Jones'  
Clinical Paediatric Surgery  
Diagnosis and Management
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By the Staff of the Royal Children’s Hospital, Melbourne

EDITED BY

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The progressive increase in the body of information relative to the surgical specialties has come to present a vexing problem in the instruction of medical students. There is not enough time in the medical curriculum to present everything about everything to them, and in textbook material, one is reduced either to synoptic sections in textbooks of surgery or to the speciality too detailed for the student or the non-specialist in complete and authoritative textbooks.

There has long been a need for a book of modest size dealing with paediatric surgery in a way suited to the requirements of the medical student, general practitioner and paediatrician. Peter G. Jones and his associates from the distinguished and productive group at the Royal Children’s Hospital in Melbourne have succeeded in meeting this need. The book could have been entitled *Surgical Conditions in Infancy and Childhood*, for it deals with the child and his afflictions, their symptoms, diagnosis and treatment rather than surgery as such. The reader is told when and how urgently an operation is required, and enough about the nature of the procedure to understand its risks and appreciate its results. This is what students need to know and what paediatricians and general practitioners need to be refreshed on.

Many of the chapters are novel, in that they deal not with categorical diseases but with the conditions that give rise to a specific symptom – Vomiting in the First Month of Life, The Jaundiced Newborn Baby, Surgical Causes of Failure to Thrive. The chapter on genetic counselling is a model of information and good sense.

The book is systematic and thorough. A clean style, logical sequential discussions and avoidance of esoterica allow the presentation of substantial information over the entire field of paediatric surgery in this comfortable-sized volume with well-chosen illustrations and carefully selected bibliography. Many charts and tables, original in conception, enhance the clear presentation.

No other book so satisfactorily meets the need of the student for broad and authoritative coverage in a modest compass. The paediatric house officer (in whose hospital more than 50% of the patients are, after all, surgical) will be serviced equally well. Paediatric surgeons will find between these covers an account of the attitudes, practices and results of one of the world’s greatest paediatric surgical centres. The book comes as a fitting tribute to the 100th anniversary of the Royal Children’s Hospital.

Mark M. Ravitch
Professor of Paediatric Surgery
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Foreword to the First Edition
Mr Peter Jones (1922–1995) MB, MS, FRCS, FRACS, FACS, FAAP. The first Australian surgeon to obtain the FRACS in paediatric surgery, and member of RACS Council (1987–1995), Vice-President of the Medical Defence Association of Victoria (1974–1988) and President of the Australian Association of Surgeons (1983–1986). He was legendary as a medical historian and in heraldry, as a great raconteur, but primarily as a great student teacher.
Preface to the Sixth Edition

The objective of the first edition of this book was to bring together information on surgical conditions in infancy and childhood for use by medical students and resident medical officers. It remains a great satisfaction to our contributors that the book has fulfilled this aim successfully, and that a sixth edition is now required. Family doctors, paediatricians and many others concerned with the welfare of children have also found the book useful.

A knowledgeable medical publisher once commented to Peter Jones that this book is not about surgery but about paediatrics, and this is what it should be, as we have continued to omit almost all details of operative surgery.

The plan for the fifth edition has been largely retained, with the inclusion of a list of key points for each chapter. Nearly half of the contributors to this edition are new members of the hospital staff, and bring a fresh outlook and state-of-the-art ideas. We have maintained the previously successful style, while including some new images.

It is now more than 10 years since Mr Peter Jones died, and this book remains as a dedication to him. Peter was a great teacher and it is a daunting task for those who follow in his footsteps. We hope this new edition will continue to honour the memory of a great paediatric surgeon who understood what students need to know.
Acknowledgements

Many members of the Royal Children’s Hospital community have made valuable contributions to this sixth edition. The secretarial staff of the Department of Surgery, and particularly Mrs Shirley D’Cruz, are thanked sincerely for their untiring support. Finally, we express our gratitude to Elisabeth Dodds of Blackwell Publishing for bringing this edition to fruition.
Introduction
Antenatal diagnosis is one of the most rapidly developing fields in medical practice. Whilst the genetic and biochemical evaluation of the developing fetus provides the key to many medical diagnoses, the development of accurate ultrasound has provided the impetus for the diagnosis of surgical fetal anomalies. At first, it was expected that the antenatal diagnosis of fetal problems would lead to better treatment and an improved outcome. In some cases, this is true. Antenatally diagnosed fetuses with gastroschisis will be delivered in a tertiary-level obstetric hospital with neonatal intensive care in order to prevent hypothermia, and the results of treatment have improved. In other cases, such as in diaphragmatic hernia, these expectations have not been fulfilled because antenatal diagnosis has opened a Pandora’s box of complex and lethal anomalies, which in the past never survived the pregnancy, and were recorded in the statistics as fetal death in utero and stillbirth.

**Indications and timing for antenatal ultrasound**

All pregnancies are now assessed with a mid-trimester ultrasound, which is usually performed at 17–18-weeks' gestation [Fig. 1.1]. The main purpose of this examination is to assess the obstetric parameters of the pregnancy, but the increasingly important secondary role of this study is to screen the fetus for anomalies. Most anomalies are picked up at 18 weeks, but some only become apparent later in the pregnancy. Renal anomalies are best seen on a 30-week ultrasound as urine flow is low before 24 weeks. Earlier ultrasound examinations may be performed with transvaginal scanning in special circumstances, such as a previous pregnancy with neural tube defect and increasingly to detect early signs of aneuploidy. Magnetic resonance imaging of the developing fetus may be another means of fetal assessment in future.

**Natural history of fetal anomalies**

Before the advent of ultrasound, paediatric surgeons saw only a selected group of infants with congenital anomalies. These babies had survived the pregnancy and lived long enough after birth to reach surgical attention. Thus the babies coming to surgical treatment were already a selected group, mostly with a good prognosis.

Antenatal diagnosis has brought surgeons into contact with a new group of conditions with a poor prognosis, and at last the full spectrum of pathology is coming to surgical attention. For example, posterior urethral valves causing obstruction of the urinary tract were thought to
be rare, with an incidence of 1:5000 male births; most cases did well with postnatal valve resection. It is now known that the true incidence of urethral valves is 1:2500 male births, and these additional cases did not come to surgical attention as they developed intrauterine renal failure with either fetal death \textit{in utero} or early neonatal death from respiratory problems such as Potter’s syndrome. It was thought that antenatal diagnosis would improve the outcome of such congenital anomalies, but the overall results have appeared to become worse with these severe ‘new’ cases being included.

There are similar problems with the antenatal diagnosis of diaphragmatic hernia [Fig. 1.2]. Congenital diaphragmatic hernia was not associated with multiple congenital anomalies when cases presented after birth. Now antenatal diagnosis of diaphragmatic hernia has uncovered a more severe subgroup with associated chromosomal anomalies and multiple developmental defects. It would seem that the earlier the diaphragmatic hernia is diagnosed, the worse is the outcome.

Despite these problems, there are many advantages in antenatal diagnosis. The outcome of many conditions is improved by the prior knowledge of ‘congenital’ anomalies.

\section*{Management following antenatal diagnosis}

\subsection*{Fetal management}
Cases diagnosed antenatally may be classified into three groups.
Good prognosis

In some cases, such as a unilateral hydronephrosis, there is no place for active management, and the main task is to track the progress of the problem through pregnancy with serial ultrasound. A detailed diagnosis is made with the more sophisticated range of tests available after birth, and UTIs are prevented with prophylactic antibiotics commenced at birth. Thus, a child with severe vesico-ureteric reflux may go through the first year of life without any UTIs. If the parents receive counselling by a surgeon experienced in the care of the particular problem, they have time to understand the condition. In the case of cleft lip seen on fetal ultrasound, the parents will have time to understand the nature of the problem by seeing photographs of the condition before and after surgery, and will have the chance to meet other families with this condition. With such preparation, the family can cope better with the birth of a baby with congenital anomalies.

The paediatric surgeon also has an important role to play in advising the obstetrician on the prognosis of a particular condition. Some cases of exomphalos are easy to repair, whereas in others, the defect may be so large that primary repair will be difficult. Moreover, in some fetuses there may be major chromosomal and cardiac anomalies associated with the exomphalos which may alter the outcome. In exomphalos, therefore, the prognosis varies from good to poor. In other conditions, the outlook for a congenital defect may change as treatment improves. Gastroschisis was a lethal condition before 1970, but now management of the disease has changed and there is a 95% survival rate. In those cases with a good prognosis, fetal intervention is not indicated and the pregnancy should be allowed to run its course. The mode of delivery will usually be determined on obstetric grounds. Babies with exomphalos may be delivered by vaginal delivery if the birth process is easy. In other obstetric circumstances, caesarian section may be indicated to prevent rupture of the exomphalos. There is evidence that spina bifida cases may undergo further nerve damage at vaginal delivery, and caesarian section may be preferred in this circumstance. If urgent neonatal surgery is required, for example, for gastroschisis, the baby should be delivered at a tertiary obstetric unit with a neonatal intensive care unit and neonatal surgical service. In other cases, for example, cleft lip and palate, where urgent surgery is not required but good family and nursing support is important, delivery close to the family’s home may be more appropriate. Antenatal planning and family counselling give us the opportunity to make the appropriate arrangements for the birth. A baby born with gastroschisis in the middle of winter in a bush nursing hospital in the mountains, many hours away from surgical care, will have a very different outlook from a baby with the same condition born at a major neonatal centre.

Poor prognosis

Anencephaly, diaphragmatic hernia with major chromosomal anomalies or urethral valves with early intrauterine renal failure are examples of conditions with a poor prognosis. These are lethal conditions, and the outcome is predetermined before the diagnosis is made.

Late deterioration

Initial assessment of the fetal anomaly indicates a good prognosis with no reason for interference, but, later in gestation, the fetus deteriorates and some action must be undertaken to prevent a lethal outcome. An example would be the lower urinary tract obstruction seen in posterior urethral valves. Early in the pregnancy, renal function may be acceptable with good amniotic fluid volumes. However, on follow-up ultrasound assessment, there may be loss of liquor with oligohydramnios as a sign of intrauterine renal failure. There are several ways to treat this problem. If the gestation is at a viable stage, for example, 36 weeks, labour could be induced and the urethral valves treated at birth. If the risks of premature delivery are higher, for example, for 28-weeks’ gestation, temporary relief may be obtained by using percutaneous transuterine techniques to place a shunt catheter from the fetal bladder into the amniotic cavity. These catheters tend to
become dislodged by fetal activity. A more definitive approach to drain the urinary tract is intrauterine surgery to perform a vesicostomy and allow the pregnancy to continue. This procedure has been performed with success in a few cases of posterior urethral valves. These patients are highly selected and only a few special centres perform intrauterine surgery. At present this surgery is regarded as experimental, and reserved for rare situations, but this may not always be the case.

Antenatal ultrasound has become the most important means of diagnosing fetal anomalies and has given us a valuable means of understanding the natural history of developmental abnormalities.

**Surgical counselling**

When a child is born with unanticipated birth defects, there is inevitably shock and confusion until the diagnosis is clarified and the family begin to assimilate and accept the information given to them, and make plans for the future. Important treatment decisions have to be made urgently while the new parents are still too stunned to play any sensible part in ongoing care of their baby. Antenatal diagnosis has changed this situation. New parents may now have many weeks to understand and come to terms with their baby’s problem. With suitable preparation, they can play an active role in the postnatal diagnosis and treatment choices for their newborn baby.

The paediatric surgical specialist who treats the particular problem uncovered by antenatal diagnosis is in the best position to advise the parents on the prognosis and further treatment of the baby. Detailed information on the management after birth, with photographs before and after corrective surgery, allows the parents to understand and come to terms with the surgical procedures. The opportunity to meet other families with a child treated for the same condition gives time for the pregnant woman and her partner to understand the problem before the birth. Handling and nurturing the baby immediately after birth is an important part of bonding. Parents and nursing staff suddenly confronted with a newborn baby with the unexpected finding of a gross anomaly, such as sacrococcygeal teratoma, may be afraid to handle the baby who is then taken away to another hospital for complex surgery. Parents in this situation may take many months to relate to the new baby and understand the nature of the problem. Prepared by antenatal diagnosis, the nursing staff and parents realise they can handle and nurture the baby. They understand the nature of the surgery and maintain their bond with the baby. Thus instead of being stunned by the birth of a malformed baby, the new parents can play an active part in the postnatal surgical management and provide better informed consent for surgery.

**Key Points**

- Antenatal diagnosis with ultrasound has revealed the natural history of some anomalies and made prognosis seem worse (e.g. diaphragmatic hernia, posterior urethra valve).
- Diagnosis before birth has allowed surgical planning (and occasional fetal intervention) as well as time for parents to be informed.

**Further reading**


The care and transport of a sick newborn baby is of critical importance to the surgical outcome. A detailed preoperative assessment is necessary to detect associated or coexistent developmental anomalies. Vital disturbances should be corrected before operation, and predictable complications of the abnormalities should be anticipated and recognised early.

Respiratory care

The aims of respiratory care are (1) to maintain a clear airway; (2) to prevent abdominal distension; (3) to avoid aspiration of gastric contents and (4) to provide supplementary oxygen if necessary. Apnoea, hyaline membrane disease, meconium aspiration and pneumothorax are common medical causes of respiratory distress in the newborn. Surgical causes of respiratory distress include oesophageal atresia and diaphragmatic hernia.

1 Placing the baby in the prone position improves the airways and reduces gastro-oesophageal reflux and the likelihood of aspiration of gastric contents.
2 Suction of the pharyngeal secretions maintains a clear airway, especially in the premature infant with poorly developed laryngeal reflexes, and in the infant with oesophageal atresia.
3 A nasogastric tube, size 8 French, will prevent life-threatening aspiration of vomitus, provided the tube is kept patent and allowed to drain freely with additional aspiration at frequent intervals. It will also reduce abdominal distension and improve pulmonary ventilation in patients with intestinal obstruction or congenital diaphragmatic hernia.
4 Oxygen therapy, endotracheal intubation and ventilation will be required in the preoperative resuscitation of some neonates with conditions such as congenital diaphragmatic hernia. However, ventilation and oxygen therapy in diaphragmatic hernia is a specialised field best left to experts as barotrauma to the poorly developed lungs will cause bronchopulmonary damage and pneumothorax.

Blood and fluid loss

The blood volume of a full term infant is 80 mL/kg. Blood loss of only 30 mL in a neonate is equivalent to losing 500 mL in an adult. Newborn babies do not tolerate blood or fluid loss well.

Fresh whole blood is cross-matched for major operations in the neonatal period. Blood loss during surgery is kept to a minimum and measured by weighing all swabs and packs used. The haemoglobin concentration in the...
first few days of life is about 19 g/dL and the haematocrit 50–70%. Blood viscosity is relatively high, and blood loss in this circumstance may be replaced in part with blood and in part with a crystalloid solution, which lowers the viscosity of the blood.

Diminished blood volume in a sick neonate with a bowel obstruction will lead to poor peripheral circulation. The baby will be lethargic and pale, with cool limbs, venoconstriction and cyanosis. Acidosis becomes a complicating factor. In this situation a ‘bolus infusion’ of a crystalloid solution such as Hartmann’s solution over 15 min at 10 mL/kg is used for resuscitation. When this is effective, the peripheral circulation will improve dramatically. If this initial infusion is not adequate, further bolus infusions of crystalloid at 10 mL/kg may be given and the clinical response monitored.

### Control of body temperature

The sick neonate with a surgical condition is prone to hypothermia, defined as a core body temperature of less than 36°C. In hypothermia, heat production is stimulated above normal metabolic requirements and may be boosted by thermogenesis from increased metabolism of brown fat deposits. However, if heat loss exceeds heat production, the body temperature will continue to fall, leading to acidosis and depression of respiratory, cardiac and nervous function.

All metabolic functions are altered by hypothermia. Newborn infants, especially the premature, are at risk of excessive heat loss because of the relatively large surface area-to-volume ratio and the lack of subcutaneous insulating fat.

Heat loss occurs from the body surface to the environment by radiation, conduction, convection and the evaporation of water. Excessive heat loss during transport, assessment and operation must be avoided, particularly in conditions such as gastrochisis where the eviscerated bowel provides a very large surface area for evaporation. Heat loss is controlled once the bowel is wrapped in domestic clear plastic wrap to prevent evaporation. Wet packs should never be applied to a neonate as they will accelerate evaporative and conductive heat losses.

Radiant overhead heaters are of particular value during procedures such as intravenous cannulation or the induction of anaesthesia, because they allow unimpeded access to the infant.

### Fluids, electrolytes and nutrition

Many infants with a surgical condition cannot be fed in the perioperative period. Intravenous fluids provide daily maintenance requirements and prevent dehydration. The total volume of fluid given must supply maintenance requirements, restore fluid and electrolyte deficits and replace ongoing losses.

**Maintenance water requirements are**
- 60–80 mL/kg on day 1 of life
- 80–100 mL/kg on day 2
- 100–150 mL/kg on day 3 and thereafter.

**Maintenance electrolyte requirements are**
- Sodium: 3 mmol/kg/day
- Chloride: 3 mmol/kg/day
- Potassium: 2 mmol/kg/day.

**Maintenance joule requirements are**
- 100–140 kJ/kg/day.

These maintenance requirements can be provided in the first week by a solution made up of 5% dextrose in 0.45% sodium chloride (sodium: 35 mmol/L) with the addition of potassium chloride at 20 mmol/L. However, this solution is inadequate for long-term maintenance of body functions as it has many deficiencies, especially in kJ.

Replacement of fluid and electrolyte deficiencies may be necessary in surgical patients, such as those with neonatal bowel obstruction. Before birth, the placenta maintains fluid and electrolyte balance. At birth, electrolyte levels are normal despite long-standing bowel obstruction, and extracellular water levels are relatively high. Persistent vomiting after birth soon causes dehydration and electrolyte imbalance. The degree of dehydration can be measured by the clinical parameters of tissue turgor, the state of peripheral circulation, depression of the fontanelle, dryness of the mouth and urine output. Bodyweight loss also gives an approximation of water loss.

The rule of thumb for estimating water loss is that dehydration of 5% or less of body mass has few clinical manifestations: 5–8% shows moderate clinical signs of dehydration; 10% shows severe signs and poor peripheral circulation. Thus a 3000 g infant who has been vomiting and has a diminished urine output, but shows no overt signs of dehydration, may have lost approximately 5% of body mass and will require $3000 \times 5\% \text{ mL} = 150 \text{ mL}$ fluid replacement to correct the deficit. Maintenance fluid requirements must be administered also.

Electrolyte estimations are most useful for identifying a deficiency of electrolytes that are distributed mainly in
the extracellular fluid, for example sodium, but will not be as reliable for electrolytes that are found mainly in the intracellular space, for example potassium. Fluid and electrolyte deficiency due to vomiting will need to be replaced with a crystalloid solution which contains adequate levels of sodium, for example 0.9% sodium chloride (sodium: 150 mmol/L).

Continuing losses of fluid and electrolytes need to be measured and replaced. Losses may arise from nasogastric tube aspirates in bowel obstruction, diarrhoea from an ileostomy, and the excessive urinary losses which may occur after the relief of urinary obstruction, for example after resection of posterior urethral valves. When the losses are high they are best measured and replaced with an intravenous infusion of electrolytes equivalent to those of the fluid being lost.

Intravenous nutrition will be required when the period of starvation extends beyond 4–5 days. Common indications for intravenous nutrition in the neonatal period include necrotising enterocolitis, extensive gut resection and gastrochisis. The aim of intravenous nutrition is to provide all substances necessary for normal growth and development. Intravenous nutrition may be maintained for weeks or months as required. Complications of prolonged nutrition include sepsis and jaundice.

Oral nutrition is preferred where possible and breast feeding is best. Surgery to the alimentary tract may make oral feeding impossible for a variable period: gut enzyme function may be poor and various substrates in the feeds may not be absorbed. Lactose intolerance is seen commonly and leads to diarrhoea with the passage of acidic fluid stools. Other malabsorptive problems relate to sugars, protein, fat and the osmolarity of the feeds. These can be handled by altering the conformation of the feeds or, in severe cases, by a period of intravenous nutrition to allow the gut enzymes time to recover.

**Biochemical abnormalities**

Important problems include metabolic acidosis, hypoglycaemia and hypocalcaemia. These are corrected before operation because they may adversely influence the infant’s response to anaesthetic agents.

**Metabolic acidosis**

Metabolic acidosis, which may result from hypovolaemia, dehydration, cold stress, renal failure or hypoxia, increases pulmonary vascular resistance and impairs cardiac output. Acidosis is corrected by fixing the underlying cause of the acidosis. In some circumstances such as renal failure sodium bicarbonate is also used.

**Hypoglycaemia**

Hypoglycaemia occurs in the sick newborn, especially if premature. Liver stores of glycogen are small, as are fat stores. Starvation and stress will use up liver glycogen rapidly, and there will be a switch to fatty acid metabolism to maintain blood glucose levels, with consequent ketoadidosis. Gluconeogenesis from amino acids or pyruvate is slow to develop in the newborn, due to the relative inactivity of liver enzymes. A point is soon reached when blood glucose levels cannot be maintained and severe hypoglycaemia will result, causing apnoea, convulsions and cerebral damage. These complications of hypoglycaemia may be prevented by intravenous dextrose infusions. Young babies should not be starved for longer than 4 h before surgery.

**Hypocalcaemia**

Hypocalcaemia may occur in infants with respiratory distress. The ionised calcium level in the blood maintains cell membrane activity. Hypocalcaemia may cause twitching and convulsions, and can be corrected by slow infusion of calcium gluconate.

**Prevention of infection**

The poorly developed immune defences of the newborn infant predispose to infection with Gram positive and Gram negative organisms. Infection may spread rapidly, and result in septicaemia. Signs of systemic infection in the neonate include hypothermia, pallor and lethargy.

Early recognition and treatment of infection is aided by bacteriological cultures from the infant’s nose, throat, umbilicus and rectum, both on admission to hospital and subsequently on a regular basis. This is important in picking up ‘marker organisms’ such as multiple antibiotic-resistant *Staphylococcus aureus*. When infection is suspected, a ‘septic workup’ is performed, taking specimens of CSF, urine and blood for culture and starting appropriate intravenous antibiotics immediately.

Infants undergoing surgery are at special risk of infection, and care must be taken not to introduce pathogenic organisms: this applies particularly to cross-infection in the neonatal ward. Handwashing with antiseptic must be performed before handling any patient. Prophylactic antibiotics may be used to cover major surgery.
Parents

An important part of the care for neonates undergoing surgery is the reassurance and support of the infant’s anxious parents. The mother may be confined in a maternity hospital while her baby is separated from her and undergoing major surgery in another institution. Close communication is important in this situation, and the mother and baby should be brought together as soon as possible. The parents should handle and fondle the baby to facilitate bonding and for the infant’s general welfare. With goodwill, gentle contact between infant and mother can be achieved, even in difficult circumstances.

General principles of neonatal transport

Transport of a critically ill neonate is a precarious undertaking, and the following principles should be followed:
1. The infant’s condition should be stabilised before embarkation.
2. The most experienced/qualified personnel available should accompany the patient.
3. Specialised neonatal ‘retrieval’ services should be used.
4. Transport should be as rapid as possible, but without causing further deterioration or incurring unnecessary risks to patient or transporting personnel.
5. Transport should be undertaken early rather than late.
6. All equipment should be checked before setting out.
7. The receiving institution should be notified early so that additional staff and equipment can be prepared for arrival.

Transport of neonatal emergencies

A list of the more common emergencies is given in Table 2.1. Most infants with these conditions should have transport arranged as soon as the diagnosis is apparent or suspected.

Some developmental anomalies do not require transportation, and specialist consultation at the hospital of birth may suffice (e.g. cleft lip and palate, orthopaedic deformities). Where doubt exists concerning the appropriateness or timing of transportation, specialist advice should be sought.

Choice of vehicle

The choice between road ambulance, helicopter or fixed-wing aircraft will depend on distance, availability of vehicle, time of day, traffic conditions, airport facilities and weather conditions. In general, fixed-wing aircraft offer no time advantages for transfers of under 160 km (100 miles).

Patients with entrapped gas (e.g. pneumothorax or significant abdominal distension) are better not to travel by air. If air travel is necessary, the aircraft should fly at low levels if it is unpressurised; otherwise expansion of the trapped gases with decrease in ambient atmospheric pressure may make ventilation difficult.

Communication

Good communication between the referring and receiving institutions can be crucial to survival and expedites treatment prior to transportation. Any change in the patient’s condition should be reported to the receiving unit in advance of arrival. Detailed documentation of the history and written permission for treatment, including surgery, should be sent with the infant. In addition, neonates require 10 mL of maternal blood to accompany them, as well as cord blood and the placenta, if available.

Details of stabilisation procedures can be discussed with the headquarters of the transport team if difficulties arise while awaiting their arrival.

Written permission for transport is required. A full explanation of what has been arranged and why, and an accurate prognosis should be given to the parents. They should be allowed as much access to the infant or child prior to transport as is possible. The parents can be given

Table 2.1 Neonatal surgical conditions requiring transportation

<table>
<thead>
<tr>
<th>Obvious malformations</th>
<th>Exomphalos/gastroschisis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Myelomeningocele/encephalocele</td>
</tr>
<tr>
<td></td>
<td>Imperforate anus</td>
</tr>
<tr>
<td>Respiratory distress</td>
<td>Chonoal atresia</td>
</tr>
<tr>
<td></td>
<td>Pierre–Robin syndrome</td>
</tr>
<tr>
<td>Upper airways obstruction</td>
<td>Emphysematous lobe</td>
</tr>
<tr>
<td>Lung compression</td>
<td>Pulmonary cyst(s)</td>
</tr>
<tr>
<td></td>
<td>Pneumothorax (should have chest drain inserted)</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>Congenital diaphragmatic hernia</td>
</tr>
<tr>
<td>Acute alimentary or abdominal emergencies</td>
<td>Oesophageal atresia</td>
</tr>
<tr>
<td></td>
<td>Intestinal obstruction</td>
</tr>
<tr>
<td></td>
<td>Necrotising enterocolitis</td>
</tr>
<tr>
<td></td>
<td>Haematemesis and/or melaena</td>
</tr>
<tr>
<td>Ambiguous genitalia</td>
<td></td>
</tr>
</tbody>
</table>
a digital photograph of the infant, taken before departure or at admission to hospital, if they are to be separated from their infant.

**Stabilisation of neonates prior to transfer**

Table 2.2 presents neonatal medical conditions that require stabilisation before transport.

**Temperature control**

An incubator or radiant warmer is used to keep the infant warm. Recommended incubator temperatures are shown in Table 2.3. The infant should be covered except for parts required for observation or access. Axillary or rectal temperatures should be taken half-hourly, or quarter-hourly if under a radiant warmer.

**Respiratory distress**

**Oxygen requirements**

Enough oxygen should be given to abolish cyanosis and ensure adequate saturation. Pulse oximeter oxygen saturation levels >97% indicate adequate oxygenation. If measurements of blood gases are available, an arterial PO$_2$ of 50–80 mm Hg is desirable. Although an excessively high PO$_2$ is liable to initiate retinopathy of prematurity, a short period of hyperoxia is less likely to be detrimental than a similarly short period of hypoxia.

**Respiratory failure**

Infants in severe respiratory failure (on clinical grounds or PCO$_2$ > 70 mmHg), or those with apnoea, may require endotracheal intubation and intermittent positive pressure ventilation. Positive pressure ventilation may cause problems in cases of diaphragmatic hernia.

**Metabolic derangements**

Hypoglycaemia should be corrected by intravenous infusion of glucose. Monitoring of babies at risk should be done with Dextrostix. Intravenous infusion may be by the umbilical, or a peripheral, route.

An infusion of blood or plasma expander at 10–20 mL/kg, i.v. over 0.5–1 h may be required to correct shock.

Acid–base balance should be estimated if facilities are available. Otherwise, a small volume of sodium bicarbonate (3 mmol/kg, slowly i.v.) may be given to an infant who has been asphyxiated severely, has had recurrent hypoxia, or shows signs of poor peripheral circulation. The best way, however, to correct acidosis is to correct the underlying abnormality.

Convulsions should be controlled with phenobarbitone (10–15 mg/kg i.v. or orally) or diphenylhydantoin (15 mg, i.v. or orally).

Specialist advice regarding management of specific conditions should be sought from the transporting agency. For example, in gastroschisis and exomphalos, the exposed viscera should be wrapped in clean plastic wrap to prevent heat loss; moist packs or gauze should never be used. A nasogastric tube with continuous drainage is required for patients with diaphragmatic hernia (Chapter 5), bowel obstruction (Chapter 7) or exomphalos (Chapter 9). In oesophageal atresia, frequent aspiration of the blind upper oesophageal pouch, at 10–15 min intervals, is essential to avoid aspiration (Chapter 6).

**Table 2.2 Neonatal medical conditions requiring stabilisation before transport**

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Prematurity</td>
</tr>
<tr>
<td>2 Temperature control problems</td>
</tr>
<tr>
<td>3 Respiratory distress causing hypoxia and/or respiratory failure</td>
</tr>
<tr>
<td>4 Metabolic derangements</td>
</tr>
<tr>
<td>- Hypoglycaemia</td>
</tr>
<tr>
<td>- Metabolic acidosis</td>
</tr>
<tr>
<td>- Hypocalcaemia</td>
</tr>
<tr>
<td>5 Shock</td>
</tr>
<tr>
<td>6 Convulsions</td>
</tr>
</tbody>
</table>

**Table 2.3 Incubator temperature**

<table>
<thead>
<tr>
<th>Baby’s weight (Gm)</th>
<th>Incubator temperature (ºC)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1000</td>
<td>35–37</td>
</tr>
<tr>
<td>1000–1500</td>
<td>34–36</td>
</tr>
<tr>
<td>1500–2000</td>
<td>33–35</td>
</tr>
<tr>
<td>2000–2500</td>
<td>32–34</td>
</tr>
<tr>
<td>&lt;2500</td>
<td>31–33</td>
</tr>
</tbody>
</table>

**Key Points**

- Sick neonates need stabilisation before transport.
- Early transport is best done by a specialised team.
- Communication with parents, and with receiving surgical centre is crucial.
Further reading

Great effort should be made to minimise psychological disturbances in children undergoing surgery. The important factors to consider are the age and temperament of the child; the site, nature and extent of the surgical procedure; the degree and duration of discomfort after operation and the time spent in hospital.

Children between 1 and 3 years of age are the most vulnerable, and many procedures should be done in the first year of life, or postponed until 4 or 6 years of age when the child can comprehend and co-operate better.

The temperament and ability of children to cope with stress are infinitely variable; the trust which children are prepared to grant those who care for them is a measure of the confidence they have in their own family circle. Major disturbances within the family may affect the patient’s equanimity and the ability of the parents to give support. Elective surgery may need to be deferred for stressful family events such as:
- the arrival of a new baby;
- a death in the family;
- shifting to a new house.

Preparation for elective admission is important for children over 4 or 5 years of age and, whether assisted by a booklet (see Further Reading) or advice, is largely in the hands of the parents whose acceptance of the situation is its endorsement in the child’s eyes.

The child needs a brief and simple description of the operation, and if something is to be removed, it should be made clear that it is dispensable. Children should also be told that they will be asleep while the operation is performed, that it will be over when they wake and that they will be ‘stiff’ and a little ‘sore’ for a day or so, and they should also be told the time when they will be able to go home. It is pointless to say that it will not hurt at all, for honesty is essential to preserve trust.

How the child’s questions are handled is just as important as the factual content of the answers; possible sources of fear should be dealt with, and the pleasant aspects suitably emphasised. The amount of information must be adjusted to the child’s age and particular needs; more detail will be expected by older children.
Effect of site of surgery

Operations on the genitalia or the body’s orifices, including circumcision after the age of 2 years, are more likely to cause emotional upset than other operations of the same magnitude. One or both parents should stay with the child and suitable occupational or play therapy is of considerable value.

Anal and oesophageal surgery should be completed shortly after birth, and subsequent dilatations performed under anaesthesia wherever possible. Many boys who have experienced both operations would prefer, in retrospect, bilateral orchidopexy to tonsillectomy by even the gentlest hands.

Day surgery

Time spent in hospital should be as short as possible. ‘Day surgery’ with admission, operation and discharge a few hours later is cost-effective, convenient and suitable for at least 80% of elective paediatric surgery.

The greatest advantage is minimizing the psychological impact on the child, which is magnified by sleeping away from home for even one night. There are many other obvious advantages, including minimal disturbances of breast feeding and reduced travelling by parents (i.e. fewer visits to the hospital).

Although surgical technique is important (haemostasis, secure dressings), day surgery has been made safe and acceptable by special anaesthetic techniques: timing and choice of premedication and general anaesthetic agents, minimal trauma during intubation (particularly the use of the laryngeal mask rather than endotracheal intubation), reversal of anaesthesia and long-acting local anaesthetic blocks or caudal analgesia in lieu of the usual postoperative injections of narcotics.

In the most vulnerable 1–3 years age group, day surgery has reduced the likelihood of behavioural disturbances. Suitable operations for day surgery depend on parental attitudes, logistics and careful selection of individual patients.

Ward atmosphere and procedures

Unlimited visiting by parents, living-in quarters for parents and a more understanding and empathetic approach by all who care for children have led to a less formal and more friendly atmosphere in hospital.

The procedures necessary for investigation, or in preparation for operation, should be scrutinised carefully to see whether they are necessary. Blood tests or x-rays are rarely required for elective day surgery.

The induction of anaesthesia is an important source of fear and distress. The presence of a parent is very helpful during most anaesthetic inductions. Effective premedication, skilful intravenous induction and the prompt administration of hypnotics and analgesics after operation keep discomfort to the absolute minimum. Again, the early presence of a parent in the postoperative recovery room will reduce the child’s stress as it wakes up from anaesthesia.

Even after major abdominal surgery, some toddlers will be walking within 48 h. They might just as well be playing on the floor or sitting at a table, and today that is where they are, with no subsequent ill effects. The playroom is not required for most postoperative patients, since once they can walk to the toilet and playroom, they can be discharged home. The child usually sets the pace of convalescence, and as a general rule will show no desire to move when they should rest, for example, during a period of paralytic ileus.

Play materials, a day room, television and bright surroundings act as constant stimuli to those who are well enough to be ‘up and doing’. Play specialists are involved in the management of children who have a longer hospital stay or require frequent dressing changes (e.g. burns patients), and they significantly reduce the amount of analgesia required.

A single, absorbable subcuticular stitch can be used to close almost all incisions, and avoids the anxiety and time spent in removing sutures. A waterproof dressing allows normal washing, and can be left on until the wound is fully healed.

Parental support

The parents always require consideration, especially when a first-born baby is transferred to a children’s hospital on the first day of life. The baby may stay there for several weeks, at precisely the time when the mother’s emotions are in turmoil and she would normally be establishing a new and unique relationship. Feelings of guilt at producing an infant with a congenital abnormality, or inadequacy following removal of the infant from her care and the lack of close physical contact, may lead her to have difficulty bonding to her baby and produce an exaggeration of the usual puerperal emotional instability. To help
 overcome this when separation is unavoidable, the mother should be given a photograph of her baby, and should see the baby again as soon as possible, to be involved in the day-to-day care of the child as much as the illness permits (Chapter 2).

Response of the child

The average child’s natural optimism, freedom from unfounded anxiety, remarkable powers of recuperation and apparently short memory for unpleasant experiences can make even major surgery a relatively short and simple matter. Most children are out of bed in 2–3 days and active for much of the day or are already at home by 5 days after many major operations.

Even when minor surgery has been uneventfully concluded, the child may have disturbed behaviour for several months after leaving hospital, and the parents should be made aware of this possibility. Signs of insecurity, increased dependency and disturbed sleep are not uncommon but fortunately are of short duration when met with warm affection, reassurance and understanding by the parents.

The undesirable psychological effects of surgery must be put in proper perspective by mentioning the beneficial effects which so often follow operation: the well-being after repair of an uncomfortable hernia, the freely expressed satisfaction at the excision of an unsightly lump or blemish.

Finally, in many older children there is a detectable increase in confidence and poise which comes from facing, and coping adequately with, an operation. This may be the first occasion on which the child has been away from home, and metaphorically at least, standing on his or her own two feet.

The timing of surgical procedures

Surgical conditions in infancy and childhood can be classified according to the degree of urgency with which treatment should be carried out. Three categories can be distinguished:

1. The immediate group – that is, conditions where immediate investigation and/or definitive operation is required; for example, intussusception; appendicitis.
2. The intermediate group – where treatment is not urgent, but should be undertaken without undue delay; for example, infant inguinal hernia.
3. The elective group – where operation is performed at an optimum age determined by one or more factors which affect the patient’s best interests, for example, undescended testis, hypospadias.

The immediate group

Trauma, acute infections, abdominal emergencies and acute scrotal conditions fall into this category. A particularly important subgroup is neonatal emergencies. Most of these are the result of developmental abnormalities causing disorders of function, some of which may threaten life. The best prognosis depends upon early diagnosis and timely transport to a hospital where the appropriate skills and equipment are available (sometimes this is best done where the infant is born, as in a congenital diaphragmatic hernia) (see Chapters 4–11).

The intermediate group

Inguinal hernias are prone to strangulation, especially in the first year of life. For this reason, herniotomy should be performed within a few days of diagnosis in those less than 1 year of age. Investigation of swellings or masses suspected to be malignant should be undertaken within a day or two of their discovery, in close consultation with the regional paediatric oncology service.

The elective group

Factors favouring deferment of operation

Factors which favour deferment of operation, and hence may determine an optimum age for surgery, include:

1. The possibility of spontaneous correction or cure. In infants, scrotal hydroceles, encysted hydroceles of the cord, true umbilical hernias and sternomastoid tumours all show a strong tendency to spontaneous resolution. Surgery is only required for those few that persist well beyond the age of natural resolution.
2. Strawberry naevi (intracutaneous capillary haemangiomas) progress and enlarge in the first year of life, but usually involute and fade spontaneously in the ensuing 2–4 years (Chapter 50). In general, they should be left alone to do so, and surgical measures are required rarely.
3. The difficulties posed by minute and delicate structures can be avoided by postponing operation until they are more robust, although this is seldom the sole reason for deferring operation: for example, an undescended testis can be repaired more easily in a 6–12-month-old boy than shortly after birth.
4. The development of co-operation and comprehension with age. Voluntary exercises are important after some
operations, and it may be desirable to defer them until the necessary degree of co-operation is forthcoming.

5 The effects of growth are important in some instances. Chest wall deformities are corrected at adolescence, when chest wall growth is almost complete.

6 Coexistent anomalies and intercurrent diseases, for example, infections, will affect the timing of operations. The situation in each patient should be assessed to establish the order of priorities when there are multiple abnormalities, and to determine whether the treatment of non-urgent conditions should be deferred temporarily.

Factors favouring early operation

Factors which favour early operation rather than deferred treatment include capacity for healing and adaptation in the very young. For example, a fracture of a long bone at birth causes such an exuberant growth of callus that clinical union occurs in 7–10 days, and the subsequent moulding will remove any residual bony deformities.

1 Stimulation of development by early treatment occurs in infants with a congenital dislocation of the hip. When splinting is commenced in the first week of life, this will prevent the secondary dysplasia of the acetabulum and femur, which in the past was thought to be the primary cause of the dislocation.

2 Malleability of infantile tissues is an advantage, for example, in talipes, in which the best results are obtained when treatment is commenced in the first few days after birth.

3 Avoidance of undesirable psychological effects. Often these can be prevented by completing treatment, including repetitive painful procedures, before the memory of past things is established (at about 18 months) or before the child goes to school, where obvious deformities or disabilities are likely to attract attention.

4 Effect on the parents. The family as a whole should be considered, and when it is not disadvantageous to the child, early operation may resolve parental anxiety and prevent rejection of the child.

Key Points

- All hospital and operative procedures are modified to reduce psychological stress in children.
- As much as possible, all painful procedures are done while the child is anaesthetised.
- Invisible stitches, waterproof dressing and local anaesthetic given before waking mean the wound can be left alone postoperatively.
- Day surgery avoids separation anxiety in older children.

Further reading

Frawley G (1999) *I'm Going to Have an Anaesthetic*. Paediatric Anaesthetic Department, Royal Children’s Hospital, Melbourne.