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Anaesthetic Management of Conjoined Twins: Experience with Six Sets of Twins

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Summary

Anaesthesia for the separation of conjoined twins requires a well-prepared, multidisciplinary team. Each patient for surgery is different and the extent of organ sharing and coexisting anomalies must be determined before surgery so that problems can be anticipated.

We report our experience of the anaesthetic management for the separation of six sets of conjoined twins. Anaesthesia and surgery were prolonged, massive blood loss and transfusion, hypothermia, electrolyte imbalance and infection being the main perioperative problems encountered.

Key Words: Conjoined twins, Anaesthetic management, Paediatric anaesthesia

Introduction

Although conjoined twins are extremely rare, they have always attracted considerable interest and continue to fascinate both the medical profession and the general public. It has been estimated that the incidence of conjoined twins ranges from 1:50,000 births to 1:200,000 births[1-5]. However, an incidence of 1:14,000 to 1:25,000 has been observed in Southeast Asia and East Africa[6-9].

The aetiology of conjoined twins is unclear. It has been postulated that environmental and maternal factors are involved[8,9]. Formation of conjoined twins is said to be due to fusion, be it between embryos on one embryonic disc or on two[10]. There is very strong evidence to suggest that conjoined twins are uniovular in origin[1-3,6,7,11]. This theory has been supported by the observation that conjoined twins nearly always unite at nearly identical parts of their external surfaces and that the sex of conjoined twins is identical in almost every reported case[1,12,13].

Conjoined twins are classified by their most prominent site of connection with the use of a Greek word "pagos" meaning 'root'[11,13]. This may be at the anterior chest wall (thoracopagus), at the xiphoid (xiphopagus) or at the lower abdomen (omphalopagus). They may be joined back-to-back (pyopagus), end-to-end or side-to-side at the pelvis (ischiopagus) or head-to-head (craniopagus). Ischiopagus twins are further classified according to the number of lower extremities present; ischiopagus bipus (two extremities), ischiopagus tripus (three extremities) and ischiopagus tetrapus (four extremities). For example, omphaloischiopagus tetrapus twins are fused at the lower abdomen and pelvis and have four legs. Such patients present complex problems in surgical separation as they often share a liver, small and large intestines, genitourinary tracts, arterial and venous circulation below the diaphragm, spinal cord and pelvic bones.

Konig probably performed the first successful surgical separation of conjoined twins in 1689[13,14]. The operation was performed in infancy by means of a

ligature which was placed around the connecting band and drawn tighter and tighter until the structure was greatly reduced in size when it was separated with a knife. This technique is still in use today particularly in the staged separation of craniopagus twins[13].

In this paper we report our experience of the anaesthetic management for the separation of six sets of conjoined twins. The operations were performed between July 1988 and June 1995.

Materials and Methods

The Patients

Six sets of conjoined twins were operated between July 1988 and June 1995 in Hospital Kuala Lumpur. The demographic data of the patients are presented in Table I.

Preoperative Management

Before scheduling the twins for operation, they were each studied to define any organ sharing and coexisting congenital anomalies (Table I). This also allowed the infants time to grow.

The date of operation was decided following consultation between the paediatric surgeons, anaesthesiologists and paediatricians. For twins 2 and 4, the orthopaedic surgeons joined in the consultation. In addition, laboratory and blood bank personnel were informed early to accommodate the many laboratory tests and requests for blood and its components.

Table I
Demographic data of the twins

Twins	1	2	3	4	5	6
Sex	female	male	female	female	female	female
Race	Chinese	Malay	Chinese	Malay	Malay	Malay
Delivery	LSCS	SVD	SVD	LSCS	Forceps	LSCS
Birth weight	5.6 kg	4.0 kg	5.0 kg	3.9 kg	4.5 kg	5 kg
Type	Omphalopa-gus	Ischiopagus tetrapus	Omphalop-agus	Omphalo-ischiopagus dipus	Xipho-omphalopagus	Omphalopagus
Pre-operative abnormal findings	Livers fused with cross circulation.	Twin A – Dextrocardia & situs invertus, hemivertebrae, scoliosis, pubic bones widely separated. Twin B – Horseshoe kidneys, posterior phallus, no anus. Both – Common term. ileum and colon.	Livers fused with cross circulation.	Livers fused with cross circulation. Pelvic bones fused. Two lower extremities.	Each has a heart with single ventricle. Twin B has a very dilated pulmonary artery. Fused liver.	Fused liver
Age (mnth) Weight at operation	7/12 7 kg	6/12 6 kg	6.5/12 7kg	4/12 5.8kg	3.5/12 7.35 kg	4.5/12 8.5 kg

Table II
Summary of intraoperative and postoperative profile of the twins

Twins	1	2	3	4	5	6
Operation date	18.7.1988	5.3.1989	27.5.1989	9.3.1990	11.2.95	24.6.95
Duration of operation	10 hours	14 hours	7 hours	16 hours	8.5 hours	9 hours
Intra-operative findings	Livers fused	Common terminal ileum, colon, pubic symphysis and part of ischium, 2 undescended testes.	Livers fused by 10 by 5 cm bar. Common apical pericardium.	Livers fused. Common joint at pubic symphysis and part of ischium. Reconstruction required, each twin received one leg.	Liver fused. Shared pericardium	Liver fused.
Blood loss	1200 mls (2.0 x bld vol)	1200 mls (2.4 x bld vol)	700 mls (1.2 x bld vol)	1500 mls (3.0 x bld vol)	800 mls (1.28 x bld vol)	1000 mls (1.38 x bld vol)
Postoperative ventilation	6 days	72 hours	48 hours	Died 1/52 postop.	Twin A – 3 days Twin B – 3 months	Twin A – 1 day Twin B – 3 days
Condition at discharge from ICU	Both well	Both well	Both well	–	Twin A – discharged well Twin B – died 3 months postoperation	Both well

A detailed rehearsal of the separation procedure was carried out one to two days before the operation, so that each team including the operating room nurses and staff, knew exactly what its responsibilities were. There were up to five anaesthesiologists, four paediatric surgeons, two orthopaedic surgeons and many nurses with two sets of surgical and anaesthetic equipment, and even an audio-visual crew with their equipment, all in one operating theatre. No other person was allowed into the operating theatre to reduce traffic and noise as well as the risk of acquiring a surgical infection. To further reduce congestion, a weekend was chosen for all the twin separations[15].

Two sets of anaesthetic and monitoring equipment were prepared. All equipment, vascular lines, drugs, laboratory sample tubes and requisitions were named (A or B) and colour coded for easy differentiation.

The first four sets of twins were admitted to the intensive care unit one to two days before the operation to facilitate familiarity, pre-operative

assessment and investigation as well as to avoid undue publicity.

Syrup trimeprazine (4 mg/kg) was given as premedication one hour before the operation to each of the twins.

Intraoperative Management

Induction of anaesthesia was started after the electrocardiographs (ECG), pulse oximeters and noninvasive blood pressure monitors of both twins were set up. There were five anaesthesiologists, two for each twin and one to coordinate and relieve.

The smaller or less active twin was induced first, with oxygen and halothane, while the larger or more active received oxygen only. A peripheral drip was then set. Atracurium (0.5mg/kg) was the muscle relaxant of choice for intubation. If difficult intubation was suspected due to the awkward position of the twins, suxamethonium was chosen. The second twin might have to be lifted up or held away from the operating theatre table to allow for intubation.

Once paralyzed, the smaller or less active twin was intubated nasally. If cross circulation was present, the other twin might also lose its airway and ventilation, hence, careful observation and monitoring were required. This sequence also prevented the overdosing of the second twin.

After both twins were intubated, the internal jugular central venous line and arterial (preferably radial) line were inserted using aseptic techniques. An adequate number of peripheral lines were also inserted. Analgesia was provided with fentanyl or morphine. Atracurium (infusion or bolus doses) or pancuronium and a volatile agent (halothane or isoflurane) were used for maintenance of anaesthesia.

Oesophageal stethoscopes, nasopharyngeal temperature probes, warming mattresses, servo-heated humidifiers, fluid and blood warmers and cotton-wrappings were used in anticipation of prolonged surgery and massive blood loss.

Close monitoring of fluid balance, urinary output, acid-base balance, arterial blood gases, blood electrolytes and glucose, haematocrit and coagulation profile (including platelet, prothrombin time and activated partial thromboplastin time) were also done hourly to two-hourly or as necessary.

The basal perioperative fluid of 4 ml/kg/hr (for maintenance and correction of pre-operative fluid deficit) was provided for in the form of 4% dextrose in 0.18% saline. To compensate for electrolyte and protein losses from exposed areas, 10ml/kg/hr of Hartmans solution and fresh frozen plasma were used. Blood loss was assessed by weighing blood-soaked gauzes, direct measurement of suction loss and estimating spillages.

Volume replacement was given according to the central venous pressure, blood pressure, heart rate, urinary output, haematocrit and blood loss estimation. An assessment of the state of peripheral perfusion was helpful.

All the twins underwent prolonged operations (7-16 hours) and had massive blood loss and transfusion (1.2-3.0 times the blood volume) (Table II). Twin 2B had hypothermia (34°C) within the first four hours of the operation and had to be actively rewarmed.

Postoperative Management

All the twins were electively ventilated in the intensive care unit postoperatively. This was to stabilize them following the prolonged surgery, correct any problems related to massive blood transfusion (such as hypothermia, dilutional coagulopathy, electrolyte imbalance and acidosis), provide pain relief, assess adequacy of ventilation following abdominal wall closure and to observe for early signs of infection.

Twins 2A and 2B both had symptomatic electrolyte imbalance. Twin 2A had seizure and multifocal cardiac ectopics with a serum sodium of 117 mmol/l while twin 2B had paralytic ileus with a serum potassium of 2.8 mmol/l. Both recovered well after these were corrected.

Twins 4A and 4B died following a Gram negative septicaemia with disseminated intravascular coagulopathy one week after the operation.

Twin 5B who had a single ventricle, had difficulty being weaned off from the ventilator. She underwent a pulmonary artery banding performed by a cardiac surgeon on the 4th post-operative day. She suffered a hypoxic episode which resulted in hypoxic encephalopathy and was ventilator dependent. She subsequently died in the paediatric intensive care unit 3 months later. Her twin sister, who also had a single ventricle, was extubated on the third post-operative day, and discharged from paediatric intensive care unit on 9th post-operative day.

All the other twins were discharged well.

Discussion

The anaesthetic management of this rare condition, though stressful, is always a very challenging and worthwhile experience.

Perioperative management of conjoined twins requires a multidisciplinary approach and teamwork[13,14]. Without rehearsal and delineation of responsibilities, there will be chaos.

Unless an emergency situation arises which jeopardizes the well-being of either twin, the surgical separation should be unhurried and well-planned. The extent of organ sharing, the coexisting congenital anomalies[13,16], the medical status as well as the ethical issues must be taken into consideration. Operation is best delayed till infants are relatively mature as operative survival is said to be 50% in those operated on in the neonatal period, but 90% in those over 4 months of age[17].

The extent and type of organ sharing, and hence, the extent and type of surgery affects the outcome as well as the anaesthetic management. Our first set of twins (1A and 1B) had to be weaned off the ventilator more slowly than the others due to the tight abdominal wall closure. Our fourth set of twins (4A and 4B) underwent extensive surgery with limb reconstruction and subsequently succumbed to severe sepsis. Others have also described difficulty in weaning and infection as major perioperative problems[18]. Surgical advancement with better equipment and staged operations[14,19] can reduce operating time and blood loss and therefore, morbidity and mortality.

At induction, it is important to be able to maintain the airway in both infants as they may be awkwardly positioned. Furthermore, the induction agent and muscle relaxant given to one infant may cross circulate to the other[21]. As such, we induced the smaller or less active one first, to avoid overdosing the second infant. Other authors[14] have induced twins simultaneously, but we prefer to do it one at a time to reduce heightened activity in an already crowded room. Nasotracheal tubes are also preferred to orotracheal tubes as they provide better stability.

Setting up the peripheral, central venous and arterial lines for two awkwardly placed babies may take hours, but adequate, well-secured lines may be life savers. Some authors recommend establishment of airways and vascular access before transport to the operating theatre[22], but we, and others[13] believe that dislodgement of endotracheal tubes and invasive lines are more likely to occur during transport.

The anaesthetic technique should be so designed as to cope with prolonged surgery and anaesthesia and massive blood loss. Intraoperative anaesthetic management is aimed at providing analgesia, muscle relaxation, adequate ventilation, maintenance of haemodynamic circulatory function and temperature stability. In prolonged surgery with extensive exposure, however, it may be impossible to maintain the core temperature[14].

The actual fluid balance and blood loss of each twin was often difficult to ascertain although Toyoshima *et al*[21] have demonstrated that acute haemorrhage from one twin results in a rapid decrease of haemoglobin in both babies equally. We found that the internal jugular venous pressure reading was the most useful index of adequate replacement of fluid and blood clinically.

None of the twins were given steroids at induction or separation. Cardiovascular collapse occurring at surgical separation has been said to be due to relative adrenal insufficiency[12], but the use of steroids remains controversial, except in twins with demonstrated absence of adrenal tissue. Others believe that hypovolemia contributes to this collapse at the point of separation[14].

Constant vigilance and monitoring for haemorrhage, hypothermia, hypoglycaemia, hypoxia, hypercarbia, acidosis, electrolyte imbalance and coagulopathies are required and should be followed through into the post-operative period.

In conclusion, the perioperative management of conjoined twins undergoing surgical separation requires a well-prepared, multidisciplinary team. Anaesthesia for the separation of conjoined twins requires a lot of vigilance and quick action, and though stressful, it is challenging and is an unforgettable experience.

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