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The anaesthetic management of conjoined twins

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ABSTRACT

The management of anaesthesia for conjoined twins poses unique anatomical, physiological and logistic challenges. Although many possible configurations of union exist and each is unique, we describe the principles of anaesthesia for conjoined twins drawing on our institutional experience of managing 26 sets for a variety of procedures including separation.

Introduction

Conjoined twins pose unique challenges to the anaesthetist in that two physically and physiologically connected infants or children, often with imprecisely understood anatomy and physiology, must be safely anaesthetised firstly for a variety of investigations and procedures, on potentially numerous occasions and ultimately for lengthy and complex surgical separation.

In this article, we will describe the anaesthetic management of conjoined twins, using examples from our experience of 26 sets over a period of 30 years at Great Ormond Street Hospital in London (Table 1). Conjoined twins present infrequently with an incidence of around 1:200,000 live births; it is therefore very rare for an individual or an institution to be able to gain extensive experience of the management of these cases. Our experience has taught us that, although each set of conjoined twins is unique, a team approach comprising individuals who have previously successfully managed many such cases is invaluable in circumventing many of the problems and pitfalls in order to achieve the best possible outcome.

Presentation and antenatal diagnosis

Modern antenatal care has enabled the early diagnosis and assessment of conjoined twins. Prenatal ultrasound, echocardiography and foetal MRI allow for both diagnosis and a reasonably accurate assessment of the type of union. It is also possible to predict the likely pathophysiology of the conjunction, including shared organs and associated co-morbidities, to a significant but not comprehensive extent. As a result, the delivery and postnatal care of conjoined twins can frequently be planned in advance. Nevertheless, for a number of reasons, conjoined infants and children may also present at any age, with potentially significantly increased anatomical, physiological, social and ethical complexity.

Antenatally diagnosed conjoined twins are usually delivered by elective caesarean section and are initially nursed in NICU prior to transfer to a neonatal surgical unit for assessment. Unless circumstances do not permit, it is essential that this occurs in an institution that is experienced in the care of conjoined twins, as only a few surgical centres possess the necessary facilities to investigate or are able to acquire the skills for their management. Access to multiple teams is needed, including neonatal intensive care; radiology and imaging; paediatric medicine including cardiology, nephrology and surgical specialities; and of course, paediatric anaesthetists who are able to meet the complex anaesthetic requirements that the care for these twins will entail.

At our hospital, a multidisciplinary team (MDT) is established under the leadership of a senior paediatric surgeon, comprising two anaesthesia leads, one for each twin, and many other professionals, as anticipated from antenatal assessment. Further expertise is often recruited into the team, as problems are identified during postnatal assessment and investigation. If newborn twins are stable, gaining weight and thriving, it is usual to wait several months before planning separation. This allows the anatomy and physiology to be more clearly defined, and surgical planning is improved as they grow.

Emergency separation

Some conjoined twins do not survive at birth or die soon after. Although the degree of connection may make any attempt at separation so dangerous as to be unjustifiable, there are occasions when emergency separation soon after birth is indicated. The death of one twin or situations where prior to separation, if one

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Table 1
Presentation and outcome data of twins separated[a] at Great Ormond Street Hospital.

Type of union	Age at separation	Emergency (E) or planned (P)	Outcome
Thoraco-omphalopagus	10 weeks	P	Both alive
Omphalopagus	2 days	E	Both alive
Omphalopagus	1 day	E	1 alive/1 died
Thoraco-omphalopagus	7 weeks	P	Both Alive
Parapagus	1 day	E	Both died
Thoracopagus	3 days	E	1 died/1 alive
Ischiopagus	8 months	P	Both alive
Parapagus	3 years	P	1 died/1 alive
Parapagus	10 months	P	1 died/1 alive
Thoracopagus	3 months	P	Both alive
Omphalopagus	6 months	P	Both alive
Thoracopagus	8 days	E	Both died
Thoracopagus	1 day	E	Both died
Omphalopagus	1 day	E	Both died
Craniopagus	14 months	P	Both alive
Craniopagus	10 months	P	Both alive
Thoracopagus	7 days	E	1 died/1 alive
Thoracopagus	6 days	E	Both died
Thoraco-omphalopagus	1 year	P	Both alive
Omphalopagus	1 day	E	Both alive
Ischiopagus	11 weeks	P	Both alive
Thoraco-omphalopagus	4 months	P	Both alive

[a] Remaining sets non–operable.

twin dies the other will also die, may mean that emergency separation must take place. Many of the principles of management are the same for both elective and emergency separation, but obviously, with less time available, emergency separation carries considerably higher risks (Table 1).

Pre-separation assessment and management

Assessment for anaesthesia should take place as soon as possible after presentation. Conjoined twins should not be scheduled for imaging studies or procedures requiring anaesthesia or sedation until this has taken place and a suitable plan of anaesthesia is made. Obviously, in making such plans, much depends on the type and extent of union. It should be appreciated that anaesthesia for even the simplest procedure will be affected by various challenges, at least two anaesthetists/anaesthesiologists will always be required, and that it is very unlikely that locations out of the OR (operating theatre) will be suitably equipped or sufficiently spacious to accommodate two patients and two teams of professionals simultaneously and will therefore need to be adapted.

Fig. 1. Setup of the operating theatre for separation of conjoined twins.

Fig. 2. Orientation of anaesthetised thoracopagus twins prior to separation.

Any configuration that reduces access to the airway, or is likely to make rapid intubation difficult, poses a problem for both anaesthesia and sedation techniques, and so careful risk-assessment of all available management options must be undertaken. A large number of procedures under GA are likely to be required in order to make a full anatomical and physiological surgical assessment prior to separation. Table 2 shows investigations and procedures that are sometimes required on at least one occasion during surgical assessment. Anaesthesia for each investigation carries its own risks and complications and so should not be undertaken lightly; however, these anaesthetics do provide the opportunity to observe the physiological behaviour of the twins while under anaesthesia and provide some insight into how they might cope with the separation procedure at a later date.

Whatever the configuration of union is, for assessment purposes, it is assumed that each twin will behave as a physiologically independent individual and is therefore assessed separately. In fact, some cases, typically where there is extensive organ sharing and particularly in older twins, may show significant circulatory inter-dependence.

Airway assessment

Management of the airways of conjoined twins can pose a challenge to the paediatric anaesthetist from a number of standpoints: it may for example be complicated by the fact that the twins are facing each other, as in the case of thoracopagus twins, or they may have distorted neck anatomy, which can occur in thoracopagus or craniopagus twins. The likely presence of difficult airway and difficult intubation should be assessed. For twins undergoing investigations, such as MRI or CT scans, an anaesthetic is usually required. Supraglottic airway devices can be used in bigger children, but for small babies, tracheal intubation is required. With one set of craniopagus twins, who had increasing torticollis and signs of upper airway obstruction, both babies needed a nasopharyngeal airway and subsequently had tracheostomies well in advance of their surgery for separation. This was in part because in addition to their airway compromise, they were scheduled to have a series of three-staged cranioplasties, and the presence of the tracheostomies meant that they could be safely managed on the neurosurgical ward in between cranioplasties and following the separation.

Vascular access

In general, for minor procedures, peripheral lines alone are used so that the central veins and peripheral arteries are preserved for the subsequent major surgery for separation of the twins.