Childs Nerv Syst (2004) 20:538–546
DOI 10.1007/s00381-004-0980-9

SPECIAL ANNUAL ISSUE

Jenny M. Thomas

Anaesthesia for conjoined twins

Received: 16 March 2004
Published online: 10 July 2004

J. M. Thomas (✉)
Department of Paediatric Anaesthesia,
Red Cross War
Memorial Children's Hospital,
Klipfontein Road, Rondebosch,
7700 Cape Town, South Africa
e-mail: jthomas@pawc.wcape.gov.za
Tel.: +27-21-6585003
Fax: +27-21-5314115

Abstract *Introduction:* Anaesthesia for conjoined twin surgery, whether prior to or for separation, is an enormous challenge to the paediatric anaesthesiologist. *Discussion:* The site and complexity of the conjunction will affect airway management, acquisition of vascular access, the extent of blood loss, and the number of surgical specialties involved. Preoperative assessment and planning, with interdisciplinary communication and cooperation, is vital to the success of the operations. These twins require a dedicated team of anaesthetists for each child, and, consequently, duplication of all monitoring and equipment in one operating room is necessary. Meticulous attention to detail, monitoring, and vigilance are mandatory. Planning for the postoperative period in the intensive care unit (ICU), as well as the babies' reconstruction and rehabilitation, is essential from the time of the initial admission.

Keywords Anaesthesia · Conjoined twins · Paediatric

Introduction

The management of conjoined twins is a multidisciplinary exercise involving many specialists and disciplines. The anaesthesiologist is an integral part of this team, and may be involved in the care of these babies from the time the initial investigations are performed, for the surgical procedures necessary before, at, and after separation, in the ICU, as well as during the period of reconstruction and rehabilitation. It is vital for the anaesthetists to establish a team approach within their own discipline. Advances in anaesthesiology and intensive care have contributed significantly to the successes worldwide of conjoined twin surgery, as have prenatal diagnosis and accurate anatomical delineation—made possible by ultrasonography, echocardiography, and foetal magnetic resonance imaging (MRI). With the recent successes with ex utero intrapartum treatment (the EXIT procedure) at the Children's Hospital of Philadelphia, foetal surgery may present a realistic option [2, 8].

Neonatal resuscitation

In developing countries where prenatal investigations may be limited, the delivery of conjoined twins may come as much of a surprise to the mother as to the attending midwife or medical practitioner. In areas where antenatal diagnosis is advanced, many aspects of the delivery and subsequent management will have been discussed, and all options presented to the parents, including the option of termination of the pregnancy.

At delivery it is optimal to have two sets of all neonatal resuscitation equipment, a paediatrician for each baby, and a large enough surface to accommodate both babies. A conventional open incubator is usually appropriate. If one twin is doing less well, it should be attended to first. In thoracopagus twins, over-vigorous bag ventilation of one baby may compromise the other twin significantly. Intubation is performed on one baby at a time, while the second twin is being supported by gentle mask ventilation.

Cardiopulmonary resuscitation (CPR) in conjoined twins may be very difficult and fraught with hazards. As one might expect, thoracopagus twins are the most diffi-

cult. Cardiac massage is unreliable, as the anatomy is distorted, and access to the heart is limited. Damage may be done to other organs, in particular the upper gastrointestinal tract (GIT) and liver.

Anaesthesia for conjoined twins

Anaesthesia may be required for procedures prior to separation, as well as the enormous challenge presented by the separation of the twins. Surgery for separation may be *emergency* or, more commonly, *elective*.

Emergency separation may be necessary in a number of situations:

- When there is damage to the connecting bridge (omphalopagus)
- Where one twin threatens the life of the other (complex congenital heart disease, cardiomyopathy, sepsis [4])
- With deterioration of both twins due to haemodynamic and respiratory compromise, typically thoracopagus
- Where the condition of one of the twins is incompatible with life (anencephalic, stillborn) and the other has a good chance of survival [3, 4]

As thoracopagus twins often require early intubation and ventilation, this group has the greatest need for separation in the neonatal period. At this stage it is necessary to realistically assess the viability of the infants. Early separation may also be considered for twins with a simple conjunction, or for a heteropagus (non-viable parasite), where no advantage is gained by waiting.

Anaesthesia prior to separation

Prior to separation, anaesthesia or sedation may be required for a variety of surgical or investigative procedures. It is advisable that, as soon as these babies are admitted to the hospital, a senior anaesthesiologist should assess them and inform other colleagues of his or her findings. This will facilitate a plan of action for emergency intubation and subsequent management, should this be necessary at a later stage. Ideally, deterioration should be anticipated, so that intubation can be performed in a controlled manner.

Diagnostic procedures that may require sedation or anaesthesia include cardiac catheterisation, endoscopy and some radiological studies—magnetic resonance imaging (MRI) and computer tomography (CT) scans in particular. The choice of agents and techniques to be used depends on the age of the twins, their airway anatomy, the duration of the procedure, whether or not it will be painful and the extent of immobility required. It is often a better option to administer general anaesthesia than to use deep sedation, especially for the thoracopagus group [13]. Chloral hydrate has been used very successfully for these babies, whether or not they are intubated. It is vital that the anaesthesiologists are aware of and present during these investigations, and are not only called in an emergency after inexperienced staff may have administered inappropriate medication. In every instance where an anaesthetic is administered to these babies, two teams should be available—one for each child.

Prior to formal separation, certain *therapeutic procedures* may need to be undertaken. These include amputations for inappropriately sited limbs, and diverting stomas for intestinal or urological obstruction—particularly in ischiopagus conjunctions. These infants are also subject to other frequently encountered infant or neonatal emergencies, and necrotising enterocolitis [7], pyloric stenosis and adenoidectomy for upper airway obstruction have all been described [3, 4]. Tissue expanders may have to be inserted to ensure adequate skin for closure after separation, and these are usually inserted 6–8 weeks prior to separation. If tissue expansion is uncomplicated, skin closure is greatly facilitated. As pressure effects from the expanders are exerted both inwards and outwards, vigilance is needed to assess possible organ compression, skin compromise, or sepsis.

When planning the separation, these preseparation procedures may provide valuable insight into potential problems that could arise. Communication with the surgeons remains an essential component of successful management. The ICU staff needs to be involved early in decision-making, and should become familiar with the babies and their parents.

Factors predicting difficult anaesthesia

Type of conjunction

Thoracopagus

Thoracopagus conjunctions are associated with a significant risk of complications.

Difficult airway management. Thoracopagus twins usually lie face to face with an exaggerated lordosis. The proximity and hyperextension of the babies make access to each baby a challenge. Intubation has to be performed on one baby at a time. It is usually easier to intubate with the infants on their sides than to hold them one above the other. This also reduces the likelihood of haemodynamic compromise. Further hyperextension of the head and neck should be avoided. Visualisation of the vocal cords is considerably improved if gentle pressure is applied to the larynx in a cephalad direction (not directly posterior). It may be necessary to ask an assistant to hold the other baby's head out of the way to avoid damage from the

laryngoscope. Awake intubation, except in moribund patients, may be very difficult. Once it has been established that ventilation is possible on both babies simultaneously with a T-piece, sedation or anaesthesia should be provided. Ketamine has proved to be a very successful agent in this situation. Vagal responses may require prophylactic atropine or glycopyrrolate, which are also useful antisialogogues. Emergency intubation of either or both twins should be avoided. Ideally, their deterioration and the need for intervention should be anticipated, and procedures carried out in a quiet, controlled way. Firm securement of their endotracheal tubes is vital, as the infants' activities may result in inadvertent extubation with potentially catastrophic results. The use of laryngeal masks (LMA) in conjoined twins depends on the type of conjunction, access to the airway, the procedure being performed, and the size of LMA used. The use of a size 1 in thoracopagus babies with their heads facing each other is hazardous, whereas a bigger LMA in ischiopagus twins with their heads at opposite ends is much more successful. Fibreoptic intubation has been used successfully, and other special laryngoscopes may be of value, but our experience with these is limited.

Ventilatory compromise. A pendleluft effect is experienced between the two infants, with their hearts and lungs moving to and fro between them with any manoeuvre, which alters their intrathoracic pressures. Their diaphragms are often shared and continuous between the babies, further contributing to the unstable respiratory dynamics. This also predisposes to gastro-oesophageal reflux (GOR), and can increase the risk of pre- and postoperative aspiration.

Cardiovascular instability. Cardiovascular instability occurs due to this same effect. Cardiac failure may be seen in either or both twins, depending on the heart conjunction and cardiac pathology.

Central vascular access. Femoral arterial and venous access is a considerable challenge, as it is necessary to rotate the babies away from each other to access the groins, thereby altering the normal relations of the vessels. Internal jugular vein anatomy may be unusual, and subclavian vein access is very difficult. The use of Doppler ultrasound may facilitate the location of vessels.

Bleeding. Bleeding may be significant but rarely exceeds 1–2 times each baby's estimated blood volume (EBV) when their hearts are joined. With complex cardiac conjunction as well as conjoined livers, 3–4 times EBV loss can be anticipated.

Postoperative problems. Postoperative problems include sternal insufficiency and instability, diaphragmatic dysfunction, inadequate skin cover, GOR and the risk of aspiration.

Omphalopagus

These range from a very simple conjunction to extremely complex conjunction. Surgery may be very difficult with major blood loss if the livers are joined. Airway access may be difficult, but is not as hazardous as in thoracopagus twins. The same principles apply to this group as to the thoracopagus group.

Ischiopagus and pygopagus

Depending on how they are joined, airway management may be a problem. Generally, their heads are at opposite ends, and access to the head and neck is straightforward. Blood loss may be significant (>1–2 times EBV). Abdominal contents can move between the babies with valsalva manoeuvres, which may compromise the ventilation and diaphragmatic excursion of the other infant.

Monitoring urine output may be complicated by the sharing and cross-over of genitourinary organs and systems: for example, a ureter belonging to one baby may cross to the other baby's bladder. Knowing the results of all investigations done is essential.

Tethering of the cord, spinal dysraphism, cord fusion and syringomyelia, in association with anorectal and urogenital abnormalities, are frequently encountered in ischiopagus, pygopagus and dipagus twins [12]. This may preclude the use of spinal or epidural anaesthesia in these twins. Preoperative documentation of neurological status is essential.

Craniopagus

Because of the babies' proximity, airway management and access to each infant is a concern. Stabilisation of the endotracheal tube may be a problem, but many of the basic principles of paediatric neurosurgery apply. Much depends on the complexity of the conjunction.

Blood loss is significant if the venous sinuses are communicating, and it is usually this factor that limits the viability of separation in craniopagus twins [10].

The preoperative neurological status needs meticulous documentation, as the degree of neurological deficit after surgery depends on the extent of brain and meningeal conjunction, and the complexity of the procedure.

Central vascular access may be limited to the femoral route.

Size and age at surgery—
for separation or other operations

As with any paediatric anaesthesia, the smaller and younger the baby, and the more immature the organs, especially the liver and kidneys, the greater the challenges of every aspect of anaesthesia. This impacts both on anaesthetic management as well as ICU care. One of the benefits of neonatal tissue is the presence of maternal hormones, which improve tissue elasticity, thereby reducing the need for tissue expanders. Prematurity adds the potential problem of hyaline membrane disease to the ventilatory dilemmas that are already encountered in thoracopagus twins. Other complications of prematurity that impact on the babies' status include barotrauma, altered response to hypoxia, and the risks of intracranial bleeds. Many investigations are more meaningful when the babies are bigger and more mature. The optimal time for separation is between 4 and 11 months [11]. Later separation presents concerns about the twins' psychological states after separation, and, in the younger group, organ immaturity and accuracy of investigations are factors to consider. Technically for anaesthesia, bigger is easier. Operative survival is 50% in the neonatal period compared with 90% if surgery is performed after 4 months of age. This may reflect the severity of the problem, the nature of the conjunction, and/or the need for urgent separation, rather than the age of the babies per se [11].

Degree of cross-circulation

The extent of shared vasculature affects drug pharmacokinetics and pharmacodynamics, as well as fluid and blood administration. Significant cross-circulation may confuse and complicate investigations and monitoring. Communication between the two anaesthetic teams is vital. Cross-circulation may be quantified by studies using contrast media, drug administration, or radioisotope injection into one baby, and measurement of the uptake in the other baby [3, 9].

Skin cover

Tight closure may compromise many organs, in particular kidneys, lungs, heart and the skin itself. Tight abdominal closure may also predispose to GOR and pulmonary aspiration, especially where leg splinting requires hip flexion (as in ischiopagus or pygopagus). For these babies, naso-jejunal feeding in the immediate postoperative period may be considered.

Duration of surgery

These are often long operations, with all the attendant challenges.

Anaesthesia for the separation of conjoined twins

Preoperative assessment and planning

Classification of twins

Important factors include the site of conjunction, the anatomy of the organs involved, the complexity of conjunctions, any associated anomalies, and the degree of cross-circulation. All these will impact on appropriate planning for anaesthesia with regard to choice of anaesthetic and analgesic agents, induction techniques and intubation, monitoring placement, vascular access, and positioning of the twins.

Airway access and fixation

In thoracopagus and some cephalopagus twins, bag and mask ventilation is easier with a T-piece rather than with an Ambu-type bag, as there is more room between the babies. The other types are less of a problem. Prior to intubation, the method used to fix the endotracheal tube needs to be decided on, as inadvertent extubation may be catastrophic for one or both of the babies. Turning them, whether in the theatre or ICU, requires thought, planning, and enough pairs of hands to execute this uneventfully.

Intubation

Intubation should always be a planned rather than a rushed, emergency event. If intubation is anticipated outside the operating theatre, the anaesthetists should be available to perform the procedure(s). Two teams must be available, one to work on each baby. Plan to intubate the sicker of the two infants first. Muscle relaxation is not advised until the airway is appropriately secured in both infants, as administration of muscle relaxants to one baby may result in paralysis of the other one. Any drug administered to one may also affect the other baby. Ketamine is the author's preferred agent for induction outside the operating room, and, in theatre, an inhalational technique—with or without ketamine. The use of topical airway anaesthesia with lignocaine can be used to facilitate intubation. Appropriate tube fixation will vary depending on the type of conjunction and the accessibility to the patient during surgery. In conjunctions where the heads are distant from each other, there is usually no problem, but where the faces are close, strapping needs to be meticu-

lously placed to avoid injury such as skin trauma or eye damage, and yet provide stability and safety.

Induction

It is preferable to have vascular access prior to induction, but this may not always be possible. In these circumstances, inhalational induction with either sevoflurane or halothane is preferred. Even with cardiopagus twins, sevoflurane has been used successfully for induction, although ketamine is preferred if there is any possibility of cyanotic heart disease in either twin. Where there is no airway compromise, any conventional paediatric intravenous induction agent is suitable. In awake patients, topical local anaesthetic creams should be applied prior to venepuncture.

Vascular access

Venous access in these patients is often limited. In the preoperative period, it is advisable to inform ward staff to avoid puncturing blood vessels that may be the only one available intraoperatively. For example, ischiopagus twins will have their legs in the operative field for separation, so intravenous cannulae cannot be placed there intraoperatively. To spare the upper limbs and neck, the lower limb vessels should be used for taking blood or placing drips in the preoperative period. Careful consideration should be given to where lines are placed, which cannulae will be used, and how many lines are required. Perioperative Doppler ultrasound may be of value in localising appropriate vessels.

General health of the babies

All the usual preoperative parameters for paediatric anaesthesia need to be assessed and documented. Especially in thoracopagus twins, features of cardiovascular and respiratory decompensation should be sought. These include loss of appetite, failure to thrive, tachypnoea, tachycardia, or decreasing saturations. In general, the two babies are more or less the same size, so it is appropriate to take their combined weight and halve it to arrive at the weight of each infant. Unless there is a significant discrepancy in their sizes, drugs should be administered on a dose/kg basis per individual. To ascertain whether the babies will fit onto the operating table, their length and width need to be measured. This is particularly relevant in bigger, older twins. It is vital that the anaesthesiologists have a comprehensive understanding of the results of the investigations, and that these are all documented on the anaesthesiology chart. Preoperative blood investigations and cross-matching need to be individualised for each set of twins.

Positioning of the babies

Positioning of the babies for surgery must be defined and accommodated, in consultation with each of the surgical specialties, especially when the position needs to be changed. Lines, monitoring and diathermy must be sited accordingly. The order in which the surgical specialties operate must be decided, and it is useful to have a written "agenda" to follow. Simple diagrams of anticipated anatomy and planned surgery are essential. Identification of each infant using colour coding of equipment, monitoring devices, and limbs and heads is very useful at this time [5].

Drugs: pharmacokinetics and pharmacodynamics

Pharmacokinetics and pharmacodynamics may be inconsistent. The degree of cross-circulation is usually indicated by contrast and radioisotope studies [3, 9], but in practical terms, thoracopagus and craniopagus twins have a considerable degree of cross-circulation, and the others have a variable degree. Very few have no cross-circulation.

Temperature control

As for any paediatric anaesthetic, all facilities to reduce heat loss and provide warmth should be used. To maintain their heat, the babies must be transported appropriately covered. Start the anaesthetic in a warm theatre (28°C) and cool down once the babies are draped. Intraoperatively, use plastic drapes to prevent the babies from getting wet and cold. Warm all fluids and blood products. Warm air convection devices are very beneficial.

Blood loss

Anticipate and monitor the loss, and have blood products available. The blood bank and laboratories should be informed, and all point-of-care blood monitors should be checked. Blood loss may be massive in craniopagus or cardiopagus twins, in sets where livers are shared, or where significant bony fusion is to be separated (osteotomies).

Tissue expanders

These are usually inserted 6–8 weeks before separation. Check the sites for infection and pressure effects, which occur both inwards and outwards. Tissue expanders are a mixed blessing; they recruit skin to facilitate skin cover, but they also contribute to the complications of organ compression, sepsis, and the breakdown of skin. If they fail and are removed, the tissue often becomes fibrous.

Cardiopulmonary bypass

Make plans for this eventuality in thoracopagus twins because it requires a lot of discussion, organisation of equipment and additional space. If extracorporeal life support systems are envisaged, plan accordingly. Where massive blood loss is anticipated, as in craniopagus twins, cardiopulmonary bypass (CPB) may also be considered.

Regional and/or local anaesthesia

Epidural and caudal anaesthesia have been utilised for surgery and postoperative pain relief [6]. There are definite pros and cons for regional anaesthesia for the separation of conjoined twins, and each set needs to be assessed individually.

Circulatory collapse at separation

Initially, adrenal suppression was cited as a common cause of circulatory collapse at separation, leading to a recommendation for perioperative steroids [1, 5]. These are no longer advised for adrenal suppression, but may be used for specific surgical specialties such as neurosurgery. Other causes of circulatory collapse to be considered include vagal response (especially with pelvic bony separation of ischiopagus or pygopagus twins), unappreciated blood loss resulting in intravascular insufficiency, and undiagnosed cardiac abnormalities.

Gastrointestinal tract prophylaxis

In the event of prolonged surgery, sucralfate via a nasogastric tube, or intravenous H_2 receptor antagonists should be considered.

Preoperative preparation of the family

Preoperative preparation of the family by the anaesthetist and other health care professionals is vital. The role of the parents at induction should be clarified, and an empathetic, understanding relationship is necessary—especially if only one survivor is anticipated. Plan to communicate with the parents during the operation.

Traffic in theatre

This operation generates a considerable amount of both media and medical interest, so it is vital for the hospital in general, and the theatre in particular, to establish a policy to reduce theatre congestion to a minimum. An overhead camera attached to the theatre lights, with cabling to a distant venue, is very useful. This allows any interested parties to watch proceedings without causing congestion. New technological equipment adds to this problem.

Anaesthesia

Plan to treat each child as a separate individual.

Premedication needs to be decided on for each set of patients. Mild sedation may be considered in older twins. Options include chloral hydrate, trimeprazine or midazolam. Oral atropine may be considered if ketamine is to be used intraoperatively, but an antisialogogue at induction is preferable.

Preparation of theatre

Equipment and monitoring. Duplicate everything: anaesthetic machines, pulse oximetry, capnography, electrocardiography, invasive and non-invasive blood pressure monitors, central venous pressure monitors, oesophageal stethoscopes, temperature probes, catheters for urine output, fluid and blood lines. Other monitoring includes arterial blood gas analysis, chemistry, haematology and the clotting profile.

Choice of endotracheal tube. To avoid intraoperative kinking, reinforced endotracheal tubes should be considered in surgery for separation of craniopagus babies. Otherwise, principles of management apply as for all paediatric anaesthesia.

Drugs.

- Routine: analgesics, anaesthetics, muscle relaxants, antibiotics, inotropes, plus the special requirements of any surgical specialty
- Emergency: epinephrine, phenylephrine, sodium bicarbonate, atropine, calcium, perioperative steroids—not for separation per se, but for any other reason (neurosurgery)

Fluids. Maintenance at 4 ml/kg/h, plus colloids as necessary for third space losses at 10–15 ml/kg/h. Blood products are administered as required.

Layout in theatre. Accommodate the two anaesthetic machines at the operating table depending on where the babies' heads will be. The heads of thoracopagus babies are both at the same end, whereas ischiopagus twins may have their heads at opposite ends. It is often easier to bring another operating table into theatre once the babies have been separated than to have it there from the beginning. Plans must be in place to move the anaesthetic

machines and diathermy away from each other once separation has occurred.

Peripheral monitoring and equipment. Blood gas machine, activated clotting time (ACT), haemoglobin, glucose, blood warming devices, and warm air convection devices should all be checked and ready.

Protection for babies and prevention of heat loss. Consider using gamgee (cottonwool and gauze), velband, plastic wrapping and plastic bags to wrap bowel contents.

Plan of action. Colour code everything to match the babies, including the anaesthetic, surgical and nursing teams. This contributes to a great team spirit. Decide on how to change positions with each surgical specialty, how to place operating tables once separation has taken place, and where to place the diathermy pads—only one being necessary until separation.

Anaesthesiologists. Two teams with one co-coordinator works well. Each team should have one paediatric anaesthetic consultant. Experienced paediatric anaesthetic nursing assistance is very valuable. For simple conjunctions, one paediatric anaesthetist per twin, and a co-ordinator, is sufficient.

Intraoperative management

Induction. The choice depends on the airway, type of conjunction, vascular accessibility, and includes any conventional induction agent used in children. Rapid sequence induction is not advisable in any twins where airway access is questionable. Ketamine has been used successfully in numerous sets, especially in thoracopagus twins. If nitrous oxide is used for induction, consider discontinuing it for twins where bowel surgery is envisaged, or where there is reduced intracranial compliance, raised intracranial pressure, or poor cardiac reserve.

Monitoring and maintenance. Placement of invasive monitoring lines may be performed in the days prior to surgery, with the risk that these may be lost before operation, or increase the risk of nosocomial infections. While induction is taking place and while lines are being placed, warming the operating room to 28°C ensures normothermia. The problems with cannulation have been mentioned. Except in the simplest conjunctions, invasive monitoring is recommended. Arterial and central venous cannulation are performed in the most appropriate vessels for the type of conjunction. No lines should be in the surgical field.

Anaesthesia maintenance and analgesia may be achieved in a variety of ways—halothane, isoflurane, sevoflurane, fentanyl, ketamine infusion, midazolam, regional anaesthesia, or morphine. Muscle relaxation can be provided by any agents commonly used in paediatric anaesthesia: pancuronium, vecuronium, or atracurium. As these are generally long procedures, pancuronium is preferable, in addition to which it has desirable sympathomimetic activity. Prophylactic antibiotics are administered at the request of the surgeons of each specialty. Aprotinin may be considered, especially with liver conjunction, massive blood transfusion, or intraoperative CPB.

Monitoring of chemistry and haematology should be planned and, depending on the need, blood samples taken every 1 or 2 h. Abnormalities should be corrected as they occur, especially when blood products are being administered.

Inotropic agents are advised for thoracopagus twins in whom cardiac function is poor. Dopamine should be considered from the onset, and phenylephrine may be beneficial where one infant has cyanotic heart disease and vasoconstriction is required. Twin-to-twin transfusion may occur if vasoconstrictors are used in one twin and not in the other.

Position change. This comprises stabilising the airway, the use of many hands, gentle handling, appropriate support and protection of limbs, freeing of diaphragms to optimise ventilation, and re-establishing lines and colour coding for each baby. Avoid holding one twin above the other for any length of time. Once the babies have been separated and moved to individual tables, sterility needs to be maintained.

Recovery. For simple conjunctions, the babies may not require postoperative ventilation. However, careful consideration should be given to factors that may be altered by the surgery, placing even these babies at risk of postoperative respiratory or cardiovascular complications. Emphasis should be placed on the need for adequate chest wall cover (both soft and bony tissue), proper diaphragmatic function, minimal abdominal distension, stable haemodynamics, and good pain management. If there is any doubt at the conclusion of surgery, the babies should be ventilated.

Postoperative considerations

Meticulous attention to fluid management, methods of ventilation, haemodynamics and low cardiac output states, and pain relief, are mandatory.

Early problems.

– Sequelae of massive blood transfusion and prolonged surgery

- Sequelae of tight closure: poor renal function with tight abdominal closure, respiratory compromise, diaphragmatic dysfunction, atelectasis and pneumonia
- Raised intra-abdominal pressure requires increased airway pressures, with all the attendant risks of ventilator-associated lung injury and decreased venous return
- Haemodynamic instability: hypotension, hypertension, arrhythmias, low cardiac output states, cardiac tamponade
- Hypothermia or hyperthermia

Later problems.

- Respiratory: compromised respiratory dynamics with diaphragmatic dysfunction, GOR, aspiration pneumonia, iatrogenic infections, incomplete sternal closure, phrenic nerve injury in chest surgery
- Poor cardiac function in thoracopagus survivors
- Inadequate skin cover
- Sepsis, especially staphylococcal. If the spleen has been removed or is missing, pneumovac should be administered
- Wound breakdown
- Nutrition requires special attention, as problems may develop with malabsorption, bowel resection, and GOR. Calories, proteins, essential fats, vitamins and trace elements may have to be supplemented
- Reconstruction and rehabilitation, for both function and cosmesis

Main causes of death. These include major congenital anomalies, especially conjoined hearts and complex congenital heart lesions; respiratory failure from poor respiratory mechanics, sternal insufficiency and aspiration; sepsis; and GOR with aspiration, pneumonia and death.

Pearls and pitfalls

- Experience helps—learn from others who have done these procedures before
- Comprehensive preoperative discussions are essential, with the results of all available investigations, all anaesthetic, medical and surgical role-players present, and ethical issues resolved, with the patients' and family's rights and wishes observed
- Early respiratory intervention should be considered for thoracopagus twins
- Rehearsals are valuable, especially in units separating twins for the first time. Meticulous attention to detail is required
- Regardless of the type of twins, airway management is a priority and requires thorough assessment, with contingency plans to cover every eventuality
- Long surgery: plan appropriately regarding fatigue, tedium, and refreshments. Despite extensive preoperative investigations, unexpected findings are common
- Monitor meticulously, from the preoperative assessment through to the ICU
- Mechanics of ventilation are considerably compromised if the anterior chest wall is not stabilised in a way that allows for normal, non-paradoxical breathing
- Have a modus operandi to deal with "visitors" entering the anaesthetic space in the operating room
- Media attention is inevitable, and each unit and hospital needs to have a protocol to attend to this
- Elect a designated person as liaison between theatre, the parents, media and other interested parties, for the length of the procedure
- A multidisciplinary approach with cooperation, communication and discussion is vital for success

Conclusion

Anaesthesia for conjoined twins may be a relatively straightforward paediatric anaesthetic, or one of the greatest challenges faced by an anaesthetist working with children. Much has been learned from other people's experiences and reports in the literature, but every case brings unique problems for consideration and solution. Multidisciplinary communication and co-operation remain the cornerstones of the management of conjoined twins.

References

1. Aird I (1954) The conjoined twins of Kano. BMJ 1:831–837
2. Bouchard S, Johnson MP, Flake AW, Howell LJ, Myers LB, Adzick NS, Crombleholme TM (2002) The EXIT procedure: experience and outcome in 31 cases. J Pediatr Surg 37:418–426
3. Cywes S, Davies MRQ, Rode H (1982) Conjoined twins—the Red Cross War Memorial Children's Hospital experience. S Afr J Surg 20:105–118
4. Cywes S, Millar AJW, Rode H, Brown RA (1997) Conjoined twins — the Cape Town experience. Pediatr Surg Int 12:234–248
5. Diaz JH, Furman EB (1987) Perioperative management of conjoined twins. Anaesthesiology 67:965–973
6. Greenberg M, Frankville DD, Hilfiker M, Frankville DD (2001) Separation of omphalopagus conjoined twins using combined caudal epidural-general anaesthesia. Can J Anaesth 48:478–482
7. Jaffray B, Russel SA, Bianchi A, Dickson AP (1999) Necrotising enterocolitis in omphalopagus conjoined twins. J Pediatr Surg 34:1304–1306

8. MacKenzie TC, Crombleholme TM, Johnson MP, Schnaufer L, Flake AW, Hedrick HL, Howell LJ, Adzick NS (2002) The natural history of prenatally diagnosed conjoined twins. J Pediatr Surg 37:303–309
9. Mann M, Coutts JP, Kaschula ROC, Fraser CB, Fisher R, Cywes S (1984) The use of radionucleotides in the investigation of conjoined twins. J Nucl Med 24:479–484
10. O'Connell JEA (1976) Craniopagus twins: surgical anatomy and embryology and their implications. J Neurol Neurosurg Psychiatry 39:1–22
11. O'Neill JA Jr, Holcomb CW, Schnaffer L, Templeton JM, Bishop HC, Ross AJ, Duckett JW, Norwood WI, Ziegler MM, Koop CE (1988) Surgical experience with thirteen conjoined twins. Ann Surg 208:299–312
12. Peter J C, Millar AJW, Rode H, Brown RA, Jee L, Cywes S (1996) Anorectal and urogenital malformations, conjoined twins and myelodysplasia. Childs Nerv Syst 12:356
13. Sury MRJ, Brown JL, Aitken K (1994) Anaesthesia for conjoined twins during magnetic resonance imaging. Eur J Anaesthesiol 11:139–142