Chapter

41

Principles of Paediatric Intensive Care*

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Introduction

Many hospitals with paediatric facilities will not have a paediatric intensive care unit (PICU) on site. Sick children presenting to these hospitals will need initial treatment and stabilisation in the emergency department (ED). Whilst in the ED, paediatric and ED staff will have already set in motion advanced life support measures, but often they will be looking for additional help.

The paediatric team will have experience in the intubation and ventilation of neonates but limited expertise in older children. The local anaesthetic team should be able to provide valuable support by understanding key elements of paediatric critical illness along with their skills in airway management. If intubated, the child will require transfer to a PICU unless potential extubation is imminent. This may be at another site, and transfer will generally involve a specialist paediatric intensive care retrieval team. For certain time-critical conditions, it may be necessary for the local anaesthetic team to undertake the transfer (see Chapter 42).

Anaesthetists are frequently involved in the management of children requiring intensive care, and this chapter will aim to cover some essential aspects of paediatric intensive care that will help the anaesthetist in these situations. There is also information on the management of the potential organ donor.

How Much Paediatric Critical Care Is Out There?

Data from the Paediatric Intensive Care Audit Network (PICANet) indicates that there were approximately 60,000 PICU admissions in the United Kingdom in the three years from 2017 to 2019. Of these, approximately 18,300 patients were transferred to a PICU, nearly 1,500 of whom were transported by non-PICU teams. The current trend is for an increasing number and longer duration of admissions to PICU of children with life-limiting conditions, accounting for 58% of PICU admissions in 2018. PICU admissions are the tip of the iceberg; paediatric admissions overall continue to increase year on year, with around 5–10% of these requiring high-dependency care at their local hospital.

In the UK population of children requiring transfer to PICU in 2016 and 2018, the most common reason for admission was a respiratory condition (29% of transfers), with bronchiolitis representing a third of these cases. The next most common cause was cardiovascular conditions (27%). The children being transported are primarily young, with 50% <1 year and 75% <5 years old. A high proportion of the cohort were critically unwell, with an overall mortality of 6% and with 3% having experienced cardiorespiratory arrest prior to transfer.

General Principles of Paediatric Intensive Care

Why Infants Readily Develop Respiratory Inadequacy

Although children show significant tolerance to physiological challenges, they have anatomical and physiological attributes that pose disadvantages when compared to adults. They have small airways, and as the Hagen–Poiseuille equation (Flow = $\Delta P \pi r^{[4]}/8\eta l$) shows, any reduction in airway radius will have a marked effect on airflow and work of breathing. Smaller volumes of secretions are more likely to completely occlude small airways, with coughing being less effective in secretion clearance.

* Many thanks to Dr Ian Jenkins, who wrote the first edition of this book chapter, much of the content of which has been used in this revision.

Table 41.1 Child Glasgow Coma Score

	1	2	3	4	5	6
Eyes	No eye opening	Opens eyes to pain	Opens eyes to speech	Opens eyes spontaneously		
Verbal	No verbal response	Inconsolable, agitated	Inconsistently consolable, moaning	Cries but consolable, inappropriate interactions	Smiles, follows objects and sounds, interacts	
Motor	No motor response	Extension to pain (decerebrate)	Flexion to pair (decorticate)	Withdraws from pain	Withdraws from touch	Infant moves spontaneously or purposefully

Note: The lowest score possible is 3; anything below 8 is associated with poor outcomes and is the accepted level for mandatory intubation and ventilation.

Infants are much more dependent on diaphragmatic and abdominal muscles to breathe as their intercostal muscles are under-developed. Their muscles also fatigue more readily than in adults. Infants have horizontal and cartilaginous ribs, causing difficulty in increasing tidal volumes via rib movement. Overall, the chest wall is much more compliant, increasing the work of breathing. In conditions such as croup and bronchiolitis, attempts to significantly increase negative intrathoracic pressure result in mechanically inefficient subcostal or sternal recession. Lacking the ability to increase tidal volumes, infants must rely on an increased respiratory rate to increase ventilation.

Clinical Indicators of Respiratory Inadequacy

- Tachypnoea
- Use of accessory muscles
- Subcostal and sternal recession
- Tachycardia
- Stridor – due to a smaller cricoid ring, which may become oedematous after tracheal intubation or in croup
- Deteriorating conscious level
- Rising $PaCO_2$
- Ultimately cyanosis

Intubation

Apart from respiratory compromise, other indications for intubation include:

- Sepsis. This may be severe, necessitating large volumes of intravenous fluids, often >60 ml kg^{-1}. Pre-emptive support with intubation and ventilation is necessary to offset the development of respiratory compromise due to pulmonary oedema. This course of action, with aggressive fluid resuscitation, control of ventilation and administration of inotropes, is often inadequately managed. Early intubation for shock improves outcomes.
- Decreased conscious level. Conventionally, intubation is indicated with a Glasgow Coma Score of 8 or less (see Table 41.1) or AVPU score of 'Unresponsive'.
- Cardiovascular instability. Low cardiac output may be due to intrinsic congenital cardiac problems, myocarditis, cardiomyopathy or dysrhythmias.
- Smoke inhalation. Even when no respiratory effects are present initially, respiratory compromise can ensue quickly from airway swelling.

Intubation and ventilatory support also provide conditions for easier insertion of central venous and arterial catheters for monitoring and the central administration of inotropes where necessary.

Drugs for Intubation in the Critically Ill Child

'Which drugs should be used?' is one of the most frequently asked questions when dealing with a seriously ill child. As in any other emergency situation, the potentially deleterious effects of anaesthetic induction agents on the circulation of a shocked patient still apply.

Agents that affect the systemic vascular resistance (SVR) the least and are less likely to

exacerbate any obvious or occult hypovolaemia are the preferred option. Even when children appear normovolaemic, intrinsic cardiovascular compromise is present in sepsis, metabolic conditions, arrhythmias, cardiomyopathies and a number of cardiac lesions.

Particular care should be exercised in patients with a failing myocardium, as a fall in SVR results in decreased aortic root pressure and decreased coronary perfusion, resulting in myocardial ischaemia. Tachycardia and arrythmias must be avoided, as they can also cause decreased coronary perfusion.

In pulmonary hypertension, a fall in SVR results in decreased coronary blood flow to the hypertrophied right ventricle, resulting in rapid ischaemia due to its exquisite sensitivity to coronary hypoperfusion. Additionally, any increase in pulmonary vascular resistance (PVR) will reduce pulmonary blood flow, reduce left-ventricular preload and reduce cardiac output. An increase in the right-ventricular end diastolic pressure causes septal deviation, impairing left-ventricle function, which may also compromise cardiac output.

Those experienced with critically ill children, particularly children with cardiovascular compromise, find that agents such as ketamine combined with small doses of fentanyl along with a muscle relaxant cause less instability. Ketamine and pancuronium should be used with caution in tachydysrhythmias as they increase heart rate; a muscle relaxant such as rocuronium should be considered instead. Suggamadex should always be available for reversal of neuromuscular blockade, in case intubation is unexpectedly difficult. Ketamine has been shown to be safe in head injury. Propofol should be used with extreme caution in any child with cardiovascular compromise or suspected hypovolaemia, as the associated hypotension can lead to arrest on anaesthetic induction. Peripheral vasoactive infusions can reduce the risk of decompensation (see the section 'Fluids and Inotropes in Critically Ill Children'). Each hospital or local retrieval team should have guidance and checklists available.

Respiratory Support in Children

There has been increasing use of high-flow nasal cannula (HFNC) oxygen therapy in the paediatric population over the last five years. Around 4 cmH_2O of positive end expiratory pressure (PEEP) may be delivered, depending on the size of the cannulae compared to the diameter of the nostrils. The humidification of the inspired gases can help loosen secretions and aid in secretion clearance, reduce the metabolic work associated with gas conditioning and improve the conductance and pulmonary compliance. The washout of the nasopharyngeal dead space leads to improved alveolar ventilation. The evidence base to determine efficacy compared to other respiratory support modalities is still being established. HFNC is usually initiated at flow rates of 2 l min^{-1} in children under 12 kg, plus 0.5 l min^{-1} for each kilogram above 12 kg (to a maximum of 50 l min^{-1}). Appropriately sized nasal prongs need to be selected for both neonates and paediatric patients; these should not occlude more than 75% of the nares. Flow can be weaned once the FiO_2 is <0.4 by halving the flow rate initially and then removing HFNC support entirely if stability is maintained.

Continuous positive airway pressure (CPAP) provides respiratory support by maintaining airway patency, increasing the functional residual capacity and preventing alveolar collapse, thereby reducing the work of breathing. CPAP is frequently used in infants with less severe forms of croup, bronchiolitis and pneumonias and may obviate the need for intubation and ventilation. Interfaces and devices exist for all weights of children down to 500 g. Many devices allow escalation to bi-level positive airway pressure (biPAP). High respiratory rates and large leak relative to paediatric tidal volumes can make patient triggering difficult to maintain reliably. CPAP is initiated at pressures of 5–8 cmH_2O and can be weaned once the FiO_2 is <0.4 by reducing the pressure to 4–6 cmH_2O and then removing CPAP support entirely if stability is maintained.

In children under two years of age that require ventilation on the PICU, nasal tracheal intubation is better tolerated than oral, with decreased requirements for sedation. Additionally, nasal tubes are more stable, easier to fix and permit easier mouth care. Nasal tracheal intubation is associated with a lower rate of unplanned extubation in children under two years. It is a technically a more challenging procedure and associated with more complications,

including bleeding and increased risk of sinusitis and ventilator-acquired pneumonia in the older patient groups.

Pressure-cycled ventilation is very commonly used in paediatric practice because leaks around traditionally used uncuffed tracheal tubes are rendered less significant, although changes in lung or thoracic compliance will alter alveolar ventilation. Volume-cycled ventilation is less vulnerable to compliance changes but is affected by the leaks around non-cuffed tubes. Current practice varies by centre but commonly includes provision of pressure support to spontaneous breaths in addition to the mandated minimal ventilation. Peripheral oxygen saturation and end-tidal CO_2 monitoring are essential, with specific paediatric devices available to minimise additional dead space.

The 'No Trace = Wrong Place' campaign from the Royal College of Anaesthetists emphasises that a capnography trace must be obtained after intubation and will be present, albeit attenuated, in cardiac arrest.

Cuffed versus Uncuffed Tracheal Tubes in PICU

Cuffed tubes decrease leak by compensating for tube and larynx size mismatch. This permits greater use of volume-cycled ventilation modes as well as protecting against aspirated fluids. Improvements in endotracheal cuff design means that cuffed tubes are available down to size 3.0 mm and are now used in term infants. Available evidence suggests that tube size selection is much more important than whether a tube is cuffed or uncuffed in determining risk of subsequent laryngeal injury. Paediatric mucosal capillary occlusion pressures are lower than in adults, and thus cuff pressures should not usually exceed 20 cmH_2O, which may pose difficulties in high-pressure ventilation scenarios. Cuff pressures should be regularly monitored, especially in higher-risk situations such as when using inhaled nitric oxide, which will diffuse into the cuff and gradually increase cuff pressures.

In children with prolonged length of stay on the PICU, tracheostomies are less readily used owing to anatomical difficulties (short necks, short tracheal length) and their use being associated with longer-term problems such as local tracheomalacia or stenosis (even with more modern surgical techniques).

High-Frequency Oscillatory Ventilation (HFOV)

The use of HFOV continues in children whilst its use in adult intensive care has declined. This decline followed evidence from the OSCAR and OSCILLATE studies, which showed HFOV use resulted in unchanged and worse mortality respectively. Its continued use in PICUs is supported mainly by evidence in neonates that HFOV reduces lung injury with no increase in mortality. Evidence in older children is less clear, but HFOV is used as a rescue therapy when conventional ventilation strategies are ineffective at the upper limit of support (e.g. peak pressures of $\geq$28 cmH_2O).

HFOV is recommended for paediatric patients with severe respiratory failure who are failing on conventional ventilation and as a second-line therapy for the management of a bronchopleural fistula. In the neonatal population, HFOV is indicated for patients with neonatal air leak syndrome, persistent pulmonary hypertension of the newborn (PPHN) and meconium aspiration syndrome (MAS).

HFOV is a mode of ventilation that delivers tiny tidal volumes (1–3 ml kg^{-1}) into the anatomical dead space at frequencies ranging from 7 Hz to 15 Hz. It is thought that the reduced tidal volumes avoid the shear stresses of pulmonary volutrauma whilst maximising alveolar recruitment and minimising atelectasis by maintaining a high mean airway pressure (MAP).

CO_2 elimination is dictated by the movement of gas in and out of the airway, driven by the action of a piston-powered diaphragm that oscillates at high frequency. This promotes gas exchange to the level of the alveoli through several mechanisms (convective streaming, radial mixing, pendelluft, asymmetric flow profiles and collateral ventilation) that differ significantly from those seen in conventional ventilation.

CO_2 elimination depends on the amplitude of the oscillation (how much the piston moves in each cycle, also referred to as ΔP) and on the frequency set. Lower frequencies allow longer periods of time for gas exchange to occur, improving CO_2 clearance. If the waveform of piston movement is plotted (Figure 41.1), it is the area under the curve where gas exchange occurs. The desired elimination of CO_2 may be more readily achieved by reducing the frequency

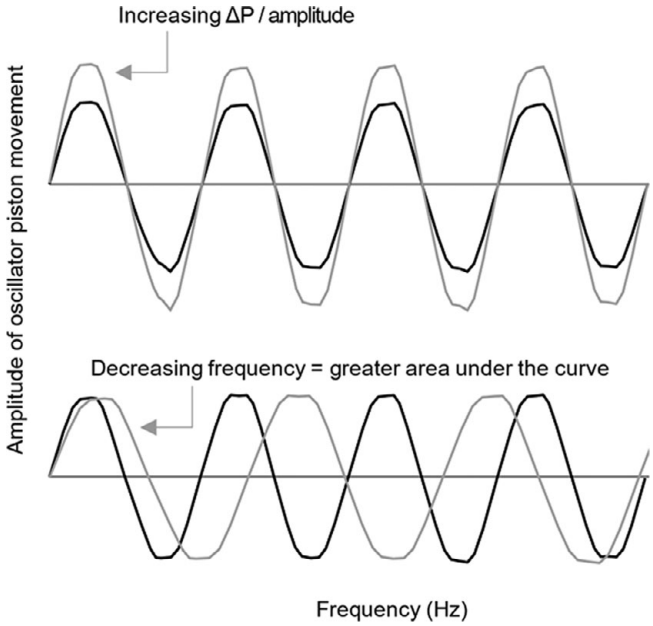

Figure 41.1 Impact of amplitude and frequency changes on the oscillation waveform.

rather than increasing the amplitude of the piston movement.

Patient oxygenation depends on delivery of adequate oxygen to recruited alveoli, dictated by the MAP and composition of the gas flow (FiO_2). As the pulmonary condition improves, a constant MAP can cause over-inflation with pulmonary capillary compression and decreased gas exchange. Routine respiratory examination is limited on HFOV, and the paucity of variables measured by the ventilator means that regular chest radiographs are necessary to avoid over-inflation. The increased pulmonary vascular resistance from alveolar distension and increased intrathoracic pressure on HFOV can reduce cardiac preload and contribute to hypotension when HFOV is initiated. Care should be taken to ensure adequate filling before starting HFOV. Limitations of HFOV include the requirement for deep sedation and paralysis as well as the need to minimise ventilator disconnection, necessitating the use of smaller-lumen inline suction catheters.

Some portable ventilators can provide HFOV, but in practice it is very rare to transfer a patient on oscillatory ventilation, and conversion to conventional ventilation will be required for transfer for imaging or procedures.

A general approach to setting initial parameters is:

- MAP: 2–6 cmH_2O above the MAP achieved using conventional ventilation.
- Frequency: set depending on age
  - Preterm infants: 15 Hz
  - Term infants: 12 Hz
  - Young children:10 Hz
  - Older child: 8 Hz
- Amplitude should be initially set at 50 cmH_2O and adjusted until the characteristic body 'wobble' that is observed stops at the level of the patient's thigh.
- Percent of inspiratory time is set at 33% and rarely changed. This corresponds to an I:E ratio of 1:2.

Table 41.2 outlines the clinical impact of changing parameters.

Extracorporeal Membrane Oxygenation (ECMO)

This comprises an extracorporeal circuit, draining blood from the venous system and returning it after gas exchange to either the arterial system (VA-ECMO, used in cardiac or cardiopulmonary failure) or the venous system (VV-ECMO, used in isolated respiratory failure). Apart from severe cardiac failure, e.g. cardiomyopathy or immediately after a cardiac surgical operation, the main indication for ECMO is severe respiratory failure. There are around 145 paediatric respiratory

Table 41.2 Adjustment of HFOV parameters

Under-oxygenation	Increase MAP by 1–2 cmH_2O (consider ECMO if >25)	Increase FiO_2 by 5%
Over-oxygenation	Decrease MAP by 1–2 cmH_2O (consider conversion to conventional ventilation if <5)	Decrease FiO_2 by 5%
Under-ventilation	Increase amplitude by 2 cmH_2O	Decrease frequency by 0.5 Hz
Over-ventilation	Decrease amplitude by 2 cmH_2O	Increase frequency by 0.5 Hz

ECMO runs and 80 cardiac ECMO runs in the United Kingdom annually.

ECMO can be considered in children over 2 kg in weight and 34 weeks of gestational age with reversible disease, for example, MAS, PPHN or congenital diaphragmatic hernia, and with no contraindication to systemic anticoagulation (such as intracranial haemorrhage), no lethal congenital abnormalities, no major immunodeficiency and no irreversible organ dysfunction including neurological injury.

Referral for ECMO should be made when one or more of the following are present:

- Severe hypoxia and/or hypercapnia with lack of response to conventional support measures (this may include high-frequency oscillation, inhaled nitric oxide, prone positioning and surfactant). Severe hypoxia in this context is usually defined as:
  - Oxygenation index (OI) >40,
    - ◇ Where OI = (FiO_2 × MAP (cmH_2O) × 100) / PaO_2 (mmHg)

or

  - P/F ratio <100
    - ◇ Ratio = PaO_2 (mmHg)/ FiO_2.
- Sustained elevated inflation pressures; for example:
  - >24 hours of MAP >20 on conventional ventilation
  - MAP >25 on HFOV
- Persistent air leak or interstitial air.

Fluids and Inotropes in Critically Ill Children

Children, particularly infants, have greater insensible fluid losses and have greater maintenance requirements than adults. However, children are also more prone to developing inappropriate antidiuretic hormone (ADH) secretion, for example in respiratory conditions such as bronchiolitis. If given full IV fluid maintenance, as calculated from the Holliday–Segar formula (Table 41.3), they can develop severe hyponatraemia. In some circumstances, maintenance fluids are reduced to 80% of normal, and hypotonic fluids are not used. Balanced crystalloids are the fluid of choice for most clinical situations (see also Chapter 13).

Table 41.3 Holliday–Segar maintenance fluid calculation

Weight (kg)	Maintenance fluid requirement per day
1–10	100 ml kg^{-1}
10–20	1,000 ml + 50 ml kg^{-1} for each kg >10
>20	1,500 ml + 20 ml kg^{-1} for each kg >20

The cardiovascular system of infants differs from older children and adults:

- The heart is less compliant, with the result that cardiac output is less responsive to volume loading and is more dependent on heart rate.
- The Starling curve is flatter with an earlier downward inflection point at lower preloads.
- After term, myocyte β-receptor populations and response to catecholamines and phosphodiesterase inhibitors are normal.
- The infant heart is particularly sensitive to serum-ionised calcium levels; hypocalcaemia is not well tolerated.
- The liver is more distensible, with the result that fluid overload rapidly leads to hepatomegaly. A rise in blood pressure in response to external hepatic pressure can be used as a marker of hypovolaemia.

The most used inotropes in children are adrenaline and noradrenaline (0–1.0 mcg kg^{-1} min^{-1} for both), selected for central inotropy and

peripheral vasoconstriction respectively. Hydrocortisone IV (1 mg kg^{-1} every six hours to a maximum 100 mg, 2.5 mg kg^{-1} every four hours in neonates) and vasopressin (0–0.002 u kg^{-1} min^{-1}) are used in catecholamine resistant shock. In complex cases, cardiac output monitoring should be used to guide inotrope titration and choice. The inodilator properties of milrinone (0–0.75 mcg kg^{-1} min^{-1}) are used in clinical situations with raised pulmonary vascular resistance, especially in children with congenital cardiac or lung disease.

There is increasing peripheral use of dilute inotrope infusions where central access is not available, with adrenaline used more frequently than noradrenaline. For both, an infusion made up with 0.3 mg kg^{-1} (maximum 4 mg) in 50 ml can be used at as low a dose and for as short a time as possible. This is of particular use in settings where staff are not experienced in inserting central access in small children.

Specific Respiratory Conditions

Croup

Croup is one of the most frequent causes of respiratory distress in young children, with viral inflammation of the upper airway leading to a barking cough and stridor as the airway becomes obstructed. Obstructive symptoms can be temporarily relieved with nebulised adrenaline (400 mcg kg^{-1} of 1:1,000, to max 5 mg) whilst allowing time for IV dexamethasone (0.15 mg kg^{-1}) to take effect. Biphasic stridor heralds severe obstruction and is one of the indications for intubation.

Bronchiolitis

Bronchiolitis is a common viral respiratory condition associated with lower airway obstruction, air trapping and atelectasis affecting infants but seen up to two years of age. It is characterised by cough, tachypnoea, nasal flaring and subcostal recession. Usually there are widespread crepitations and a coarse wheeze. Patients are escalated through HFNC and then CPAP and infrequently require intubation. High-risk groups include those affected by prematurity, pre-existing respiratory conditions, neuromuscular conditions and immune deficiency. There is no evidence of benefit from any combination of inhaled or parenteral medication. Intubation and ventilation may be necessary for actual or impending respiratory fatigue, characterised by:

- SpO_2 <92% (despite O_2)
- Respiratory rate >70 bpm
- Signs of respiratory distress
- Apnoea
- Decreased level of consciousness

Pulmonary compliance is often very poor, so 'permissive' hypoventilation with modest hypercapnia, respiratory acidosis (pH $\geq$ 7.25) and SpO_2 92–95% should be accepted. Current research is examining whether a conservative oxygen saturation target of 88–92% may be beneficial.

Life-Threatening Asthma

Children usually present with respiratory distress, cough and wheeze. Life-threatening asthma is characterised by the following:

- Silent chest
- Cyanosis
- Hypotension
- SpO_2 <92% (despite high-flow O_2 therapy)
- Peak expiratory flow <33% best or predicted
- Poor respiratory effort
- Exhaustion or confusion
- Coma

Medical management of severe asthma includes:

- Nebulised bronchodilators:
  - Mixed ipratropium 250 mcg (125 mcg <2 years) every four to six hours and salbutamol nebulisers 2.5–5mg every 20–30 minutes.
- There is increasing use of nebulised magnesium sulphate in severe asthma:
  - 150 mg magnesium sulphate nebulised every 20 minutes for the first hour only (maximum of three doses) in patients two years or older.
- Steroids should be administered early, given an effect onset of three to four hours. In severe asthma, give hydrocortisone at 4 mg kg^{-1} (max 100 mg).
- There is differing practice in the use of IV bronchodilators, with no clear superiority demonstrated for one approach:
  - Boluses of IV magnesium sulphate may be used at doses of 40–50 mg kg^{-1}, repeated

after one to two hours with monitoring of serum magnesium.
- IV salbutamol, loading dose 15 mcg kg^{-1} given over 20 minutes (max 250 mcg), followed by infusion at 1 mcg kg^{-1} min^{-1} (max 20 mcg min^{-1}). Monitor for hypokalaemia and side effects such as tachycardia and lactic acidosis, which increase at higher doses.
- IV aminophylline, 5 mg kg^{-1} loading dose (max 500 mg) over 20 minutes with electrocardiogram (ECG) monitoring, followed by infusion at 1 mg kg^{-1} h^{-1}. Levels must be monitored, as there is a narrow therapeutic index.

Intubation and ventilation are indicated in the event of life-threatening asthma unresponsive to the aforementioned measures. Care needs to be taken during intubation, as there may be occult hypovolaemia secondary to decreased fluid intake. Fentanyl should be used in place of morphine, as it triggers less histamine release. Ketamine is useful in preserving blood pressure and for its bronchodilator action. A cuffed tube is essential, given the anticipated high ventilator inflation pressures. It should be remembered that the presence of the tracheal tube may be an ongoing stimulus for bronchospasm, and so the tube should be removed at the earliest opportunity, even if some wheeze persists.

For ventilation, adopt the following strategy:

- Pressure control mode with muscle relaxation
- Modest hypercapnia, accepting an arterial pH >7.2 (acceptable $PaCO_2$ 7–14 kPa)
- Tidal volume 5–7 ml kg^{-1}
- Inspiratory plateau pressures <35 cmH_2O
- Slow rate, 10–15 bpm (expiratory flow should reach zero before the next breath)
- Short inspiratory times to keep an adequate respiratory rate and avoid air trapping; I:E ratio of at least 1:2.
- Low PEEP (5 cmH_2O)

Cardiovascular

Children may present to the ED or PICU in a low cardiac output state secondary to an undiagnosed cardiac disorder, an acute dysrhythmia or cardiomyopathy. The lack of out-of-hours paediatric echocardiography expertise in many centres can make management challenging.

In a baby who has been discharged entirely well from the maternity unit but re-presents to the hospital in a collapsed state in the first few days of life, congenital cardiac disease should be suspected. Most severe congenital cardiac lesions are identified on antenatal scanning, but occasionally these are missed. In some conditions, the patency of the ductus arteriosus compensates for the congenital abnormality immediately after birth by providing a shunt pathway. As the duct closes over the next few days, the change in haemodynamics can lead to a profound low-output state with severe metabolic acidosis and organ dysfunction. This can mimic severe sepsis.

Administering prostaglandins in this situation is essential. This may not re-open the duct fully but may allow time for transfer to a surgical centre for definitive diagnosis and management. During the infusion of prostaglandin, the newborn requires careful monitoring of heart rate, blood pressure, respiratory rate and core body temperature. In the event of a complication such as apnoea, profound bradycardia or severe hypotension, the infusion should be temporarily stopped and the complication dealt with; the infusion should be restarted at a lower dose. Recurrent or prolonged apnoea may require ventilatory support to allow the prostaglandin infusion to continue.

Total anomalous pulmonary venous drainage (TAPVD) is a rare congenital defect in which the pulmonary veins do not connect to the left atrium. Pulmonary venous blood instead drains into the right atrium via systemic veins and then to the left side via an atrial septal defect (ASD) and/or ventrical septal defect (VSD). It is frequently undiagnosed on antenatal scans. If pulmonary venous return is obstructed, infants will present within the first hours of life with tachypnoea, profound cyanosis, a plethoric chest X-ray and low cardiac output, and can be mis-diagnosed as sepsis, PPHN or MAS, especially if they have been compromised in utero and passed meconium during delivery. Obstructed TAPVD is a surgical emergency.

PIMS-TS

Paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) is a novel condition that was identified early

in the SARS-CoV-2 pandemic. It has many similarities to Kawasaki disease, but also shares features of toxic shock syndrome, macrophage activation syndrome and haemophagocytic lymphohistiocytosis. The syndrome usually develops within four weeks of SARS-CoV-2 infection.

Common presenting features include fever, abdominal pain, vomiting, diarrhoea and shock, with shock necessitating PICU admission for vasoactive infusions in around two thirds of cases. Respiratory symptoms are not normally significant in these children. The mainstays of therapy are intravenous immunoglobulin and steroids, with some children requiring additional anti-inflammatory biologic therapies. The inflammation in PIMS-TS is prothrombotic, requiring prophylactic anti-coagulation in most cases.

Early echocardiography is mandatory, with the frequency of coronary artery abnormalities and left-ventricular impairment necessitating the close involvement of cardiology throughout the PICU admission. The shock is presumed to be secondary to a transient inflammatory vasoplegia, often requiring invasive blood pressure monitoring. Noradrenaline is used to counter the vasoplegia, with the addition of adrenaline if cardiac function is impaired.

Gastrointestinal

Intussusception can present with lethargy and intermittent pain rather than the classical vomiting and redcurrant jelly stools, which are seen in only a third of cases. It can develop into a time-critical emergency where the viability of the intestine is at stake, with a marked systemic inflammatory response and circulatory failure.

Diarrhoeal conditions may go on to develop into haemolytic uraemic syndrome (HUS), particularly those associated with Shiga toxin-producing *Escherichia coli*, but also with *salmonella* and *campylobacter*. HUS is a condition caused by diffuse small vessel injury from thrombotic microangiopathy with a triad of haemolysis, thrombocytopenia and renal failure. Complications include seizures, coma, cardiac failure, pancreatitis and ischaemic colitis, where frank necrosis and perforation can occur. The treatment is to support the failing kidney with peritoneal dialysis or haemofiltration and, where possible, avoid platelet transfusion and systemic antibiotics, which may fuel the condition.

Renal Failure and Support

The past few years have seen refinement in the equipment available for paediatric renal replacement therapy (RRT) in the PICU, and haemofiltration and haemodiafiltration have become relatively commonplace. Mortality is increased if the patients are hypotensive on initiation of RRT, particularly if receiving inotropic support or in secondary, rather than primary, renal failure.

RRT is also used as specific therapy in metabolic disease, particularly when there is marked hyperammonaemia, which is treatable by haemofiltration or haemodialysis. In certain cases, RTT may have to be used in neonates and is technically challenging.

Problems seen particularly in smaller patients include:

- Acute haemodynamic changes, exacerbated by the low circulating volume of neonates compared with circuit volumes. The smallest children may require initiation of peritoneal dialysis. There are current trials of devices designed for haemodialysis at weights <8 kg.
- Thrombocytopenia due to adsorption of platelets to the filter.
- A tendency to under-estimate insensible fluid losses.
- Hypothermia due to the patient's blood travelling through a circuit with a very high relative surface area.

Neurology

Seizures and Status Epilepticus

In a previously well child, the potential causes of seizures include infection, trauma, electrolyte derangement and tumours. Meningitis must be considered if the history is suggestive, but the most common cause of seizures in the six-month to six-year-old child is so-called febrile convulsions. These are classified as 'simple' if they are generalised tonic-clonic seizures lasting less than 15 minutes with recovery to baseline within one hour. Complex features include partial onset or focal features, duration >15 minutes, recurrence within 24 hours and incomplete recovery after one hour.

The trigger is cerebral irritation from inflammatory cytokines crossing the immature blood–brain barrier and the seizures are not secondary to fever itself. Control of temperature is not a primary treatment goal.

Should a presumed febrile convulsion last longer than five minutes, meeting the definition of status epilepticus, it is treated as for any other seizure. Children with complex febrile seizures are likely to be observed in hospital. CT imaging may be undertaken for focal seizures or persistent neurological deficit.

In status epilepticus, the usual cascade is:

- Two doses of benzodiazepines:
  - Midazolam 500 mcg kg^{-1} buccally, or
  - Diazepam 500 mcg kg^{-1} rectally, or
  - Lorazepam 100 mcg kg^{-1} IV, then
- Phenytoin 20 mg kg^{-1} IV over 20 minutes.
  - If already taking regular phenytoin, phenobarbital 20 mg kg^{-1} IV.
- Paraldehyde 0.4 ml kg^{-1} can be instilled rectally whilst preparing or infusing phenytoin.

Levetiracetam has been demonstrated to be as effective as phenytoin and is increasingly used in the management of status epilepticus. If these measures do not halt the seizure after 20 minutes, then intubation using IV thiopental should be undertaken. It may be necessary to continue a thiopental infusion at 1–8 mg kg^{-1} h^{-1} and proceed to continuous EEG recordings to detect ongoing seizure activity.

Neonatal seizures are managed with phenobarbital as the first line treatment.

Traumatic Brain Injury (TBI)

Many patients with TBI will require time-critical transfer for imaging or to another centre for neurosurgery. This is likely to be undertaken by the local anaesthetic team.

Neuroprotection measures should be initiated as soon as possible:

- Keep the head in midline position with the bed 30° head up.
- Ventilation
  - Maintain oxygenation: PaO_2 >10–13 kPa.
  - Maintain $PaCO_2$ at 4.7 kPa (range: 4.5–5.0 kPa).
  - PEEP: 5 cmH_2O.
- Adequate sedation (fentanyl plus dexmedetomidine +/– vecuronium infusion initially)
- Normoglycemia: 3–12 mmol l^{-1} .
- Haemoglobin: >70 g l^{-1}.
- Fluid restriction: 60%, maintain Na: 142–150 mmol l^{-1}.
- Maintain normothermia: 36–38°C.
  - 36.5°C if muscle relaxed and using a cooling blanket.
- Discuss with neurosurgery the initiation of prophylactic anti-epileptics.

Most centres that insert intracranial pressure (ICP) monitoring aim for an ICP of <20 mmHg with a minimal cerebral perfusion pressure (CPP) of at least 50 mmHg:

CPP = mean arterial blood pressure – ICP

For situations without ICP monitoring, empiric targets for MAP are used:

0–2 years >60 mmHg
2–6 years >70 mmHg
>6 years >80 mmHg

If there are difficulties maintaining ICP <20, then the initial approach is:

- Ensure sedation is adequate (avoiding the risk of hypotension with bolus sedation).
- Initiate the neuromuscular blockade.
- Administer bolus ± infusion of hypertonic saline.
- Consider CSF drainage via ventriculostomy.

If these interventions are insufficient, then surgical intervention for external ventricular drainage (EVD) or decompressive craniectomy may be required. Hyperventilation to $PaCO_2$ 3.7–4.5 kPa can be used as a temporising measure as can a barbiturate infusion or increasing hypertonic therapy (avoiding serum Na >160 mmol l^{-1} or osmolality >360).

EVDs are inserted to decrease ICP by controlled release of cerebrospinal fluid and also for draining obstructive hydrocephalus in haemorrhagic brain injury. When transporting patients with EVDs the level of the drain must be controlled carefully. These are usually set 5–10 cm above a zero point (the midpoint between the lateral edge of the orbit and the tragus of the ear). Dropping the drain level may cause precipitous and excessive drainage. If the drain is lifted, then there may be some reflux back into the patient, leading to a rise in ICP and increasing danger of infection. Generally, it is safer to clamp these drains briefly for transfer but, once this is completed, to replicate their setting in PICU and maintain controlled drainage. Above all, it is

important to remember to unclamp the drain and then ensure that free drainage has returned. If not, the drain may be blocked and will need attention.

Metabolic Diseases

Inborn Errors of Metabolism

These usually declare themselves in early childhood but may not manifest until young adulthood. Many conditions are insidious with 'failure to thrive', but there are a few that can present as medical emergencies, sometimes catastrophically. A trigger, such as infection, precipitates an acute collapse in a previously well child, with variable manifestations: lactic acidosis, hypoglycaemia and/or hyperammonaemia. The presentation is of a sick, drowsy, poorly perfused and sometimes encephalopathic child. Failure of energy regulation may cause single- or multi-organ impairment necessitating PICU admission. Where doubt exists, it is important to measure blood glucose, lactate and ammonia (which must go to the lab on ice).

Adequate carbohydrate administration will switch off protein and lipid metabolism, which may ameliorate the root cause of the crisis:

- A 10% glucose infusion must be started and run at a minimum of 5 ml kg^{-1} h^{-1} to give glucose delivery of 6–10 mg kg^{-1} min^{-1}.
- All feeds must be stopped, as they may be substrates for toxic by-products that the child is failing to metabolise.

Once this is all accomplished, there will be time to set in motion more detailed metabolic investigations and to start ammonia scavenging medications and coenzymes. However, very high ammonia levels (>500 mcmol l^{-1}) will require haemofiltration as an emergency to clear this neurotoxic substance.

Diabetic Ketoacidosis (DKA)

DKA in children differs significantly from in adults because of the increased risk of cerebral oedema. This disastrous complication is due to the rapidity with which deranged biochemistry is corrected, such as rapid changes in serum osmolality caused by the fluids used in rehydration, rapid control of hyperglycaemia or aggressive correction of the acidosis, particularly through use of bicarbonate, or any combination of these.

Consequently, paediatric management differs from adults and specifics vary between guidelines. Constant themes include:

- Cautious administration of fluids.
- Very gradual correction of glucose.
- No initial insulin bolus, with infusion rates kept to 0.05–0.1 units kg^{-1} h^{-1}.
- Avoidance of bicarbonate.

Every patient should be given a 10 ml kg^{-1} fluid bolus over one hour, with shocked patients given 20 ml kg^{-1} over 15 minutes. Estimated fluid deficit (with the 10 ml kg^{-1} bolus subtracted) should be corrected over the next 48 hours (body weight × % dehydration × 10). After an hour, a fixed-rate insulin infusion can be commenced. Thereafter, the glucose and potassium content of fluids is altered according to changes in serum values to achieve slow normalisation of acidosis and hyperglycaemia. The fluid calculations are complex and are frequently a source of errors. It is essential to refer to current guidelines for each patient.

If cerebral oedema is suspected, and there is a deteriorating Glasgow Coma Scale (GCS) rating, then mannitol 20% (2.5–5 ml kg^{-1}) or hypertonic saline 2.7% or 3% (2.5–5 ml kg^{-1}) should be given, fluids slowed to half maintenance and the deficit replaced over 72 hours. If this does not procure a lasting improvement, the child may need to be intubated.

Intubation in DKA can pose a significant risk from worsening acidosis secondary to an abrupt rise in $PaCO_2$. The decision to intubate should be discussed with the most experienced clinician available and is usually only undertaken in cases of ventilatory failure, airway compromise or decompensated shock. A CT scan should be performed to exclude cerebral venous thrombosis, a recognised complication of profound dehydration.

Sepsis

In severe sepsis, where fluid volumes administered equal or exceed 60 ml kg^{-1}, it is advisable to consider intubation. Not only does this counteract the effects of the pulmonary oedema that often develops in these situations, but it also presents an opportunity to insert central venous and arterial catheters for more accurate haemodynamic monitoring and to facilitate the use of inotropes. It is a common mistake to underestimate

the amount of fluid a child needs to maintain cardiac output in sepsis and not to intervene early enough with inotropes. See the section 'Fluids and Inotropes in Critically Ill Children'; for more detail, see the 'Further Reading' section.

Non-accidental Injury and Child Protection ('Safeguarding')

Non-accidental injury may present as overt trauma but may 'masquerade' with seizures as 'failure to thrive' or as a 'collapse' in the emergency department. Anaesthetists and general intensivists must be alert to these possibilities, and all now have an obligation to have at least basic training in the recognition of abuse and in knowing what subsequent course of action to take. An ever-present index of suspicion is unfortunately necessary. See Chapter 5.

Managing the Potential Organ Donor

When a child or young person has been subjected to a catastrophic brain injury with incipient brain death, it may not be in the child's (or the family's) best interests to move the child from a district general hospital (DGH) to a regional PICU. Good intensive care practice includes end-of-life care. All units have agreed triggers for referral to the specialist nurse for organ donation (SN-OD) team as part of end-of-life care, enabling assessment for organ and tissue donation in a timely manner. The triggers follow best practice as identified and supported in National Institute for Health and Care Excellence (NICE), United Kingdom Post-Intensive Care Syndrome (UKPICS) and National Health Service Blood and Transplant (NHSBT) guidance and strategies for organ donation. If donation is considered an option, the specialist team are then on hand to give all the information the family requires to make an informed choice and offer support through that process.

Donation of solid organs can occur following determination of death using either a neurological (donation after brainstem death, or DBD) or cardiopulmonary (donation after circulatory death, or DCD) determination. In the United Kingdom, for infants between 37 weeks corrected gestational age and two months of life, there is an agreed code of practice for the confirmation of neurological death (see 'Further Reading').

The majority of transplanted organs come from donors after neurological determination of death. Once neurological death is declared, a period of donor optimisation is carried out. This is important, as the potential organ donor is at high risk of instability due to the loss of CNS-dependent homeostasis. Haemodynamic instability and cardiac arrest after brain death accounts for the loss of up to 25% of potential donors; loss of hormonal and metabolic equipoise also contribute to physiological derangements.

The intensivist's role is to optimise potential organs for donation along with supporting the family. The goals for organ support are well established (see 'Further Reading'); Donor Optimisation Care Bundles include:

Respiratory:

- Lung protective strategies:
  - TV 8–10 ml kg^{-1}
  - PEEP 5–10 cm H_20
  - PIP <30 mmHg
  - FiO_2 <0.4, if able, keeping $PaO_2 \geq$ 10kPa
  - pH >7.25, $PaCO_2$ 5–6.5 kPa
- Fully inflated cuffed endotracheal tube.
- Measures to prevent ventilator-associated pneumonia (VAP).
  - Bed >20° head-up, lansoprazole, etc.

Cardiovascular:

- Normal blood pressure for age.
  - Use of inotropes or vasopressors as needed.
- CVP 4–10 mmHg.
- SvO_2 >70%.
- Cardiac output measurement where appropriate and possible.

Fluids and metabolic management:

- Urine output 1–2 ml kg^{-1} hr^{-1}:
  - If >4 ml kg^{-1} hr^{-1}, consider diabetes insipidus, and treat with DDAVP or vasopressin.
- Correct electrolyte abnormalities:
  - Maintain Na <150 mmol l^{-1}.
- Start insulin if necessary to keep blood sugar 4.0–12 mmol l^{-1}.
- Methylprednisolone and thyroid hormone replacement may be necessary.
- Continue NG feeding, if appropriate.

In recent years, there has been an increase in DCD organ donation. This involves the donation of solid

organs, including heart and lungs, following active withdrawal of life-sustaining treatment and determination of death using standard cardiopulmonary criteria. The clinical team caring for the child must reach a consensus opinion that the withdrawal of life-sustaining treatment is indicated. It is important to note that imminent death should be expected at the time of treatment withdrawal. Patients who currently are expected to have a prolonged death or one way wean are not currently deemed potentials for organ donation. Potentially all patients that die can donate tissues, and this should be considered as part of normal end of life care.

Key Points

- It is important that 'generalist' anaesthetists understand the principles of care of the critically ill child, as they are best placed to provide immediate care to a child presenting to the emergency department, working in conjunction with paediatricians.
- Ketamine maintains cardiovascular stability in the shocked child. Propofol should be used with caution in cardiovascular compromise.
- Cuffed tracheal tubes should be used in children over 28 days old, and the cuff pressure should be monitored.
- The need for fluid replacement and inotropic support is frequently underestimated in children with shock.
- Non-accidental injury should be considered in children presenting with trauma and in neonates with unexplained collapse.

Further Reading

Academy of Medical Royal Colleges. A Code of Practice for the Diagnosis and Confirmation of Death. 2008. Available at: www.aomrc.org.uk/wp-content/uploads/2016/04/Code_Practice_Confirmation_Diagnosis_Death_1008.pdf. Accessed 25 April 2024.

Davis AL, Carcillo JA, Aneja RK et al. American College of Critical Care Medicine clinical practice parameters for hemodynamic support of pediatric and neonatal septic shock. *Critical Care Medicine* 2017 June; 45(6):1061–93.

Heddy N, British Society for Paediatric Endocrinology and Diabetes. Guideline for the Management of Children and Young People under the Age of 18 Years with Diabetic Ketoacidosis. 2021. Archives of Disease in Childhood – Education and Practice.

Healthcare Improvement Scotland. British Guideline on the Management of Asthma: A National Clinical Guideline. 2019.

Kotloff RM, Blosser S, Fulda GJ et al. Management of the potential organ donor in the ICU: Society of Critical Care Medicine/American College of Chest Physicians/Association of Organ Procurement Organizations consensus statement. *Critical Care Medicine* 2015; 43(6): 1291–325.

Murphy PJ, Marriage SC, Davis PJ. *Case Studies in Pediatric Critical Care*. Cambridge University Press. 2009.

Nichol DG, ed. *Rogers' Textbook of Pediatric Intensive Care*, 5th ed. Lippincott, Williams & Wilkins. 2016.

Pearson GA. Intensive care: because we can or because we should? *Archives of Disease in Childhood* 2018; 103(6):527–8.

PICANet, Universities of Leeds and Leicester. Paediatric Intensive Care Audit Network Annual Report 2020. 2021.

Royal College of Paediatrics and Child Health. The Diagnosis of Death by Neurological Criteria in Infants Less than Two Months Old. 2015.