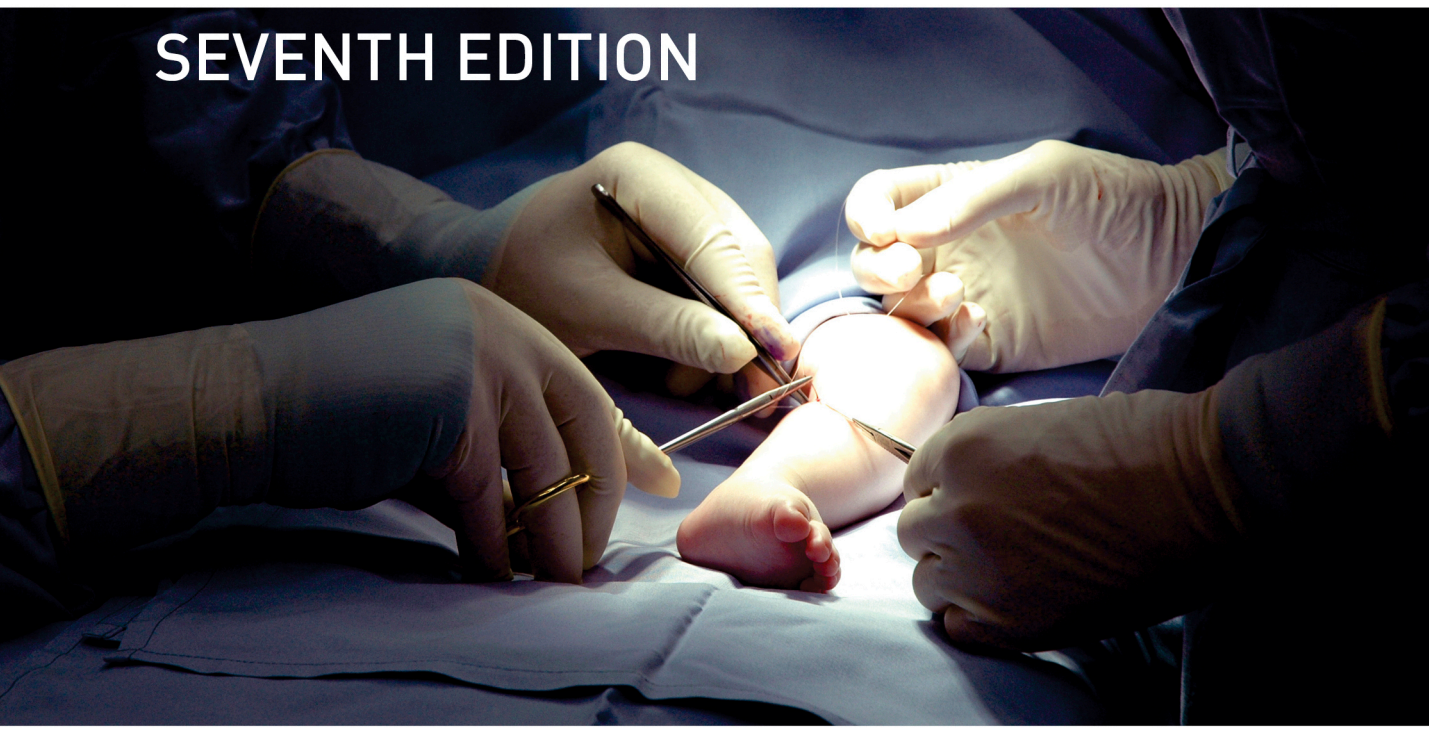
JONES' CLINICAL PAEDIATRIC SURGERY

SEVENTH EDITION



Edited by:
John M. Hutson
Michael O'Brien
Spencer W. Beasley
Warwick J. Teague
Sebastian K. King

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EDITED BY

John M. Hutson

AO, MD, DSc (Melb), MD (Monash), FRACS, FAAP
Department of Paediatrics, University of Melbourne, Melbourne, Victoria, Australia
Chair of Paediatric Surgery, Royal Children's Hospital, Parkville, Victoria, Australia

Michael O'Brien

PhD, FRCSI (Paed)
Department of Paediatric Urology, Royal Children's Hospital, Parkville, Victoria, Australia
Chief of Division of Surgery, Royal Children's Hospital, Parkville, Victoria, Australia

Spencer W. Beasley

MS, FRACS
Professor of Paediatric Surgery, Christchurch School of Medicine, University of Otago, Christchurch, New Zealand
Clinical Director, Department of Paediatric Surgery, Christchurch Hospital, Christchurch, New Zealand

Warwick J. Teague

DPhil (Oxon), FRACS
Department of Paediatric and Neonatal Surgery, Royal Children's Hospital, Parkville, Victoria, Australia

Sebastian K. King

PhD, FRACS (Paed)
Department of Paediatric and Neonatal Surgery, Royal Children's Hospital, Parkville, Victoria, Australia

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Contents

Contributors

Spencer W. Beasley, MS, FRACS
Professor of Paediatric Surgery, Christchurch School of Medicine, University of Otago
Clinical Director, Department of Paediatric Surgery, Christchurch Hospital
Christchurch, New Zealand

Robert Berkowitz, MD, FRACS
Department of Otolaryngology
Royal Children's Hospital
Parkville, Victoria, Australia

Thomas Clarnette, MD, FRACS
Department of Paediatric and Neonatal Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Joe Crameri, FRACS
Department of Paediatric and Neonatal Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

James E. Elder, FRACO, FRACS
Department of Ophthalmology
Royal Children's Hospital
Parkville, Victoria, Australia

Kerr Graham, MD, FRCS (Ed)
Professor of Orthopaedics
Royal Children's Hospital
Parkville, Victoria, Australia

Anthony Holmes, FRACS
Diplomate, American Board of Plastic Surgery;
Plastic and Maxillofacial Surgery Department
Royal Children's Hospital
Parkville, Victoria, Australia

John M. Hutson, AO, MD, DSc (Melb), MD (Monash), FRACS, FAAP
Department of Paediatrics
University of Melbourne
Melbourne, Victoria, Australia
and
Chair of Paediatric Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Bruce R. Johnstone, FRACS
Department of Plastic and Maxillofacial Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Sebastian K. King, PhD, FRACS
Department of Paediatric and Neonatal Surgery
Royal Children's Hospital,
Parkville, Victoria, Australia

Wirginia J. Maixner, FRACS
Neuroscience Centre
Royal Children's Hospital
Parkville, Victoria, Australia

Michael O'Brien, PhD, FRCSI (Paed)
Department of Paediatric Urology;
Chief of Division of Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Anthony J. Penington, FRACS
Professor of Plastic Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Russell G. Taylor, FRACS
Department of Paediatric and Neonatal Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Warwick J. Teague, DPhil (Oxon), FRACS
Department of Paediatric and Neonatal Surgery
Royal Children's Hospital
Parkville, Victoria, Australia

Foreword to the First Edition

The progressive increase in the body of information relative to the surgical specialities has come to present a vexing problem in the instruction of medical students. There is only enough time in the medical curriculum to present an overview 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 children and their 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
University of Pennsylvania

Tribute to Mr. Peter Jones

Mr Peter Jones (1922–1995) MB, MS, FRCS, FRACS, FACS, FAAP. The first Australian surgeon to obtain the FRACS in paediatric surgery, 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 Seventh 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 seventh edition is now required. Family doctors, paediatricians and many others concerned with the welfare of children have also found this 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 sixth edition has been largely retained but with the addition of new coloured photographs. Mr Alan Woodward has retired as an editor, and we have added two new editors, Mr Warwick Teague and Mr Sebastian King. 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.

It is now about 20 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 seventh edition. The secretarial staff of the Department, and particularly Mrs Shirley D'Cruz, are thanked sincerely for their untiring support.

PART I
Introduction

CHAPTER 1
Antenatal Diagnosis: Surgical Aspects

CASE 1

At 18 weeks' gestation, right fetal hydronephrosis is diagnosed on ultrasonography.

Q 1.1 Discuss the further management during pregnancy.

Q 1.2 Does antenatal diagnosis improve the postnatal outlook for this condition?

CASE 2

An exomphalos is diagnosed on the 18-week ultrasound scan.

Q 2.1 What further evaluation is required at this stage?

Q 2.2 Does this anomaly influence the timing and mode of delivery?

Antenatal diagnosis is one of the most rapidly developing fields in medical practice. While 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 to the diagnosis of surgical fetal anomalies. At first, it was expected that 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 are now routinely delivered in a tertiary-level obstetric hospital with neonatal intensive care in order to prevent hypothermia and delays in surgical treatment, and the results of treatment have improved. In other cases, such as congenital diaphragmatic hernia, these expectations have not been fulfilled because antenatal diagnosis has revealed a number of complex and lethal anomalies which in the past never survived the pregnancy and were recorded in the statistics of fetal death in utero and stillbirth.

Indications and timing for antenatal ultrasound

Most pregnancies are now assessed with a mid-trimester morphology ultrasound scan, which is usually performed at 18–20 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 fetal anomalies can be diagnosed at 18 weeks, but some only become apparent later in the pregnancy. Renal anomalies are best seen on a 30-week ultrasound scan, as urine flow is low before 24 weeks. Earlier transvaginal scanning may be performed in special circumstances, such as a previous pregnancy with neural tube defect, and increasingly to detect early signs of aneuploidy. Fetal magnetic resonance imaging is increasingly being used to assess the developing fetus in cases of suspected or confirmed fetal anomalies without exposing the fetus or mother to ionising radiation.

Natural history of fetal anomalies

Before the advent of ultrasonography (as earlier), 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.

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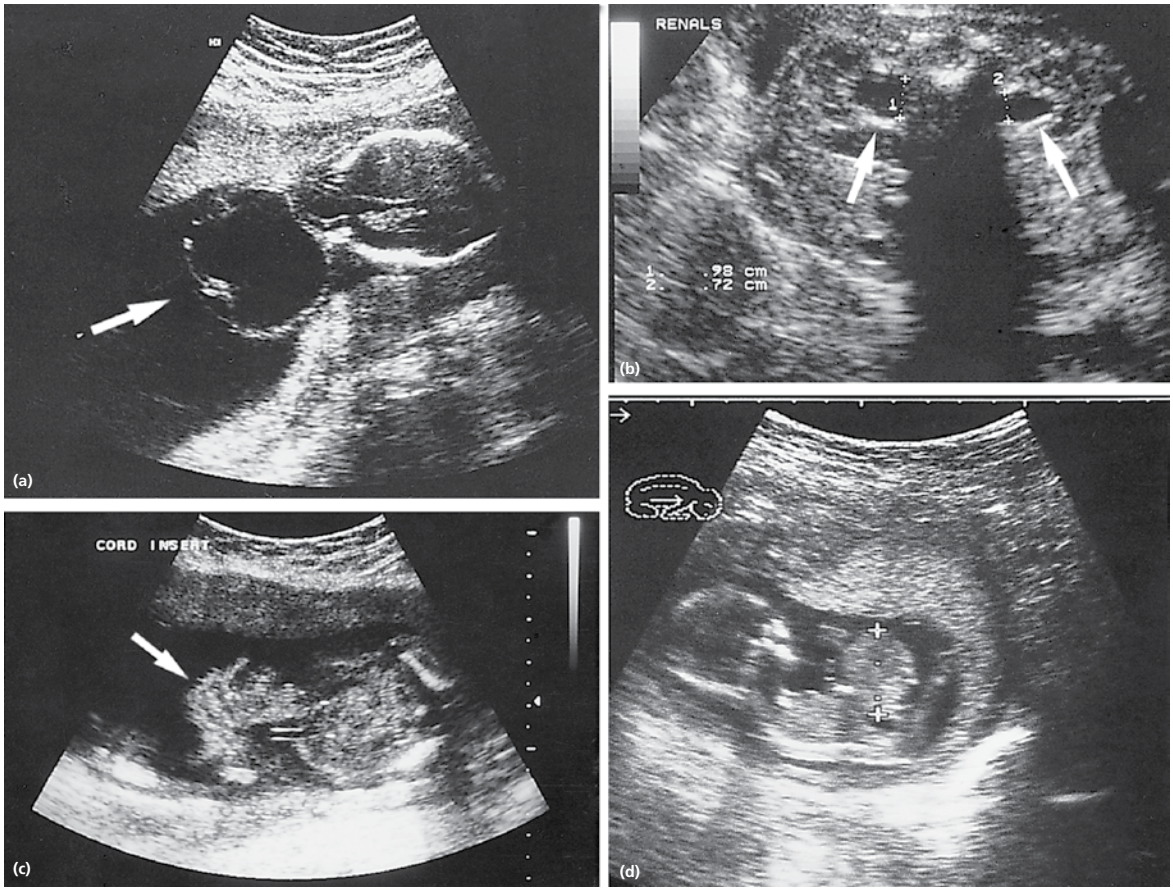

Figure 1.1 (**a**) Encephalocele shown in a cross section of the fetal head. The sac protruding through the posterior skull defect is arrowed. (**b**) Bilateral hydronephrosis shown in an upper abdominal section. The dilated renal pelvis containing clear fluid is marked. (**c**) The irregular outline of the free-floating bowel in the amniotic cavity of a term baby with gastroschisis. (**d**) A longitudinal section through a 14-week fetus showing a large exomphalos. The head is seen to the left of the picture. The large sac (marked) is seen between blurred (moving) images of the arms and legs.

Antenatal diagnosis has exposed surgeons to 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 valve causing obstruction of the urinary tract was 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 valve 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 or early neonatal death from respiratory failure because of Potter 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 the inclusion of these severe *new* cases.

In the same way, antenatal diagnosis has exposed the significant *hidden mortality* of congenital diaphragmatic hernia [Fig. 1.2]. Previously, congenital diaphragmatic hernia diagnosed after birth was not commonly associated with multiple congenital anomalies, but now, antenatal diagnosis has uncovered a more severe subgroup with associated chromosomal anomalies and multiple developmental defects. It is now apparent that the earlier the congenital diaphragmatic hernia is diagnosed in utero, the worse the outcome.

Despite these problems, there are many advantages in antenatal diagnosis. The outcomes of many congenital

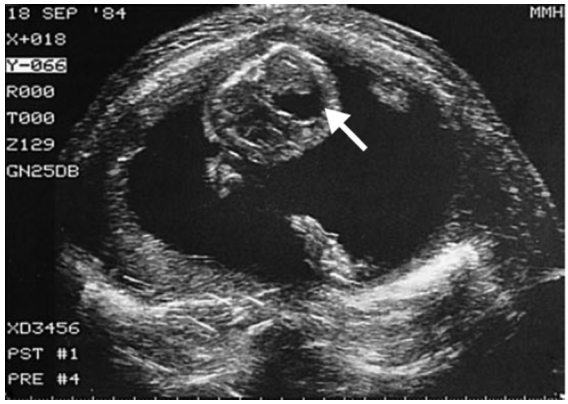

Figure 1.2 Cross section of a uterus with marked polyhydramnios. The fetal chest is seen in cross section within the uterus. The fluid-filled cavity within the left side of the chest is the stomach protruding through a congenital diaphragmatic hernia (arrow).

anomalies are improved by prior knowledge of them before birth.

Management following antenatal diagnosis

Fetal management

Cases diagnosed antenatally may be classified into three groups:

Good prognosis

In some cases, such as a unilateral hydronephrosis, there is no role for active antenatal management, and the main task is to document the progress of the condition through pregnancy with serial ultrasound scans. The detailed diagnosis is made with the more sophisticated range of tests available after birth, and the incidence of urinary tract infections (UTIs) may be reduced with prophylactic antibiotics commenced at birth. Thus, a child with severe vesicoureteric reflux may go through the first year of life without any UTIs. If the parents receive counselling by an experienced surgeon, they have time to understand the condition, its treatment and prognosis. With such preparation, the family may cope better with the birth of a baby with a congenital anomaly.

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. In addition, there may be major chromosomal and cardiac anomalies, which may alter the outcome. In other conditions, the outlook for a congenital defect may change as treatment improves. Gastroschisis was a lethal condition before 1970, but now, management 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 continue to close to term. 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. Primary caesarean section may be indicated for major exomphalos to prevent rupture of the exomphalos and damage to the organs such as the liver, as well as for obstetric indications. There is evidence that in fetuses with large neural tube defects, further nerve damage may occur at vaginal delivery, and caesarean section may be preferred in this circumstance. If urgent neonatal surgery is required, for example, in gastroschisis, the baby should be delivered at a tertiary obstetric unit with appropriate neonatal intensive care. In other cases (e.g. cleft lip and palate), where urgent surgery is not required but good family and nursing support is important, delivery closer 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, may have a very different prognosis from a baby with the same condition born at a major neonatal centre.

Poor prognosis

Anencephaly, congenital diaphragmatic hernia with major chromosomal anomalies or urethral valve 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

In most cases, initial assessment of the fetal anomaly will indicate a good prognosis with no reason for interference. However, later in gestation, the fetus may deteriorate, and some action must be undertaken to prevent

a lethal outcome. An example would be posterior urethral valve causing lower urinary tract obstruction. Early in the pregnancy, renal function may be acceptable with good amniotic fluid volumes, but on follow-up ultrasound assessment, there may be loss of amniotic fluid with oligohydramnios as a sign of renal failure. There are several approaches to this problem. If the gestation is at a viable stage, for example, 36 weeks, labour may be induced, and the urethral valve treated at birth. If the risks of premature delivery are higher, for example, at 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 valve. These patients are highly selected, and only a few special centres are able to perform intrauterine surgery. At present, this surgery is regarded as experimental and reserved for rare situations, but this may not always be the case.

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 begins to assimilate and accept the information given to them. Important treatment decisions may have to be made urgently while the new parents are still too stunned to play any sensible part in the 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 condition. With suitable preparation, they may play an active role in the postnatal treatment choices for their newborn baby.

The paediatric surgeon 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 the operative procedures. The opportunity to meet other families with a child treated for the same condition may give time for the pregnant woman and her partner to understand the problem prior to 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 an unexpected anomaly, such as sacrococcygeal teratoma, may be afraid to handle the baby prior to the baby being taken away to another hospital for complex surgery. Parents in this situation may take many months to bond with the new baby and to understand fully the nature of the problem. Prepared by antenatal diagnosis, parents realise they may handle and nurture the baby, understand the nature of the surgery and form a bond with the baby. Thus, instead of being stunned by the birth of a baby with a significant malformation, the new parents may play an active part in the postnatal surgical management and provide better informed consent for surgery.

KEY POINTS

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

Further reading

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CHAPTER 2
The Care and Transport of the Newborn

CASE 1

A 30-week gestation neonate is born with gastroschisis.

Q 1.1 What advice would you give the referring institution about the management of this infant prior to transport to a tertiary institution?

CASE 2

A 40-week gestation neonate develops respiratory distress shortly after birth. A left congenital diaphragmatic hernia (CDH) is diagnosed.

Q 2.1 List two iatrogenic problems that may occur with positive-pressure ventilation.

Q 2.2 How do you avoid these iatrogenic problems?

The initial care and transport of a sick newborn baby is critically important 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 (i) to maintain a clear airway, (ii) to prevent abdominal distension, (iii) to avoid aspiration of gastric contents and (iv) to provide supplementary oxygen if necessary. Various manoeuvres and adjuncts are commonly used in neonatal respiratory care to achieve these aims, including:

1 *Suctioning* of the pharyngeal secretions maintains a clear airway. This is especially important in the premature neonate with poorly developed laryngeal reflexes, and will need to be repeated regularly in neonates with oesophageal atresia.

2 *Prone positioning* improves the airways, assists ventilation and reduces the risk of aspiration of gastric contents with gastro-oesophageal reflux or vomiting. Importantly, this positioning applies to monitored neonates in an intensive care setting and does not contradict the *back to sleep* public health advice pertaining to prevention of sudden infant death syndrome (SIDS).

3 *Nasogastric tube insertion*, size 8 French, will minimise the risk of 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 ventilation in patients with intestinal obstruction or congenital diaphragmatic hernia (CDH).

4 *Supplementary oxygen therapy* with or without endotracheal intubation and ventilation is provided as required for respiratory distress. Common medical causes of the breathless neonate include transient tachypnoea of the newborn, meconium aspiration, pneumothorax, hyaline membrane disease and apnoea. Surgical causes of respiratory distress include oesophageal atresia and CDH. Ventilation strategies in CDH are complex and require input of specialist and experienced practitioners, who may be neonatologists, intensive care physicians or surgeons. These strategies seek to minimise barotrauma to the poorly developed lungs, which may cause bronchopulmonary damage, pneumothorax and death.

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Blood and fluid loss

Newborn babies do not tolerate blood or fluid loss well. The blood volume of a full-term neonate is 80 mL/kg. Therefore, a loss of only 30 mL blood constitutes a loss of approximately 10% of blood volume, which is equivalent to 500 mL loss in an adult. For this reason, it is routine to crossmatch whole blood prior to neonatal surgery. Blood loss is strictly kept to a minimum and measured by weighing all swabs used. Neonatal blood is relatively concentrated; haemoglobin concentration in the first days of life is about 19 g/dL and the haematocrit 50–70%. In this circumstance, blood loss may be replaced in part with blood and in part with a crystalloid solution, which lowers the viscosity of the blood.

Neonatal bowel obstruction is another common setting resulting in fluid depletion due to vomiting and nasogastric losses. Hypovolaemia is manifest with lethargy, pallor, prolonged capillary return, cool limbs, venoconstriction and cyanosis. Acidosis becomes a complicating factor. In this situation, the baby is fluid resuscitated with an initial *bolus infusion* of 10 mL/kg crystalloid solution of normal saline (0.9% NaCl) over 15 min. Effectiveness of resuscitation is indicated by improved peripheral circulation in response to the bolus. If the response is not adequate or not sustained, further 10 mL/kg bolus infusions of crystalloid may be given and circulatory status monitored.

Control of body temperature

Newborn infants, especially the premature, are at risk of excessive heat loss because of their relatively large surface area-to-volume ratio, lack of subcutaneous insulating fat and immature thermoregulation. The sick neonate with a surgical condition is prone to hypothermia, defined as a core body temperature of less than 36 °C. Neonates counteract hypothermia by increasing metabolic activity and thermogenesis by brown fat metabolism. 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.

Heat loss occurs from the body surface by radiation, conduction, convection and the evaporation of water. Excessive heat loss during assessment, procedures, transport and operation must be avoided. 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. Neonates with gastroschisis are at superadded risk of heat loss as the eviscerated bowel provides increased surface area for evaporation. Heat loss during transport and assessment is minimised by enclosing the bowel with plastic kitchen wrap or a bowel bag to prevent evaporation. Wet packs should never be applied to a neonate as they will accelerate evaporative and conductive heat losses.

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 restore fluid and electrolyte deficits, supply maintenance requirements and replace ongoing losses.

Maintenance fluid requirements are:
60–80 mL/kg on day 1 of life
80–100 mL/kg on day 2 of life
100–150 mL/kg on day 3 of life and thereafter

Maintenance electrolyte requirements are:
Sodium: 3 mmol/kg/day
Chloride: 3 mmol/kg/day
Potassium: 2 mmol/kg per day

Maintenance joule requirements are:
100–140 kJ/kg/day

In the first 2–3 days of life, maintenance requirement for sodium, potassium and chloride is minimal due to a low glomerular filtration rate and low urine output at birth. Therefore, 10% dextrose solution alone is typically sufficient for maintenance needs. Beyond 2–3 days of age, a dextrose–saline solution is required, for example, 10% dextrose in 0.18–0.225% sodium chloride (sodium: 30 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 kilojoules.

In addition to maintenance fluids, many surgical neonates will require replacement of excess fluid and electrolyte losses, especially those with neonatal bowel obstruction. Useful clinical signs of dehydration include prolonged capillary return (>2 seconds), depression of the

fontanelle, dryness of the mucous membranes, reduced tissue turgor and cool peripheries. Reduced urine output and bodyweight loss may precede these findings.

The rule of thumb for estimating fluid 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 150 mL (3000 × 5% mL) fluid replacement to correct the deficit. Maintenance fluid requirements must be administered also in addition.

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 fluid, for example, potassium. Fluid and electrolyte deficiency due to vomiting needs to be replaced with a crystalloid solution that contains adequate levels of sodium, for example, 0.9% sodium chloride (sodium:150 mmol/L).

Continuing fluid and electrolyte losses need to be measured and replaced. Losses may arise from nasogastric aspirates in bowel obstruction, diarrhoea from an ileostomy or diuresis after the relief of urinary obstruction, for example, after resection of posterior urethral valve. 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 (parenteral) nutrition will be required when starvation extends beyond 4–5 days. Common indications for parenteral nutrition in the neonate include necrotising enterocolitis, extensive gut resection and gastroschisis. The aim of parenteral nutrition is to provide all substances necessary to sustain normal growth and development. Parenteral nutrition may be maintained for weeks or months as required, although complications include sepsis and jaundice.

Oral nutrition is preferred where possible and breastfeeding is best. Gastrointestinal surgery may make oral feeding impossible for a while: gut enzyme function may be poor, and various substrates in the feeds may not be absorbed. Lactose intolerance is common and leads to diarrhoea with acidic, fluid stools. Other malabsorptive problems relate to sugars, protein, fat and osmolarity of the feeds. These may be managed by changing the formula or, in severe cases, by a period of parenteral nutrition to allow the gastrointestinal tract to recover.

Biochemical abnormalities

Important problems include metabolic acidosis, hypoglycaemia and hypocalcaemia. These must be minimised prior to an operation as they may adversely influence the neonate'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, and in renal failure, sodium bicarbonate may also be 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 consume liver glycogen rapidly, resulting in a need for fatty acid metabolism to maintain blood glucose levels, with consequent ketoacidosis. Gluconeogenesis from amino acids or pyruvate is slow to develop in the newborn, due to the relative inactivity of liver enzymes. Eventually, blood glucose levels cannot be maintained, and severe hypoglycaemia results, causing apnoea, convulsions and cerebral damage. These complications of hypoglycaemia may be prevented by intravenous dextrose infusions. Neonates should not be starved for longer than 3 h prior to an operation.

Hypocalcaemia

Hypocalcaemia may occur in neonates with respiratory distress. The ionised calcium level in the blood maintains cell membrane activity. Hypocalcaemia potentially causes twitching and convulsions but may be corrected by slowly infusing calcium gluconate.

Prevention of infection

The poorly developed immune defences of neonates 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 are often non-specific, but may include hypothermia, pallor and lethargy.

Early recognition and treatment of infection is aided by microbiological cultures from the neonate's nose and umbilicus, and in select cases groin and rectum, both on admission to hospital and while in the hospital. 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 the cerebrospinal fluid, urine and blood for culture and starting appropriate intravenous antibiotics immediately.

A neonate undergoing an operation is at a significantly increased risk of infection, and care must be taken not to introduce pathogenic organisms: this applies particularly to cross infection in the neonatal ward. Handwashing or antiseptic gel must be applied before and after handling any patient. Prophylactic antibiotics may be used to cover major operations.

Parents

An important part of care for a neonate undergoing an operation is reassurance and support for the neonate's anxious parents. The mother may be confined in a maternity hospital, while her baby is separated from her and undergoing a major operation 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. With goodwill and planning, gentle contact between neonate and mother may 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 neonate'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 may be prepared for arrival.

Transport of neonatal emergencies

A list of the more common surgical emergencies is given in Table 2.1. Most neonates 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.

Table 2.1 Neonatal surgical conditions requiring emergency transport

Obvious malformations	Exomphalos/gastroschisis
	Myelomeningocele/ encephalocele
	Anorectal malformation
Respiratory distress	
Upper airway obstruction	Choanal atresia
	Pierre Robin sequence
Lung dysplasia/compression	Congenital diaphragmatic hernia
	Emphysematous lobe
	Pulmonary cyst(s)
	Pneumothorax (insert chest drain first)
Congenital heart disease	
Acute alimentary or abdominal emergencies	Oesophageal atresia
	Intestinal obstruction
	Necrotising enterocolitis
	Haematemesis and/or melaena
Disorders of sex development (DSD)	

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, 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 is 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 neonate. 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 may be discussed with the transport team, or receiving institution, if difficulties arise while awaiting the transport team's 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 as is possible to the neonate prior to transport. The parents may be given a digital photograph of their child, taken before departure or at admission to hospital, if they are to be separated.

Stabilisation of neonates prior to transfer [Table 2.2]

Temperature control

An incubator or radiant warmer is used to keep the neonate warm. Recommended incubator temperatures are shown in Table 2.3. The neonate should remain 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.

Table 2.2 Neonatal medical conditions requiring stabilisation before transport

1. Prematurity
2. Temperature control problems
3. Respiratory distress causing hypoxia and/or respiratory failure
4. Metabolic derangements
   - Hypoglycaemia
   - Metabolic acidosis
   - Hypocalcaemia
5. Shock
6. Convulsions

Table 2.3 Incubator temperature

Neonate's weight (g)	Incubator temperature (°C)
<1000	35–37
1000–1500	34–36
1500–2000	33–35
2000–2500	32–34
>2500	31–33

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 mmHg 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

Neonates 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. Special attention must be paid to those neonates with CDH.

Metabolic derangements

Hypoglycaemia should be corrected by intravenous glucose. Monitoring of neonates at risk should be done with Dextrostix, with intravenous access by the umbilical or a peripheral vein.

An infusion of blood or plasma expander at 10–20 mL/kg over 30–60 min 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 IV) may be given to an infant with severe asphyxia, has had recurrent hypoxia or has 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, IV or orally) or diphenylhydantoin (15 mg, IV or orally).

Specialist advice regarding management of specific conditions should be sought from the transport 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 CDH (Chapter 5), bowel obstruction (Chapter 7) or gastroschisis (Chapter 9). In oesophageal atresia, frequent aspiration of the blind upper oesophageal pouch, at 10–15 min intervals, is essential to minimise the risk of aspiration (Chapter 6).

KEY POINTS

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

Further reading

Pierro A, DeCoppi P, Eaton S (2012) Neonatal physiology and metabolic considerations. In: Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger RC, Caldamone AA (eds) *Pediatric Surgery*, 7th Edn. Elsevier Saunders, Philadelphia, pp. 89–108.

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CHAPTER 3
The Child in Hospital

CASE 1

Erin, aged 2 years, is seen in the surgical clinic because of an inguinal hernia. During the explanation prior to filling out the consent form, the surgeon describes the use of 'invisible stitches', a waterproof dressing and local anaesthetic.

Q 1.1 Will the operation be done under local anaesthetic?

Q 1.2 Why are 'invisible stitches' important?

Q 1.3 Why should the dressing be waterproof?

CASE 2

Jacob, aged 6 years, attends the surgical clinic very reluctantly because he is apprehensive about an upcoming epigastric hernia repair.

Q 2.1 What are his major fears likely to be?

Great effort should be made to minimise psychological disturbances in children undergoing surgery. The important factors to consider are the child's age and temperament, the site, nature and extent of the operation, the degree and duration of discomfort afterwards, and the time spent in hospital. Children between 1 and 3 years of age are the most vulnerable and do not like to be separated from their parents. For this reason a parent is encouraged to be with their child during induction of the anaesthetic and to be present in the recovery room as the child awakes from the anaesthetic.

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 parents to give support. Sometimes, elective operations may need to be deferred for stressful family events, such as the following:

- The arrival of a new baby
- A death in the family
- Shifting to a new house

Preparation for admission

Preparation for elective admission is important for children over 4 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. If the parents are calm the child too is usually calm, but if the parents are highly anxious, it is likely their child will be fearful and uncertain – and difficult to manage.

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 they will not wake during the operation and that it will be already over when they do wake up. They also will want to know when they will be able to go home, and whether they will be 'stiff' and a little 'sore' for a day or so. It is counter-productive 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

Jones' Clinical Paediatric Surgery, Seventh Edition. Edited by John M. Hutson, Michael O'Brien, Spencer W. Beasley, Warwick J. Teague and Sebastian K. King.

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. Many hospitals have 'play specialists' who are expert in addressing children's anxieties and provide distractions for those who are particularly anxious.

Effect of site of operation

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 can be of considerable value. Most inguinoscrotal operations (e.g. herniotomy or orchidopexy) are well tolerated and the use of local anaesthesia infiltration during surgery means that they have little discomfort afterwards. Many boys who have experienced both operations would prefer, in retrospect, bilateral orchidopexy to tonsillectomy.

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 about 80% of elective paediatric surgery.

The greatest advantage is minimising 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) and less nosocomial infection, alongside reduced burden on healthcare resources and budget.

Although operative 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), quick reversal of anaesthesia and long-acting local anaesthetic blocks or caudal analgesia in lieu of the usual post-operative injections of narcotics.

In the most vulnerable 1–3 year old 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 an understanding and empathetic approach by all staff lead to an informal and friendly atmosphere in hospital. The procedures for investigations or preparation for operation should be scrutinised carefully to see whether they are really necessary. Blood tests or x-rays are rarely required for elective day surgery.

Anaesthesia is an important source of fear and distress. The presence of a parent is very helpful during most anaesthetic inductions. Anaesthetic rooms often have large television screens or electronic games which act as a distraction during induction. 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 recovery room may reduce the child's stress as they wake from anaesthesia.

Even after major abdominal operations, some toddlers will be walking within 24 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. A play room is not required for most post-operative patients, since once they can walk to the toilet and play room, they may 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 admission or require frequent dressing changes (e.g. burns patients) and may significantly reduce the amount of analgesia required.

A single, absorbable subcuticular suture may be used to close almost all incisions, which avoids the anxiety and time spent in removing sutures. It also gives an excellent cosmetic result. A waterproof dressing allows normal washing and may 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 a neonate with a congenital abnormality, or inadequacy following removal of the neonate 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, and be involved in the day-to-day care, of her 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 may make recovery from even major operations a relatively short and simple matter. Most children are out of bed in 2–3 days and active for much of the day, or already at home by 5 days after many major operations.

Even with minor operations the child may have disturbed behaviour for several months after leaving hospital, and 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 an operation 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 operative procedures

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

1. The immediate group – conditions where immediate investigation and/or definitive operation is required, for example, torsion of the testis, intussusception, appendicitis.
2. The expedited 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 testes, 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 functional disorders, some of which may be life-threatening. 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 before the neonate is born, as in a congenital diaphragmatic hernia and gastroschisis (see Chapters 4–11); fortunately, most of these conditions are easily diagnosed on antenatal ultrasonography.

The expedited group

Inguinal herniae are prone to strangulation, especially in the first year of life. For this reason, herniotomy should be performed promptly: for those less than 1 year of age, this usually means the operation is performed in the coming days or weeks on the next semi-urgent or elective list (e.g. '6–2 rule': for a baby <6 weeks, herniotomy within 2 days; for infants 6 weeks to 6 months, herniotomy within 2 weeks; for children 6 months to 6 years, herniotomy within 2 months). 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. For many malignancies, several cycles of chemotherapy are given before definitive surgery is undertaken.

The elective group

Factors favouring deferment of operation

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

1 The possibility of spontaneous correction or cure. In infants, scrotal hydroceles, encysted hydroceles of the cord, true umbilical herniae and sternomastoid tumours all show a strong tendency to spontaneous resolution. An operation is only required for those few that persist well beyond the age of natural resolution.
2 Infantile haemangiomas (Strawberry naevi) 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 or treated medically. Operative intervention is rarely required and only in specific circumstances, such as a haemangioma which obstructs the visual axis, or has failed to respond to medical management.
3 The difficulties posed by delicate structures may 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 may be repaired more easily in a 6–12-month-old boy than shortly after birth.
4 The development of cooperation and comprehension with age. Voluntary exercises are important after some operations and it may be desirable to defer them until the necessary degree of cooperation is forthcoming.
5 The effects of growth are important in some instances. Chest wall deformities are corrected at adolescence, once 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 thus determine whether the treatment of non-urgent conditions should be deferred temporarily.

Factors favouring early operation

Factors which favour early operation 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 neonates with a developmental dysplasia 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 once was thought to be the primary cause of the dislocation.
2 Malleability of neonatal tissues is an advantage, for example, talipes, where the best results are obtained when treatment is commenced immediately after birth.
3 Avoidance of undesirable psychological effects. Often these may be prevented by completing treatment, including repetitive painful procedures, before the memory of things past 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 when children are anaesthetised.
- Invisible stitches, waterproof dressing and local anaesthetic given before waking mean the wound may be left alone post-operatively.
- Day surgery avoids separation anxiety in older children.

Further reading

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