-natal depression) or had learning disability?
  - parents with inadequate social or problem solving skills?
- Has the family any support network (e.g. grandparents)?
- Social isolation?
- Is there a lack of money in the home or unemployment?
- Other sources of stress (e.g. divorce)?
- Substance abuse?
- Domestic violence?

Measurements

Measurements must be carried out properly and checked if there is doubt

- Record birth weight and gestation
  - some 'light-for-dates' infants fail to catch up, and grow parallel but below the 2nd percentile
- Measure and plot
  - weight (unclothed)
  - head circumference
  - length or height
  - body mass index and plot on appropriate chart (useful if height or weight below 0.4th centile)

- Infant may be a small, normal child growing below but parallel to the 2nd percentile
- parents are often also small
- record height of parents and grandparents

Single set of measurements of limited value and does not justify complex investigations. Serial measurements of more value and should be plotted on percentile charts

Investigations

Routine tests

- Faeces: culture and sensitivity, microscopy for ova, cysts and parasites
- Urinalysis for protein, nitrites and blood
- Haemoglobin, blood film (for signs of iron deficiency), WBC and ESR
- Biochemical profile including creatinine, bicarbonate, calcium and albumin
- Coeliac screen (anti-tTG and IgA)

Further tests

- If underlying pathology indicated by history, clinical examination or results of routine investigations, request further tests, such as:
- chest X-ray
- bone age
- sweat test/cystic fibrosis (CF) gene
- Further gastrointestinal investigation or management of malabsorption disorders should be undertaken by referral to specialist gastroenterology team as appropriate:
- endoscopy
- gastrointestinal imaging

Differential diagnosis

- Low genetic growth potential:
- familial
- 'light-for-dates' baby
- genetic syndrome
- Social factors:
- maternal depression
- poor parenting skills
- abuse
- Malabsorption:
- pancreatic insufficiency: CF, Swachman-Diamond syndrome
- enteropathy: coeliac, cow's milk protein allergy
- inflammatory bowel disease (IBD)
- carbohydrate intolerance: lactose, sucrose, post-enteritis syndrome
- infective: Giardia, bacterial overgrowth
- others (rarer): abetalipoproteinaemia, lymphangiectasia
- Vomiting/severe regurgitation
- Any chronic underlying disorder:
- renal failure
- liver disease
- congenital heart disease
- severe asthma
- immunodeficiency
- other rare conditions e.g. chromosomal or metabolic conditions if dysmorphic features present

MANAGEMENT

- Most patients can be managed as out-patient
- record height and weight at each visit
- seek dietitian opinion
- seek child psychologist opinion and evaluation
- if treatable cause identified, treat appropriately

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- If social problems responsible, consider:
- admission to ward to demonstrate good weight gain out of home environment
- significant weight gain after admission (>180 g/week in infant) supports parenting issues as cause
- health visitor support
- social work support
- child psychology consultation, referral and/or intervention (evaluation of: child's cognitive development, food refusal etc; parents' perception of the child; family/child disturbances of affect expression and family dynamics)
- day care and nursery provision
- case conference
- care proceedings

JAUNDICE • 1/3

Jaundice in neonates after discharge from maternity unit

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Yellow colouration of skin in a pale-skinned infant observed in natural light
- Yellow conjunctivae in dark-skinned infants

Assess

- Pallor (haemolysis)
- Poor feeding, drowsiness (neurotoxicity)
- Hepatosplenomegaly (blood-group incompatibility or cytomegalovirus)
- Splenomegaly (spherocytosis)
- Stools (pale, chalky) and urine colour (dark, stains nappy: conjugated hyperbilirubinaemia)

Causes

- Physiological
- Prematurity
- Increased bilirubin load (e.g. bruising, blood group incompatibility)
- G6PD deficiency and other red cell enzyme deficiencies
- congenital spherocytosis
- cephalhaematoma
- Rarely infection (e.g. UTI, congenital infection)
- Metabolic disorder

Persistent jaundice after 14 days of age

- Breast milk jaundice
- Hypothyroidism
- Liver disease (e.g. extra hepatic biliary atresia and neonatal hepatitis)
- Alpha-1-antitrypsin deficiency
- Galactosaemia
- TPN-induced cholestasis

Investigations

All

- If bilirubinometer ≥250 micromol/L, total bilirubin
- Conjugated bilirubin on all babies with very light yellow/pale stools

Jaundice in first 24 hours of life or requiring treatment

- Urgent bilirubin (result within 2 hr)
- Full blood count and film
- Baby's blood group and direct Coombs test
- Mother's blood group and antibody status (should be available from maternal case notes)
- Full infection screen (in an ill baby)
- G6PD concentration (if indicated by ethnic origin: Mediterranean, Middle Eastern, South East Asian, and local hospital policy)

Persistent jaundice aged >14 days old term/preterm infants

- Total and conjugated bilirubin
- Liver function test (ALT, albumin, GGT)
- FBC, blood group, direct Coombs test and coagulation profile
- Urine MC&S
- Document stool colour
- Check routine metabolic screening has been performed

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Second line investigations if indicated by associated problems

- If conjugated bilirubin >25 mmol/L seek specialist advice
- G6PD screen in African, Asian or Mediterranean patients
- Thyroid function tests: ask for 'FT_4 priority and then TSH'
- Congenital infection screen:
  - CMV PCR: in urine first 2 weeks life, later test newborn blood spot card
  - toxoplasma ISAGA-IgM and
  - throat swab for HSV PCR
- Metabolic investigations:
  - blood galactose-1-phosphate
  - urine for reducing substances
  - urine for amino acid and organic acid
  - alpha-1-antitrypsin

If conjugated bilirubin elevated (>20% of total or >20 micromol/L), discuss with consultant urgently

Limits (micromol/L) for phototherapy and exchange transfusion for infants ≥38 weeks gestation

Age (hours)	Repeat transcutaneous bilirubin/serum bilirubin (6–12 hours)*	Consider phototherapy #	Phototherapy	Exchange transfusion
0			>100	>100
6	>100	>112	>125	>150
12	>100	>125	>150	>200
18	>100	>137	>175	>250
24	>100	>150	>200	>300
30	>112	>162	>212	>350
36	>125	>175	>225	>400
42	>137	>187	>237	>450
48	>150	>200	>250	>450
54	>162	>212	>262	>450
60	>175	>225	>275	>450
66	>187	>237	>287	>450
72	>200	>250	>300	>450
78	>212	>262	>312	>450
84	>225	>275	>325	>450
90	>237	>287	>337	>450
96+	>250	>300	>350	>450

* Result in this category repeat transcutaneous measurement in 6–12 hr

\# Result in this category repeat serum bilirubin measurement in 6 hr whether or not phototherapy started

TREATMENT <7 DAYS

- Adequate fluid and energy intake
- Phototherapy

Jaundice presenting in first 24 hours of life

- Visible jaundice can be treated with phototherapy after sample taken for bilirubin measurement
- Bilirubin >100 micromol/L: repeat in 6–12 hr

After first 24 hours

- Commence phototherapy according to following equation:
- for infants <37 weeks, start if serum bilirubin (micromol/L) ≥ phototherapy level [(gestational age in completed weeks x 10) – 100]
- for infants ≥37 weeks, start if serum bilirubin >340 micromol/L (phototherapy level)

Phototherapy

- If bilirubin near exchange threshold or still rising:
- increase power number of lights
- increase area exposed (e.g. biliblanket and overhead)

Exchange transfusion

- See **Exchange transfusion** in **Neonatal** guidelines

IVIG

- Use as an adjunct to multiple phototherapy in rhesus disease when bilirubin continues to rise by >8.5 micromol/L/hr

MONITORING TREATMENT

- If haemolysis present, check bilirubin 4–6 hrly until rate of rise flattens
- If bilirubin concentration approaching threshold for exchange transfusion, or rising rapidly (>10 micromol/hr), check 4-hrly

SUBSEQUENT MANAGEMENT

- When bilirubin concentration has fallen below threshold for phototherapy (see above), discontinue phototherapy
- If jaundice persists after 14 days of age, review and treat cause

DISCHARGE AND FOLLOW-UP

- GP follow-up with routine examination at 6–8 weeks
- If exchange transfusion necessary or considered, request development follow-up and hearing test
- In babies with positive Coombs test who require phototherapy, check haemoglobin at 2 and 4 weeks of age because of risk of continuing haemolysis and give folate

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VITAMIN D DEFICIENCY • 1/1

Serum 25-OHD concentration	Vitamin D status	Manifestation	Management
<25 nmol/L (<10 ug/L)	Deficient	Rickets Osteomalacia	Treat with high-dose vitamin D
25–50 nmol/L (10–20 ug/L)	Insufficient	Associated with disease risk	Vitamin D supplementation
50–75 nmol/L (20–30 ug/L)	Adequate	Healthy	Lifestyle advice
>75 nmol/L (>30 ug/L)	Optimal	Healthy	None

Treatment for deficiency

For 12 weeks

- Aged <6 months: 3000 units daily
- Aged 6 months–12 yr: 6000 units daily OR 20,000 units weekly
- Aged >12 yr: 10,000 units daily OR 40,000 units weekly

OR

Modified Stoss regime

- Aged >1 yr: as a one off high dose
  - <40 kg: 160,000 units oral stat
  - ≥40 kg: 300,000 units oral stat or 40,000 units once/day for 10 days
- Malabsorption or chronic liver disease
  - aged 1–12 yr: 10,000–25,000 units daily
  - aged >12 yr: 10,000–40,000 units daily

Maintenance treatment or treatment for insufficiency

For 6 months

- Aged <1 month: 200 units daily (400 units if exclusively breast fed or maternal vitamin D deficiency)
- Aged 1 month–2 yr: 400 units daily
- Aged >2 yr: 800 units daily or 20,000 units colecalciferol weekly
- Malabsorption or chronic liver disease
  - aged <1 yr: 800 units daily
  - aged >1 yr: 800–1600 units daily

Administration

- All children who can swallow normal food can take the small colecalciferol capsules (e.g. Dekristol)
- Children who have swallowing difficulties (aged <1 yr or disabled) a liquid preparation may be used but is unpalatable
- Colecalciferol and ergocalciferol liquid preparation doses are equivalent
- Colecalciferol capsules may be initiated by a paediatrician in children aged <6 yr
- GP can continue prescribing the maintenance

Preparations

Liquid

- Healthy Start vitamin drops 300 units colecalciferol per daily dose
- Abidec® (contains peanut oil) and Dalivit® 0.6 mL dose 400 units ergocalciferol

Colecalciferol capsules

- Fultium-D3 800 unit capsule (contains peanut oil)
- Dekristol 20,000 unit capsule (contains peanut oil) unlicensed
- ProD3 10,000, 20,000 and 30,000 unit capsules unlicensed

Other unlicensed preparations

- May be very much more expensive: check with pharmacy (use brand name to ensure correct formulation)
- Zymad 10,000 units/mL (300 units colecalciferol per drop) 5 drops 1500 units unlicensed
- Uvesterol 1500 units/mL ergocalciferol unlicensed
- Sterogyl 20,000 units/mL ergocalciferol unlicensed
- Vigantol 20,000 units/mL colecalciferol

Always check front sheet in patient notes before prescribing any blood product

Before transfusion

- Explain indications for blood products to parents
- Document indications and verbal consent
- If previous reactions to blood products have occurred, pre-medicate with chlorphenamine (oral or IV), if severe with hydrocortisone 4 mg/kg IV

BLOOD TRANSFUSION

When to transfuse

Oncology children

- If haemoglobin ≤80 g/L or if >80 g/L and symptomatic, transfuse
- If having radiotherapy, transfuse if Hb <100 g/L

Non-oncology children

- If haemoglobin <60 g/L or >60 g/L and symptomatic

Target Hb and volume to be transfused

- Aim for target haemoglobin of 120 g/L or for 100 g/L if initial haemoglobin <60 g/L
- In newly diagnosed patients with leukaemia/profound anaemia, aim for target Hb 80–90 g/L
- Calculate volume to be given as: (round to nearest unit)

  [Target Hb – actual Hb (g/L)] x weight (kg) x 0.4 mL
- **Total volume should not exceed 20 mL/kg**

Rate of infusion

- Give **total** over 3–4 hr. **Max rate 5 mL/kg/hr**
- If Hb <60 g/L, give blood over 4–8 hr (each unit must be used **within 4 hr** once removed from fridge)
- Give furosemide 1 mg/kg oral if tolerated, or IV half-way through

Use irradiated blood if

- Allogenic bone marrow transplant (BMT) from start of conditioning regimen
- Allogenic BMT donors
- If <7 days pre-harvest for autologous BMT and stem cell transplant patients (e.g. stage IV neuroblastoma)
- Hodgkin's disease or if patient has received fludarabine
- Children with severe immunodeficiency (e.g. SCID)
- HLA-matched platelets
- For high risk neonates e.g. post intrauterine transfusion

Leucodepleted blood

- All packed cells are leucodepleted

CMV negative blood

- All the packed cells are leucodepleted and therefore CMV negative
- For neonates aged <28 days post expected date of delivery and for intrauterine transfusions CMV serology negative blood requested

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PLATELET TRANSFUSION IN ONCOLOGY CHILDREN

Transfuse platelets if platelet level

- <10 x 10^9/L oncology children except brain tumour
- <20 x 10^9/L oncology children except brain tumour and unwell
- <30 x 10^9/L brain tumour
- <50 x 10^9/L brain tumour and unwell
- <50 x 10^9/L for lumbar puncture

Dosage and rate

- <15 kg: 15 mL/kg round off the nearest unit
- ≥15 kg: one pack
- Transfuse within 15–30 min

Immune thrombocytopenic purpura (ITP) – transfuse platelets only if bleeding and see **Immune thrombocytopenic purpura (ITP)** ***guideline***

Frequent clinical re-assessment of patients is a vital part of effective management of febrile neutropenia in children

RECOGNITION AND ASSESSMENT

Definition

- Temperature: ≥38°C at any time
- Neutrophils <1x10^9 cells/L

IMMEDIATE TREATMENT

See **Figure 1** (see **BNFc** for dose reduction in renal impairment)

ALL PATIENTS – with central venous access

- Culture both lumens/portacath. Take FBC, group and save, coagulation screen, U&E, Cr, LFTs, CRP
- Urinalysis in all children aged <5 yr
- CXR only if respiratory signs
- **Do not wait for results, administer antibiotics**
- 'Door to needle time' must be within 1 hr
- Follow individual trust antibiotic policy or individual patient plan if resistant organisms

No haemodynamic compromise

- Start piperacillin with tazobactam (Tazocin®) 90 mg/kg 6-hrly (maximum single dose 4.5 g) unless penicillin allergy or previous Tazocin® resistant gram negative infection:
  - then use meropenem 20 mg/kg 8-hrly (maximum single dose 1 g)
- If previous documented MRSA infection, **add** vancomycin 15 mg/kg 6-hrly (maximum single dose 700 mg) until levels available. Aim 10–15 mg/L
- Pre-dose vancomycin level before third dose, and no post-dose sample required
- Adjust pre-dose concentration (mg/L) dose as follows:
  - <10 give 6-hrly and recheck level before dose 4 or 5
  - 10–15 continue current dose and recheck concentration in 3–5 days
  - 15–20 reduce frequency of dosing and recheck level before dose 4 or 5 (unless higher levels advised by microbiology)
  - >20 stop vancomycin and recheck level next day to see if therapy can be restarted

Haemodynamic compromise

- Check A, B, C and initiate appropriate resuscitation
- Give sodium chloride 0.9% 20 mL/kg bolus
- Start meropenem 20 mg/kg 8-hrly

LOW RISK PATIENTS

- No central access **and**
- Neutrophils >0.5x10^9 cells/L **and**
- Clinically well
- consider discharge on oral antibiotics after discussion with oncology team

SUBSEQUENT TREATMENT

- Reassess at 24 hr and chase blood cultures
- **Positive cultures:** Discuss patients with microbiologist or paediatric oncology team for advice on appropriate treatment. Where blood cultures positive for yeast in presence of suspected line infection, remove suspected lines promptly
- Give culture-positive patients at least 7 days treatment

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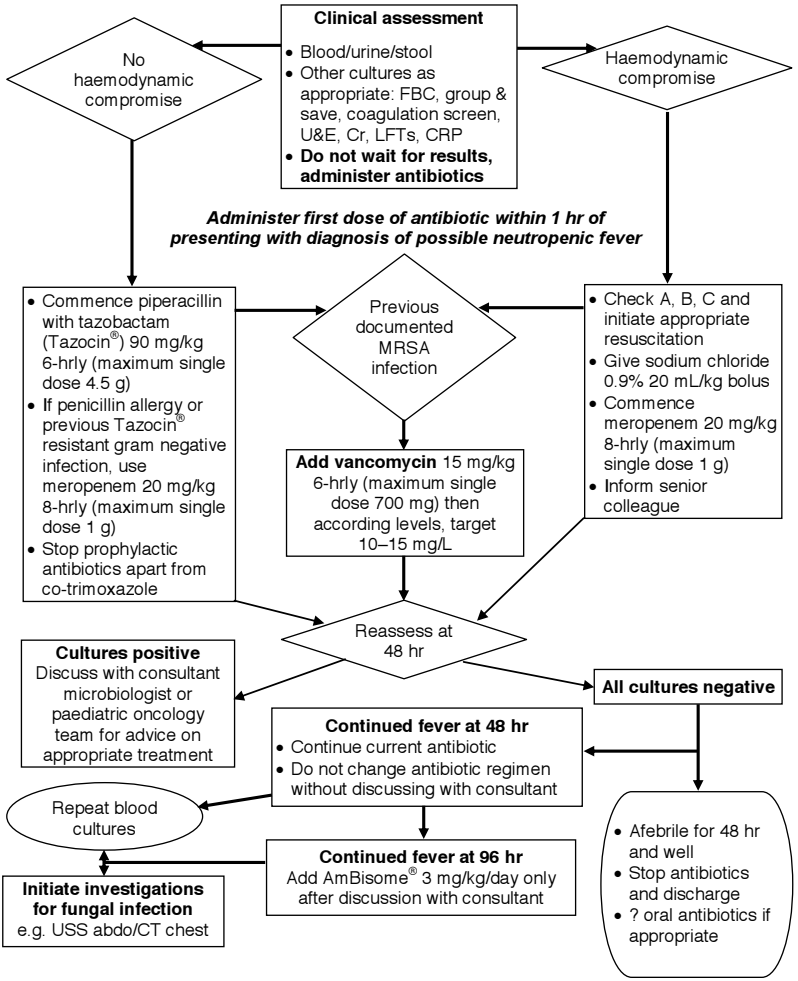
- **Negative cultures:** Do not switch initial empiric antibiotics with unresponsive fever unless there is clinical deterioration or a microbiological indication
- If febrile after 48 hr:
  - repeat blood cultures and discuss with on-call consultant/paediatric oncology team
- Initiate investigations for fungal infection e.g. US abdo/CT chest
- If febrile after 96 hr or clinically unstable between 48 and 96 hr:
  - repeat blood cultures
  - add **liposomal amphotericin** (AmBisome®) **3 mg/kg/day** (give test dose 100 microgram/kg (max 1 mg)

When to discharge

- If clinically well and afebrile for 48 hr, and no growth in blood cultures after 48 hr:
  - stop antibiotics
  - no need for routine in-patient observation after stopping antibiotics

Figure 1: Management of fever in neutropenic/immunocompromised child



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RECOGNITION AND ASSESSMENT

Definition

- Vasculitic condition of unknown aetiology
- 50% have a preceding upper respiratory tract infection
- Affects skin, gastrointestinal tract, joints and renal tract
- Typical age group 2–8 yrs old

Symptoms and signs

Rash

- Purpuric, raised on extensor surfaces of legs, buttocks and arms, with surrounding erythema

Gastrointestinal tract

- Abdominal pain mostly idiopathic, typically resolves in 72 hr
- if severe or persistent, exclude intussusception, testicular torsion or pancreatitis (rare)
- Nausea and vomiting
- Intestinal haemorrhage: haematemesis, melaena, bloody stools (rare)

Joints

- Arthralgia and swelling of large joints, especially ankles and knees. Pain typically resolves in 24–48 hr

Renal

- Microscopic haematuria (common)
- Proteinuria can present 4–6 weeks after initial presentation
- Hypertension
- Nephritic syndrome: haematuria with at least one of following:
  - raised urea and creatinine
  - hypertension
  - oliguria
- Nephrotic syndrome: proteinuria +/- oedema and hypoalbuminaemia
- Oedema of hands, feet, sacrum and scrotum

Neurological

- Headache (common)
- Seizures, paresis, coma (rare)

Differential diagnosis

- Purpuric rash:
  - meningococcaemia – clinical diagnosis
  - thrombocytopenia – FBC (rash looks different, ITP not vasculitic)
  - rarer vasculitides – more difficult to exclude; differentiation requires review over a period of time
  - pancreatitis – suspect in abdominal pain lasting >3 days

Investigations

All patients

- BP
- Urine dipstick
  - if proteinuria, send urine for early morning protein:creatinine ratio
  - if haematuria, send urine for microscopy

Additional investigations

Blood tests if urinalysis abnormal or diagnosis uncertain

- FBC+ film
- U&E
- Albumin
- If fever, blood culture and ASO titre
- Coagulation
- Throat swab

IMMEDIATE TREATMENT/SUBSEQUENT MANAGEMENT

- Condition is self-limiting, symptomatic relief only
- Mortality <1% usually related to kidneys
- Long-term morbidity related to renal disease
- 1% of those with renal involvement progress to end stage renal failure
- HSP accounts for 5–15% of patients with end stage renal failure in children

Joint pain

- NSAIDs (ibuprofen first-line, indometacin or diclofenac second-line. Use with caution if renal involvement)

Abdominal pain

- Give prednisolone 1 mg/kg/day for 2 weeks
- Renal involvement not a contraindication
- If severe and persists, exclude pancreatitis, intussusception or spontaneous bowel perforation

MONITORING

Uncomplicated HSP (e.g. urine analysis ≤1+ blood and protein, and normal BP)

- No hospital follow-up required but GP to follow-up as below

HSP with haematuria or proteinuria >1+ and normal renal function

- GP follow-up in 1–2 weeks. Monthly BP for 6 months and weekly urine dipsticks at home until urine clear
- If blood or protein >1+, routine follow-up in children's out-patients

Refer to nephrologist if

- Urinalysis blood or early morning protein >1+ after 6 months
- Macroscopic haematuria or heavy proteinuria at presentation
- Hypertension (see **Hypertension** guideline)
- Significant proteinuria (early morning urine protein:creatinine ratio >100 g/mmol or 3+ proteinuria for 3 days)
- Impaired renal function

Refer to rheumatologist if

- Atypical or rapidly evolving rash

DISCHARGE AND FOLLOW-UP

- Inform parents condition may fluctuate for several months but recurrence rare once settled properly
- Very rare risk of renal failure, hence importance of monitoring urine
- Seek medical advice if child develops headache, PR bleeding or severe abdominal pain

Uncomplicated HSP

- GP follow-up in 1–2 weeks. Monthly BP for 6 months and weekly urine dipsticks at home until urine clear

Discharge from GP follow-up

- If urine analysis is normal **and**
- If BP normal at 6 months of symptoms onset

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IMMUNE THROMBOCYTOPENIC PURPURA (ITP) • 1/2

RECOGNITION AND ASSESSMENT

Definition

- Platelets <100 x 10^9/L, usually <20 x 10^9/L
- Self-limiting disease with shortened platelet survival and increased megakaryocytes
- Good prognosis
- Acute 0–3 months
- Persistent 3–12 months
- Chronic >12 months

Symptoms and signs

- Acute onset bruising, purpura and petechiae
- serious mucosal bleeding unusual, look for other causes
- Preceding infection
- Absence of:
- hepatosplenomegaly
- lymphadenopathy
- evidence of serious cause/chronic underlying illness

Investigations

- FBC, blood film and clotting
- Blood group
- CMV and EBV IgM
- Consider HIV, Hepatitis B and C if risk factors
- If ITP, headache and neurological signs, urgent CT scan of head
- Bone marrow aspiration unnecessary unless:
- neutropenia or severe anaemia
- hepatosplenomegaly
- lymphadenopathy
- pallor and lassitude
- pain limb/abdomen
- limp

IMMEDIATE TREATMENT

- None regardless of platelet count, unless life-threatening owing to significant bleeding
- If significant bleeding (e.g. uncontrollable epistaxis, GI haemorrhage, intracranial bleed), give:
- platelets (see **Blood and platelet transfusions** guideline)
- immunoglobulin 1 g/kg (see local policy) can be repeated once within 3 days if required
- If moderate bleeding e.g. prolonged mucosal bleeds, give prednisolone 4 mg/kg (max for 4 days)
- Consider tranexamic acid for small bleeds
- Avoid NSAIDs e.g. ibuprofen
- Reassure parents
- Discuss newly diagnosed ITP with paediatric haematologist
- Discuss treatment with platelets with paediatric haematologist in event of:
- essential operations
- emergency dental extractions

SUBSEQUENT MANAGEMENT

- 75–80% resolve in 6 months
- favourable outcome irrespective of treatment
- Avoid contact sports
- impossible to prevent fighting/rigorous knockabout games at home
- Parents can find additional information from ITP support association: www.itpsupport.org.uk

MONITORING TREATMENT

- FBC and film monthly until diagnosis clear or recovery
- Repeat sooner if bleeding or increased bruising

DISCHARGE AND FOLLOW-UP

- Discharge from long-term follow-up when platelets >100 x 10^9/L and asymptomatic
- Advise of risk of relapse (20%)
- Note that mothers with history of ITP (even if they have normal platelet counts) can give birth to thrombocytopenic babies

CHRONIC IMMUNE THROMBOCYTOPENIC PURPURA

- Avoid NSAIDs
- Avoid contact sports
- Investigate for autoimmune disease (ANA antinuclear antibody; APLA, antiphospholipid antibodies; ACA, anticardiolipin antibody; and LAC, lupus anticoagulant) and immune deficiency (HIV)
- Treat only:
  - profound thrombocytopenia (<10 x 10^9/L) with repeated mucosal bleeding
  - older girls with menorrhagia
  - trauma
  - acute neurological signs
- If treatment indicated, give prednisolone 1–2 mg/kg/day until count responds
  - reduce gradually
  - must have bone marrow aspirate before treatment
- If unresponsive, discuss with paediatric haematologist about treatment with rituximab

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INTRODUCTION

- Haemophilia is a serious disease. Each child with haemophilia must:
  - have open access
  - be treated within 30 min of attending the ward
  - be registered with designated tertiary haemophilia unit
  - be registered locally (if shared care appropriate) – for local registration, there will be two copies of the treatment sheet, one copy at front of patient notes and a second copy on ward

Inform haemophilia nurse of any patient attending for treatment

INDICATIONS FOR ADMISSION

- Bleeding in mouth, neck, respiratory passages or gastro-intestinal tract
- Suspected internal bleeding (intracranial, intra-thoracic or intra-abdominal)
- Haemorrhage endangering a nerve (e.g. carpal tunnel – median nerve, iliopsoas – femoral nerve) or other vital structure
- Requiring surgical treatment, including dental surgery
- Haemarthrosis, especially weight-bearing joints (e.g. hips and knees)
- Any lesion requiring 12-hrly or more frequent replacement therapy

MANAGEMENT OF ACUTE BLEEDING

- Patients present for treatment, particularly when developing a haemarthrosis before any physical signs are present
  - give **immediate** replacement therapy as haemarthroses are very painful and any delay may increase severity of bleed and risk of joint damage
  - if any doubt, contact haemophilia nurse or haematologist

Replacement therapy dosage

- When deciding dose, consider:
  - type of lesion
  - time of onset of symptoms
  - factor level required to sustain haemostasis
  - half-life of therapy (varies with each concentrate)

Type of lesion	Level of factor desired
● Uncomplicated bleeding into joints and muscles	● Non-weight bearing joint 30% ● Weight bearing joint 50% (may need twice daily infusion)
● Haematoma in potentially serious situations: ○ bleeding in mouth ○ neck ○ respiratory passages ○ endangering nerves	● 30–50%
● Pre-dental extraction	● 50%
● Major surgery ● Serious accident ● Head injury	● 80–100%

Calculation of replacement factor

- Most boys with haemophilia receive recombinant factor
- Calculate units of factor needed, X, using following formula:
- $X = \frac{\%\ \text{rise in factor required} \times \text{wt (kg)}}{K}$

  (where K is the recovery constant)
- Recovery constants vary. Common factors used are:
- **haemophilia A:** Factor VIII concentrates are: Advate, Kogenate, ReFacto, Helixate and Recombinate, with recovery constant (K) = 2
- **haemophilia B:** Factor IX concentrate BeneFIX, with recovery constant (K) = 0.8. It often has a short half-life in children (aim 60% rise)
- **Factor X deficiency:** Beriplex blood product, with recovery constant (K) = 2.2
- **von Willebrand's disease:** use Haemate P, with recovery constant (K) = 1.5
- For any other Factor concentrate, contact on-call haematologist to discuss treatment and ascertain correct recovery constant

Other treatment

- On advice of consultant haematologist for those with inhibitors to factors VIII or IX
- Factor VIIa (recombinant: Novoseven) 90 microgram/kg 2-hrly with frequent review

Administration of factor concentrate

- Give intravenously over about 3 min
- adverse reactions rare but include anaphylactic shock
- During prolonged treatment screen for inhibitors every 5 doses

Duration of treatment

- Decided by local on-call haematologist or designated tertiary haemophilia unit (on-call haematologist). **If in doubt, ask**

DESMOPRESSIN IN MILD HAEMOPHILIA A AND VON WILLEBRAND'S DISEASE

- Subcutaneously, intranasally or IV
- may be used to raise Factor VIII concentration
- response usually fourfold rise (IV) or twofold rise (intranasal) in Factor VIII and von Willebrand's antigen concentration

Patient selection

- Consider **only** in mild (**NOT** severe) haemophilia A
- **Not** appropriate in Factor IX deficiency (haemophilia B)
- Do not use in child aged <1 yr
- caution in children aged <2 yr
- Check notes for outcome of previous desmopressin challenge

Administration of desmopressin

- Intravenously: 0.3 microgram/kg IV in sodium chloride 0.9% 50 mL over 20 min. May be repeated after 12 hr
- Intranasally: 4 microgram/kg once
- **Side effects** include hypertension
- measure pulse and BP every 5 min during infusion. If either rises unacceptably, reduce rate of infusion
- Ensure blood samples taken before and after infusion to measure Factor VIII level and ensure therapeutic level reached
- tachyphylaxis can occur with depletion of stored Factor VII with consecutive days. After 3 days there may be an inadequate rise of Factor VIII
- Restrict patient's fluid intake to 50% of maintenance (max 1 L/day) over the following 24 hr

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VON WILLEBRAND'S DISEASE

- More common than haemophilia
- caused by deficiency (qualitative or quantitative) of vWF protein, which binds to Factor VIII (prolonging half-life) and platelets
- Can present with acute episodes of mucosal bleeding, helping to form initial clot
- Before treatment, consider:
- von Willebrand's disease (vWD) sub-type
- bleeding history, including previous response to any treatment
- nature of haemostatic challenge
- Treatment is often a combination of tranexamic acid and desmopressin or Haemate P

Tranexamic acid

- Anti-fibrinolytic agent
- Contraindicated in presence of frank haematuria (>2+ blood)
- Decrease dose in renal failure
- For minimal mucosal bleeding, tranexamic acid mouth wash may be sufficient to stop initial bleeding
- Oral tranexamic acid alone can be used to treat minor problems such as recurrent epistaxis, but main use is in combination with desmopressin if appropriate
- oral dose 15–25 mg/kg 8-hrly (max dose 1.5 g) for max 5 days
- Intravenous tranexamic acid 10 mg/kg (max 1 g) 8-hrly over 10 min

Desmopressin

- Treatment of choice in responsive patients for spontaneous bleeding, trauma and minor surgery
- For administration – see **Administration of desmopressin**

vWD Type	Advice
Type 1	● Most patients responsive
Type 2A	● Some patients responsive ○ ask about previous challenge
Type 2B	● DO NOT GIVE desmopressin ○ it causes platelet agglutination and thrombocytopenia
Type 3	● Not all responsive and some can be severe ○ ask about previous challenge

Haemate P (blood product)

- Avoid if at all possible
- Use in patients not responsive to, or unsuitable for, desmopressin (e.g. aged <2 yr)
- See above for administration of replacement factor. Recovery constant for Haemate P = 1.5

EMPIRICAL ANTIBIOTICS

Once organism identified, change antibiotic to narrowest spectrum appropriate for site of infection

Oral unless unavailable or IV stipulated; if not tolerating oral fluids use same antibiotic IV

Pneumonia:	**Mild/Moderate**	**Severe respiratory distress**
Community acquired	Amoxicillin	
1st line	(vomiting: benzylpenicillin IV)	Co-amoxiclav IV + azithromycin
2nd line	Amoxicillin **and** azithromycin	Piperacillin/tazobactom + azithromycin
Flu	Oseltamivir	Oseltamivir + co-amoxiclav
Empyema	Co-amoxiclav IV + clindamycin	
Hospital acquired	Piperacillin/tazobactam	

Allergy

- azithromycin instead of amoxicillin
- clindamycin instead of co-amoxiclav or piperacillin/tazobactam

		Penicillin allergy
Meningitis	Cefotaxime or ceftriaxone (high dose) + amoxicillin IV (aged <3 months) + teicoplanin IV (multiple antibiotics in last 3 months or recent travel outside UK)	Cefotaxime or ceftriaxone IV If confirmed severe anaphylaxis chloramphenicol IV or teicoplanin + gentamicin
Sepsis from: **Community** **Hospital**	Cefotaxime or ceftriaxone (high dose) Piperacillin/tazobactam	
Encephalitis	Aciclovir IV (high dose)	
UTI aged <3 months **aged >3 months:** Cystitis Pyelonephritis	Cefotaxime or ceftriaxone Cefalexin Co-amoxiclav	Gentamicin Gentamicin
Osteomyelitis and septic arthritis	Cefotaxime or ceftriaxone aged <5 yr Flucloxacillin (high dose) IV aged >5 yr	Cefotaxime or ceftriaxone Clindamycin
Endocarditis	Flucloxacillin IV + gentamicin (low dose)	Vancomycin + gentamicin (low dose)
Prosthesis or ?MRSA	Vancomycin + rifampicin + gentamicin (low dose)	
GI surgical prophylaxis **Peritonitis treatment**	Co-amoxiclav IV Piperacillin/tazobactam	Gentamicin + metronidazole
Tonsillitis	Penicillin V (if can swallow tabs) Amoxicillin (suspension)	Azithromycin
Otitis media	Amoxicllin (1st line) Co-amoxiclav (2nd line)	Azithromycin
Otitis externa	Flucloxaxillin (if can swallow tabs) Co-amoxiclav (suspension)	Azithromycin
Impetigo	Fusidic acid 2% ointment	
Erysipelas	Co-amoxiclav	Azithromycin
Cellulitis	Co-amoxiclav Flucloxacillin IV (if severe)	Clindamycin
Periorbital cellulitis	Co-amoxiclav	Azithromycin
Orbital cellulitis	Cefotaxime	Chloramphenicol IV

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LOCAL ANTIBIOTIC POLICY

- Follow your local Trust antibiotic formulary as appropriate

Prevention of infection after bites from humans and other animals

Give prophylactic antibiotics to

- All human bite wounds ≤72 hr old, even if no sign of infection
- Animal bite wounds if wound ≤48 hr old and risk of infection high as follows:
- bites to hand, foot, and face; puncture wounds; wounds requiring surgical debridement; crush wounds with devitalised tissue; wounds in genital areas; wounds with associated oedema; wounds involving joints, tendons, ligaments, or suspected fractures
- wounds that have undergone primary closure
- patients at risk of serious wound infection (e.g. immunosuppressed)
- asplenic patients, even after trivial animal bites
- patients with prosthetic implants e.g. heart valve, VP shunt
- antibiotics are not generally needed if wound ≥2 days old and no sign of local or systemic infection
- Advise patient and carers of signs of developing infection and to attend urgently for review should this happen

Source is known or suspected to be positive for HIV, hepatitis B or C, or rabies

- Ask vaccination and HIV/Hep B and C status of person bitten
- Ask if biter is willing to be tested
- Risk of HIV transmission is extremely small. See **HIV and hepatitis B post-exposure prophylaxis (PEP)** guideline
- Seek advice from consultant microbiologist or consultant in infectious diseases
- If significant risk of blood borne virus transmission, offer to test person bitten:

Time	Hepatitis C	Hepatitis B	HIV
clotted sample for archiving at time of incident			
6 weeks	PCR	HBsAg	Antigen/antibody combined test
3 months	PCR and antibody	HBsAg	Antigen/antibody combined test
6 months	Antibody	HBsAg Anti-HBc antibody (anti-HBs antibody)*	Antigen/antibody combined test if PEP was given

* Anti-HBs only needed at 6 months if vaccination only started at injury

Antibiotic prophylaxis

Type of bite	Human, dog or cat†	
Specimen	If clinical infection present, send tissue, aspirate or swab for bacterial culture If patient systemically unwell, blood cultures	
Treatment	Tetanus vaccine [e.g. combined diphtheria (low dose), tetanus, and poliomyelitis] and immunoglobulin if indicated (see current BNFc)	
	First line Co-amoxiclav	**Alternative (penicillin allergy)** Clindamycin and cotrimoxazole
Duration	5 days	

† For bites from other mammals, contact consultant microbiologist or consultant in infectious diseases

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CERVICAL LYMPHADENOPATHY • 1/4

Enlargement of cervical lymph nodes >1 cm

Acute lymphadenitis

- Short history (usually <2 weeks)
- Neck mass with features of acute inflammation

Subacute lymphadenopathy

- History variable
- Often non-tender but with overlying erythema

Chronic lymphadenopathy

- Longer history (usually >1 month)
- No feature of acute inflammation

HISTORY

Symptoms

- Duration
- Symptoms of URTI
- Fever
- Weight loss
- Night sweats
- Eczema/skin infection
- Bruising
- Pallor
- Bone pain
- Pruritis

Social

- Contact with TB or cats
- Travel

EXAMINATION

- Site of node(s) (see **Figure 1**)
- Size of node(s)
- ENT examination
- Skin – especially eczema
- Axillae, supraclavicular and groin for other nodes
- Abdomen for hepatosplenomegaly

DIFFERENTIAL DIAGNOSIS

Acute unilateral

- Reactive
- URTI (*Strep. pneumoniae*)
- skin infection (*Group A Strep, Staph. aureus*)
- dental infection (anaerobes)
- Kawasaki (see **Kawasaki disease** guideline)
- Cat scratch disease (Bartonella: tender, axillary lymphadenopathy)
- Kikuchi-Fujimoto disease (histocytic necrotising lymphadenitis)

Acute bilateral

- Reactive
- viral URTI
- EBV, CMV (generalised lymphadendopathy, hepatosplenomegaly)

Subacute

- Non-tuberculous mycobacteria (aged <5 yr, unilateral, non-tender, purple, systemically well)
- Mycobacterium tuberculosis
- Toxoplasma gondii (generalised lymphadenopathy, fatigue, myalgia)

Chronic

- Reactive
- Neoplasia
- lymphoma
- leukaemia
- soft tissue tumours
- juvenile chronic arthritis
- SLE

INVESTIGATIONS

- See **Flowchart**
- Serology for Bartonella, toxoplasma, CMV and EBV
- CXR
  - Hilar lymphadenopathy significantly increases likelihood of neoplastic disease
- Ultrasound
  - high sensitivity and specificity for abscess formation in acute lymphadenitis
  - value in chronic lymphadenopathy for assessing size, site, shape and vascularity
- CT only if suspected deep neck space infection

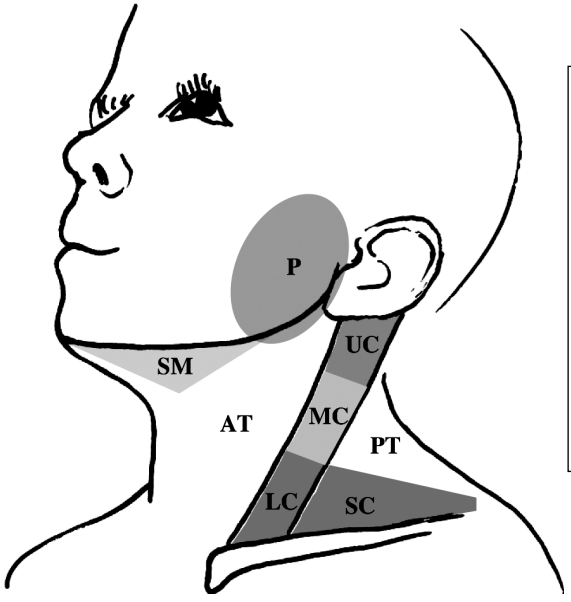
Surgical excision biopsy

- Atypical mycobacterial infection
- Features highly suggestive of neoplasia:
  - lymph nodes >3 cm diameter
  - all supraclavicular nodes
  - constitutional symptoms
  - hepatosplenomegaly
  - generalised lymphadenopathy

Children undergoing surgical biopsy for suspected neoplastic disease

- FBC and film
- U&E, LDH, uric acid, LFTs
- CXR

Figure 1 Node sites



AT:	Anterior triangle
PT:	Posterior triangle
SM:	Submandibular triangle
SC:	Supraclavicular triangle
LC:	Lower cervical chain
MC:	Mid cervical chain
UC:	Upper cervical chain
P:	Parotid

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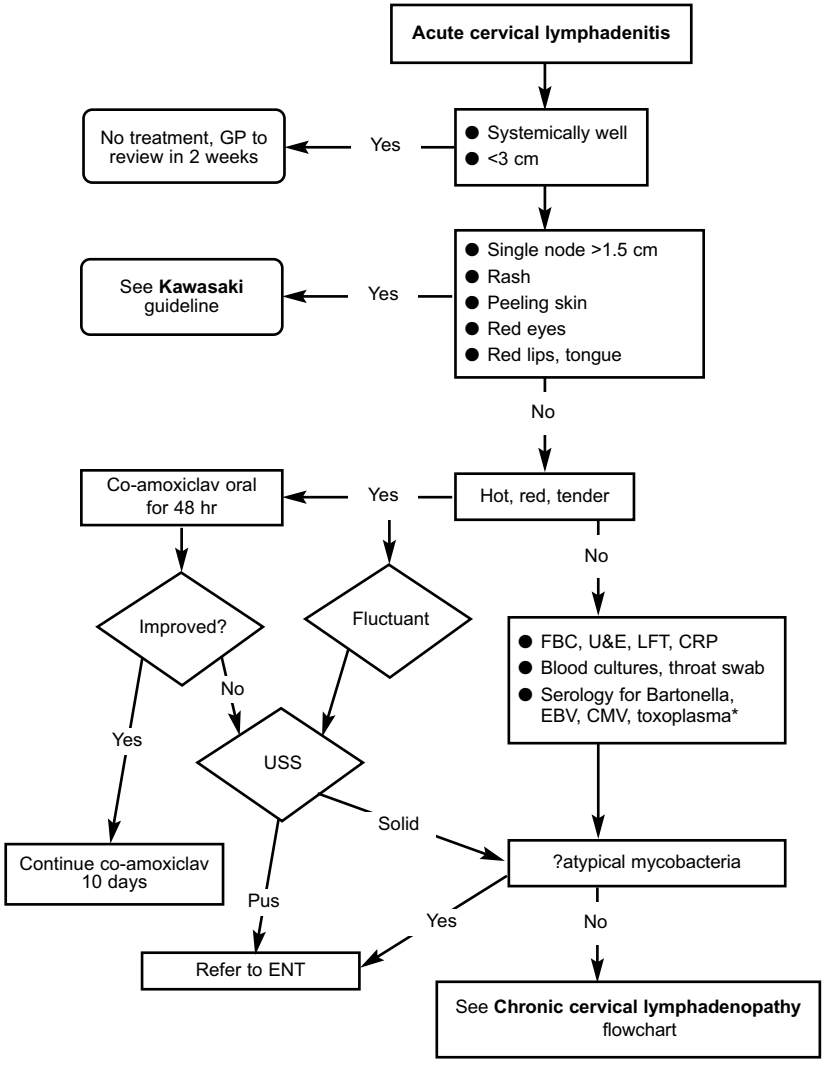
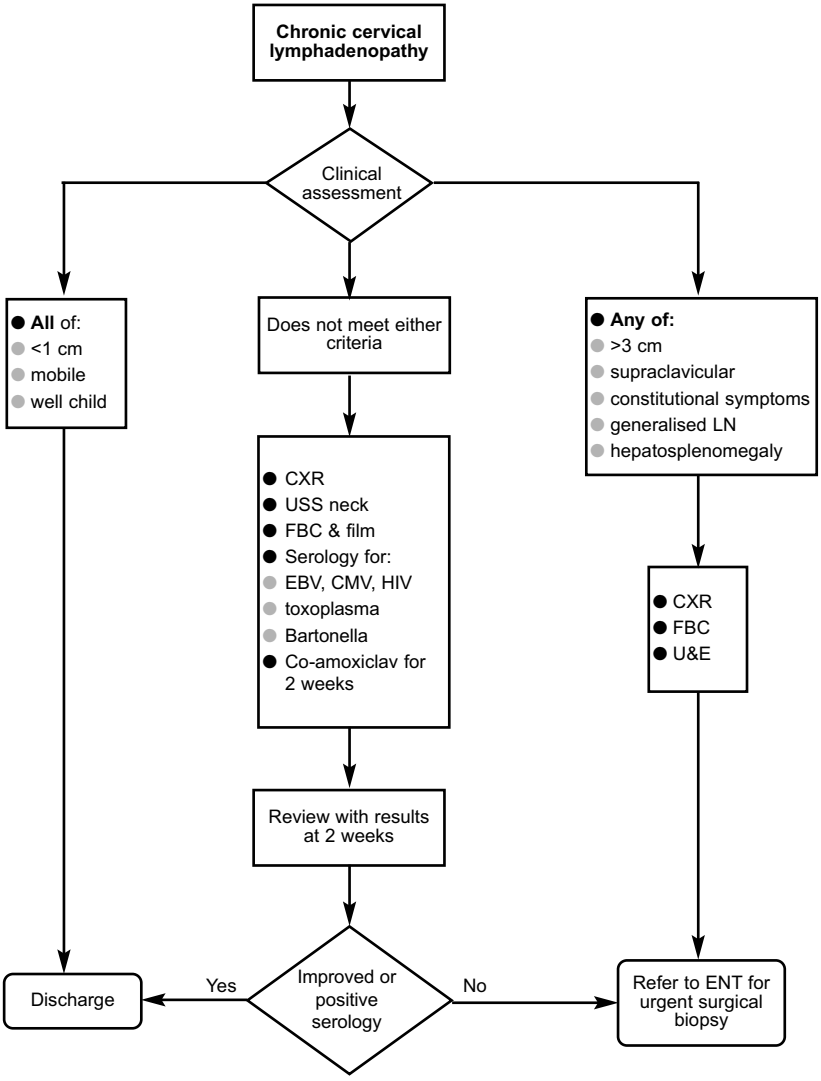


* For storage pending repeat titre in chronic course



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This guideline applies until underlying condition diagnosed, at which point treat child according to the guidance for that condition

ASSESSMENT AND INITIAL MANAGEMENT

- Fever, in child aged <5 yr, usually indicates underlying infection
- Parental perceptions of fever are usually accurate and must be taken seriously

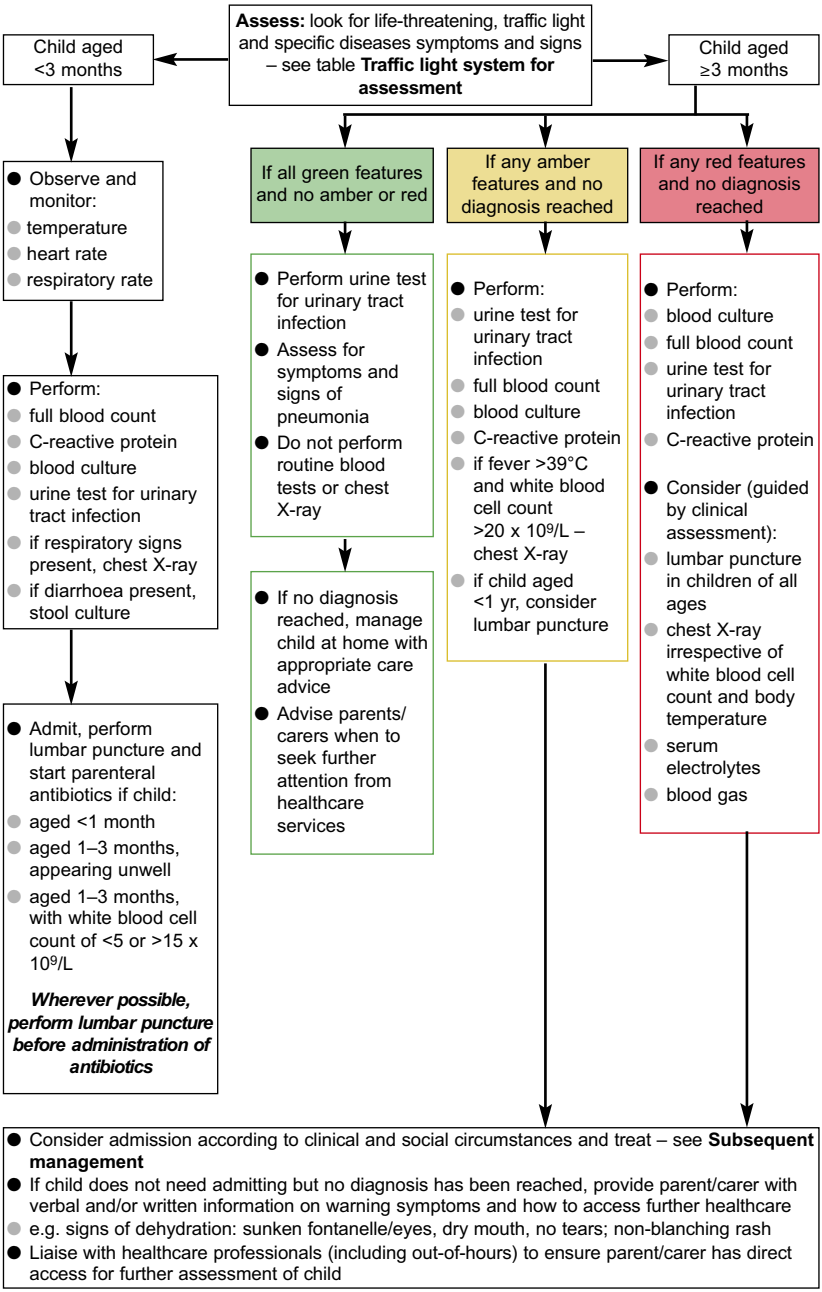
IDENTIFYING RISK OF SERIOUS ILLNESS

Three stages of clinical assessment

1. Identify life-threatening features (utilising Airway, Breathing, Circulation and Disability assessment)
2. Assess risk of serious illness (see **Traffic light system for assessment**)
3. Attempt to identify source of infection/features of specific serious conditions

Traffic light system for assessment

	Low risk	Intermediate risk	High risk
Colour	● Skin, lips and tongue normal	● Pallor reported by carer	● Pale, mottled, ashen or blue
Activity	● Responds to normal social cues ● Is content or smiles ● Stays awake/wakes quickly ● Strong normal cry/settled/smiles	● Not responding normally to social cues ● Wakes only with prolonged stimulation ● Decreased activity ● No smile	● No response to social cues ● Looks ill ● Unrousable/doesn't stay awake after rousing ● Weak, high pitched or continuous cry
Respiration	● Normal	● Nasal flare ● Tachypnoea ● respiratory rate ≥50/min (aged <1 yr) ● respiratory rate ≥40/min (aged >1 yr) ● Oxygen saturation ≤95% ● Crackles on auscultation	● Grunting/nasal flare ● Tachypnoea ● respiratory rate >60/min (any age) ● Chest wall recession (moderate/severe)
Circulation and Hydration	● Normal skin and eyes ● Moist mucous membranes	● Dry mucous membranes ● Poor feeding (infants) **Age** / **Heart rate** (bpm): <1 yr >160; 1–2 yr >150; 2–5 yr >140 ● CRT ≥3 sec ● Reduced urine output	● Reduced skin turgor
Other	● No amber/red features	● Temperature ≥39ºC (aged 3–6 months ● Fever ≥5 days ● New lump >2 cm diameter ● Swelling of joint/limb ● Not using a limb/weight bearing	● Temperature ≥38ºC (aged <3 months) ● Non-blanching rash ● Bulging fontanelle ● Neck stiffness ● Status epilepticus ● Focal neurological signs ● Focal seizures ● Bilious vomiting



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Observations

- Measure and record in **all** febrile children:
  - temperature
    aged <4 weeks: electronic thermometer in the axilla
    aged >4 weeks: infrared tympanic or electronic thermometer in the axilla
  - respiratory rate, heart rate, capillary refill time
  - signs of dehydration: skin turgor, respiratory pattern, weak pulse, cool extremities
  - travel history
- Re-assess all children with amber or red features within 1–2 hr

IMMEDIATE TREATMENT

Antipyretic treatment

- Tepid sponging not recommended
- Do not over or under dress a child with fever
- If child appears distressed or unwell, consider **either** paracetamol or ibuprofen
- Do not routinely administer both drugs at the same time with the sole aim of reducing fever or preventing febrile convulsions
- Alternate if distress persists or recurs before next dose due

Antibiotics

- Do not prescribe oral antibiotics to children with fever without apparent source
  - if aged >3 months consider admission and observation

Signs of shock

- Increased respiratory and heart rate, cold peripheries, prolonged capillary refill time, pallor/mottled, drowsy/agitated/confused
- Give immediate IV fluid bolus of sodium chloride 0.9% 20 mL/kg. Give additional boluses as necessary
- If signs of shock, SpO_2 <92% or clinically indicated, give oxygen
- Urgent senior support: discuss with PICU
- See **Septicaemia** guideline

SUBSEQUENT MANAGEMENT

- Serious bacterial infection suspected:
  - shock
  - unrousable
  - meningococcal disease
  - aged <1 month
  - aged 1–3 months with a white blood cell count <5 or >15 x 10^9/L
  - aged 1–3 months appearing unwell
- Ceftriaxone: <50 kg body weight or aged <12 yr, 50 mg/kg once daily; >50 kg or aged >12 yr, 1 g once daily
  - if ceftriaxone contraindicated (<41 weeks postmenstrual age; neonates with jaundice, hypoalbuminaemia or acidosis; or on IVI calcium) give cefotaxime (aged <1 month see BNFc for neonatal doses)
- If no evidence of bacterial sepsis, stop antibiotics 36 hr after time blood put in culture bottle
- Decreased level of consciousness: consider meningitis and herpes simplex encephalitis
  - give aciclovir: aged <3 months 20 mg/kg IV 8-hrly; aged >3 months–12 yr 500 mg/m^2; aged >12 yr 10 mg/kg IV 8-hrly
- RSV/flu: assess for serious illness/UTI
- If rates of antibacterial resistance are significant, refer to local policy
- See **Septicaemia** and **Meningitis** guidelines

Symptoms and signs of specific diseases

Meningococcal disease

- Non-blanching rash with one or more of the following:
  - ill-looking child
  - lesions >2 mm in diameter (purpura)
  - CRT ≥3 seconds
  - neck stiffness

Meningitis

- Neck stiffness
- Bulging fontanelle
- Decreased level of consciousness
- Convulsive status epilepticus

Herpes simplex encephalitis

- Focal neurological signs
- Focal seizures
- Decreased level of consciousness

Pneumonia

- Tachypnoea, measured as:
  - aged 0–5 months: respiratory rate >60 breaths/min
  - aged 6–12 months: respiratory rate >50 breaths/min
  - aged >12 months: respiratory rate >40 breaths/min
- Crackles in the chest
- Nasal flaring
- Chest indrawing
- Cyanosis
- Oxygen saturation ≤95%

Urinary tract infection

- Vomiting (in children aged >3 months)
- Poor feeding
- Lethargy
- Irritability
- Abdominal pain or tenderness
- Urinary frequency or dysuria
- Offensive urine or haematuria

Septic arthritis/osteomyelitis

- Swelling of a limb or joint
- Not using an extremity
- Non weight bearing

Kawasaki disease

- Fever lasting >5 days and at least 4 of the following:
  - bilateral conjunctival injection
  - change in upper respiratory tract mucous membranes (e.g. injected pharynx, dry cracked lips or strawberry tongue)
  - change in peripheral extremities (e.g. oedema, erythema or desquamation)
  - polymorphous rash
  - cervical lymphadenopathy

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RECOGNITION AND ASSESSMENT

Fever

- Type of thermometer used, site, user (factitious)
- Duration, height
- Pattern:
  - intermittent [pyogenic, TB, lymphoma, juvenile idiopathic arthritis (JIA)]
  - baseline raised (viral, endocarditis, lymphoma)
  - sustained (typhoid)
  - days between (malaria, lymphoma)
  - weeks between (metabolic, CNS, cyclic neutropenia, hyperIgD)
- Circumstances when fever (e.g. exercise)
- Appearance
  - when fever: well (factitious)
  - between fever: ill (serious)
- Response to paracetamol and or NSAID (no response: dysautonomia)

Symptoms

- Red eyes (Kawasaki)
- Nasal discharge (sinusitis)
- Recurrent pharyngitis with ulcers (periodic fever)
- GI: salmonella, intra-abdominal abscess, inflammatory bowel disease (IBD)
- Limb pain (leukaemia, osteomyelitis)

Contact

- Human illness
- Animals

Travel

- Years ago (histoplasmosis)
- Part of country
- Prophylaxis and immunisations
- Contaminated water/food
- Bites (tick: arbovirus, malaria)
- Meat: undercooked (brucella, toxoplasma, hepatitis)
- Pica (visceral larva migrans, toxoplasmosis)

Medical history

- Operations

Drug history

- All, including any non-prescription

Ethnic group

- Sephardic Jew, Armenian, Turkish, Arab (Familial Mediterranean Fever)
- Ashkenazi Jew (familial dysautonomia)

Examination

- Sinuses
- Lymph nodes
- Chest: murmur, crackles
- Abdominal: hepato/spleno-megally (salmonella, cat scratch, endocarditis, malaria)
- Genito-urinary: girls – pelvic tenderness (child sex abuse – STI)

Skin

- Rash only during fever (JIA)
- No sweat (familial dysautonomia)
- Petechiae (endocarditis, rickettsia)
- Papules (cat scratch)
- Eschar (tularaemia)
- Erythema migrans (Lyme)
- Malar (SLE)
- Palpable purpura [polyarteritis nodosa (PAN)]
- Erythema nodosum (JIA, SLE, malignancy, IBD, TB)
- Seborrheic (histiocytosis)
- Sparse hair (ectodermal dysplasia)
- Scars (dysautonomia)

Eyes

- Conjunctivitis:
  - palpebral (infectious mononucleosis)
  - bulbar (Kawasaki)
  - phlyctenular (TB)
- Retinopathy (PAN, miliary TB, toxoplasmosis, vasculitis)
- Pupil dilation (hypothalamic or autonomic dysfunction)

Oropharynx

- Red, no exudates (EBV)
- Dental abscess
- Conical teeth (ectodermal dysplasia)
- Smooth tongue (dysautonomia)
- Gum hypertrophy, tooth loss (leukaemia, histiocytosis)

Musculoskeletal

- Tender:
  - bone (osteomyelitis, malignancy)
  - muscle (trichinella, arbovirus, dermatomyositis, PAN)
- Trapezius (subdiaphragmatic abscess)
- Reflexes
  - brisk (hyperthyroid)
  - absent (dysautonomia)

Investigations

All

- FBC, ESR, CRP, U&E, LFT, blood culture, HIV antibody, urinalysis, urine culture, CXR

- FBC:
  - low Hb (malaria, endocarditis, IBD, SLE, TB)
  - high platelets (Kawasaki)
  - blasts (leukaemia)
  - eosinophils (fungal, parasites, neoplastic, allergic, immune deficiency)
- ESR/CRP: normal (factitious, dysautonomia, drug fever)
- LFTs: abnormal (EBV, CMV)
- Blood cultures: several times (endocarditis)
- Urine: pyuria (Kawasaki, intra-abdominal infection, GU, TB)

Selective

- Stool (if loose)
- Bone marrow (leukaemia, histiocytic haemophagocytosis)
- Serology (syphilis, brucella, EBV, CMV, toxoplasma, bartonella)
- Auto-antibodies (rheumatoid arthritis, SLE)
- IgG, A & M (recurrent infections)
- IgE (allergy, eosinophilia)
- IgD (periodic fever)

Imaging (selective)

- Sinuses
- US/CT/MR abdo (IBD, abscess, lymphadenopathy)
- White cell scan (abscess)
- Bone scan (osteomyelitis)
- PET scan (abscess)

Other investigations (selective)

- Echo (endocarditis)
- Ophthalmologist (uveitis, leukaemia)
- Biopsy (lymph node, liver)

EMPIRICAL TREATMENT

- Critically ill: no focus – ceftriaxone or cefotaxime (after blood and urine specimens taken)
- TB treatment: after induced sputum, lymph node biopsy, TB blood culture
- Otherwise avoid antibiotics until organism isolated

REFERRAL

- Rheumatology (JIA, connective tissue disorder)
- Gastroenterology (IBD)
- Cardiology (endocarditis/Kawasaki)

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Discuss all children with hepatitis B or C with infectious diseases team or regional liver unit for counselling, information, consideration for anti-viral therapy and need for referral

HEPATITIS B

Diagnostic tests

- HBsAg (Hepatitis B surface antigen) and HBcAb (IgM and IgG)
- HBsAb (anti-HBs: antibody) indicates previous immunisation or infection
- If HBsAg positive then check HBeAg, HBeAb, genotype and refer to regional liver unit

Yearly follow-up

- Clinical assessment
- Serology (clotted specimen)
  - HBsAg
  - if previously HBeAg positive, HBeAg
  - HBeAb
- Hepatitis B DNA PCR viral load (EDTA)
- LFT (bilirubin, ALT/AST, GGT, albumin)
- FBC
- Coagulation (INR, PT, PTT)
- Alpha-fetoprotein
- Abdominal ultrasound and fibroscan if available (yearly if eAg +ve; 5 yearly if eAb +ve or if rise in alpha-fetoprotein)

Action

- If LFT or alpha-fetoprotein abnormal, or viral titres are rising, inform regional liver unit

HEPATITIS C

Diagnostic tests

(For neonates see **Neonatal guidelines**)

- Hepatitis C Virus (HCV) antibody (ab) aged >18 months old
- HCV PCR if HCV ab +ve

Action

- If HCV ab negative not infected. Discharge
- If HCV ab positive and HCV PCR negative in two samples taken 6 months apart, not infected (resolved infection or maternal antibody if aged <18 months). Discharge
- If HCV PCR positive, check genotype and yearly bloods below, refer to regional liver unit

Yearly follow-up

- Clinical assessment
- HCV PCR viral blood
- LFT (bilirubin, ALT/AST, GGT, albumin)
- FBC
- Coagulation (INR, PT, PTT)
- Alpha-fetoprotein
- Abdominal ultrasound (and fibroscan if available)

HIV AND HEPATITIS B POST-EXPOSURE PROPHYLAXIS (PEP) • 1/2

INDICATIONS FOR PEP

	Source HIV status			
	HIV positive Viral load detectable	**HIV positive Viral load undetectable**	**Unknown high prevalence group/area**	**Unknown low prevalence group/area**
Receptive anal sex	Recommended	Recommended	Recommended	Not recommended
Insertive anal sex	Recommended	Not recommended	Consider	Not recommended
Receptive vaginal sex	Recommended	Not recommended	Consider	Not recommended
Insertive vaginal sex	Recommended	Not recommended	Consider	Not recommended
Fellatio with ejaculation	Consider	Not recommended	Not recommended	Not recommended
Fellatio without ejaculation	Not recommended	Not recommended	Not recommended	Not recommended
Splash of semen into eye	Consider	Not recommended	Not recommended	Not recommended
Cunnilingus	Not recommended	Not recommended	Not recommended	Not recommended
Sharing of injective equipment	Recommended	Not recommended	Consider	Not recommended
Human bite	Not recommended	Not recommended	Not recommended	Not recommended
Needle-stick from a discarded needle in the community			Not recommended	Not recommended

Consider: recommend PEP if additional high risk factor for HIV

PEP

- <40 kg: zidovudine, lamivudine and Kaletra®
- ≥40 kg: Truvada® 1 tab daily and Kaletra® 2 x (200/50 tab) 12-hrly
- If source has drug-resistant virus, seek expert help
- If patient known to have HIV do not give PEP
- Start as soon as possible up to 72 hr after exposure

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HIV AND HEPATITIS B POST-EXPOSURE PROPHYLAXIS (PEP) • 2/2

HIV PEP drugs

Drug	Dose	Formulation	Side effects	Intake recommendation
Zidovudine (ZDV or AZT)	180 mg/m^2/dose (max 250 mg) 12-hrly	Caps. 100, 250 mg Susp. 10 mg/mL	Neutropenia +/- anaemia, nausea, headache, hepatitis myopathy, neuropathy	Can be given with food; capsules can be opened and dissolved in water
Lamivudine (3TC)	4 mg/kg/dose 12-hrly; max dose 150 mg 12-hrly	Tab. 100, 150 mg; Susp. 10 mg/mL; 5 mg/mL (room temp)	Peripheral neuropathy, nausea, diarrhoea, headache	Can be given with food
Truvada® (TDF and FTC)	>40 kg 1 tab daily	Tab. 300 mg Tenofovir (TDF) 200 mg Emticitabine (FTC)	Headache, diarrhoea, nausea, renal tubular dysfunction	Can be given with or without food
Kaletra® [lopinavir (LPV)/ ritonavir (RTV)]	300 mg LPV/m^2 + 75 mg RTV/m^2 12-hrly 15–25 kg: 2 x 100/25 tab 12-hrly 25–35 kg: 3 x 100/25 tab 12-hrly >35 kg: 2 x 200/50 tab 12-hrly	Tab 200 mg LPV/ 50 mg RTV Paed tab 100 mg LPV/25 mg RTV Liq 5 mL = 400 mg LPV/100 mg RTV	Diarrhoea, headache, nausea, vomiting Caution in liver disease	Give with or after food

HEPATITIS PEP

If HIV PEP indicated give

- Hepatitis B vaccine accelerated course (0, 1, 2 and 12 months)
- Hepatitis B immunoglobulin only if source known to be HBsAg +ve

FOLLOW-UP

- Before discharge, provide families embarking on HIV PEP with:
  - appointment to see a paediatrician with experience in antiretroviral drugs or member of ID/GUM team the same day or next working day
  - contact telephone number in case of concerns about any aspect of HIV PEP
  - enough antiretroviral medication to last until clinic appointment
  - letter for GP
- After sexual exposure consider emergency contraception and screen for other sexually transmitted infections
- Arrange HBV, HCV and HIV antibody test baseline and 3 months after exposure
- Check FBC, U&Es and LFTs if starting PEP
- Check need for tetanus immunisation
- If source is HCV RNA PCR positive, arrange the following enhanced HCV follow-up:
  - at 6 weeks: EDTA blood for HCV PCR
  - at 12 weeks: EDTA blood for HCV PCR and clotted blood for anti-HCV antibodies
  - at 24 weeks: clotted blood for anti-HCV antibodies

HIV TESTING • 1/2

INTRODUCTION

- HIV is a treatable medical condition
- The majority of those living with the virus are well
- Many are unaware of their HIV infection
- Late diagnosis is life-threatening
- HIV testing can be done in any medical setting and health professionals can obtain informed consent for an HIV test in the same way they do for any other medical investigation

HOW

Who can test?

- Doctor, nurse, midwife or trained healthcare worker

Who should be offered a test?

- First-line investigation for suspected immune deficiency: unusual type, severity or frequency of infection. See **Table 1**
- Take a sexual history in post-pubertal children

Primary HIV infection

- Symptoms typically occur 2–4 weeks after infection:
  - fever
  - rash (maculopapular)
  - myalgia
  - pharyngitis
  - headache/aseptic meningitis
- Resolve spontaneously within 2–3 weeks

Source patient in a needlestick injury or other HIV risk exposure

- Consent must be obtained from source patient before testing
- The person obtaining consent must be a healthcare worker, other than person who sustained the injury

Pre-test discussion with parents and children able to give consent

- Purpose of pre-test discussion is to establish informed consent and should cover:
  - benefits of testing
  - details of how result will be disclosed
- Lengthy pre-test HIV counselling is not a requirement
- Document patient's consent to testing
- If patient refuses test, explore why and ensure decision has not resulted from incorrect beliefs about the virus or consequences of testing
- Advise that, if negative, testing will not affect patient's insurance
- Some patients, (e.g. those whose first language is not English) may need additional help to reach a decision
- Document the offer of an HIV test in medical notes, together with any relevant discussion and reasons for refusal
- Written consent not necessary but **record on laboratory request form that consent has been obtained**
- Arrange appointment for result to be disclosed personally by testing clinician

POST-TEST

HIV negative result: post-test discussion

- If still within window period after a specific exposure, discuss need to repeat test
- for definitive exclusion of HIV infection a further test after three months is recommended
- If reported as reactive or equivocal, refer to infectious diseases (may be seroconversion)

HIV positive result: post-test discussion

- For all new HIV positive diagnoses, carry out appropriate confirmatory assays and test a second sample
- Testing clinician must give result personally to patient in a confidential environment and in a clear and direct manner
- arrange follow-up programme with infectious diseases before informing patient of positive result

Table 1: Clinical indicator diseases for HIV infection

	AIDS-defining conditions	Others where testing should be offered
Respiratory	Pneumocystis pneumonia Tuberculosis	Bacterial pneumonia Aspergillosis
Neurology	Cerebral toxoplasmosis Primary cerebral lymphoma Cryptococcal meningitis Progressive multifocal leucoencephalopathy	Aseptic meningitis Space occupying lesion of unknown cause Guillain-Barré syndrome Transverse myelitis Peripheral neuropathy Dementia Leucoencephalopathy
Dermatology	Kaposi's sarcoma	Severe/recalcitrant seborrhoeic dermatitis/ psoriasis Multidermatomal or recurrent herpes zoster
Gastroenterology	Persistent cryptosporidiosis	Persistent/recurrent oral candidiasis Oral hairy leukoplakia Chronic diarrhoea/weight loss of unknown cause Salmonella, shigella or campylobacter Hepatitis B/C infection
Oncology	Non-Hodgkin's lymphoma	Hodgkin's lymphoma Castleman's disease
Haematology		Any unexplained blood dyscrasia
Ophthalmology	Cytomegalovirus retinitis	Infective retinal diseases
ENT		Lymphadenopathy of unknown cause Chronic parotitis Lymphoepithelial parotid cysts
Other		Mononucleosis-like syndrome Pyrexia of unknown origin Anyone with a mother who is HIV +ve no matter what age Anyone who has a partner who is HIV +ve Men who have sex with other men Female sexual contacts of men who have sex with men Patients reporting use of injecting drugs Anyone from a country of HIV prevalence >1% Anyone who has had sex in a country of HIV prevalence >1% Anyone who has had sex with someone from a country of HIV prevalence >1% All pregnant women

IMMUNODEFICIENCY • 1/2

RECOGNITION AND ASSESSMENT

- SPUR to recognition: Serious, Persistent, Unusual, or Recurrent infections
- The younger the onset, the more life-threatening the immune defect likely to be
- bacterial infection; early presentation: antibody defect
- viral/fungal infection; later presentation: cellular defect

Warning signs of primary immunodeficiency:

- ≥8 new bacterial ear infections
- ≥2 serious sinus infections
- ≥2 months on antibiotics without resolution of symptoms
- ≥2 episodes of pneumonia
- Failure to thrive with prolonged or recurrent diarrhoea
- Recurrent, deep skin or organ abscess
- Persistent candida in mouth or napkin area
- Failure of IV antibiotics to clear infections
- ≥2 severe infections (e.g. meningitis, osteomyelitis, cellulitis or sepsis)
- Family history of primary immunodeficiency

Symptoms of immune deficiency

- Delayed umbilical cord separation of ≥3 weeks, omphalitis
- Delayed shedding of primary teeth
- Severe adverse reaction to immunisation e.g. BCGitis
- Unusually severe course of measles or chickenpox
- Family history of any syndrome associated with immunodeficiency, (e.g. DiGeorge anomaly or Wiskott-Aldrich syndrome); or of death during early childhood
- High risk group for HIV and no antenatal HIV test (a –ve antenatal HIV test does not exclude HIV in the child)
- Autoimmune liver disease, diabetes, vasculitis, ITP
- Poor wound healing
- Unexplained bronchiectasis or pneumatoceles
- >1 unexpected fracture

Signs of immune deficiency

- Congenital abnormalities: dysmorphic features, congenital heart disease, situs inversus, white forelock, albinism, microcephaly
- Children who appear chronically ill
- Scarring or perforation of tympanic membranes from frequent infection
- Periodontitis
- Enlargement of liver and spleen
- Hypoplastic tonsils and small lymph nodes
- Lymphadenopathy
- Skin: telangiectasia, severe eczema, erythroderma, granuloma, acneiform rash, molluscum, zoster
- Ataxia

Other investigations suggestive of immune deficiency

- Haemolytic anaemia
- Neutropenia
- Eosinophilia
- Hypocalcaemia

Unusual organisms suggestive of immune deficiency

- Viruses: CMV, EBV, VZV, warts
- Fungi: candida, aspergillus, cryptococcus, pneumocystis, nocardia
- Protozoa: cryptosporidium, toxoplasma
- Bacteria: salmonella, giardia, mycobacterium (inc BCG), serratia
- Recurrent infection with common organisms: *H. influenzae, S. pneumoniae, N. meningitidis, S. aureus*

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Table 1: Investigations

Investigations	Sample	Volume	
		Minimum	Ideal
Initial tests (complete all tests for any suspected immune deficiency)			
FBC (note absolute lymphocyte count) and ESR	EDTA	1.3 mL	4 mL
IgG, IgM, IgA	Clotted	0.5 mL	4 mL
IgG function (antibody response to tetanus, Hib +/- pneumococcus) Retest 4 weeks after vaccination	Clotted	0.5 mL	4 mL
HIV antibody	Clotted	0.5 mL	4 mL
Second-line tests (with immunology advice)			
Lymphocyte subsets	EDTA	1 mL	4 mL
		Haematology	
Lymphocyte proliferation	Lithium heparin	Discuss with local immunology centre	
Enzyme assay (ADA, PNP)	EDTA	3.5 mL	1 mL
		Discuss with Guy's Hospital	
Neutrophil function test for CGD	EDTA or lithium heparin	0.25 mL	4 mL
		Discuss with local immunology centre	
Adhesion molecule assay	EDTA	0.25 mL	4 mL
		Discuss with local immunology centre	
CH50 & Complement components (if recurrent or case with family history of meningococcal disease)	Clotted	1 mL to reach lab within 2 hr	4 mL to reach lab within 2 hr or separate and freeze immediately

SUBSEQUENT MANAGEMENT

- Avoid live vaccines (e.g. BCG, MMR and varicella)
- Ensure that any blood products given to patients with suspected or proven T-cell immunodeficiency are irradiated and CMV negative
- For specific infections, use same antibiotics as in immunocompetent patients, at higher recommended dosage
- Obtain throat, blood and other culture specimens before starting treatment
- Treat infectious episodes for longer than usually recommended (approximately double)
- In patients with B-cell, T-cell or phagocytic defects, request regular pulmonary function tests and home treatment plan of physiotherapy and inhalation therapy similar to that used in cystic fibrosis
- In children with significant primary or secondary cellular (T-cell) immunodeficiency (e.g. aged <1 yr CD4 <25%, aged 1–5 yr CD4 <15% or aged >5 yr <200 CD4 cells/mm^3), give *Pneumocystis jiroveci* (PCP) prophylaxis with co-trimoxazole

Early treatment reduces mortality from coronary artery aneurysms

RECOGNITION AND ASSESSMENT

Symptoms and signs

- Fever for at least 5 days and 4 of the following:
  - bilateral non-exudative conjunctival infection
  - oral changes (red lips/pharynx/tongue)
  - peripheral oedema followed by desquamation 10–15 days after onset of fever
  - polymorphous rash
  - acutely enlarged cervical lymph nodes with individual node(s) >1.5 cm diameter
  - absence of another diagnosis e.g. group A streptococcal infection (GAS), measles
- The presence of a coronary artery aneurysm with any one of the above features is diagnostic

Other features

- Most common in children aged <5 yr
- Atypical cases may not fulfil all the above criteria
- Fever usually precedes the other signs and is characteristically unresponsive to antipyretics
- Other symptoms include irritability, aseptic meningitis, uveitis, cough, vomiting, diarrhoea, abdominal pain, urethritis, arthralgia and arthritis

Investigations

None is diagnostic

- FBC: platelets often high for 10–15 days after onset of fever
- ESR
- LFT's, CRP
- ECG
- If full diagnostic criteria found, echocardiogram **not** required until 6 weeks from onset of signs and symptoms
- If criteria incomplete or presentation atypical, aneurysms on echo are diagnostic: discuss with cardiologist
- Throat swab for Gp A strep
- Anti-streptolysin O titre (ASOT) or anti-DNase B for evidence of streptococcal infection
- Blood culture
- Urinalysis, microscopy and culture
- If rash present, serology for enterovirus, parvovirus, EBV, CMV; if features of measles urine or throat swab in viral transport medium for PCR

IMMEDIATE TREATMENT

- Aspirin 7.5–12.5 mg/kg oral 6-hrly until afebrile or a minimum of 2 weeks
- Intravenous immunoglobulin (IVIG) 2 g/kg
  - check concentration (g/mL) for preparation used in your Trust

Administration of 100 mg/mL (e.g. Flebogamma® DIF)

Rate*	Duration
0.6 mL/kg/hr = ...mL/hr	30 min
1.2 mL/kg/hr = ...mL/hr	30 min
2.4 mL/kg/hr = ...mL/hr	30 min
3.6 mL/kg/hr = ...mL/hr	30 min
4.8 mL/kg/hr = ...mL/hr	To completion

*** up to a maximum rate of 180 mL/hr**

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Product	Infusion rates		Infusion time of 70 g in minutes at maximum rate
	Initial	Maximum	
Baxter Kiovig	0.5 mL/kg/hr for 30 min	6 mL/kg/hr (8 mL/kg/hr in PID)	100
BPL Gammaplex	0.01–0.02 mL/kg/min for 15 min	0.04–0.08 mL/kg/min	250
BPL Vigam	0.01–0.02 mL/kg/min for 30 min	0.04 mL/kg/min (max 3 mL/min)	500
Biotest Intratect	1.4 mL/kg/hr for 30 min	1.9 mL/kg/hr	640
CSL Privigen	0.3 mL/kg/hr	4.8 mL/kg/hr (7.2 mL/kg/hr in PID)	125
Grifols Flebogamma 5	0.01–0.02 mL/kg/min for 30 min	0.1 mL/kg/min	200
Grifols Flebogamma 10	0.01 mL/kg/min for 30 min	0.08 mL/kg/min	125
Octapharma Octagam 5	1 mL/kg/hr for 30 min	5 mL/kg/hr	241
Octapharma Octagam 10	0.01–0.02 mL/kg/min for 30 min	0.12 mL/kg/min	83

Start IVIG as soon as possible (delayed treatment increases risk of aneurysm)

MONITORING IVIG INFUSION

- Monitor temperature, heart rate, BP and respiratory rate:
  - every 5 min for first 15 min
  - then every 15 min for first hour
- Anticipate anaphylaxis, flushing, fever, headache, shivering
- If tolerated, increase infusion rate to give total dose over remaining 10 hr and monitor hourly
- If mild reaction, stop infusion for 15 min then restart at slower rate

SUBSEQUENT MANAGEMENT

- If fever persists 36 hr after completion of IVIG, consider a single repeat dose of IVIG
- If fever persists after second dose IVIG give intravenous pulse methylprednisolone, 30 mg/kg over 2–3 hr, administered once daily for 1–3 days with BP monitoring 4-hrly
- After 2 weeks, reduce dose of aspirin to 5 mg/kg oral as single daily dose for 6 weeks (until result of echocardiogram known)

DISCHARGE AND FOLLOW-UP

- Discharge when fever settles
- Echocardiogram at 6 weeks from onset of signs and symptoms
- Out-patient appointment 1 week after echocardiogram
- Advise to avoid excessive strenuous activity until out-patient appointment after echocardiogram
- Advise to avoid all live vaccines (e.g. MMR) for 3 months following IVIG therapy

OUT-PATIENT MANAGEMENT

- No aneurysms at 6 weeks echocardiogram
  - stop aspirin
  - no restriction on activity
- Single aneurysm <7 mm diameter
  - aspirin 3–5 mg/kg (max 75 mg) once daily until aneurysm disappears
  - cardiologist will advise on limitation of activity
  - annual echocardiogram
- Multiple or giant aneurysm
  - avoid strenuous activity
  - discuss need for anticoagulation, stress test and repeat echocardiogram with cardiologist

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Falciparum is a medical emergency: immediate treatment is essential

- Test for malaria in anyone with fever
- who has travelled to a malarial area within last 12 months
- or febrile infant whose mother has travelled to a malarial area in pregnancy

Clinical features

Non-specific	Severe (complicated) malaria
● Fever	● Persistent vomiting, severe dehydration
● Malaise	● Shock, renal failure (oliguria <0.5 mL/kg/hr)
● Headache	● Depressed conscious state, seizures
● Sweating	● Tachypnoea or increased work of breathing
● Diarrhoea	● Hypoxia (SpO_2 <95%)
● Vomiting	● Metabolic acidosis (base deficit >8)
● Abdominal pain	● Severe hyperkalaemia (K >5.5 mmol/L)
● Splenomegaly	● Hypoglycaemia <3 mmol/L
● Anaemia	● Severe anaemia (<80 g/L)
● Thrombocytopenia	● Unable to walk
● Jaundice	● Parasitaemia >2% or schizonts on film

Investigations

- EDTA blood sample sent to haematology for an urgent thick blood film
- 3 blood films 12 hr apart
- Negative malaria ICT (stix test) does not exclude malaria
- Do not treat unless proven on blood test
- Admit all patients with falciparum to a unit with experience in managing severe malaria (e.g. infectious disease unit)
- Opportunistic screen for other imported diseases: hepatitis B, HIV

If malaria is diagnosed on blood film, but type unclear, treat as falciparum malaria

SEVERE (COMPLICATED) MALARIA

Anti-malaria treatment

- Quinine dihydrochloride IV diluted to 2 mg/mL with sodium chloride 0.9% or glucose 5%
- loading dose 20 mg/kg max 1.4 g as infusion over 4 hr (NEVER as IV bolus)
- omit loading dose if mefloquine or quinine used in previous 24 hr
- glucose stix 2-hrly during IV quinine, cardiac monitor and daily ECG (check QTc)
- then 8 hr after start of loading dose, 10 mg/kg infusion (max 700 mg) over 4 hr every 8 hr
- when able to swallow give Malarone® (see **Treatment of uncomplicated falciparum malaria** below)
- daily FBC, U&E and blood films as in-patient until asexual parasites undetectable

- If parasitaemia >15% or from area of quinine resistance (Thai/Cambodia border, Papua New Guinea) or history of arrhythmias, discuss with ID specialist about artesunate instead of quinine
- Artesunate 2.4 mg/kg IV [in 1 mL sodium bicarbonate (vial provided with drug), dilute further in 5 mL glucose 5% and inject over approximately 2 min] at 0, 12 and 24 hrs and then daily
- When parasitaemia resolving and patient improving, switch to oral agent:
- Malarone®, or if resistance suspected Riamet®, or oral quinine (if neither other agent available)

Complications

- Renal failure: discuss early filtration/dialysis with PICU
- Hypovolaemia: cautious rehydration (high risk pulmonary oedema)
- Shock: add cefotaxime
- Hypoglycaemia: common, give glucose 10% 2 mL/kg IV bolus then glucose 10% 5 mL/kg/hr with sodium chloride 0.45%
- Anaemia: common, transfuse if Hb <80 g/L
- Thrombocytopenia: expected, transfuse only if bleeding and platelets <20 x 10^9/L

CEREBRAL MALARIA

Impaired level of consciousness

- Correct hypoglycaemia
- Monitor GCS, reflexes, pupils
- Plan for intubation and transfer to PICU if:
- signs of raised ICP
- persisting shock after 40 mL/kg fluid
- or pulmonary oedema

TREATMENT OF UNCOMPLICATED FALCIPARUM MALARIA (no clinical features of severe malaria)

- If child can tolerate oral intake (can be crushed):
- **Malarone®** (proguanil with atovaquone) once a day for 3 days

Weight (kg)	5–8	9–10	11–20	21–30	31–40	>40
Dose	2 paed tablets	3 paed tablets	1 adult tablet	2 adult tablets	3 adult tablets	4 adult tablets

Paediatric tablet contains proguanil 25 mg and adult tablet 100 mg

No second agent required

Or

Riamet® (artemether with lumefantrine)

Weight (kg)	Dose
5–15	1 tablet initially followed by 5 further doses of 1 tablet each given at 8, 24, 36, 48 and 60 hrs (total 6 tablets over 60 hr)
16–25	2 tablets initially followed by 5 further doses of 2 tablets each given at 8, 24, 36, 48 and 60 hrs (total 12 tablets over 60 hr)
26–35	3 tablets initially followed by 5 further doses of 3 tablets each given at 8, 24, 36, 48 and 60 hrs (total 18 tablets over 60 hr)
>35 (12–18 yr)	4 tablets initially followed by 5 further doses of 4 tablets each given at 8, 24, 36, 48 and 60 hrs (total 24 tablets over 60 hr)

No second agent required

Or

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Quinine sulphate

- 10 mg/kg (max 600 mg) oral 8-hrly
- Reduce to a 12-hrly regimen if severe cinchonism (severe tinnitus, deafness, unsteadiness)
- Mild tinnitus and feeling of 'blocked' ears are expected on quinine and resolve once therapy completed
- Continue until blood films negative or for a 7 day course (whichever is the longer). A shorter course may be possible but only at infectious diseases consultant's discretion

Weight (kg)	Paediatric dosing of oral quinine sulphate
5–7	50 mg ($^1/_4$ x 200 mg tablet)
8–12	100 mg ($^1/_2$ x 200 mg tablet)
13–17	150 mg ($^3/_4$ x 200 mg tablet)
18–22	200 mg (1 x 200 mg tablet)
23–27	250 mg ($^1/_2$ x 300 mg + $^1/_2$ x 200 mg tablet)
28–39	300 mg (1 x 300 mg tablet)
40–49	400 mg (2 x 200 mg tablet)
50–60	500 mg (1 x 200 mg tablet and 1 x 300 mg tablet)
>60	600 mg (2 x 300 mg tablet)

- With quinine give second agent
- aged >12 yr doxycycline 200 mg once/day for 7 days
- aged <12 yr clindamycin 7–13 mg/kg (max 450 mg) 8-hrly for 7 days

If in doubt treat as severe (complicated) malaria

NON-FALCIPARUM MALARIA

- Complications are rare
- Usually sensitive to chloroquine (chloroquine-resistant *P. vivax* reported in Indonesia, New Guinea and some adjacent islands)

Treatment of non-falciparum malaria

- If chloroquine resistance suspected then refer to non-complicated falciparum management
- Chloroquine 10 mg (base)/kg oral initial dose (max 620 mg)
- then 5 mg/kg (max 310 mg) after 6 hr, then once daily for 2 days
- Then give primaquine 250 microgram/kg oral (max 15 mg) daily for *P. ovale* and 500 microgram/kg (max 30 mg) daily for *P. vivax* for 14 days
- Liquid nivaquine 68 mg/5 mL is equivalent to 50 mg/5 mL chloroquine base
- Itch is common, does not respond to antihistamines, if severe give quinine

Before giving primaquine, check and review G6PD concentration, as severe haemolysis can occur if G6PD-deficient

G6PD-deficient patients

- In mild G6PD-deficiency, primaquine 750 microgram/kg (max 30 mg) once a week for 8 weeks
- Otherwise contact ID specialist

RECOGNITION AND ASSESSMENT

If aged <28-days-old see **Neonatal meningitis** in **Neonatal Guidelines**

Symptoms may be non-specific: if meningitis considered, LP

Symptoms and signs

- Pyrexia
- Petechial rash
- Evidence of raised intracranial pressure in the older child
  - disc oedema (often late sign), any localising neurological features, reduced conscious level
- Neck stiffness
- Kernig's sign positive
- Irritability
- Focal neurological signs including squints
- Infants:
  - poor feeding
  - vomiting
  - irritability
  - fever
  - fits
  - full fontanelle (unless dehydrated)
- Older child may also have:
  - severe headache
  - photophobia
  - confusion
  - lower backache

Differential diagnosis

- If rash or severely ill, see **Septicaemia (including meningococcal)** guideline
- Look for signs of viral meningitis e.g. resolving mumps
- It is not possible to differentiate viral from bacterial meningitis clinically
- Other intracranial sepsis
- Encephalitis
- Systemic sepsis
- Malaria in travellers
- Other causes of confusion or raised intracranial pressure

INVESTIGATIONS

Lumbar puncture

If any doubt about need for lumbar puncture (LP) discuss with consultant

- Perform LP before giving antibiotics if child stable, do not delay antibiotics by >1 hr
- Discuss with consultant if any of following:
  - signs suggesting raised intracranial pressure
    - GCS <9 or drop ≥3
    - relative bradycardia and hypertension
    - focal neurological signs
    - abnormal posture or posturing
    - unequal, dilated or poorly responsive pupils
    - papilloedema
    - abnormal 'doll's eye' movements
  - shock
  - extensive or spreading purpura
  - after convulsion until stabilised
  - coagulopathy: on anticoagulants or if already obtained platelets <100 x 10^9/L or INR >1.4 or suspected (e.g. purpuric rash)
  - local infection over lumbar spine
  - respiratory insufficiency
- If LP initially contraindicated, perform LP as soon as no longer contraindicated to confirm diagnosis

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- Repeat LP if:
  - no clinical response after 48 hr of therapy
  - re-emergent fever
  - deterioration
  - persistent abnormal inflammatory markers

Specimens

- One fluoride tube (and 4 CSF bottles)
- If tap traumatic, may need more samples
- If insufficient CSF discuss priorities with microbiology

Table 1: Collection of specimens (stated volumes represent minimum required)

Department	Specimens (6 drops = approx 0.2 mL)
Biochemistry	● 0.2 mL in a fluoride tube for glucose (also send blood glucose) ● 0.2 mL in a CSF bottle for protein ● 0.2 mL for lactate if metabolic disorder suspected
Microbiology	● 0.2 mL in a CSF bottle for MC&S ● 0.5 mL for meningococcal and pneumococcal PCR ● 1 mL for TB culture if high clinical suspicion of TB meningitis
Virology	If possible viral meningitis or encephalitis: ● 0.5 mL for herpes simplex virus, enterovirus and VZV PCR ● 0.3 mL for Human Herpes Virus 6 if rash, high temperature or rapid recovery
Save	● 0.5 mL plain bottle for additional neurology tests (e.g. oligoclonal bands) depending on other results and progress

Interpretation of cerebrospinal fluid results

- White cell count showing polymorphonuclear leucocytosis usually indicative of bacterial meningitis but can also be seen in viral meningitis
  - lymphocytosis can occur in partially-treated pyogenic, TB and viral meningitis and inflammatory conditions
  - in neonates with no specific signs of meningitis up to 20 cells/microlitre may be normal (but treat as meningitis if symptoms suggestive of meningitis e.g. fitting if any neutrophils)
  - older children treat as meningitis if >5 white cells or >1 neutrophil/microlitre
- Protein usually elevated in bacterial meningitis (upper limit of normal is 20 mg/L in infants and children, and up to 60 mg/L in neonates)
  - very high levels of CSF protein are sometimes seen in TB meningitis
- CSF glucose normally about 1 mmol/L below serum level
  - CSF glucose likely to be depressed in bacterial meningitis including TB (lymphocytosis) or
  - herpes or mumps encephalitis
  - CSF glucose usually normal in viral meningitis

CT scan

- Not routine, does not exclude raised intracranial pressure
  - indicated if GCS <9 or focal neurological signs: if no space occupying lesion or sign of raised intracranial pressure then LP if no other contraindication
  - do not delay treatment for CT
  - stabilise before, and monitor closely during CT

Other

- FBC and differential WBC
- Blood cultures before start of antibiotics

- U&E, glucose and CRP
- If meningococcal disease suspected, refer to **Septicaemia (including meningococcal)** guideline; send EDTA blood for meningococcal DNA PCR
- Viral titres plus mycoplasma IgM if indicated or store to assay if other results negative
- Stool or rectal swab and throat swab for enteroviruses, if viral meningitis suspected

IMMEDIATE TREATMENT

Corticosteroids

- Give in children aged >3 months with:
  - frankly purulent CSF
  - CSF WBC count >1000/microlitre
  - raised CSF WBC count and protein greater than 1 g/L
  - bacteria on Gram stain
- Do not give in septic shock, meningococcal disease, immunocompromised patients or if post-operative
- If TB meningitis suspected, discuss with infectious diseases team before giving
- Dexamethasone sodium phosphate 150 microgram/kg (max 10 mg) IV 6-hrly for 4 days
  - first dose before antibiotics or as soon as possible up to 12 hr afterwards

Antibiotics

Start immediately without waiting for identification of organisms or sensitivities

- Ceftriaxone or cefotaxime
  - infants aged <3 months, add amoxicillin to cover listeriosis
  - infants aged <3 months, ceftriaxone may be used but not in premature babies or in babies with jaundice, hypoalbuminaemia or acidosis
  - if suggestive of herpes encephalitis (period of decreased level of consciousness with fever or focal signs) add aciclovir IV 8-hrly: aged <3 months 20 mg/kg; aged 3 months–12 yr 500 mg/m^2; aged >12 yr 10 mg/kg
- If suggestive of TB (contact with TB, other features of TB, long history), discuss with infectious diseases team
- If recent travel outside UK, or prolonged or multiple antibiotics in last 3 months add vancomycin
- If definite history of anaphylaxis to cephalosporin give chloramphenicol IV or vancomycin/teicoplanin and gentamicin if chloramphenicol not immediately available
- If unusual cause suspected, contact infectious diseases team/microbiologist

Anticonvulsants

- Drugs of choice if child has seizures (prophylaxis not recommended):
  - phenytoin
  - lorazepam for acute control

Other supportive measures

- If child shocked, give human albumin 4.5% or if not immediately available sodium chloride 0.9%:
  - initial dose 20 mL/kg over 5–10 min and reassess

Do not give excessive fluid boluses: risk of cerebral oedema

Intensive care

- Inform PICU if:
  - depressed conscious level
  - shock does not respond to initial resuscitation

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MONITORING TREATMENT

- In a semi-conscious patient, monitor hourly until improvement evident:
  - respiratory rate
  - pulse and BP
  - level of consciousness and pupils
  - in young infants, measure head circumference daily
- If persistent pyrexia look for other foci
  - repeat blood cultures and other investigations according to signs
  - CT scan at 10 days for microabscess or hypodensity
  - if CT normal, repeat LP

SUBSEQUENT MANAGEMENT

Length of antibiotic course

- Meningococcus: 7 days
- *Haemophilus influenzae*: 10 days
- Pneumococcus or Group B Streptococcus: 14 days
- Gram-negatives: 21 days
- Listeria: 21 days (with gentamicin for first 7 days)
- No organism identified: aged >3 months, 10 days; aged <3 months, 14 days
- Other, discuss with microbiologist

Steroids

Continue dexamethasone if:

- CSF WCC >1000/microlitre
- CSF protein >1 g/L
- Bacteria on Gram stain or culture

Fluid restriction

- Maintenance fluids: sodium chloride 0.9% with glucose 5% with potassium chloride 10 mmol/500 mL if not hyperkalaemic
- Restrict fluid to 80% maintenance if:
  - severe illness
  - hyponatramia
  - raised intracranial pressure
- Measure urine and plasma osmolalities daily whilst severely ill

Public health

- Inform Public Health consultant of a case of suspected meningitis
  - Public Health Department will arrange prophylaxis for close contacts
- Meningococcal meningitis
  - if ceftriaxone given as treatment, eradication treatment not required for patient
- Close contacts (all ages): ciprofloxacin single dose
- *Haemophilus influenzae*
  - close contact aged <10 yr, give rifampicin 20 mg/kg oral once daily for 4 days

DISCHARGE AND FOLLOW-UP

- Organise formal hearing test 6 weeks after discharge from hospital
- If severely ill during admission, discuss with consultant about follow-up to monitor developmental progress
- If viral cause unconfirmed but still possible, repeat viral titres 6 weeks after day of admission
- If >1 episode of meningococcal disease, not serogroup B, recurrent serious bacterial infections or family history of meningococcal disease or immune deficiency, refer to immunology or infectious diseases

NOTIFIABLE INFECTIOUS DISEASES AND FOOD POISONING • 1/2

URGENT NOTIFICATION

- Urgent out-of-hours notifications (to be followed by normal paper notification later)
- meningitis (suspected bacterial)
- meningococcal infection (clinical diagnosis)
- haemolytic uraemic disease (suspected)
- infectious bloody diarrhoea

NOTIFIABLE DISEASES

Admitting doctor is required to notify **suspected** or confirmed cases of the following to Health Protection Unit:

- Cluster or outbreak suspected (two or more cases epidemiologically linked)
- Any other case where the potential for transmission is significant (e.g. highly infectious)
- Where contacts are particularly susceptible (e.g. healthcare worker, school)
- Where public health action is known to be effective (e.g. prophylaxis, immunisation)
- Other infections or contaminations (e.g. chemical) not listed below if potential risk of further harm

- Anthrax
- Botulism
- Brucellosis
- Cholera
- Diphtheria
- Diarrhoea, infectious bloody
- Encephalitis
- Food poisoning*
- Group A streptococcal invasive disease
- Haemolytic uraemic syndrome
- Hepatitis (viral)
- Legionnaires'
- Leprosy
- Malaria
- Measles*
- Meningitis (viral, bacterial or fungal)
- Meningococcal disease
- Mumps
- Paratyphoid fever
- Plague
- Poliomyelitis
- Rabies
- Rubella*
- Severe acute respiratory syndrome (SARS)
- Scarlet fever*
- Smallpox
- Tetanus
- Tuberculosis*
- Typhoid fever
- Typhus
- Viral haemorrhagic fever
- Whooping cough*
- Yellow fever

***Definitions**

- **Food poisoning or suspected food poisoning:** inform public health if acquired abroad or if family member is a food handler or healthcare worker
- **Measles:** fever, maculopapular rash for ≥3 days and two or more of following: Koplik's spots, coryza, conjunctivitis, raised measles IgM, measles encephalitis or pneumonitis. Inform public health of MMR or measles vaccination history. Do not bring children with suspected measles in primary care to hospital for diagnosis, only if hospital based treatment required or if immunocompromised: arrange for immediate isolation on arrival

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- **Rubella:** rash and occipital lymphadenopathy or arthralgia (if not parvovirus), **or** congenital rubella or raised IgM to rubella. Inform public health of MMR vaccine history
- **Scarlet fever:** tonsillitis, fever, rash with either culture of *Streptococcus pyogenes* from throat or raised ASO or anti-DNaseB titre
- **Tuberculosis:** diagnosed clinically, not just microbiologically (atypical mycobacterial infection or patients given chemoprophylaxis but not thought to have TB are not notifiable)
- **Whooping cough:** cough with a whoop, with history of contact with similar illness **or** positive pernasal swabs for *Bordetella pertussis* or raised IgM to *B. pertussis* in an adult or child. Inform public health of pertussis immunisation history

Non-statutory notifiable diseases

It has been agreed that, although they are not statutorily notifiable, the following diseases will nevertheless be reported to the consultant in communicable disease control:

- AIDS/HIV infection
- Legionnaires' disease
- Listeriosis
- Psittacosis
- Cryptosporidiosis
- Giardiasis
- Creutzfeldt-Jakob disease and other prion diseases

CONTACT DETAILS

- Contact details for your nearest HPU can be found on the Health Protection Agency website (www.hpa.org.uk/ProductsServices/LocalServices)
- Template for reporting procedures under topics/notifiable/reporting procedures

ORBITAL CELLULITIS • 1/1

RECOGNITION AND ASSESSMENT

Preseptal	Orbital
Facial erythema and tenderness	Painful eye movements
Normal eye movements	Orbital pain and tenderness
Normal vision	Visual impairment (red-green colour differentiation lost early)
Preceding superficial trauma	Proptosis
Eye pain	Chemosis
Periorbital swelling	Ophthalmoplegia
Fever	Preceding sinusitis

Definition

- Infection of soft tissues surrounding the eye

Complications

- Intracranial abscess
- Meningitis
- Cavernous sinus thrombosis
- Periorbital abscess

Investigations

- Eye swab (send pus if present)
- FBC
- Blood culture
- CT scan if:
  - orbital involvement suspected
  - central neurological signs
  - unable to assess eye movements/vision or if eyelid cannot be opened
  - bilateral oedema
  - deterioration despite treatment

MANAGEMENT

Preseptal peri-orbital cellulitis

- Co-amoxiclav oral
- Review eye movements and red-green colour vision twice daily
- If no improvement in 48 hr IV antibiotics for 48 hr:
  - if aged ≤4 yr or no Hib vaccination, cefuroxime
  - if aged >4 yr and has received Hib vaccination, benzylpenicillin and flucloxacillin
  - if improving, convert to oral high dose co-amoxiclav
  - if penicillin allergy give clindamycin
- Total duration of treatment (including IV) 14 days

Orbital cellulitis

- Urgent ophthalmology review within 4 hr
- ENT review
- IV cefotaxime or ceftriaxone
- If toxaemic add clindamycin
- If history of anaphylaxis to penicillin give ciprofloxacin and clindamycin
- Consider surgical drainage
  - if improving, convert to oral high dose co-amoxiclav
  - if penicillin allergy give clindamycin
- Total duration of treatment (including IV) 21 days (up to 6 weeks if bone involvement)

Intracerebral complications

- Urgent neurosurgical review

Sinusitis

- URTI symptoms ≥10 days and ≥1 of:
  - nasal congestion and discharge
  - persistent cough (often nocturnal)
- Treat with co-amoxiclav oral high dose
- Severe if:
  - ill, temperature >39ºC, purulent discharge
- Urgent CT, ENT, neurosurgical review

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RECOGNITION AND ASSESSMENT

Symptoms and signs

- Fever
- Loss of function e.g. limp
- Pain in bone or joint
  - localised, constant, increasing
- Restricted range of movement
- Soft tissue swelling
- Point tenderness of bone
- Effusion

The above symptoms and signs are indicative of osteomyelitis or septic arthritis (in absence of clear history of obvious trauma) irrespective of WBC, CRP, ESR and fever or radiological appearance

Previous history

- Ask about:
  - duration of symptoms
  - injuries
  - fever
  - antibiotics
  - antipyretics/anti-inflammatories

Urgent investigations

- FBC
- ESR
- CRP
- Blood culture **(before antibiotics)**
- If cause of fever uncertain, collect other specimens (e.g. urine) for culture before antibiotics

Osteomyelitis

- Plain X-ray AP and lateral of the affected part
- Tissue or pus for Gram stain and culture if surgically explored or needle aspiration

Septic arthritis

- Aspiration of joint for Gram stain and culture
  - interventional radiologist or orthopaedic registrar
  - for sedation and analgesia contact paediatric registrar or on-call paediatric anaesthetist

Further investigations

Perform as soon as possible (must be within 36 hr)

- If plain X-ray normal, infection clinically localised and urgent MR is available:
  - consultant paediatrician or orthopaedic surgeon to authorise urgent MR of bone
  - if deep sedation or general anaesthetic required, contact on-call paediatric anaesthetist
- If plain X-ray normal, and infection clinically localised and MRI not available, request ultrasound scan of bone
- If localising signs poor or possible multifocal infection, request isotope bone scan
- If cardiac murmur or multifocal *Staph. aureus,* request echocardiogram

IMMEDIATE TREATMENT

- Admit
- Nil-by-mouth and maintenance fluids IV
- Bed rest
- Refer immediately to orthopaedic and paediatric registrar on-call
  - confirm they will assess child within 4 hr of admission
- Early involvement of on-call consultant orthopaedic surgeon

Antibiotics (see BNFc for neonatal doses)

- Severe sepsis with organ dysfunction (e.g. hypotension, oxygen requirement, GCS <12, platelet <80, creatinine x 2 normal, abnormal LFTs)
  - after blood and urine cultures taken, start cefotaxime 50 mg/kg 6-hrly (high dose; max 12 g/day) IV over 3–4 min
- No organ dysfunction; as soon as possible (must be within 4 hr):
  - if aged <5 yr: cefotaxime 50 mg/kg 6-hrly or ceftriaxone 80 mg/kg daily and flucloxacillin 50 mg/kg IV (max 2 g/dose)
  - if aged >5 yr: flucloxacillin 50 mg/kg IV (max 2 g/dose)
- Targeted antibiotic therapy
- If organism identified, use narrowest spectrum possible with good bone/joint penetration
  - *Staph aureus* sensitive to flucloxacillin 50 mg/kg 6-hrly IV (high dose max 2 g/dose)
- Penicillin allergy, substitute flucloxacillin for:
  - history of rash: cefuroxime
  - history of anaphylaxis: clindamycin

Analgesia

- If necessary initially, to allow splintage, use morphine IV (see **Analgesia** guideline)
- Elevate and splint affected limb
  - plaster backslab for peripheral joints
  - rest in skin traction on a pillow for central joints

Surgery

Ask parent(s) to stay with child until consent obtained

- Resuscitate if severe sepsis
- Emergency theatres to be alerted as soon as possible (must be within 36 hr of admission)
- Contact:
  - anaesthetic office to arrange paediatric anaesthetist
  - orthopaedic RSO to book patient onto planned emergency list
  - consultant paediatrician and orthopaedic surgeon
  - transfer to Trauma Theatre (nurse escort)

SUBSEQUENT MANAGEMENT

Inform paediatric orthopaedic surgeon and paediatrician

Uncomplicated septic arthritis (not complicated by associated osteomyelitis)

- Aspirate or drain joint in theatre
- Request long line insertion under GA and repeat any blood tests required
- If discharged for hospital at home IV treatment, change to ceftriaxone
- If treatment started within **24 hr** of first symptoms and clinically improving, discuss with consultant about changing IV to oral antibiotics after 72 hr if:
  - recovery of joint movement
  - absence of pyrexia after 4-hrly monitoring for 48 hr
  - WCC <11, CRP and ESR falling on two successive specimens ≥24 hr apart
- If agreed by consultant, give oral antibiotic to complete treatment
  - no organism identified: co-amoxiclav (high dose)
  - organism identified: narrowest spectrum with good bone penetration
    - if *Staph. aureus* sensitive to flucloxacillin: flucloxacillin oral (high dose) if capsules tolerated; or co-amoxiclav (high dose) if can only take suspension
  - allergic to penicillin: clindamycin oral

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- Stop treatment only if CRP is normal: agree duration of treatment with orthopaedic consultant depending on individual case

Early-presenting osteomyelitis

- If IV antibiotics started within 24 hr of onset of symptoms with a good clinical response as above, follow **Uncomplicated septic arthritis**

Established osteomyelitis or complicated septic arthritis

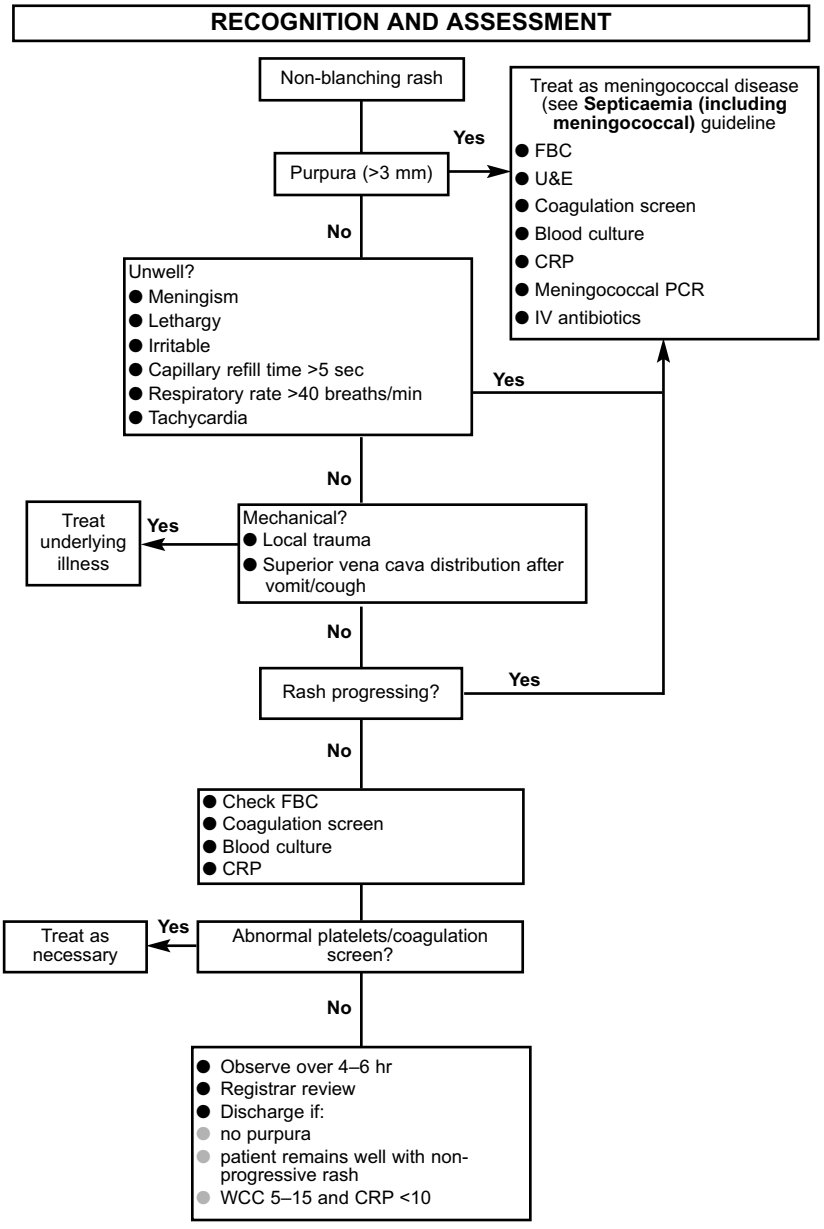
- Presentation >24 hr after onset of symptoms or partial treatment (e.g. oral antibiotics)
- Formal debridement in theatre with insertion of Hickman line
- Antibiotics IV as above. Discuss with consultant about switch to oral antibiotics after 14 days, if afebrile, pain free for 48 hr and CRP <20
- Continue antibiotics until ESR <20 (minimum 6 weeks)
- Continue oral antibiotics until all inflammatory markers are normal and clear evidence of healing established on radiographs
- Discuss with orthopaedic consultant duration of antibiotics on individual case

Septic arthritis or osteomyelitis (deteriorating condition/failure to improve within 48 hr)

- Inform orthopaedic team for exploration to drain pus
- Review culture result
- Discuss with consultant microbiologist and paediatrician
- Arrange for repeat blood cultures
- consider a change of antibiotic therapy or targeted antibiotic therapy
- Complete or repeat any investigations listed above
- Consultant paediatric medical and orthopaedic review
- Exclude important differential diagnoses
- systemic inflammatory response as seen in juvenile chronic arthritis
- transient synovitis, associated with intercurrent infection
- acute leukaemia, septicaemia, multifocal disease, endocarditis
- Continuing problems with local sepsis
- return to theatre for further debridement and insertion of Hickman line

MONITORING TREATMENT

- Peripheral colour, warmth, movement of affected limb: hourly for first 4 hr then 4-hrly for 24 hr
- Respiratory rate, pulse, temperature 4-hrly



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SEPTICAEMIA (INCLUDING MENINGOCOCCAL) • 1/4

Treat IMMEDIATELY (<1 hr) as delay increases mortality

RECOGNITION AND ASSESSMENT

- Assess **A**irway, **B**reathing, **C**irculation and resuscitate as required
- **D**isability: be alert to coexisting meningitis, see **Meningitis** guideline
- Core Temp >38.5ºC or <36ºC
- Mean HR >2 SD for age or persistent elevation over 0.5–4 hr
- If aged <1 yr: bradycardia HR <10th centile for age
- Mean RR >2 SD above normal for age

Meningococcal septicaemia

- Assess severity of disease on **Glasgow Meningococcal Septicaemia Prognostic Score:**
  - on admission and at least hourly for first 4 hr then if improving at least 4-hrly for next 24 hr
  - a score >8 indicates high risk of mortality: refer to PICU

Glasgow Meningococcal Septicaemia Prognostic Score (GMSPS)

Criteria	Score
Systolic BP (cuff width >2/3 upper arm length) if <75 mmHg in child aged <4 yr or <85 mmHg in child aged >4 yr	3
Skin/rectal temperature difference (measure for 2 min) if axilla/rectal temperature difference >3ºC	3
Modified coma scale (see **Glasgow coma score** guideline**)** if initial score <8 or deterioration of ≥3 points at any time	3
Deterioration in last hour (subjective) ask parents or nurse; if yes, score	2
Neck stiffness if **no** neck stiffness, score	2
Extent of purpura widespread ecchymoses or extending lesions on review	1
Base deficit if deficit >8 mmol/L	1
Maximum score	15

Investigations

- FBC and differential
- Blood culture (important to put in maximum amount of blood bottle designed to take)
- Blood gas and lactate
- Blood glucose
- Meningococcal PCR
- Group and save
- Clotting profile
- U&E, LFT, Ca^{++}, Mg^{++}, PO_4^{-}, CRP
- Save for serum cortisol

If purpuric rash

- See **Petechial/purpuric rashes** guideline
- If septicaemic treat immediately

If macular rash

- Be alert for septicaemia in any child presenting with a purpuric rash

- In the early stages of meningococcal septicaemia, a macular rash that DOES blanch on pressure is often present first. When in doubt, seek an experienced opinion urgently

If no rash

- Chest X-ray
- Urine culture (in severe sepsis, catheterise)
- Lumbar puncture if not contraindicated and where cardiovascularly and haematologically stable

Differential diagnosis

- Toxic shock syndrome
- Malaria

IMMEDIATE TREATMENT

- Ensure patent airway and adequate breathing
- Administer 15 L/min oxygen through a reservoir mask
  - if airway and breathing remain compromised despite simple airway manoeuvres and 100% oxygen, contact consultant and on-call anaesthetist
- Assess circulation, if severe sepsis or extensive rash, insert two large IV cannulae or establish intraosseous access and give 20 mL/kg sodium chloride 0.9% over 5–10 min

Antibiotics

- Give IV cefotaxime 50 mg/kg (max 3 g 6-hrly) over 20 min or ceftriaxone 80 mg/kg (max 4 g) daily over 30–60 min (not with calcium IV; not <41 weeks postmenstrual age or neonate with jaundice, hypoalbuminaemia or acidosis) see BNFc for neonatal doses
  - if documented history of definite anaphylaxis to cephalosporin discuss with consultant microbiologist
  - if MRSA suspected, add vancomycin
  - if anaerobic infection suspected, add metronidazole
  - if pseudomonas suspected give piperacillin with tazobactam (Tazocin)
  - if multiple-resistant organisms suspected (e.g. previous culture results, hospital acquired) give meropenem
- If hypotension continues, peripheries remain cool, rash continues to evolve, and capillary refill >2 sec, give sodium chloride 0.9% or human albumin 4.5% 20 mL/kg over 5–10 min

SUBSEQUENT MANAGEMENT

Circulation still compromised

- Contact on-call paediatric consultant and anaesthetist and inform PICU
- If hypotension continues give human albumin 4.5% or sodium chloride 0.9% 20 mL/kg boluses and start inotropes
- Dopamine 5–20 microgram/kg/min (7.5 mg/kg in 50 mL sodium chloride 0.9% at 2–8 mL/hr peripherally)
  - start at 5 microgram/kg/min
  - increase in 5 microgram/kg/min increments every 5 min
  - up to 20 microgram/kg/min as required
- If still hypotensive, start adrenaline 0.1 microgram/kg/min (0.3 mL/kg of 1:1000 in 50 mL sodium chloride 0.9% at 1 mL/hr)
- If still hypotensive, give hydrocortisone 1 mg/kg 6-hrly

Reassess ABC

- If still unstable (requiring >40 mL/kg fluid resuscitation), arrange immediate intubation with senior anaesthetist
  - prepare atropine 20 microgram/kg (max 600 microgram)

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- if no neck stiffness, ketamine 1 mg/kg; if neck stiffness and BP stable, thiopental sodium 3 mg/kg
- suxamethonium 2 mg/kg aged >1 yr; 1 mg/kg aged <1 yr
- then morphine 20 microgram/kg/hr (1 mg/kg in 50 mL sodium chloride 0.9% at 1 mL/hr)
- and midazolam 1–2 microgram/kg/min (6 mg/kg in 50 mL sodium chloride 0.9% at 0.5–1 mL/hr)
- and vecuronium 1 microgram/kg/min (1.5 mg/kg in 25 mL sodium chloride 0.9% at 1 mL/hr)
- or other muscle relaxant as per local practice
- Site nasogastric tube and urinary catheter
- Prepare for central venous line with portable ultrasound
- Monitor blood glucose hourly for first 6 hr: if <3 mmol/L give glucose 10% 2 mL/kg bolus and start maintenance fluids with glucose
- If glucose >3 mmol/L give sodium chloride 0.9% at 100% maintenance requirement
- If passing urine, give IV fluids with potassium 10 mmol/L, if hypokalaemic give 0.2 mmol/kg over 1 hr **(use commercial pre-mixed bags)**
- If hypocalcaemic, give calcium gluconate 10% (0.22 mmol/mL) 0.5 mL/kg (max 20 mL) IV over 5–10 min (do not give ceftriaxone in same line, give cefotaxime until stable)
- If INR >2, give fresh frozen plasma (FFP) 10 mL/kg
- After 60 mL/kg, give packed cells 20 mL/kg

Patient stabilises with <40 mL/kg bolus fluids

- Inform on-call consultant paediatrician
- Admit to general paediatric ward for monitoring and continue treatment
- Administer oxygen via face mask or nasal cannula to maintain continuous oxygen saturation >95%
- Treat poor perfusion and hypotension with aliquots of human albumin 4.5% 20 mL/kg solution

MONITORING

- Monitor the following every 30 min for first 2 hr, hourly for next 2 hr, then 4-hrly:
  - conscious level
  - temperature
  - respiratory rate
  - heart rate
  - BP
  - capillary refill time
- Monitor urine output hourly
- Monitor blood glucose and electrolytes 6-hrly until stable. Treat hypoglycaemia with bolus IV glucose
- Monitor clotting screen 12-hrly. Treat deranged clotting with FFP 10 mL/kg IV

SUBSEQUENT MANAGEMENT

- Adjust antibiotic treatment once culture results available
  - if meningococcal or no organism identified give 7 days: cefotaxime 50 mg/kg 6-hrly IV
  - or ceftriaxone 80 mg/kg IV daily, over 30–60 min (for cautions see **Antibiotics**)
- Treat *S. aureus* sepsis for 2 weeks
- Give antibiotics to treat carrier states in *Haemophilus* sepsis (see **Meningitis** guideline)
- Avoid enteral feeds until acute shock has resolved

Public health

- Meningococcal: inform Public Health (see **Notifiable infectious diseases** guideline)
- Public Health Department will arrange prophylaxis for close contacts

DISCHARGE AND FOLLOW-UP

- Organise formal hearing test after discharge from hospital
- Arrange appointment in follow-up clinic in 8–12 weeks to review problems with:
  - hearing loss
  - orthopaedic complications
  - scarring
  - psychosocial
  - neurology and development
  - renal function
- Test for complement deficiency if:
  - >1 episode meningococcal disease
  - meningococcus other than type B
  - other recurrent or serious bacterial infections
  - family history immune deficiency

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RECOGNITION AND ASSESSMENT

History is most important factor in diagnosing tuberculosis

Symptoms

Suspect TB when following symptoms persist for weeks:

- Persistent, non-remitting cough for 2–4 weeks
- Weight loss
- Failure to thrive
- Lack of energy
- Fever and sweats
- Lymph nodes, especially if painless and matted
- Headache or irritability for >1 week
- Limp, stiff back
- Joint swelling
- Abdominal distension

Signs

- Delayed growth: plot weight and height on growth chart and compare with earlier records
- Fever
- Wasting
- Lymphadenopathy
- Chest signs
- Cardiac tamponade
- Ascites
- Meningism
- Conjunctivitis
- Limited flexion of spine
- Kyphosis
- Swollen joint
- Cold abscess

Family and social history

- Ask about recent contact with any family member (specifically grandparent or parent) who has:
  - chronic cough
  - previous treatment for TB especially multi-drug resistant (MDR) TB, failed/defaulted TB treatment, or recurrent TB
  - travelled to regions/countries with a high prevalence of TB/MDR TB
  - recently died

INVESTIGATIONS

- Tuberculin skin test (Mantoux): does not exclude TB, can be negative in miliary TB
  - avoid if other tests positive or clinical diagnosis of TB
- Rapid diagnostic test on primary specimen for rifampicin resistance if contact with multi-drug resistant (MDR) TB
- IGRA (interferon-gamma release assay, e.g. QuanitFERON® TB Gold or T-SPOT® TB) are not recommended by NICE for diagnosis of active TB

Pulmonary TB

- Chest X-ray: look for hilar lymphadenopathy, apical consolidation, pleural effusion, miliary nodules
- Sputum: send at least 3 (1 early morning) for AFB and TB culture in cooperative child, expectoration may require physio +/- nebulised sodium chloride 0.9% as necessary (with FFP3 mask and HEPA filtered ventilation if available)

- If unable to provide sputum specimen, send gastric aspirate (for TB culture only as microscopy is unreliable) early morning before feed, daily for 3 days
- if no aspirate, rinse stomach with small volumes of sodium chloride 0.9% (5 mL aliquots maximum 20 mL)
- do not send saliva
- Discuss broncho-alveolar lavage for AFB and TB culture via bronchoscopy with respiratory consultant

Pleural effusion

- Pleural tap +/- biopsy for histology and microbiology (AFB and TB culture)
- Discuss with cardiothoracic surgeons about pleural biopsy

Lymphadenopathy

- If single node, excision biopsy
- If large matted nodes, ultrasound scan +/- simultaneous guided aspiration (discuss before scan)
- Lymph node aspirate: fine needle aspiration biopsy (FNAB; 23 G needle)
- low risk, high yield with sedation and local anaesthetic
- Send aspirate in two separate bottles:
- one to microbiology for TB culture with no preservative
- one to histology in 10% formalin
- If atypical mycobacterial infection suspected, prefer complete excision biopsy, if possible, to aspiration biopsy

Meningism

- CSF: request staining for acid-fast bacilli (AFB) **AND** culture and sensitivity for TB (Note: TB meningitis extremely rare in the UK; often AFB negative)
- PCR: expensive, consider for CSF if highly suspicious of TB meningitis, poor sensitivity
- If tuberculoma suspected, CT or MR brain

Bone/joint pain

- Plain X-ray and CT or MR if available
- Biopsy/aspiration important for diagnosis and sensitivities

Abdominal distension

- Ultrasound then CT abdomen
- Culture ascites/bowel biopsy

Pyuria

- Urinalysis: if blood and leucocytes present, send for smear and culture
- non-tuberculous acid-fast bacteria common in urine
- Ultrasound kidneys
- Early morning urine culture

Pericardial effusion

- Echocardiogram
- Pericardial fluid

Disseminated (inc. miliary)

- CT thorax and ultrasound abdomen
- LP (CT or MR first if CNS signs or symptoms)
- Bone marrow biopsy if diagnosis uncertain

IMMEDIATE MANAGEMENT

Discuss treatment with local TB team and lead paediatrician for TB

- Inform Public Health through TB clinic, who will organise chest X-ray and Mantoux for all close and visiting contacts
- Inform infection prevention and control team: advise anyone with cough to avoid visiting ward
- Admission not mandatory but useful to ensure adherence with treatment. If supervision can be guaranteed, allow treatment at home

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- If sputum +ve and hospitalisation necessary, strict nurse in single room for 2 weeks or discharge
- Patient should wear a surgical mask if leaves room
- Masks, gowns and barrier nursing unnecessary unless MDR TB or aerosol generating procedure
- Negative pressure room for aerosol generating procedure if TB considered (e.g. nebuliser)

Drugs

- Isoniazid (H): 10 mg/kg once daily up to max 300 mg
- (suspension; 50 mg, 100 mg tab)
- Rifampicin (R): 15 mg/kg once daily up to max 450 mg if <50 kg; 600 mg ≥50 kg
- (suspension; 150 mg, 300 mg capsule)
- Pyrazinamide (Z): 35 mg/kg once daily up to max 1.5 g if <50 kg; 2 g ≥50 kg
- (500 mg tablets can be crushed)
- Ethambutol (E): 15 mg/kg once daily (100 mg, 400 mg tablets can be crushed)
- check renal function first, **do not round dose up**
- Round up doses of HRZ to give easily measured volumes of syrup or appropriate strengths of tablet. Re-calculate doses with weight gain
- Use drug combinations if possible
- Rifater®: H 50 mg; R 120 mg; Z 300 mg
- Rifinah® 150/100: H 100 mg; R 150 mg
- Rifinah® 300/150: H 150 mg; R 300 mg

Presentation	Treatment
Respiratory TB: lungs, pleural cavity, mediastinal lymph nodes or larynx	Rifampicin and isoniazid for 6 months Pyrazinamide and ethambutol for first 2 months
Meningeal TB	Rifampicin and isoniazid for 12 months Pyrazinamide and ethambutol for first 2 months Prednisolone 1–2 mg/kg (max 40 mg), with gradual withdrawal, starting within 2–3 weeks of initiation
Other extra pulmonary lesions	As for respiratory TB

- Add pyridoxine to prevent isoniazid neuropathy in malnourished or breastfed infants, diabetes, HIV or renal failure
- Pericardial TB: add prednisolone 1 mg/kg/day (max 40 mg/day)
- Inform patient/parents of both common (gastrointestinal upset, rash) and rare but important side effects (staining of secretions, signs of hepatotoxicity)
- Advise patient/parents and GP of indications for seeking advice: fever, malaise, vomiting, jaundice or unexplained deterioration. Consider co-existent viral hepatitis. If AST/ALT level rises to 5x normal, stop treatment and seek advice re: alternate regimen

SUBSEQUENT MANAGEMENT

- HIV test
- Other drugs may be necessary once sensitivities available: if resistant, seek specialist advice

MONITORING TREATMENT

- If baseline ALT/AST raised but ≤2x normal, repeat at 2 weeks; if falling only recheck if fever, malaise, vomiting, jaundice or unexplained deterioration
- If ALT/AST >2x, monitor weekly for 2 weeks then 2-weekly until normal, check viral hepatitis serology

- Stop treatment only if ≥5x normal
- If on ethambutol and unable to report visual problems, check visual evoked response

DISCHARGE AND FOLLOW-UP

- Discharge if tolerating treatment and adherence guaranteed
- If concerns about adherence, will need direct observed therapy, organised through TB team
- Review to ensure adherence:
  - at least monthly for first 2 months
  - 2-monthly until treatment complete
  - for 3 months after end of treatment
  - further as clinically indicated

LATENT TB

Close contact with sputum +ve TB or new entrant from high-incidence country

- If symptomatic refer to TB team
- If asymptomatic neonate contact with smear +ve case on treatment <2 weeks, treat with isoniazid for 3 months then do Mantoux. If +ve: CXR and refer to TB team; if –ve repeat Mantoux and do IGRA: if both –ve stop isoniazid and give BCG, if either +ve do CXR and refer to TB team
- If asymptomatic aged >4 weeks do Mantoux
- If Mantoux +ve (≥15 mm with BCG/≥6 mm no BCG) request CXR and refer to TB team for chemoprophylaxis
- If Mantoux –ve (<15 mm with BCG/<6 mm no BCG):
  - if index case smear +ve, after 6 weeks do IGRA and if aged 2–5 yr repeat Mantoux: if Mantoux or IGRA +ve refer to TB team
  - if index case smear –ve, advise to see GP if symptomatic
  - if aged 4 weeks–2 yr and no BCG, start isoniazid and repeat Mantoux and do IGRA after 6 weeks: if both –ve, stop isoniazid, if either +ve, do CXR and refer to TB team
  - if aged 4 weeks–2 yr and BCG, repeat Mantoux and do IGRA after 6 weeks: if both –ve, discharge, if either +ve, do CXR and refer to TB team

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RECOGNITION AND ASSESSMENT

Definition

- Bell's palsy: idiopathic lower motor neurone facial nerve palsy
- Facial nerve palsy secondary to infection, inflammation, tumour, trauma, vascular event

Symptoms and signs

- Asymmetry of face or smile and loss of nasiolabial fold on same side
- Increased or decreased lacrimation
- Hyperacusis
- Altered taste
- Facial pain
- Difficulty in closing eye

History

- History of prior viral infection may be present
- Abrupt onset with no progression
- No history of preceding seizure or head injury

Examination

- Full neurological examination, including other cranial nerves, and fundoscopy
- demonstrable weakness in lower motor neurone distribution (includes loss of wrinkles on forehead)
- Ears, nose and throat to exclude cholesteatoma, mastoiditis or herpes infection
- Blood pressure to exclude hypertension
- Check for lymphadenopathy, hepatosplenomegaly, pallor or bruising to exclude malignancy

INVESTIGATIONS

- If all history/examination unremarkable, no investigations needed
- If difficulty in closing eye, ophthalmology referral
- Bilateral facial palsy – consider Lyme disease, Guillain-Barré syndrome, brain stem pathology: discuss further investigations with senior
- Recurrent facial palsy: discuss with senior
- Recurrent infections: first line immune deficiency investigations (including HIV)
- Severe pain associated with VZV

IMMEDIATE TREATMENT

- If difficulty in closing eye, provide eye patch and hypromellose eye drops
- If no other signs, no other treatment necessary
- If vesicles suggest HSV, prescribe aciclovir, test for immune deficiency
- Within 72 hr prednisolone 1 mg/kg/day for 10 days discuss with a senior

DISHCHARGE AND FOLLOW-UP

- 4 weekly GP follow-up until symptoms and signs have resolved (95% by 1 yr)
- If facial palsy does not improve considerably within 4 weeks discuss imaging

DEFINITIONS

- Seizures/convulsions: paroxysmal disturbance of consciousness, behaviour, motor function, sensation – singly or in combination
- Epilepsy: recurrent seizures without any provoking factor and happening in different situations
- Seizure type: (focal, generalised or any other type) based on history and EEG
- Try to categorise into one of epilepsy syndromes

RECOGNITION AND ASSESSMENT

- Detailed and accurate history from an eyewitness (beginning, middle and end of the episode)
- When history unclear, recording the episode with a camcorder/mobile phone can be very useful
- Episodes occurring only in certain situations with certain provoking factors (such as fall, emotions, certain posture etc., except photosensitive stimuli) are likely to be non-epileptic
- Any underlying problem: learning difficulties, cerebral palsy, HIE, head injury or other CNS insult
- Look for any co-morbidity
- Family history may be positive in certain idiopathic generalised epilepsies, some symptomatic epilepsies (tuberous sclerosis), autosomal dominant frontal epilepsies
- Genetic conditions (e.g. Angelman's syndrome)
- Neurocutaneous syndromes, café-au-lait spots/depigmented patches, use Woods Light
- Neurological examination
- If in doubt about diagnosis, do not label as epilepsy but watch and wait or refer to specialist

Diagnosis of epilepsy is clinical

Seizure types

Generalised

- Tonic-clonic/tonic
- Clonic
- Atonic
- Absence
- Myoclonic

Focal

- Without impairment of consciousness (focal motor, sensory or other types)
- With impairment of consciousness (previously known as complex partial)
- Focal with secondary generalisation (clinically look like generalised seizures) – history of aura, Todd's paralysis, focal features on EEG

Underlying cause

- In most cases epilepsy is idiopathic but a few cases have an underlying cause
- actively look for the cause to guide prognosis, other treatment and recommendation for epilepsy surgery

EPILEPSY SYNDROMES

Identification

- Based on:
- seizure type
- age of onset
- neurodevelopmental status
- appearance of EEG (ictal and interictal)

Childhood absence epilepsy

- Usually presents aged 3–8 yr
- More common in girls
- Several (up to 100) brief episodes in a day
- Very quick recovery
- Typical EEG 3 per sec spike and wave

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- 10–30% of children have generalised seizures at some stage, usually in teenage years

Juvenile absence epilepsy

- Usually presents after age 9–10 yr
- Absence frequency is less than in childhood absence epilepsy
- Cluster after awakening
- 90% of children have generalised seizures in the same period while they have absences
- EEG generalised spike and wave

Juvenile myoclonic epilepsy (JME)

- Usually presents between ages 12–18 yr
- Myoclonic jerks are hallmark of this syndrome
- Jerks after awakening (myoclonic jerks), common and often go unrecognised
- 90% of children have generalised seizures at some stage
- 15–30% of children will have absences

Benign epilepsy of childhood with rolandic spike

- Usually nocturnal seizures
- Unilateral focal motor seizures of face and arm with gurgling and salivation
- May become secondary generalised
- May present with nocturnal generalised seizures
- Spikes in one or the other centro temporal areas
- Awake interictal EEG could be normal and sleep EEG would usually show the abnormality

Panayiotopoulos syndrome

- Younger children (peak age 5 yr)
- usually nocturnal and happens in sleep
- Usually starts with vomiting and child initially conscious
- Child continues to vomit repeatedly and becomes unresponsive
- Subsequent deviation of eyes to one side or may end in hemiclonic seizure or (rarely) generalised seizure
- Other autonomic features very common (e.g. dilated pupils, pale skin or flushing, incontinence)
- Usually lasts for a few to 30 min, occasionally for several hours

Temporal lobe epilepsy (TLE)

- Focal seizures with impaired consciousness and complex automatism
- Aura is common before the seizure, which could be a sense of fear, abnormal abdominal sensation or any other
- Children are very tired and sleepy after episode
- Children with history of prolonged febrile seizure in the early years of life may have mesial temporal sclerosis as a cause of their seizures
- Other known causes: cortical dysplasia, gliomas, disembryonic neuroectodermal tumour
- Some patients can be a candidate for epilepsy surgery

Frontal lobe epilepsy

- Usually focal motor seizures
- Either tonic or clonic seizures – may have speech arrest and head rotation or complex partial seizures or focal with secondary generalisation
- Multiple brief seizures in the night
- Repeated/multiple brief nocturnal seizures are a characteristic feature of frontal lobe epilepsy
- Ictal EEG can be normal
- Can mimic pseudo seizures

Other epilepsy syndromes

- Epileptic encephalopathy in newborns (myoclonic or Ohtohara syndrome)
- West's syndrome (infantile spasms)
- Severe myoclonic epilepsy of infancy (Dravet's syndrome)
- Lennox-Gastaut syndrome
- Laundau-Kleffner syndrome
- For other epilepsy syndromes see International League against Epilepsy website (www.ilae-epilepsy.org)

INVESTIGATIONS

Indications for EEG

- Clinically diagnosed epilepsy
- After an episode of status epilepticus
- Unexplained coma or encephalopathy
- Suspicion of non-convulsive status in children with learning difficulties and epilepsy
- Acquired regression of speech or language function
- Developmental regression suspected to have neurodegenerative condition
- To monitor progress in West's syndrome and non-convulsive status

EEG not indicated

- Funny turns, apnoeic attacks, dizzy spells, strange behaviour
- Non-convulsive episodes [e.g. syncope, reflex anoxic seizures, breath-holding episodes (ECG more appropriate)]
- Febrile seizures
- Single uncomplicated generalised tonic-clonic seizures
- To monitor progress in well-controlled epilepsy
- Before stopping treatment

Indications for MRI of brain

- Focal epilepsy (including TLE) except rolandic seizures
- Epilepsy in children aged <2 yr
- Myoclonic epilepsy
- Intractable seizures
- Loss of previous good control
- Seizures continuing in spite of first line medication
- Associated neurological deficits or appearance of new neurological signs
- Developmental regression in children with epilepsy
- Infantile spasms (West's syndrome)

Other investigations

- Sleep or sleep-deprived EEG useful in all children in whom there is a high clinical suspicion but awake EEG normal
- sleep EEG useful to pick up some focal/generalised epilepsies and sleep-deprived EEG useful in generalised epilepsies in young adults including JME. Perform sleep EEG with melatonin
- Video telemetry useful if diagnostic dilemma, pseudo seizures or before surgery
- Drug levels: phenytoin, phenobarbitone (other anticonvulsants only if concerns about compliance and overdose)
- Biochemistry: glucose, calcium, LFT, lactate, ammonia; metabolic and genetic investigations where suspicion of metabolic disorder (e.g. progressive developmental delay)
- Epileptic encephalopathies, such as West's Syndrome, need a series of investigations (discuss with paediatric neurologist)

TREATMENT

General guidelines

- Discuss treatment with a consultant before starting
- Start anti-epileptic only if diagnosis certain (two or more unprovoked seizures)

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- Preferably after initial EEG results obtained
- Start with small dose and build up to half maintenance. If seizures continue, increase to full maintenance
- Increase dose stepwise every 2–3 weeks

First line drugs

- See **Table** for choice of anti-epileptic drug
- Carbamazepine: start with 2.5–5 mg/kg/day in two divided doses gradually increasing to 20 mg/kg/day

OR

- Sodium valproate: start with 5–10 mg/kg/day in two divided doses gradually increasing to 40 mg/kg/day
- Avoid polypharmacy; do not add a second medication unless the full or maximum tolerated dose of the first medication has been reached (discuss with a paediatrician with special interest or paediatric neurologist before adding second drug)
- Aim to switch to monotherapy after a period of overlap
- Give liquids as sugar-free preparations
- Make sure you discuss potential adverse effects with parents and document these in notes
- If child develops adverse effects, discuss and reduce dose

Discussion with child and parents

- Provide additional advice regarding safety (e.g. supervision when swimming) and document discussion in notes
- Discuss and prescribe rescue treatment, especially in generalised epilepsy, with training for parents
- Provide written information, including information about national or local epilepsy associations and website for Epilepsy Action (www.epilepsy.org.uk)
- Explain how to gain access to epilepsy specialist nurse
- Allow parents and children to ask questions, especially about sensitive issues such as sudden death

Table 1: Drugs of first, second and third choice in treatment of seizure types

Seizure type	First	Second	Third
Generalised epilepsy	Sodium valproate OR Carbamazepine†	Carbamazepine† OR Sodium valproate	Lamotrigine* Levetiracetam Topiramate
Childhood absence epilepsy	Sodium valproate Ethosuximide	Lamotrigine*	Levetiracetam Topiramate
Focal epilepsy including TLE	Carbamazepine	Sodium valproate Lamotrigine* Topiramate Levetiracetam	Clobazam Phenytoin
Infantile spasms	Prednisolone/tetracosactide OR Vigabatrin	Sodium valproate Nitrazepam	Trial of pyridoxine

* Lamotrigine can increase myoclonic seizures in some myoclonic epilepsy syndromes

† Carbamazepine should be avoided in childhood absences, juvenile absences and juvenile myoclonic epilepsy and can increase seizures in some epileptic encephalopathies and primary generalised epilepsies

Epilepsy in adolescence – additional factors to be considered

- Compliance
- Career choices
- Driving
- Contraception and pregnancy, including pre-pregnancy counselling
- Alcohol and drugs

SUBSEQUENT MANAGEMENT

- Increase dose of anti-epileptic gradually towards full dose or maximum tolerated dose until control good
- If control suboptimal with one drug or unacceptable side effects, start second-line drug

OUT-PATIENT MANAGEMENT

- Initial follow-up at 6–8 weeks
- Subsequent follow-up/structured review every 3–12 months based on clinical need

FURTHER OPINION/REFERRAL TO SPECIALIST SERVICE OR TERTIARY CENTRE (NICE GUIDELINES)

Refer immediately

- Behavioural or developmental regression
- Epilepsy syndrome cannot be identified

Refer soon

- When one or more of the following are present:
  - child aged <2 yr
  - seizures continuing despite being on anti-epileptic drug (AED) for 2 yrs
  - 2 AEDs have been tried and are unsuccessful
  - risk of unacceptable side effects of medication
  - unilateral structural lesion
  - psychological or psychiatric co-morbidity
  - diagnostic doubt about seizure type and/or syndrome

Refer

- Refer specific syndromes such as:
  - Sturge-Weber syndrome
  - Rasmussen's encephalitis
  - hypothalamic hamartoma

WITHDRAWAL OF ANTI-EPILEPTIC DRUGS

- Consider when child has been seizure free for 2 yrs
- Discuss the risks of recurrence (25–30%), if this occurs, recommence treatment
- Recurrence is very high in some syndromes (e.g. juvenile myoclonic epilepsy, 70–80% usually requires lifelong treatment)
- Postpone withdrawing anti-epileptic medication if important events such as GCSEs are looming
- Gradual withdrawal over 2–3 months usual
- Some drugs (phenobarbital or benzodiazepines) need very slow withdrawal over 6–12 months

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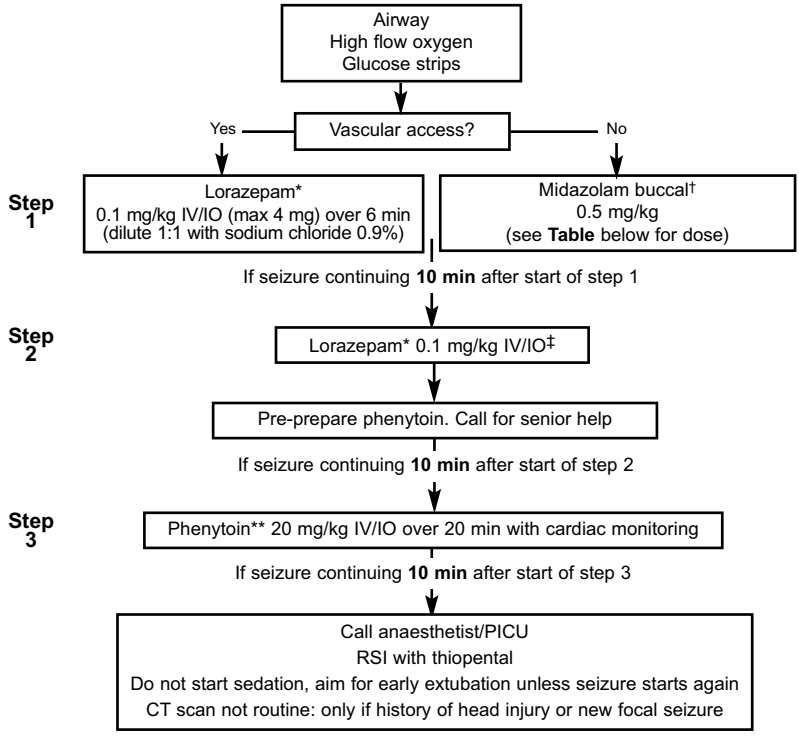
STATUS EPILEPTICUS • 1/1

- Follow each step until fits resolve, but do not treat post-ictal posturing as seizure
- Prepare next step in algorithm immediately after previous one administered
- Do not give more than 2 doses of benzodiazepine, including any pre-hospital doses



* If lorazepam not available give IV diazepam

† If buccal midazolam not available give rectal diazepam

‡ If vascular access and intraosseous still not obtained give paraldehyde PR: 0.8 mL/kg ready mixed solution or 0.4 mL/kg diluted with equal volume of olive oil

** If already taking phenytoin, give phenobarbital 20 mg/kg IV/IO over 20 min diluted 1:1 with water for injection

Diazepam (IV)	Diazepam (rectal)	Midazolam (buccal)	Paraldehyde (rectal) volume of 50:50 diluted
Aged 1 month–12 yr 300 microgram/kg (max 10 mg)	Aged 1 month–2 yr: 5 mg	Aged <6 months: 300 microgram/kg (max 2.5 mg)	Aged 1–3 months: 0.5 mL
Aged >12 yr: 10 mg	Aged 2–12 yr: 5–10 mg	Aged 6 months–1 yr: 2.5 mg	Aged 3–6 months: 1 mL
	Aged >12 yr: 10 mg	Aged 1–5 yr: 5 mg	Aged 6 months–1 yr: 1.5 mL
		Aged 5–10 yr: 7.5 mg	Aged 1–2 yr: 2 mL
		Aged >10 yr: 10 mg	Aged 2–5 yr: 3–4 mL
			Age 5–18 yr: 5–10 mL

ON ADMISSION

- Ask parents if they have a copy of a care plan
- Inform child's long-term consultant

CLINICAL HISTORY

- Adequacy of cough and swallowing
- Previous sleep difficulties, wakefulness at night (nocturnal hypoventilation)
- Difficulty waking in morning, early morning headache (nocturnal hypoventilation)
- Poor appetite, weight loss (chronic respiratory failure)
- Learning or behavioural problems, school attendance (chronic respiratory failure)
- Palpitations, breathlessness, chest pain (cardiomyopathy)
- Muscle cramps, skeletal pain, back pain (for fractures)
- Abdominal pain, distension, melaena (GI perforation)

ASSESSMENT

- May not show overt signs of respiratory distress such as tachypnoea, recessions and use of accessory muscles even in respiratory failure
- Assess adequacy of chest wall excursion and cough
- Look for pallor, tachycardia, signs of circulatory compromise
- Assess for abdominal signs (GI bleed, perforation, gastritis)
- Measure:
  - SpO_2 in air
  - CO_2 by blood gas, transcutaneous CO_2 or end-tidal CO_2, especially if on oxygen
  - spirometry: FVC most useful if previous readings available
- ECG
- Blood gas for cardiac status
- Chest X-ray: clinical signs can fail to detect collapse/consolidation/cardiomegaly
- Consider skeletal/spinal X-rays for possible fractures

Medical problems commonly found in children with myopathy

- Respiratory failure (hypoxaemia and hypercapnia) without signs of respiratory distress. Susceptibility to respiratory failure due to:
  - muscle weakness (upper airway, intercostals, diaphragm)
  - scoliosis
  - poor secretion clearance
  - aspiration, chest infections
  - sleep disordered breathing
  - cardiac failure
- Lower respiratory infection, aspiration pneumonitis
- Cardiomyopathy and cardiac decompensation
- Gastro-oesophageal reflux, gastritis and gastric ulceration (especially if on corticosteroids)
- Adrenal insufficiency (if on corticosteroids)
- Fractures, especially vertebral, if on long-term corticosteroids
- Malignant hyperthermia following anaesthesia in certain muscular dystrophies and myopathies

MANAGEMENT

- If unwell, on **long-term corticosteroids**, double usual daily dose of steroids for 2–3 days. If unable to tolerate oral steroids, use IV hydrocortisone

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Respiratory failure

- Carefully titrated administration of oxygen by mask/nasal cannulae to achieve SpO_2 between 94–98%. Monitor CO_2 and respiratory effort as risk of rising CO_2 and respiratory failure (despite normal oxygen saturations) if hypoxic respiratory drive overcome by oxygen therapy
- Mask ventilation (bi-level positive airway pressure, BIPAP)
- Chest physiotherapy and postural drainage
- Use in/ex-sufflator (e.g. Cough Assist) if patient has one
- Suction
- if copious loose secretions use glycopyrrolate 40–100 microgram/kg oral max 2 mg 6-hrly (use 200 microgram/mL IV solution if specials manufacturer solution not available)
- Antibiotics
- obtain cough swab or sputum specimen, ideally before starting treatment
- check previous culture results
- choice same as for community acquired pneumonia
- if bronchiectasis use broad spectrum for 14 days to cover pseudomonas (discuss with senior)
- if not improving on 1st line antibiotics add macrolide for atypical pneumonia
- Consult senior to discuss need for ITU care, escalation of respiratory support

Cardiac failure

- Fluid restriction
- Diuretics
- Oxygen and respiratory support
- Cardiology consultation

GI tract bleed: prevention and treatment

- Nil-by-mouth and IV fluids
- Ranitidine (omeprazole if reflux)
- Senior advice

Fractures

- Analgesia
- Orthopaedic consultation
- IV biphosphonates for vertebral fractures, discuss with metabolic bone expert

Malignant hyperthermia

Malignant hyperthermia is a medical emergency

- Occurs following general anaesthesia and may be first presentation of a neuromuscular disorder
- Check creatine kinase, calcium, renal function, urine output and for myoglobinuria: dialysis may be needed
- In addition to temperature control and general life support measures, use IV dantrolene to control excessive muscle contraction
- Obtain senior anaesthetic advice and liaise with PICU

GLASGOW COMA SCORE • 1/1

Response aged ≥4 yr

Eye opening	Score
Spontaneously	4
To speech	3
To pain	2
Never	1

Best motor response	Score
Obeys commands	6
Localises pain	5
Withdrawal	4
Flexion to pain	3
Extension to pain	2
None	1

Best verbal response	Score
Orientated and converses	5
Disorientated and converses	4
Inappropriate words	3
Incomprehensible sounds	2
None	1

Response aged <4 yr

Eye opening	Score
Spontaneously	4
To speech	3
To pain	2
Never	1

Best motor response	Score
Obeys commands (or spontaneous, purposeful <2 yr)	6
Localises pain or withdraws to touch	5
Withdrawal	4
Flexion to pain	3
Extension to pain	2
None	1

Best verbal response	Score
Alert and babbles, words to usual ability	5
Less than usual words, spontaneous irritable cry	4
Cries only to pain	3
Moans to pain	2
None	1

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RECOGNITION AND ASSESSMENT

Definition

- Acute inflammatory process affecting the glomeruli leading to haematuria, proteinuria, oedema, hypertension and renal insufficiency

Symptoms and signs

- Reduced urine output
- Macroscopic haematuria, coca-cola coloured urine
- Headache/breathlessness, indicative of severe disease
- History of sore throat in preceding 2–3 weeks
- Oedema variable, periorbital/pedal
- check weight, trend is useful
- check jugular venous pressure (JVP), if raised, indicates volume overload (cardiac failure)
- Oliguria (urine output: infant/child <0.5 mL/kg/hr, neonate <0.6 mL/kg/hr)
- Hypertension +/- features of encephalopathy (headache, nausea, vomiting, visual disturbance, restlessness, confusion)
- Signs of cardiac failure (tachypnoea, raised JVP, gallop rhythm, basal crackles, enlarged liver)

Investigations

- Urine dipstick: >3+ blood and protein
- Urine microscopy: red cell casts
- U&E
- sodium may be low (dilutional effect)
- potassium, urea and creatinine
- bicarbonate may be low
- phosphate, uric acid
- albumin usually normal
- FBC: low haemoglobin (dilutional effect)
- Immunology: complement C3, anti-nuclear antibodies (ANA) and IgG, A and M
- Serology: hepatitis B
- Antistreptolysin O titres (ASOT) and Anti-DNAase B
- Throat swab for Group A streptococcus
- Renal ultrasound scan for evidence of pre-existing disease

Differential diagnosis

- Sequelae of other bacterial/viral infections
- Chronic renal failure with acute exacerbation
- Henoch-Schönlein purpura
- IgA nephropathy
- Alport hereditary nephritis
- ANCA positive vasculitis, anti GBM disease

IMMEDIATE TREATMENT

- Admit
- Strict fluid balance monitoring and management
- see **Renal failure** guideline
- Treatment of volume overload/hypertension
- furosemide
- see **Hypertension** guideline
- severe cases of fluid overload will require dialysis
- Treatment of abnormal chemistry consequent to renal failure
- see **Renal failure** guideline
- Oral antibiotics: phenoxymethyl penicillin if able to take tablets or amoxicillin suspension (if penicillin allergy azithromycin) for 10 days
- Nutrition: encourage high carbohydrate intake

DISCHARGE FROM HOSPITAL

- BP under good control
- Passing urine normally on free fluids
- Renal function improving
- Normal serum potassium

SUBSEQUENT MANAGEMENT

Follow-up/progress

- Gross haematuria, oliguria and abnormal chemistry usually resolves by 2–3 weeks
- BP usually normal by 3–4 weeks
- Serum C3 usually normal by 4–6 weeks
- Proteinuria resolves by 6 months
- Microscopic haematuria usually resolves by 12 months

Tertiary referral

Refer to nearest paediatric renal centre if:

- Atypical presentation
- Evidence of serious degree of renal failure requiring dialysis
- Poorly-controlled hypertension/cardiac failure/encephalopathy
- Heavy or persistent proteinuria leading to hypo-albuminaemia
- Normal serum C3 at presentation (i.e. not post-streptococcal)
- Failure of normalisation of C3 by 6 weeks, positive ANA, ANCA or anti GBM
- Associated vasculitis
- Delay in recovery as indicated by timescales above
- Recurrent episodes

DISCHARGE FROM FOLLOW-UP

- Normal BP (when not receiving antihypertensive treatment)
- Normal renal function
- Normal urinalysis

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RECOGNITION AND ASSESSMENT

Definition

- Triad of features
  - haemolytic anaemia
  - thrombocytopenia
  - renal insufficiency

Symptoms and signs

- Diarrhoea with blood and mucus (HUS can occur in absence of diarrhoea)
- Vomiting
- Abdominal pain
- Pallor, lethargy
- Bleeding tendency
- Reduced urine output/facial puffiness
- Pallor
- Mucocutaneous bleeding
- Tachycardia
- Reduced consciousness: consider cerebral oedema, intracranial haemorrhage
- Convulsions: consider hyponatraemia, cerebral oedema, intracranial haemorrhage
- Paralysis: consider intracranial haemorrhage
- Over-hydration
  - oedema (periorbital/pedal) variable
  - weight gain, observe trend
  - raised jugular venous pressure (JVP) indicates volume overload (cardiac failure)
  - oliguria (urine output <1 mL/kg/hr)
  - tachypnoea
  - liver enlargement
  - dehydration if diarrhoea has been severe, see **Diarrhoea and vomiting** guideline
  - check BP: hypotension

Investigations

- FBC and blood film (look for fragmented red cells)
  - low Hb and platelets
- Clotting studies
- U&E, creatinine
- Bicarbonate
- Calcium, phosphate, uric acid
- Glucose
- Liver function tests
- *E. coli* 0157 serology acute and convalescent (10 days after onset of symptoms)
- Urine stick test for significant blood and protein (indicating glomerular damage) and leucocytes
- Stool culture for *E. coli* (and typing for 0157 strain)

IMMEDIATE TREATMENT

- Admit, discuss with regional paediatric nephrology team in all cases
- Strict fluid balance monitoring and management
  - see **Renal failure** guideline
- Dehydration
  - if signs of hypovolaemic shock give circulatory support (sodium chloride 0.9% 20 mL/kg IV immediately)
  - correct dehydration, see **Diarrhoea and vomiting** guideline
- Over-hydration
  - if signs of overload/cardiac failure, furosemide IV 2–4 mg/kg (max rate 4 mg/min), repeated 6-hrly if response obtained
  - if furosemide ineffective, discuss dialysis with regional paediatric renal centre
- Hypertension – see **Hypertension** guideline

- Anaemia
  - daily FBC: only transfuse after discussion with regional paediatric nephrology team as may require dialysis. If asymptomatic, Hb can drop as low as 60 g/L
- Thrombocytopenia
  - do not transfuse platelets unless there are life-threatening bleeds
- AVOID antibiotics
- Observe for non-renal complications e.g. encephalopathy and seizures, cardiomyopathy

DISCHARGE FROM HOSPITAL

- Patient may be discharged when:
  - diarrhoea/abdominal pain resolved
  - Hb stable (haemolysis ceased)
  - drinking fluids freely and passing normal amounts of urine
  - urea and electrolytes improving with serum potassium normal

SUBSEQUENT MANAGEMENT

Tertiary referral

- If significant renal impairment (anuria, rising creatinine) dialysis required (see **Renal failure** guideline), refer to regional paediatric renal centre

Follow-up

- Weekly until renal function stable
  - if impaired renal function or proteinuria persists, arrange paediatric renal follow-up
- Once renal function normal, arrange GP or general paediatric follow-up every year to check BP and early morning urine (protein:creatinine ratio) with a detailed renal specialist review every 5 yrs
- Advise that women with history of haemolytic uraemic syndrome require close monitoring during pregnancy

DISCHARGE FROM FOLLOW-UP

- Renal function normal
- No proteinuria
- Renal growth and function satisfactory at 5-yrly review for 15 yr

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RECOGNITION AND ASSESSMENT

Diagnosis is difficult because symptoms can be minimal and often go unrecognised

- Severe hypertension can cause:
  - loss of consciousness
  - convulsion
  - hemiplegia

Definition

- Depends on age, sex and height of child
- Measure on at least 3 separate occasions with ausculatory method
- Normal: systolic and diastolic BP <90th centile for age, sex and height
- High normal: systolic and diastolic BP between 90th and 95th centile for age, sex and height (>120/80 even if below 90th centile in adolescents)
- Stage 1 hypertension: 95th–99th centile PLUS 5 mmHg
- Stage 2 hypertension: >99th centile PLUS 5 mmHg

Symptoms and signs

Hypertension

Listed in order of frequency with common presenting features first:

- Infants
  - congestive cardiac failure
  - respiratory distress
  - failure to thrive, vomiting
  - irritability
  - convulsions
- Older children
  - headaches
  - nausea, vomiting
  - hypertensive encephalopathy (see below)
  - polydipsia, polyuria
  - visual problems
  - tiredness, irritability
  - cardiac failure
  - facial palsy
  - epistaxis
  - poor growth, weight loss
  - cardiac murmur
  - abdominal pain
  - enuresis

Hypertensive encephalopathy (accelerated hypertension)

- Any neurological sign associated with grossly elevated blood pressure, most commonly:
  - severe generalised headache
  - visual disturbance (+/- retinal changes)
  - seizure

Do not delay initiation of treatment pending investigations once diagnosis has been made

History

- Family history of hypertension, diabetes, cardiovascular and cerebrovascular disease, obesity, hereditary renal and endocrine disease
- Past history of renal, cardiac, endocrine or neurological problems
- Presenting complaints as listed above
- Drug intake such as corticosteroids, ciclosporins, tacrolimus, antidepressants

Examination

- Detailed clinical examination of all systems
- **Do not** forget fundoscopy

Investigations

- Check for evidence of renal disease
  - serum creatinine, urea and electrolytes, calcium
  - urinalysis for blood and protein
  - if urine dipstick positive for protein send early morning urine for protein:creatinine ratio
  - renal ultrasound scan
  - DMSA scan may be required to exclude scarring
- Check for cardiovascular causes
  - check femoral pulses
  - right arm and leg blood pressure
  - ECG for left ventricular hypertrophy (LVH)
  - echocardiogram
- Check for endocrine causes
  - fasting plasma glucose
  - lipid profile
  - plasma renin and aldosterone concentration
  - urine catecholamines (contact biochemistry department for details of how to perform test)
  - urine metadrenalines (performed at Manchester Children's Hospital)
  - 24 hr urinary free cortisol and/or discuss with endocrinologist for further investigations

Differential diagnosis

- Incorrectly sized (too small) or placed BP cuff
- Transient hypertension secondary to pain, anxiety, distress

IMMEDIATE TREATMENT

Hypertensive encephalopathy (accelerated hypertension)

Urgent treatment necessary but bring BP under control slowly

Abrupt BP reduction can result in cerebral ischaemia with the risk of permanent neurological sequelae, owing to failure of cerebral auto-regulation after sustained elevation of BP

- Excess BP = actual BP – acceptable BP (**Table 1 and 2**)
  - 'acceptable BP' given by the 90th percentile according to height
- Reduce BP gradually. Aim to reduce 'excess BP' by $^1/_3$ in first 8 hr, another $^1/_3$ in next 12 hr, and final $^1/_3$ in next 48 hr
- Mark target BP ranges on chart so nurses know when to ask a doctor to review
- Monitor perfusion: may need volume expansion in first 12 hr if rapid BP drop
- Discuss choice of drug treatment with **consultant**
- Options comprise in following order: (**Table 3**)
  - **sodium nitroprusside** infusion
    - give in high dependency or intensive care unit as close monitoring required
    - starting dose 500 nanogram/kg/min
    - increase in increments of 200 nanogram/kg/min
    - maximum 8 microgram/kg/min for first 24 hr, reducing to 4 microgram/kg/min thereafter
    - only effective whilst infused as short half-life
    - stop infusion slowly over 15–30 min to avoid any rebound effects

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- **labetalol** infusion
  - starting dose 0.5–1 mg/kg/hr
  - increase by 1 mg/kg/hr every 15–30 min until effective
  - maximum dose 3 mg/kg/hr (max 120 mg/hr)
  - stop infusion when effective
  - restart as BP starts to rise again
  - normally lasts 4–6 hr
- **nifedipine** oral
  - quick acting: use modified release to prevent large drop in BP
  - can be crushed but may have more rapid onset
  - may be used to clip peaks of BP
  - dose varies with product: check with pharmacy

SUBSEQUENT MANAGEMENT

Essential hypertension

- High normal BP
- non pharmacological measures such as weight loss, dietary modification, exercise
- medication (**Table 3**) only if compelling indications such as if symptomatic, diabetes mellitus, heart failure, left ventricular hypertrophy
- Stage 1 hypertension
- non pharmacological measures
- give medications (**Table 3**) if symptomatic, presence of end organ damage, diabetes, persistent hypertension despite non pharmacological measures
- Stage 2 hypertension
- non pharmacological measures
- start medications (**Table 3**)
- add drug therapy only after discussion with a consultant

Renal hypertension

- In children with impaired renal function, keep BP within same target range as for children with normal renal function

OUT-PATIENT MANAGEMENT

Table 1: Blood pressure (BP) for boys by age and height percentiles

		Systolic (mmHg) percentile of height							Diastolic (mmHg) percentile of height						
Age (years)	BP percentile	5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	94	95	97	99	100	102	103	49	50	51	52	53	53	54
	95th	98	99	101	103	104	106	106	54	54	55	56	57	58	58
	99th	105	106	108	110	112	113	114	61	62	63	64	65	66	66
2	90th	97	99	100	102	104	105	106	54	55	56	57	58	58	59
	95th	101	102	104	106	108	109	110	59	59	60	61	62	63	63
	99th	109	110	111	113	115	117	117	66	67	68	69	70	71	71
3	90th	100	101	103	105	107	108	109	59	59	60	61	62	63	63
	95th	104	105	107	109	110	112	113	63	63	64	65	66	67	67
	99th	111	112	114	116	118	119	120	71	71	72	73	74	75	75
4	90th	102	103	105	107	109	110	111	62	63	64	65	66	66	67
	95th	106	107	109	111	112	114	115	66	67	68	69	70	71	71
	99th	113	114	116	118	120	121	122	74	75	76	77	78	78	79
5	90th	104	105	106	108	110	111	112	65	66	67	68	69	69	70
	95th	108	109	110	112	114	115	116	69	70	71	72	73	74	74
	99th	115	116	118	120	121	123	123	77	78	79	80	81	81	82
6	90th	105	106	108	110	111	113	113	68	68	69	70	71	72	72
	95th	109	110	112	114	115	117	117	72	72	73	74	75	76	76
	99th	116	117	119	121	123	124	125	80	80	81	82	83	84	84
7	90th	106	107	109	111	113	114	115	70	70	71	72	73	74	74
	95th	110	111	113	115	117	118	119	74	74	75	76	77	78	78
	99th	117	118	120	122	124	125	126	82	82	83	84	85	86	86
8	90th	107	109	110	112	114	115	116	71	72	72	73	74	75	76
	95th	111	112	114	116	118	119	120	75	76	77	78	79	79	80
	99th	119	120	122	123	125	127	127	83	84	85	86	87	87	88
9	90th	109	110	112	114	115	117	118	72	73	74	75	76	76	77
	95th	113	114	116	118	119	121	121	76	77	78	79	80	81	81
	99th	120	121	123	125	127	128	129	84	85	86	87	88	88	89
10	90th	111	112	114	115	117	119	119	73	73	74	75	76	77	78
	95th	115	116	117	119	121	122	123	77	78	79	80	81	81	82
	99th	122	123	125	127	128	130	130	85	86	86	88	88	89	90
11	90th	113	114	115	117	119	120	121	74	74	75	76	77	78	78
	95th	117	118	119	121	123	124	125	78	78	79	80	81	82	82
	99th	124	125	127	129	130	132	132	86	86	87	88	89	90	90
12	90th	115	116	118	120	121	123	123	74	75	75	76	77	78	79
	95th	119	120	122	123	125	127	127	78	79	80	81	82	82	83
	99th	126	127	129	131	133	134	135	86	87	88	89	90	90	91
13	90th	117	118	120	122	124	125	126	75	75	76	77	78	79	79
	95th	121	122	124	126	128	129	130	79	79	80	81	82	83	83
	99th	128	130	131	133	135	136	137	87	87	88	89	90	94	94
14	90th	120	121	123	125	126	128	128	75	76	77	78	79	79	80
	95th	124	125	127	128	130	132	132	80	80	81	82	83	84	84
	99th	131	132	134	136	138	139	140	87	88	89	90	91	95	92
15	90th	122	124	125	127	129	130	131	76	77	78	79	80	80	81
	95th	126	127	129	131	133	134	135	81	81	82	83	84	85	85
	99th	134	135	136	138	140	142	142	88	89	90	91	92	93	93
16	90th	125	126	128	130	131	133	134	78	78	79	80	81	85	82
	95th	129	130	132	134	135	137	137	82	83	83	84	85	86	87
	99th	136	137	139	141	143	144	145	90	90	91	92	93	94	94
17	90th	127	128	130	132	134	135	136	80	80	81	82	83	84	84
	95th	131	132	134	136	138	139	140	84	85	86	87	87	88	89
	99th	139	140	141	143	145	146	147	92	93	93	94	95	96	97

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Table 2: Blood pressure (BP) for girls by age and height percentiles

		Systolic (mmHg) percentile of height							Diastolic (mmHg) percentile of height						
Age (years)	BP percentile	5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	97	97	98	100	101	102	103	52	53	53	54	55	55	56
	95th	100	101	102	104	105	106	107	56	57	57	58	59	59	60
	99th	108	108	109	111	112	113	114	64	64	65	65	66	67	67
2	90th	98	99	100	101	103	104	105	57	58	58	59	60	61	61
	95th	102	103	104	105	107	108	109	61	62	62	63	64	65	65
	99th	109	110	111	112	114	115	116	69	69	70	70	71	72	72
3	90th	100	100	105	103	104	106	106	61	62	62	63	64	64	65
	95th	104	104	105	107	108	109	110	65	66	66	67	68	68	69
	99th	111	111	113	114	115	116	117	73	73	74	74	75	76	76
4	90th	101	102	103	104	106	107	108	64	64	65	66	67	67	68
	95th	105	106	107	108	110	111	112	68	68	69	70	71	71	72
	99th	112	113	114	115	117	118	119	76	76	76	77	78	79	79
5	90th	103	103	105	106	107	109	109	66	67	67	68	69	69	70
	95th	107	107	108	110	111	112	113	70	71	71	72	73	73	74
	99th	114	114	116	117	118	120	120	78	78	79	79	80	81	81
6	90th	108	105	106	108	109	110	111	68	68	69	70	70	71	72
	95th	104	109	110	111	113	114	115	72	72	73	74	74	75	76
	99th	115	116	117	119	120	121	122	80	80	80	81	82	83	83
7	90th	106	107	108	109	111	112	113	69	70	70	71	72	72	73
	95th	110	111	119	113	115	116	116	73	74	74	75	76	76	77
	99th	117	118	119	120	122	123	124	81	81	82	82	83	84	84
8	90th	108	109	110	111	113	114	114	71	71	71	72	73	74	74
	95th	112	112	114	115	116	118	118	75	75	75	76	77	78	78
	99th	119	120	121	122	123	125	125	82	82	83	83	84	85	86
9	90th	110	110	112	113	114	116	116	72	72	72	73	74	75	75
	95th	114	114	115	117	118	119	120	76	76	76	77	78	79	79
	99th	121	121	123	124	125	127	127	83	83	84	84	85	86	87
10	90th	112	112	114	115	116	118	118	73	73	73	74	75	76	76
	95th	116	116	117	119	120	121	122	77	77	77	78	79	80	80
	99th	123	123	125	126	127	129	129	84	84	85	86	86	87	88
11	90th	114	114	116	117	118	119	120	74	74	74	75	76	77	77
	95th	118	118	119	121	122	123	124	78	78	78	79	80	81	81
	99th	125	125	126	128	129	130	131	85	85	86	87	87	88	89
12	90th	116	116	117	119	120	121	122	75	75	75	76	77	78	78
	95th	119	120	121	123	124	125	126	79	79	79	80	81	82	82
	99th	127	127	128	130	131	132	133	86	86	87	88	88	89	90
13	90th	117	118	119	121	122	123	124	76	76	76	77	78	79	79
	95th	121	122	123	124	126	127	128	80	80	80	81	82	83	83
	99th	128	129	130	132	133	134	135	87	87	88	89	89	90	91
14	90th	119	120	121	122	124	125	125	77	77	77	78	79	80	80
	95th	123	123	125	126	127	129	129	81	81	81	82	83	84	84
	99th	130	131	132	133	135	136	136	88	88	89	90	90	91	92
15	90th	120	121	122	123	125	126	127	78	78	78	79	80	81	81
	95th	124	125	126	127	129	130	131	82	82	82	83	84	82	85
	99th	131	132	133	134	136	137	138	89	89	90	91	91	92	93
16	90th	121	122	123	124	126	127	128	78	78	79	80	81	81	82
	95th	125	126	127	128	130	131	132	82	82	83	84	85	85	86
	99th	132	133	134	135	137	138	139	90	90	90	91	92	93	93
17	90th	122	122	123	125	126	127	128	78	79	79	80	81	81	82
	95th	125	126	127	129	130	131	132	82	83	83	84	85	85	86
	99th	133	133	134	136	137	138	139	90	90	91	91	92	93	93

Table 3: Drugs commonly used for management of hypertension in children

Drug	Mechanism of action	Advice
Atenolol	Beta-adrenoceptor blocker	● Reduces heart contractility – contraindicated in early stages of hypertensive heart failure ● Avoid in confirmed asthmatics
Labetalol	Non-cardioselective beta-blocker with additional alpha-blocking properties	● Combining alpha- and beta-blockade reduces tachycardia that can be a problem without beta-blockade ● Contraindicated in asthmatics and in heart failure
Nifedipine	Calcium channel blocker	● Can be used in heart failure as any negative inotropic effect offset by a reduction in left ventricular work
Enalapril	Angiotensin-converting enzyme (ACE) inhibitor	● Recommended in children with renal hypertension. First dose should be given at night to prevent transient hypotension ● In children with impaired renal function, check serum creatinine and potassium 2–3 days after starting treatment and consider withdrawal if they have risen ● Contraindicated in bilateral renal artery stenosis
Losartan	Angiotensin II receptor blocker	● Second line if enalapril contraindicated or not tolerated ● In children with impaired renal function, check serum creatinine and potassium 2–3 days after starting treatment and consider withdrawal if they have risen ● Contraindicated in bilateral renal artery stenosis
Sodium nitro-prusside	Vasodilator	● Use for hypertensive emergencies ● Avoid in hepatic or renal impairment ● Monitor blood cyanide if used >3 days

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RECOGNITION AND ASSESSMENT

Definition

A triad of features:

- Oedema
- Hypoalbuminaemia: plasma albumin <25 g/L
- Heavy proteinuria, defined as:
  - dipstick 3+ or more, or
  - urinary protein >40 mg/m^2/hr, or
  - early morning protein:creatinine ratio >200 mg/mmol

Symptoms and signs

Oedema

- Peri-orbital, pedal, sacral, scrotal
- Also ascites or pleural effusion

Cardiovascular

Difficult to assess due to oedema

Assess for hypovolaemia carefully

- Child with diarrhoea and vomiting and looks unwell
- Abdominal pain: strongly suggestive
- Poor peripheral perfusion and capillary refill >2 sec
- Pulse character: thready, low volume, difficult to palpate
- Tachycardia or upward trend in pulse rate
- Hypotension: a late sign
- Jugular venous pressure (JVP) low

Muffled heart sounds suggest pericardial effusion

Respiratory

- Tachypnoea and recession: suggest pleural effusion

Abdomen

- Swelling and shifting dullness: suggest ascites
- Tenderness with fever, umbilical flare: suggest peritonitis
- Scrotal oedema: stretching can cause ulceration or infection

Investigations

Femoral blood sampling is contraindicated because of risk of thrombosis

Urine

- Urinalysis
- Early morning urine protein:creatinine ratio first morning after admission
  - normal value <20 mg/mmol; nephrotic >200 mg/mmol, usually >600 mg/mmol

Baseline bloods

- U&E and creatinine
- Albumin
- FBC
- Immunoglobulins G, A and M
- Complement C3 and C4
- Zoster immune status: as a baseline
- Hepatitis B and C serology

Second-line tests

Request only if features suggestive of more aggressive nephritis (hypertension, macroscopic haematuria, high creatinine, no response to corticosteroids)

- Anti-streptolysin O titre and anti-DNase B
- Antinuclear antibodies
- Anti-ds DNA antibodies

Interpretation

- High haematocrit suggests hypovolaemia
- Raised creatinine or urea suggests hypovolaemia, tubular plugging or other nephritis
- Serum cholesterol and triglycerides: often elevated
- IgG usually low
- C3 normal

Differential diagnosis

- Minimal change disease (95%)
- Focal segmental glomerular sclerosis (FSGS)
- Multisystem disorders (e.g. HSP, diabetes mellitus, SLE, and malaria)
- Congenital nephrotic syndrome very rare

IMMEDIATE TREATMENT

General

- Admit
- Strict fluid balance monitoring
- daily weight: **mandatory**
- Avoid added salt, but a low salt diet not indicated
- Manage hypovolaemia – see **Complications**
- seek **senior advice before volume resuscitation,** as a risk of volume overload

Fluid restriction

- Restrict to usual maintenance intake (insensible losses plus output)
- If not tolerated, aim for:
- 600 mL/day in children aged <5 yr
- 800 mL/day in children aged 5–10 yr
- 1000 mL/day in children aged >10 yr

Medication

- Prednisolone 60 mg/m^2 oral once daily (maximum 80 mg), in the morning (see **BNFc** for surface area)
- Phenoxymethylpenicillin (Penicillin V) for pneumococcal prophylaxis
- If oedema upsetting to patient or causing breathlessness, add furosemide 1–2 mg/kg oral or 1 mg/kg IV over 5–10 min
- may intensify hypovolaemia, in which case use 20% albumin: discuss with **consultant**
- If disease severe, especially with hypovolaemia, as judged by poor perfusion, high haemoglobin, thrombophilia, or abdominal pain **or** if relapse for >2 weeks, treat with dipyridamole to reduce risk of thrombotic complications. Discuss need for heparin/warfarin with specialist

COMPLICATIONS

Acute hypovolaemia

- Abdominal pain, looks unwell, tachycardia, poor perfusion, high Hb
- Seek **senior advice before volume resuscitation,** as a risk of volume overload
- give human albumin **4.5%** (if available) 10 mL/kg immediately or sodium chloride 0.9% 10 mL/kg immediately

Do not confuse 4.5% albumin with 20% as the latter is hyperosmolar and can easily cause fluid overload

- Start dipyridamole

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Chronic hypovolaemia

- More common in corticosteroid-resistant disease
- Looks unwell, abdominal pain and vomiting
- Low JVP, rising urea and creatinine, and poor response to diuretics
- Treatment: check with consultant first
- salt-poor hyperosmolar albumin **20%** 0.5–1.0 g/kg (2.5–5.0 mL/kg) over 2–4 hr with furosemide 1–2 mg/kg IV midway through infusion
- regular observations for signs of circulatory overload (e.g. raised JVP, tachycardia, gallop rhythm, breathlessness)
- often required daily or twice daily: liaise with a specialist centre
- Start dipyridamole

Peritonitis

- Difficult to recognise
- steroids may mask signs, including fever, or cause leucocytosis
- Abdominal pain
- consider hypovolaemia and appendicitis: request an early surgical opinion
- Obtain blood culture and peritoneal fluid (for gram stain and culture) if possible, then start piperacillin with tazobactam (Tazocin®) IV pending culture results

Cellulitis

- Commonly caused by haemolytic streptococci and pneumococci – treat promptly

Thrombosis

- Renal vein: an important differential in abdominal pain
- Cerebral vasculature
- Pulmonary vein
- Femoral vein: femoral blood sampling contraindicated
- A fall in platelets, rise in D-Dimers and reduced PTT are suggestive
- USS with Doppler study to look at perfusion and to image renal vein and IVC can be helpful. If in any doubt, seek advice from nephrologist regarding investigation/management

DISCHARGE POLICY AND SUBSEQUENT MANAGEMENT

- Discharge once in remission
- defined as trace/negative urine protein for 3 days
- patients with normal BP and stable weight who are well may be allowed home on ward leave with **consultant approval.** Normally twice weekly review will be required until in remission
- Arrange plan of care with patient and carers – see **below**
- Out-patient review in 4 weeks

New patients

- Prednisolone 60 mg/m^2 (maximum 80 mg) once daily for 4 weeks
- Then 40 mg/m^2 (maximum 60 mg) alternate days for 4 weeks
- gradually reduce dose
- Response usually apparent in 7–10 days
- No response after 4 weeks suggests corticosteroid resistance

Relapsing patients

- Three consecutive days of 3+ or more early morning proteinuria, having previously been in remission
- Start prednisolone 60 mg/m^2 (maximum 80 mg) once daily
- continue until nil or trace proteinuria for 3 days
- then 40 mg/m^2 (maximum 60 mg) alternate days for a further 4 weeks
- If relapses frequent despite alternate-day prednisolone, discuss with a paediatric nephrologist

Oral prednisolone

- While on prednisolone 60 mg/m^2 once daily advise to:
- carry a corticosteroid card
- seek prompt medical attention for illness, especially zoster contacts

Other management

- Urine testing
- teach technique and provide appropriate dipsticks
- test only first daily urine sample
- keep a proteinuria diary
- Corticosteroid diary with instructions regarding corticosteroid dosage

Infectious precautions

- Avoid live immunisations for 3 months after period of treatment with high-dose corticosteroids
- Benefit of inactivated vaccines can be impaired by high-dose corticosteroids and so a similar delay advisable where possible
- where not possible because of frequent relapse, give INACTIVATED vaccines after a shorter delay and check for an antibody response
- Continue penicillin prophylaxis until oedema has resolved
- If zoster non-immune (VZV IgG negative) and on high-dose corticosteroids, give intramuscular zoster immunoglobulin:
- after definite zoster contact, a contact will be infectious 2 days before onset of rash, and cease when all lesions are crusted over
- can be given up to 10 days after exposure. Contact consultant microbiologist on duty for release of VZIG
- at first sign of illness give aciclovir IV
- varicella vaccine (live vaccine) available and should be given if a suitable opportunity arises between relapses
- If child has not received pneumococcal conjugate vaccine – see BNFc for schedule

Refer for specialist advice if:

- Corticosteroid-resistant disease
- non-responsive after 4 weeks of daily prednisolone, but start discussions with specialist centre in third week
- Corticosteroid-dependent disease
- two consecutive relapses during corticosteroid treatment or within 14 days of cessation
- Significant corticosteroid toxicity
- Aged <1 yr or >12 yr at first presentation
- Mixed nephritic/nephrotic picture: macroscopic (not microscopic) haematuria, renal insufficiency or hypertension
- Low complement C3/C4

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RECOGNITION AND ASSESSMENT

Definition

- Presence of crystalline material within urinary tract

Symptoms and signs

- Non-specific recurrent abdominal pain
- Dysuria or painful micturition
- Classical renal colic
- Urinary infection (particularly *Proteus* spp)
- Persistent pyuria
- Macroscopic or microscopic haematuria
- Passage of stones
- Renal failure

Initial investigations

- Renal ultrasound scan
- Urine microscopy and culture

Further investigations

- DMSA scan
  - to determine function when calculi multiple or large
- Repeat renal ultrasound scan
  - to see if stones have been passed
  - to monitor progress of stones
  - six weeks after treatment (see below)

IMMEDIATE TREATMENT

- Analgesia for severe pain
- If obstruction is present, urgent referral to urology at renal specialist centre
- Cefalexin oral if symptomatic for urinary tract infection, adjusted once sensitivities available
  - antibiotic treatment unlikely to eradicate organism in presence of stones

OUT-PATIENT MANAGEMENT

Investigations in patients with proven renal calculi

- Fasting (before breakfast) blood sample for:
  - creatinine
  - calcium
  - phosphate
  - uric acid
  - venous bicarbonate
  - pH (warm arterialised capillary sample to coincide with urine pH)
- Random mid-stream urine
  - microscopy, culture and sensitivity
- Early morning urine (first voided specimen) and 24 hr collection (request 'urinary stone screen' and record height and weight on request form) for:
  - calcium
  - oxalate
  - citrate
  - uric acid
  - cystine
  - creatinine
  - pH (to coincide with blood pH)

Stone analysis

- May give useful information about aetiology, discuss with biochemistry department first
- If stone passage is frequent or associated with symptoms, ask parents to strain urine

Table 1: Characteristics of urinary stones

Type	Appearance	Causes	Radio-opaque*
Magnesium ammonium phosphate	Very soft, white, toothpaste consistency or gravel fragments	● Infection with urea-splitting organisms, especially in children with urinary stasis	No
Calcium oxalate	Hard grey-brown rough surface	● Hypercalciuria (any cause) ● Hyperoxaluria	Yes
Calcium phosphate	Large, smooth, pale, friable	● Infection ● Renal tubular acidosis ● Vitamin D toxicity ● Idiopathic hypercalciuria ● Immobilisation ● Hyperparathyroidism ● Sarcoidosis	Yes
Cystine	Pale-yellow, crystalline Maple syrup	● Cystinuria	Yes
Uric acid	Hard, yellow	● Lesch-Nyhan syndrome ● Dietary ● Induction in haematological malignancies	No
Xanthine	Smooth, soft, brown yellow	● Xanthinuria	No
Dihydroxyadenine	Friable, grey-blue	● Adenine phosphoribosyl transferase deficiency	No

* Radiolucency depends on amount of calcium in the stone and individual patient can have more than one type of stone, each with different radiolucencies

Interpretation of results

- Urinary pH
  - pH <5.3 in presence of normal capillary pH and bicarbonate excludes distal renal tubular acidosis
  - when above criteria not met, a more formal test of renal acidification required in those with nephrocalcinosis or in recurrent stone formers
  - pH >6 with capillary bicarbonate <18 mmol/L is seen in mild distal tubular acidosis
- Calcium:creatinine (mmol/mmol) ratio consistently >0.2 indicates hypercalciuria
- Oxalate:creatinine (mmol/mmol) ratio is age-dependent, and suggestive of hyperoxaluria if it exceeds following thresholds:
  - aged <6 months: 0.35
  - aged 6–11 months: 0.2
  - aged 1–2 yr: 0.18
  - aged 3–6 yr: 0.11
  - aged 7–14 yr: 0.08
  - aged >14 yr: 0.065
- Uric acid/creatinine (mmol/mmol) ratio is age-dependent, and suggestive of hyperuricaemia if it exceeds following thresholds:
  - aged <1 yr: 1.5
  - aged 1–2 yr: 1.26
  - aged 3–6 yr: 0.83
  - aged 7–10 yr: 0.67
  - aged 11–14 yr: 0.45
  - aged >14 yr: 0.4
- Magnesium:creatinine ratio <0.2 may increase stone formation

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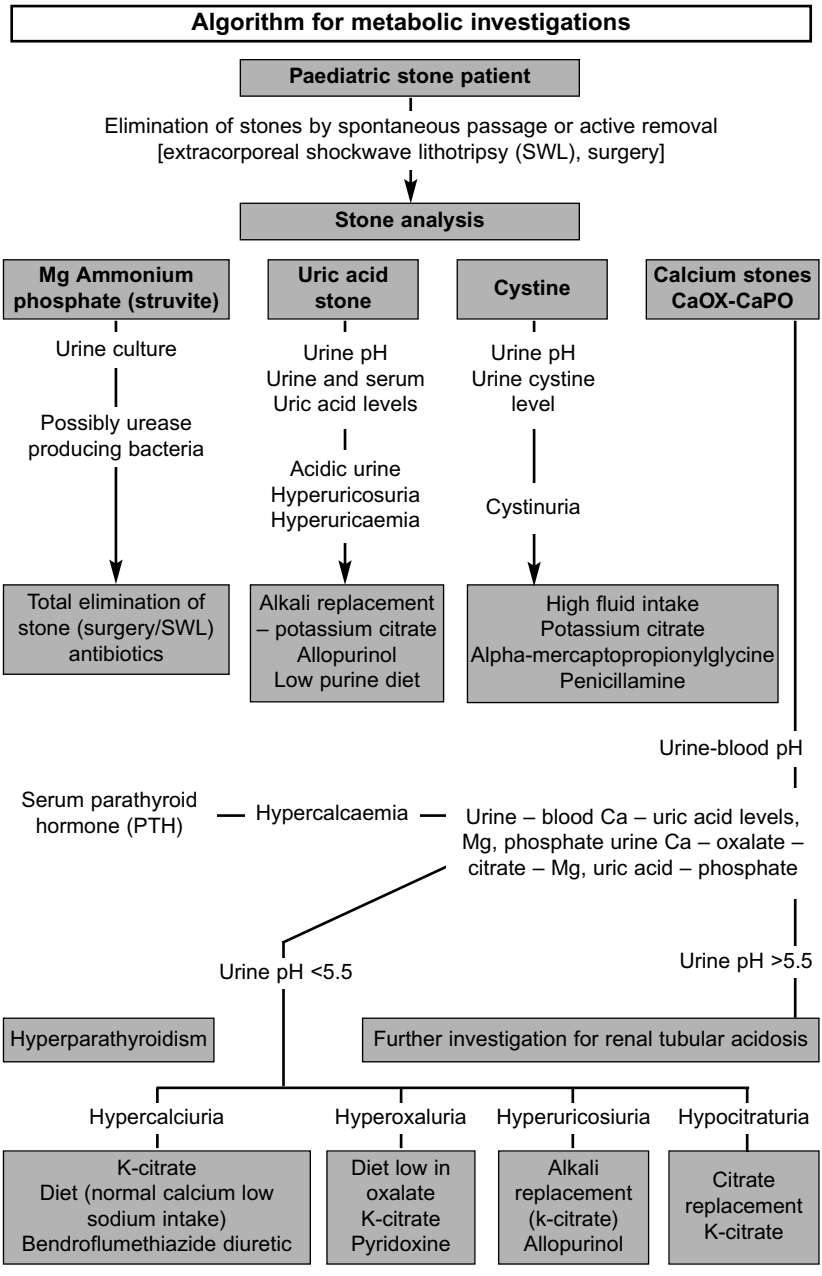
- Calcium:citrate ratio <0.6 may increase stone formation
- Cystine, if present, is indicative of cystinuria
- Overall solubility index (RS value)
  - negative value: stable urine
  - value 0–1: metastable (liable to precipitate if seeded)
  - value >1: spontaneous precipitation

TREATMENT

- Treat any metabolic disorder identified by above investigations, seek advice from regional nephrology service
- Keep urine free from infection, particularly in those with history of Proteus infection by prompt treatment if symptomatic
- Advise liberal fluid intake
  - adolescent 3 L/day
  - pre-puberty (school age) 1.5 L/day
- Additional measures for recurrent stone formation or idiopathic hypercalciuria (in order):
  - dietary assessment to optimise oxalate, vitamin C, calcium, and vitamin D intake
  - reduced sodium intake in idiopathic hypercalciuria, if sodium excretion >3 mmol/kg/day
  - high fibre diet with cellulose or whole wheat flour to reduce calcium and oxalate absorption
  - potassium citrate at a starting dose of 0.5 mEq/kg 12-hrly in patients with low urinary citrate
  - bendroflumethiazide 1–2 mg/kg/day to reduce calcium and oxalate excretion (unlicensed). Usual dose 50–100 microgram/kg/day if aged <2 yr, 50–400 microgram/kg/day if aged >2 yr; then maintenance of 50–100 microgram/kg up to max 10 mg, aged 12–18 yr 5–10 mg/day
  - pyridoxine can be used in hyperoxaluria
  - alpha-mercaptopropionylglycine or penicillamine may be useful for cystine stones under specialist recommendation as can cause bone marrow suppression and nephrotic syndrome
- For large stones that are unlikely to pass, surgical removal or lithotripsy may be required
  - modality of treatment determined by location and size of stone
  - generally, stones <2 cm suitable for lithotripsy
  - larger stones treated by percutaneous nephrolithotomy (PCNL) or by open operation
  - nephrectomy may be advised where kidney function poor



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RECOGNITION AND ASSESSMENT

Definition

- Acute renal failure: sudden deterioration in renal function associated with retention of nitrogenous waste and acute disturbance of water and electrolyte balance

Presentation

- Poor/absent urine output (oliguria) with puffiness/oedema:
  - neonates <0.6 mL/kg/hr
  - infant/child <0.5 mL/kg/hr

Differential diagnosis

Pre-renal

- Secondary to hypotension (e.g. hypovolaemia from gastroenteritis or septicaemia)
- Urine osmolality >300 mOsm/kg
- Urine:plasma urea ratio >5
- Urine sodium <20 mmol/L
- Good response to diuretics after correction of hypovolaemia

Renal

- Haemolytic uraemic syndrome – see **Haemolytic uraemic syndrome** guideline
- Acute nephritis – see **Glomerulonephritis** guideline
- Acute tubular necrosis or renal vein thrombosis
- Unrecognised chronic renal failure (oliguria usually not a feature)
- Acute-on-chronic renal failure (e.g. dehydration or infection)

Post-renal

- Urinary tract obstruction (rare)

Assessment

- Hydration (under/over)
- Weight
- Skin (turgor/oedema)
- Ascites
- BP/capillary refill
- Jugular venous pressure (JVP)
- Urine output

Immediate investigations

- See separate **guidelines for specific causes**
- Blood
  - U&E, creatinine, calcium, phosphate, uric acid, magnesium, LFT's and bicarbonate
  - FBC
  - venous blood gas
- Urine
  - urinalysis for blood, protein, nitrites and leucocytes
  - osmolality
  - electrolytes
- Renal ultrasound scan
  - size and appearance of kidneys
  - inflammation and swelling
  - evidence of obstruction

IMMEDIATE TREATMENT

- Correct volume status and maintain fluid and electrolyte balance
- Prevent hyperkalaemia
- Treat underlying cause where appropriate
- Maintain adequate nutrition

Fluid and sodium balance

Initial correction

- Dehydration
  - for shock, give sodium chloride 0.9% 20 mL/kg immediately
  - for correction of dehydration – see **Diarrhoea and vomiting** guideline
- Volume overload/hypertension
  - low serum sodium usually indicates fluid overload
  - furosemide 1 mg/kg IV immediately (max rate: 500 microgram/kg/min up to 4 mg/min): if no urine output after 30 min, give a further 1 mg/kg and if still no urine a third 1 mg/kg after 30 min

Metabolic acidosis

- Sodium bicarbonate may be required – discuss with on-call consultant

Potassium

- Hyperkalaemia can lead to cardiac arrest or serious arrhythmias
- severely restrict potassium intake by introducing low potassium diet and avoiding potassium in IV fluids unless serum potassium <3.5 mmol/L or there are ongoing losses
- If potassium >6.0 mmol/L, ECG monitoring essential
- watch for development of prolonged P-R interval and/or peaked T wave
- as toxicity worsens, P wave is lost, QRS widens and S-T depression develops
- Once toxicity develops, the following (see **Table 1**) are holding measures whilst dialysis is set up
- give salbutamol IV or by nebuliser if no IV access as first-line emergency treatment, followed by oral/rectal calcium polystyrene sulphonate (even if salbutamol effective) to start to reduce potassium load
- if ECG still unstable, give calcium gluconate by slow IV injection
- if patient acidotic pH <7.30, give sodium bicarbonate
- if further reduction required after other measures implemented, use insulin and glucose
- After starting treatment discuss with on-call consultant

Table 1: Emergency treatment of hyperkalaemia

Treatment	Dose	Onset	Mode of action
Salbutamol nebuliser	2.5 mg (<25 kg) 5 mg (>25 kg)	5 min. Lasts up to 2 hr; repeat as necessary	Shifts potassium into cells
Salbutamol infusion	4 microgram/kg over 5 min repeat as necessary. Limited by tachycardia	Immediate. Effect maximal at 60 min	Shifts potassium into cells
Calcium gluconate 10%	0.5 mL/kg IV (max 20 mL) over 5–10 min. Monitor ECG Do NOT administer through same line as bicarbonate	1 min Repeat after 5 min if ECG changes persist	Antagonises effect of high potassium
Sodium bicarbonate 4.2% infusion (only if patient acidotic)	1 mmol/kg IV over 15 min (2 mL/kg of 4.2% diluted 1 in 5 with sodium chloride 0.9%) Do NOT administer through same line as calcium	1 hr Effect may last 2 hr	Shifts potassium into cells
Glucose/insulin infusion	Glucose 10% 0.5 g/kg/hr (5 mL/kg/hr) and when blood glucose >10 mmol/L infuse insulin 0.1 units/kg/hr stop when glucose stops when K^+ falls by 0.5 mmol/L	15 min. Effect may last several hours Frequent glucose stick checks	Shifts potassium into cells
Furosemide	1 mg/kg IV over 5 min	May not be effective in chronic renal failure	Potassium excreted in urine
Polystyrene sulphonate resins	Calcium polystyrene sulphonate 250 mg/kg 6-hrly (max 15 g/dose) oral/rectal 6–8 hrly	Oral 2 hr Rectal 30 min (irrigate to remove residue before next dose)	Removes potassium from body

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- Hypokalaemia is also dangerous
  - if patient becomes potassium depleted from heavy ongoing losses (fistula or diuretic phase), it is most important that replacement is given
  - amount and rate of replacement depend on estimation of losses and response to initial supplementation. If in doubt, discuss with **on-call consultant**

SUBSEQUENT MANAGEMENT

Fluid and sodium balance

- Once normal hydration restored, aim to replace insensible loss (300–400 mL/m^2/day) + urine output + other losses
- In anuric patients (as opposed to oliguric), give fluids that are free of electrolytes to compensate for insensible loss; in patients having IV fluids, glucose 5% is most appropriate initially, although glucose 4%/sodium chloride 0.18% may be required later to compensate for sodium loss from sweat
- Replace sodium losses in urine and in other fluids (diarrhoea, gastric aspirate, fistula)
  - in most patients, dietary sodium will suffice
  - in those with large fluid losses, consider IV sodium to match losses

Nutrition

- Involve a paediatric dietitian
- A low-protein high-energy diet is ideal (aim for energy intake of 400 kcal/m^2)
- Avoid high potassium foods
- Be realistic about what a child will take

Indications for dialysis

- Fluid overload
- Uncontrolled hypertension (for height-related 97th centiles – see **Hypertension** guideline)
- Potassium toxicity (as indicated by features listed previously)
- Metabolic acidosis (pH <7.2) unresponsive to base supplementation
- Convulsions
- Loss of general well being +/- alteration in conscious level – see **Glasgow coma score** guideline
- Spontaneous resumption of renal function likely to be delayed
  - acute-on-chronic renal failure
  - haemolytic uraemic syndrome

MONITORING TREATMENT

- Accurate fluid balance – maintain strict input-output chart
- Re-assess fluid intake at least 12-hrly
- Record weights, plasma sodium and PCV as indicators of hydration
- Check K^+ hourly if >6 or <3 mmol/L
- Check U&E 12-hrly if K^+ 3–6 mmol/L in renal failure
- Respond promptly to increase in urine volume, fall in serum creatinine and increase in urine osmolality by increasing fluid intake to prevent prolonged oliguria
- Once diuresis begins, increase electrolyte replacement, including potassium
  - once stable, reduce fluid intake gradually to avoid prolonged diuretic phase

PROTEIN EXCRETION

- As a diagnostic indicator in any child thought to have an underlying renal disorder
- To monitor progress in renal disorders
- Normally glomerular, rarely tubular in origin
- Investigate as below in patients with persistent proteinuria where cause is unknown
- Request protein:creatinine ratio (**must** be first urine specimen voided in the morning); elevation confirms glomerular proteinuria

Protein:creatinine ratio

- Best performed on first urine specimen voided in the morning
- Upper limit of normal <20 mg/mmol
- Significant proteinuria >100 mg/mmol
- Heavy proteinuria (nephrotic) >200 mg/mmol

Timed urine collection

- Only appropriate for older patients (out of nappies)
- Night-time collection to rule out orthostatic proteinuria
- empty bladder at bedtime and discard sample
- collect all urine passed during the night
- empty bladder on rising in morning and collect urine
- record time from bladder emptying at night to bladder emptying in morning
- Calculate protein output as mg/m^2/hr (see BNFc for surface area)
- Upper limit of normal = 2.5 mg/m^2/hr
- Heavy proteinuria >40 mg/m^2/hr

Tubular proteinuria

- Request retinol binding protein (RBP):creatinine ratio, elevation confirms tubular proteinuria

OSMOLALITY

- Used to exclude urinary concentrating disorders
- patients with polyuria (may present as wetting or excessive drinking)
- Test early morning urine after overnight fast >870 mOsm/kg virtually excludes a concentrating defect
- if concern re diabetes insipidus, do water deprivation tests during the day

SODIUM EXCRETION

- Fractional sodium excretion (FE_{Na}) assesses capacity to retain sodium
- ensure normal sodium intake (dietitian to advise)
- stop any existing supplements 6 hr before taking samples
- document weight loss after supplements stopped, may provide useful supporting evidence
- random urine sample for urinary sodium (U_{Na}) and creatinine (U_{cr})
- blood sample immediately after voiding for plasma sodium (P_{Na}) and creatinine (P_{cr})
- enter results into equation (using same units for U and P; 1000 micromol = 1 mmol)

- $FE_{Na} = \dfrac{U_{Na} . P_{Cr}}{P_{Na} . U_{Cr}} \times 100$
- normal values for FE_{Na}
  aged 0 to 3 months <3
  aged >3 months <1

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PLASMA CREATININE

- Mean and upper limit dependent on height but can be determined roughly from child's age if height not available

Table 1

Age	Height (cm)	Mean (µmol/L)	Upper limit
Up to 2 weeks		66	87
2 weeks to 6 months	50	44	58
6 months to 1 yr	60	34	49
2 yr	87	28	39
4 yr	101	33	43
6 yr	114	38	52
8 yr	126	42	57
10 yr	137	46	62
12 yr	147	52	69
Adult female	163	68	89
Adult male	174	86	108

GLOMERULAR FILTRATION RATE (GFR)

- Serial measurements of glomerular filtration rate (in mL/min/1.73 m^2) predict rate of deterioration when renal function impaired

Table 2

Age	Mean GFR (mL/min/1.73 m^2)	Range (2 SD)
Up to 1 month	48	28–68
1–6 months	77	41–103
6–12 months	103	49–157
1–2 yr	127	63–191
2–12 yr	127	89–165

Plasma creatinine method

- Estimates GFR in children with reasonable accuracy from P_{Cr} and height, using following formula:

  GFR (mL/min 1.73 m^2) =
  $$\frac{40 \times \text{height (cm)}}{P_{Cr}\ (\mu\text{mol/L})}$$
- Not suitable for children:
  - aged <3 yr
  - with muscle disease/wasting

^{51}Cr-EDTA slope clearance

- Use only when GFR needs to be determined very accurately
- Request via nuclear medicine
- Provide height and weight of child
  - 'correct' result for surface area and expressed as per 1.73 m^2
  - if result expressed as mL/min 'correct' for surface area

ULTRASOUND

Indications

- To indentify structural abnormalities of urinary tract

Table 3: Normal values for renal ultrasound measurement

Age	Length (mm)	Range (mm)
Up to 3 months	45	35–60
3–6 months	50	50–60
6–9 months	55	52–60
9–12 months	58	54–64
1–3 yr	65	54–72
3–6 yr	75	64–88
6–9 yr	80	73–86
9–12 yr	86	73–100

- Measurements of pelvicalyceal size at hilum of kidney (during 3rd trimester):
  - <9 mm: mild (do not need any intervention/follow-up)
  - 9–15 mm: moderate
  - >15 mm: severe

ISOTOPE SCANS

Dynamic imaging (MAG3)

Indications

- To assess obstruction in dilated system
- To assess drainage 6 months after pyeloplasty
- Indirect cystography in older children before and/or after surgical correction of reflux

Operational notes

- Request via nuclear medicine
- SHO or nurse required to insert venous cannula in young children
- Consider sedation if child has had previous problems lying still during examinations
- Maintain good hydration
- When assessing obstruction in dilated system or outcome of pyeloplasty, give furosemide 0.5 mg/kg slow IV bolus over 3–10 min (max rate 4 mg/min) 15 min before giving isotope. Helps to differentiate genuine obstruction from isotope pooling, provided function of affected kidney not severely impaired
- Do not use furosemide for indirect cystography

Static imaging (^{99m}Tc-DMSA)

Indications

- To assess differential function between kidneys and within duplex kidneys
- To locate an ectopic kidney
- To identify renal scars after recovery from acute infection
  - atypical UTI aged <3 yr or recurrent UTI any age

Operational notes

- Request via nuclear medicine
- Scan kidney 2–6 hr after injection
- Sedation rarely required
- Delay DMSA for 3–6 months after infection to avoid false positive

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X-RAY IMAGING

Micturating cystourethrogram (MCUG)

To assess bladder for vesicoureteric reflux

Indications

- Atypical or recurrent UTI aged <6 months
- Recurrent or atypical UTI in children aged >6 months, but <3 yr if:
  - dilatation on ultrasound
  - poor urine flow
  - non-*E. coli* infection
  - family history of VUR

Operational notes

- Give prophylactic antibiotics oral for 3 days with MCUG taking place on the second day
- Urethral catheter will need to be passed in X-ray dept

RECOGNITION AND ASSESSMENT

Treat symptomatic urinary tract infection (UTI) in infants promptly to reduce risk of renal scarring

Symptoms and signs

Age group		Most common	Intermediate	Least common
Infants aged <3 months		● Fever ● Vomiting ● Lethargy ● Irritability	● Poor feeding ● Failure to thrive	● Abdominal pain ● Jaundice ● Haematuria ● Offensive urine
Infants and children aged ≥3 months	**Pre-verbal**	● Fever	● Abdominal pain ● Loin tenderness ● Vomiting ● Poor feeding	● Lethargy ● Irritability ● Haematuria ● Offensive urine ● Failure to thrive
	Verbal	● Frequency ● Dysuria	● Dysfunctional voiding ● Changes to continence ● Abdominal pain ● Loin tenderness	● Fever ● Malaise ● Vomiting ● Haematuria ● Offensive urine ● Cloudy urine

Risk factors for UTI and serious underlying pathology

- The following should always be recorded in suspected cases of UTI:
  - poor urine flow in males
  - history suggesting recurrent UTI
  - recurrent fever of uncertain origin
  - antenatally diagnosed renal or urinary tract abnormality
  - family history of vesico-ureteric reflux (VUR) or renal disease
  - constipation
  - dysfunctional voiding
  - enlarged bladder
  - abdominal mass
  - evidence of spinal lesion
  - poor growth
  - high blood pressure

Investigations

- Dipstick test fresh urine for leukocytes and nitrites in:
  - all symptomatic children (see **Table** above)
  - all unexplained febrile admissions with temp >38ºC
  - with an alternate site of infection but who remain unwell
- Culture urine if:
  - aged <3 yr
  - a single positive result for leukocyte esterase or nitrite
  - recurrent UTI
  - infection that does not respond to treatment within 24–48 hr
  - clinical symptoms and dipstick tests do not correlate
  - suspected pyelonephritis
- If child seriously unwell, measure serum electrolytes, take blood cultures and insert cannula

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Collection of specimens

- Do not delay treatment if a sample cannot be obtained and child at high risk of serious illness
- **Clean catch** in sterile container is recommended method:
  - in babies too young to co-operate, eliciting lateral abdominal reflex may provoke micturition
- Collect mid-stream urine in those old enough to co-operate
- Pad urine specimens can be used in babies and young children (only useful if negative)
  - make sure napkin area thoroughly cleaned before applying pad
  - urine extracted from specially designed pads with a syringe
  - always follow manufacturer's instructions
  - do not use cotton wool balls or 'home made' equipment
  - for urinalysis (do not send for culture: if +ve nitrites and +ve leukocytes collect another urine sample by clean method)
- Suprapubic aspiration only required to obtain urgent specimens in very ill child or where there is continuing diagnostic uncertainty
  - always check there is urine in bladder by ultrasound first
  - lie patient supine with legs held in frog position by assistant
  - cleanse suprapubic skin with alcohol
  - use 21G 3.5 cm needle
  - insert midline 1–2 cm above symphysis pubis, with needle perpendicular to skin
  - advance needle whilst applying gentle suction, urine aspirated on insertion

Handling specimens

- Use plain white-topped sterile bottles for hospital-collected samples
- Use borate (red top) only when child large enough to fill bottle
- Keep specimen in fridge at 4ºC until transfer to laboratory
- During working hours, transfer specimens to laboratory within 2 hr
  - out-of-hours, keep specimen in fridge until laboratory open
- State date and time of collection on specimen bottle

Interpretation of results

Always take clinical symptoms into account when interpreting results

- **Children aged ≥3 yr:** use dipstick to diagnose UTI
- **Both leukocyte esterase and nitrite positive:** start antibiotic treatment for UTI
- **Leukocyte esterase negative and nitrite positive:** start antibiotic treatment, if fresh sample was tested. Send urine sample for culture
- **Leukocyte esterase positive and nitrite negative:** only start antibiotic treatment for UTI if there is good clinical evidence of UTI. Send urine sample for microscopy and culture
- **Both leukocyte esterase and nitrite negative:** do not send urine sample for culture unless recommended in indications for culture. Do not start treatment for UTI

Microscopy of fresh sample

- Indications:
  - aged <3 yr with fever
  - aged >3 yr, fever with:
    - specific urinary symptoms
    - history of recurrent UTI
    - seriously ill
    - leukocyte esterase or nitrite on urinalysis (see **Interpretation of results**)

- Very useful method of confirming acute infection
- bacteria and leukocytes (UTI)
- bacteria only (UTI or contaminant)
- leukocytes only (treat if symptomatic)
- no bacteria or leukocytes (no UTI)
- Pyuria
- normal <10 x 10^6/L
- vulvitis, vaginitis or balanitis can also give rise to high counts
- viruses (echovirus, adenovirus and CMV) can cause sterile pyuria
- Colony counts
- organism count >10^5 organisms/mL pure growth confirms infection in properly collected and stored mid-stream sample
- certainty reduced to 80% with pad urine
- low counts do not exclude infection

IMMEDIATE TREATMENT

If child systemically unwell, do not delay treatment while trying to obtain urine specimen

- Ensure good hydration with maintenance fluids
- Empiric antibiotics (narrow spectrum as soon as organism and sensitivities known)
- If pyelophephritis: systemic illness (fever >38ºC or loin pain/tenderness)
- aged <3 months: cefotaxime, aged >3 months: co-amoxiclav oral if tolerated or IV for 7 days
  - if penicillin allergy give gentamicin IV (once daily dosage regimen) over 30 min for 48 hr minimum
  - if shocked refer to **Septicaemia** guideline
  - ongoing treatment depends on response
- if cystitis: minor systemic disturbance, give cefalexin oral for 3 days
- high rates of trimethoprim resistance (no longer first line)
- when child on prophylaxis already, always give an alternative antibiotic for acute infection
- Imaging: urgent ultrasound imaging is only indicated in 'atypical' cases with:
- seriously ill child
- poor urine flow
- abdominal or bladder mass
- raised creatinine
- septicaemia
- failure to respond to treatment within 48 hr
- infection with organisms other than *E. coli*

SUBSEQUENT MANAGEMENT

Imaging

Dependent of age and type of infection

- Simple UTI: responds within 48 hr
- Atypical UTI:
- seriously ill child
- poor urine flow
- abdominal or bladder mass
- raised creatinine
- septicaemia
- failure to respond to treatment within 48 hr
- infection with organisms other than *E. Coli*
- Recurrent UTI:
- 2 or more episodes of UTI with acute pyelonephritis/upper urinary tract infection
- one episode of UTI with acute pyelonephritis/upper urinary tract infection **plus** one or more episode or UTI with cystitis/lower urinary tract infection
- 3 or more episodes or UTI with cystitis/lower urinary tract infection

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Test	Simple UTI	Atypical UTI	Recurrent UTI
Aged 0–6 months			
US during acute infection	No	Yes	Yes
US within 6 weeks	Yes	No	No
DMSA	No	Yes	Yes
MCUG	No	Yes	Yes
Aged 6 months–3 yr			
US during acute infection	No	Yes	No
US within 6 weeks	No	No	Yes
DMSA	No	Yes	Yes
MCUG	No	No	No
Aged >3 yr			
US during acute infection	No	Yes	No
US within 6 weeks	No	No	Yes
DMSA	No	No	Yes
MCUG	No	No	No

- US 6 weeks after infection when not indicated urgently (see above)
- Bladder scan pre/post micturition helpful to exclude incomplete bladder emptying
- DMSA (dimercaptosuccinic acid) scan 6 months after infection
- If child has subsequent UTI while awaiting DMSA, review timing of test and consider doing it sooner
- MCUG (micturating cysto-urethrography) 6 weeks after infection
  - also required where there are voiding problems or abnormalities on US scan requiring further investigation (discuss with consultant)
  - requires 3 days of prophylactic antibiotics, usually nitrofurantoin aged >3 months or cefalexin aged <3 months at night (max 100 mg) according to previous culture sensitivities, with test on middle day; MCUG for neonates with hydronephrosis give a single dose of IV gentamicin 5 mg/kg over 3–5 min just before MCUG (avoid MCUG in neonates with UTI)

DISCHARGE AND FOLLOW-UP

- Home when:
  - symptoms mild, or severe symptoms controlled
  - taking oral antibiotics and tolerating them
  - discuss and advise to avoid risk factors at discharge:
    - constipation
    - poor perineal hygiene
    - low fluid intake
    - infrequent bladder emptying
- Repeat urine test not required on asymptomatic children
- Prompt treatment of recurrences with co-amoxiclav
- Out-patient review
  - not required for simple UTI
  - in 8–10 weeks where ultrasound imaging has been indicated
- Prophylactic antibiotics
  - not required following first simple UTI

- Required for:
  - proven grade 3+ reflux until aged 2 yr (provided infections well controlled)
  - urinary tract obstruction pending surgical management
  - any child with frequent symptomatic infections (>3 urinary tract infections per year)
  - aged >3 months: nitrofurantoin 1 mg/kg oral at night (max 100 mg)
- Surgical management
  - antireflux surgery not routinely indicated in VUR
  - refer for antireflux surgery for obstructive mega-ureters with reflux
  - refer for antireflux surgery if failure to control infections with prophylaxis in grade 3+ reflux
  - refer all neuropathic bladder patients
- Circumcision may be considered for recurrent UTI in children with structurally abnormal urinary tracts

Management of children with renal scars

- No follow-up for minor unilateral parenchymal defect unless recurrent UTI or family history or lifestyle risk factors for hypertension
- In cases of significant scarring:
  - annual BP measurement
  - females must book early when pregnant and inform obstetric team
- Where scarring bilateral:
  - annual BP measurement
  - assessment of urinary protein excretion and renal function every 3–4 yr
  - long-term follow-up in the renal clinic
  - transfer to adult service

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RECOGNITION AND ASSESSMENT

Definition

- Acute, chronic or recurrent inflammation of a one or more joints

Symptoms and signs

- Pain
- Stiffness
- Refusal to participate in usual activities
- Swollen, hot, red and/or tender joint
- Reduced range of movement

Differential diagnosis

- Juvenile idiopathic arthritis (JIA):
  - arthritis of unknown aetiology before age 16 yr (peak aged 1–5 yr)
  - persisting for ≥6 weeks
  - morning irritability, stiffness, gradual refusal to participate in usual activities
  - relatively little pain
  - any or multiple joint (rarely hip initially)
- Reactive arthritis
  - history of diarrhoea (salmonella, shigella, campylobacter)
  - viral illness (parvo, EBV, mumps, rubella)
  - monoarthritis of large joint
  - 7–14 days after acute illness
  - self-limiting in response to an infection
- Reiters syndrome: conjunctivitis, sterile urethritis
- Rheumatic fever (migratory arthritis, history of tonsillitis)
- Non-accidental injury (NAI)
- Systemic rheumatic diseases, such as SLE, dermatomyositis, vasculitis (including HSP and Kawasaki disease)
- Arthritis associated with inflammatory bowel disease
  - monoarthritis in large joint or peripheral arthritis associated with bowel disease activity
- Malignancy, especially leukaemia or neuroblastoma
  - bone pain, lymphadenopathy, hepatosplenomegaly
- Rickets and other endocrine disease (e.g. type 1 DM, thyroid disease)
- Acute septic arthritis (if fever even if history of chronic arthritis see **Osteomyelitis and septic arthritis** guideline)
- Infectious causes (e.g. TB, rheumatic fever, Lyme disease)
- Serum sickness following drug ingestion associated with urticarial rash

Rarer causes

- Inherited metabolic diseases and other genetic disorders
- Chronic recurrent multifocal osteomyelitis
- Auto-inflammatory diseases, including chronic infantile neurological cutaneous and arthritis syndrome
- Haemophilia

INVESTIGATIONS

- X-rays of joints most affected if child has features of other differential diagnoses that have radiological changes and, if severe, as a baseline assessment to look for erosions
- FBC, ESR, CRP, ASOT, rheumatoid factor, ANA and if SLE suspected, ds-DNA auto-antibodies to exclude differential diagnoses or for JIA classification purposes (not useful to confirm the diagnosis of JIA)

ACUTE MANAGEMENT

- **Telephone** local paediatric rheumatology team for advice for management of musculoskeletal conditions and assessment of pyrexia of unknown origin
- **Analgesia**/anti-inflammatory medications vary in individual side effects and clinical effectiveness
- Use with caution in asthma, angioedema, urticaria, rhinitis, coagulation defect, cardiac, hepatic or renal impairment
- Contraindicated in gastro-intestinal ulceration or bleeding
- Give a proton pump inhibitor if taking other medicines that increase the risk of upper GI side-effects or with serious co-morbidity

If JIA is a possible diagnosis, arrange early referral to local ophthalmologist to start screening program for uveitis, chronic anterior uveitis can be asymptomatic initially and can progress to irreversible loss of vision if referral delayed

DISCHARGE AND FOLLOW-UP

- **Refer** all children with suspected JIA and autoinflammatory connective tissue diseases (e.g. SLE, dermatomyositis, scleroderma and sarcoidosis) to paediatric rheumatology service for urgent appointment
- Management will involve:
  - exploring differential diagnoses
  - disease education
  - physiotherapy and rehabilitation
  - optimising medical treatment including corticosteroid joint injections (nearly always under general anaesthetic), methotrexate, and the institution of shared care monitoring

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INTRODUCTION

Differential diagnosis varies with age (see also **Arthritis** guideline)

Common/important diagnoses

Any age	● Trauma (including NAI) ● Septic arthritis ● Reactive arthritis ● Juvenile idiopathic arthritis (JIA) ● Malignancy ● Referred pain (e.g. from hip to knee)
Aged 0–4 yr	● Developmental dysplasia of hip (DDH) ● Transient synovitis ● Non-accidental injury (NAI)
Aged 4–10 yr	● Perthe's ● Transient synovitis
Aged 11–16 yr	● Slipped upper femoral epiphysis (SUFE) ● Gonococcal septicaemia

Irritable hip (transient synovitis)

- Commonest reason for a limp in the pre-school age group
- Usually occurs aged 3–8 yr
- History of recent viral URTI (1–2 weeks)
- Child usually able to walk but with pain
- Child otherwise afebrile and well
- Mild-moderate decrease in range of hip movement, especially internal rotation
- Severe limitation of hip movement suggests septic arthritis
- Exclude septic arthritis: discuss with orthopaedics

Perthes disease

- Avascular necrosis of the capital femoral epiphysis
- Age range 2–12 yr (majority 4–8 yr)
- 20% bilateral
- Present with pain and limp
- Restricted hip motion on examination

Slipped capital femoral epiphysis

- Late childhood/early adolescence
- Weight often >90th centile
- Presents with pain in hip or knee and associated limp
- The hip appears externally rotated and shortened
- Decreased hip movement, especially internal rotation
- May be bilateral

HISTORY

Ask about:

- Trauma
- Fever: shivering/sweating
- Recent viral illness
- Swollen joints
- Stiff joints
- Sickle cell
- Delayed presentation

EXAMINATION

General

Look for:

- Fever
- Rash
- Pallor
- Bruising
- Impaired growth

Musculoskeletal

Check:

- Gait
- Joint swelling
- Range of movement
- Leg lengths
- Weakness
- Spinal configuration/movement

INITIAL INVESTIGATIONS

- FBC and film
- ESR
- CRP
- If febrile, blood cultures
- X-ray symptomatic joint (and 'normal' side) and, if origin of pain uncertain, request X-rays of adjacent joints
  - where SUFE a possibility, request AP and frog views of hips
  - if effusion suspected, confirm with ultrasound scan
- Other investigations (e.g. muscle enzymes, bone scan) may be useful – dependent on clinical assessment

MANAGEMENT

- If there are any features consistent with septic arthritis:
  - severe pain or local tenderness
  - range of movement <75% normal
  - temperature >37.5ºC
  - WBC >13 x 10^9/L
  - ESR >20 mm/first hr
  - CRP >10 mg/L
  - effusion on USS

OR

  - X-ray abnormal or suggests orthopaedic problem (e.g. Perthe's, SUFE)
- Refer to orthopaedics for diagnostic aspiration/washout – **before** starting antibiotics (see **Osteomyelitis and septic arthritis** guideline)

DISCHARGE AND FOLLOW-UP

- If blood tests and X-ray normal, irritable hip (reactive arthritis) likely
  - discharge with analgesia and reassurance
  - advise return if fever occurs or problem becomes worse

Review after 5 days

- If worse, refer for orthopaedic opinion
- If no worse, review after a further 5 days
- If still no better, arrange joint orthopaedic/paediatric review, and consider referral for paediatric rheumatology opinion
- If normal at 5 or 10 days, discharge

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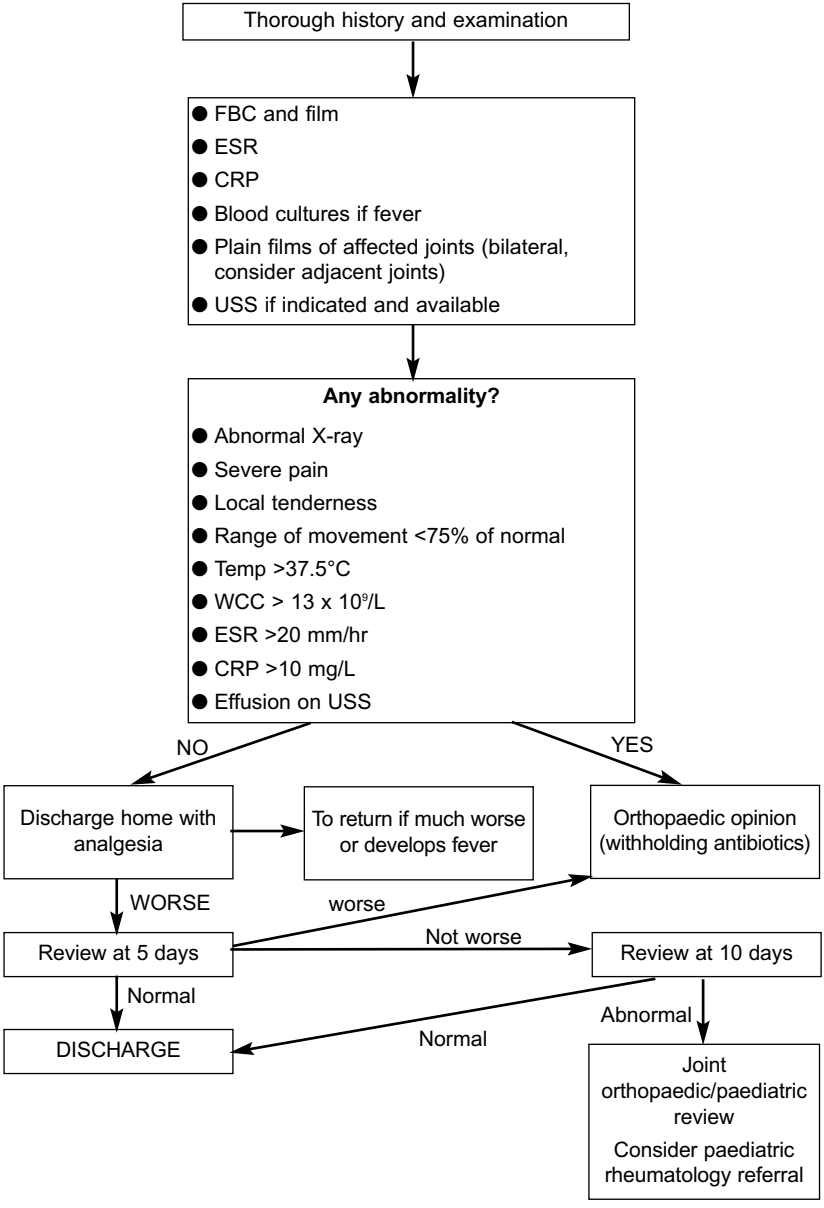
Thorough history and examination
FBC and film
ESR
CRP
Blood cultures if fever
Plain films of affected joints (bilateral, consider adjacent joints)
USS if indicated and available
Any abnormality?
Abnormal X-ray
Severe pain
Local tenderness
Range of movement <75% of normal
Temp >37.5°C
WCC > 13 x 10⁹/L
ESR >20 mm/hr
CRP >10 mg/L
Effusion on USS
NO
YES
Discharge home with analgesia
To return if much worse or develops fever
Orthopaedic opinion (withholding antibiotics)
WORSE
worse
Review at 5 days
Not worse
Review at 10 days
Normal
Abnormal
DISCHARGE
Normal
Joint orthopaedic/paediatric review
Consider paediatric rheumatology referral

Always follow the Child Safeguarding Policy and Procedures in your Trust. It is everyone's responsibility

- 4 recognised categories of abuse (rarely seen in isolation)
  - physical abuse (non-accidental injury)
  - emotional abuse
  - neglect
  - sexual abuse

NON-ACCIDENTAL INJURY (NAI)

Definition

Physical abuse may involve hitting, shaking, throwing, poisoning, burning or scalding, drowning, suffocating or otherwise causing physical harm to a child. Physical harm may also be caused when a parent fabricates the symptoms of, or deliberately induces, illness in a child

Recognition and assessment

Unless there is an appropriate explanation, discuss injuries with a senior doctor. All child protection cases must be dealt with by an SpR (minimum ST4) or above

There may be direct information from the child or carer. The following presentations need to be considered

- Delay in seeking medical attention following an injury
- History incompatible with injury seen
- Numerous explanations suggested for injury
- Changes in the history
- Parents 'shopping around' for medical help (e.g. from GP, A&E, different hospitals)
- History of domestic violence
- Odd or aggressive parental behaviour
- Any fracture in an infant without a satisfactory explanation
- Any bruise on a child aged <6 months old or pre-mobile
- Patterns of bruising, injury or explanation not compatible with child's development
- Recurrent injuries
- Evidence of other forms of abuse (e.g. failure to thrive, neglect)
- Previous evidence of injury or neglect (check if child known to local authority children's social care or is the subject of a child protection plan)

Referrals

- Discuss referrals by GP with consultant before arranging medical assessment by on-call team
  - consultant will decide whether referral should be made to the child protection agencies first
- Referrals from A&E or surgical wards should be taken by a doctor SpR grade or above
  - discuss with a consultant first to determine who should carry out initial examination and whether social care or police should be present

Always discuss referrals with the on-call consultant for child protection duties

Immediate action

- If there is an urgent or life-threatening situation, start necessary emergency treatment
- Refer to your Trust on-call child protection arrangements
  - if you suspect harm, refer to social care, and police if they are not already involved

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- if recommended by police, examine child jointly with the Forensic Medical Examiner (FME)
- Keep any social worker or police officer present informed
- Always consider potential risks to siblings or other children

History

- Where a referral is made from social care and/or the police, the child may have given a full history, often a visual recording
- ask for this information from social worker or police officer at beginning of examination. It may not be necessary to repeat this information unless further detail is required
- If child first presents in a health setting, particularly if story unclear, SpR (ST4 or above) or consultant should take history and examine child before discussing with social care or police

How

- Record findings accurately during or immediately after examination, using a dedicated child protection proforma with body charts if available
- Complete and sign each page and include:
- full family history
- persons present at interview
- source of your information (including the child)
- person giving consent
- date and time of start and finish

Take care when talking to the child not to ask leading questions or make suggestions that could contaminate evidence in a subsequent trial, document clearly what is said in child's own words

Examination

- There should be only one examination. Repeated examinations are not in the child's interest
- Keep your immediate senior informed

If this is a planned medical assessment at the request of child protection agencies, carers with parental responsibility and the child (depending on age and understanding) must give their consent (usually written) for examination to take place. If consent not forthcoming, social care may obtain a legal order giving permission for the child to be examined. This does not apply where a child needs urgent assessment and treatment

- Must include:
- state of child: cleanliness, appropriate clothing, etc
- **all** body areas
- accurate description of all injuries (size, colour, position and pattern) on body charts
- mouth (torn frenulum of lip and tongue especially)
- fundi: look particularly for haemorrhages. With small children, especially where head injuries are suspected, this is usually the role of the paediatric ophthalmologist
- a note of any birth marks, etc
- a full paediatric systemic examination
- plotting height and weight and head circumference on growth charts
- child's emotional state, demeanour and degree of co-operation
- a comment on the developmental state (or school progress)
- observations on relationships or behaviour between parents and child

Investigations

A selection of the following tests will usually be necessary; seek advice from consultant as to which are appropriate:

- If personal history of abnormal bleeding or concerning family history, discuss with a paediatric haematologist first as other tests may be indicated
- Bone biochemistry (including vitamin D) if there are unexplained fractures
- Investigations into other suspected abuse (e.g. failure to thrive)
- Skeletal survey in children aged <2 yr with unexplained injuries, ask radiology if repeat views after 11–14 days required. Head CT scan in children aged <12 months and in older children if focal encephalopathic features, focal neurology or haemorrhagic retinopathy
- Further neuroimaging according to RCR/RCPCH guidelines
- Document in notes if decision made not to proceed with imaging
- Photographs (often a police photographer is used)

Haematological investigations

When a bleeding diathesis suspected or needs to be ruled out, perform following:

- Initial baseline investigations
- FBC and film (EDTA up to 1 mL)
- APTT and PT (not INR)
- thrombin time
- fibrinogen levels
- if thrombocytopenic, mean platelet volume
- send 2 or 3 sodium citrate bottles, filled to appropriate fill line level

Subsequent investigations

- Identify all requests as NAI investigations
- Interpret all test results with age appropriate reference values
- If significant bruises, before further investigations, discuss with a paediatric haematologist:
- von Willebrand Factor antigen and activity
- Factor 8, 9 and 13 assay
- blood group
- child aged <2 yr: platelet function assay
- child aged <3 months, delayed cord separation, slow healing, bleeding after surgery or after cord separation: Factor 13 assay

EMOTIONAL ABUSE

Recognition and assessment

Definition

- Habitual harassment of a child by disparagement, criticism, threat and ridicule
- Present in most cases of physical and sexual abuse, and neglect
- presents difficulties in definition, recognition and management
- long-term consequences upon social, emotional and cognitive development can be more harmful than other forms of abuse

Presentation

- Part of the differential diagnosis if a child presents with the following non-specific behaviours:
- unhappy
- disturbed
- poor concentration leading to learning difficulties/school failure
- poor social interactions
- unable to play

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- problems with attachment to parents or caretakers
- over-friendly or craving affection from strangers

Assessment

- Assessment is complex and requires a multidisciplinary approach
- Social care take the investigative lead

NEGLECT

Neglect may not always be intentional (e.g. parental mental health problems)

Recognition and assessment

Definition

- Neglect is persistent failure to meet a child's physical and/or psychological needs
- Lack of care of physical needs that can result in failure to thrive
- important to eliminate organic causes
- neglect of physical care most likely to come to Child Health attention along with developmental delay

Presentation

- Child's appearance
- note condition of clothing, hair, skin
- Growth
- height, weight, serial measurements to check growth rate
- head circumference
- mid-upper arm circumference
- Non attendance (or repeat alterations) of appointments

Physical examination

- Signs of medical problem not appropriately treated
- Evidence of other forms of abuse
- Development
- gross motor skills, fine motor skills, vision, hearing, language, behaviour, play

SEXUAL ABUSE

Recognition and assessment

Definition

- Forcing or enticing a child or young person to participate in sexual activities, whether or not the child is aware of what is happening
- may involve physical contact, including penetrative (e.g. rape or buggery) or non-penetrative acts
- may include non-contact activities (e.g. involving children in looking at, or in production of, pornographic material, watching sexual activities, or encouraging them to behave in sexually inappropriate ways)

Presentation

- Information given by child
- Symptoms resulting from local trauma or infection (e.g. bruises, bleeding, discharge)
- Symptoms resulting from emotional effects (e.g. behavioural changes, enuresis, encopresis, self-harming, eating disorders or psychosomatic symptoms)
- Sexualized behaviour or sexual knowledge inappropriate to age
- Under-age pregnancy
- Sexually transmitted infections

Referrals

- Referrals usually come from local authority children's social care or the police
- refer to your departmental child protection rota

If a child presents in a medical setting and there are concerns about sexual abuse, call the on-call consultant for child protection immediately. Depending on any urgent medical needs e.g. bleeding; child protection agencies may need to be contacted before medical assessment

IMMEDIATE ACTION – HISTORY AND EXAMINATION

Preparation

- Where sexual abuse suspected, whoever examines the child **MUST** have training and experience in this field
- In exceptional cases, particularly where there is acute trauma and bleeding that may require surgical management, it may be appropriate for the examination to be carried out under anaesthetic by a gynaecologist after discussion with the FME

Examination

- Purpose of medical examination is to:
- detect traumatic or infective conditions that may require treatment
- evaluate the nature of any abuse
- secure forensic evidence
- reassure the child
- start process of recovery

Initial management

- If penetration and/or passage of bodily fluids are suspected consider sexually transmitted diseases and pregnancy
- pregnancy test
- if assault within 72 hr, offer post-coital contraception (ideally <12 hr) – usually levonorgestrel 1.5 mg stat dose
- Contact genito-urinary medicine department
- Post exposure prophylaxis should be started within 1 hr of assault if indicated (can be given up to 72 hr after assault). See **HIV PEP** guideline

Investigations

- Mid-stream urine
- Forensic tests (FME to determine)
- Photos/video recordings obtained with a colposcope, stored in accordance with local policy

Always follow the Child Safeguarding Policy and Procedures in your Trust

SUBSEQUENT MANAGEMENT

- Majority of children seen will be allowed home if it is safe and after discussion with social care
- some children who have been abused will be admitted while problems are investigated
- Always keep parents and children informed of concerns and what next actions will be
- Be open and honest with parents where possible unless this could put child (or others) at risk of further harm

Keeping children safe

- If there is clear evidence of child abuse and parents attempt to remove child there are two courses of action:
- in an emergency, dial 999, the police can use police protection powers to keep child safe
- if there is time, a social worker can obtain an Emergency Protection Order from Court (Section 44, Children Act 1989)
- Put the child's safety first

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- Communicate with other staff involved (e.g. nursing staff) so that situation can be supervised
- Consider the safety of siblings
- usual for siblings to be examined at same time as index child

DISCHARGE AND FOLLOW-UP

Only a consultant may allow child to go home

- Consultant should make decision regarding discharge, usually after discussion with the police and social care

Communication is vital

- Send written report to GP without delay, with a copy for social care and the police
- If child referred from A&E, send copy of report to them for feedback
- Ensure notes and dictation is available to secretary, marked 'for urgent attention'
- Ensure report is signed in a timely manner
- Complete ward discharge forms
- Check with consultant if follow-up is required

Child protection conference

- May be convened following a child protection investigation to consider:
- whether child needs to be the subject of a child protection plan
- Medical and nursing staff will be invited if child has been admitted
- expected to contribute, usually in person, or via a written report
- ensure reports are available for future reference

SELF HARM • 1/2

- Self harm can take a number of forms, including:
  - cutting or burning
  - self poisoning with medicines or tablets
  - punching
  - self strangulation
  - pulling out hair or eyelashes
  - scratching or picking at skin
  - inhaling or sniffing harmful substances
  - swallowing non-food substances
  - inserting objects into the body either through orifices or the skin
  - head banging

ASSESSMENT

- Identifying behaviour, intended behaviour or self-harming thoughts
- Who knows about the behaviour
- How often this occurred
- If at risk from others
- Stressors e.g. bullying, bereavement, relationships
- Difficulties, abuse, sexuality issues
- General health
- Use of drugs and alcohol
- Education
- Family and social issues
- Support network available
- Child protection issues

MANAGEMENT

- Patients who have self harmed, admit overnight
- See **Poisoning** guidelines
- Advise carers to remove all medications or other means of self harm
- Manage child protection issues according to local policy and procedures. On-call consultant available 24 hr for child protection advice
- Assess risk/need for ongoing psychological treatment or support. The Connect Child and Adolescent Mental Health Services (CAMHS) and First Steps Priority Referral Team (PRT) provides service for patients aged up to 18 yr. Obtain valid consent for a referral to CAMHS from parent/other adult with parental responsibility or the young person if they are deemed to have capacity (Gillick competence). Clearly document in medical records who obtained consent, who consent was taken from and when it was obtained i.e. date and time

Documentation

- Clearly document assessment in notes with any decisions made and reasons

REFERRALS

Criteria for referral to PRT

- Deliberate self harm (e.g. overdose, self strangulation, serious cuts)
- Deliberate harm from substance misuse (e.g. poisoning from excessive alcohol and/or illicit drugs if **intention** was to self harm)
- Mental health symptoms:
  - depression/low mood with active suicidality
  - psychotic symptoms.
  - low weight anorexia nervosa i.e. BMI <15 or accompanied by rapid weight loss
- Referrals must be phoned through to PRT before 1000 hr to be seen that day. Where there are exceptional circumstances (as determined by the ward), referrals will be accepted up to 1230 hr

DISCHARGE AND FOLLOW-UP

- Discharge when medically fit **and** have been assessed by PRT
- Discuss with CAMHS to ensure child has an agreed plan in place
- If there are safety concerns, refer to children's social care
- Ensure health professionals i.e. GP and school nurse are aware of admission and management plan

Local contact:

UHNS: PRT are based at the Ashlands and can be contacted via 0300 7900 235

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These guidelines are advisory, not mandatory.
Every effort has been made to ensure accuracy.
The authors cannot accept any responsibility for
adverse outcomes.

Suggestions for improvement and additional guidelines
would be most welcome by Partners in Paediatrics,
please contact via http://www.networks.nhs.uk/nhs-networks/
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