Case Report

Anaesthetic management of thoracopagus twins with complex cyanotic heart disease for cardiac assessment: special considerations related to ventilation and cross-circulation

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We describe the anaesthetic management of a pair of thoracopagus twins of 14 months of age undergoing complex cardiac evaluation. Synchronous ventilation of the twins, needed for the ECG-gated magnetic resonance imaging-angiography, was achieved through a Carlens (Y) adaptor during procedures and transport. The complex logistical implications are obvious. We also describe the first use of bispectral index monitor for detection of cross-circulation in conjoint twins.


Keywords: anaesthesia, paediatric; conjoint twins; monitoring, bispectral index

Accepted for publication: November 30, 2005

The incidence of conjoint twinning is rare, with an occurrence of 1 in 50 000–200 000 live births.¹ The most common type of conjoined twins is termed thoracopagus, in which the twins are joined at the thorax.² Although many are still-born,³ successful separation of thoracopagus twins, without sacrificing either of them, is largely dependent on the severity of cardiac involvement. To determine the feasibility of cardiac separation, it is essential that the intra-cardiac anatomy be defined accurately. Depending on the extent of thoracic communication between the twins, the usual transthoracic echocardiographic (TTE) windows may be limited, necessitating other diagnostic modalities including transoesophageal ECG (TOE), ECG-gated magnetic resonance imaging (MRI)-angiography (ECG-g-MRI-A), and cardiac catheterization with angiography. To perform these semi-invasive diagnostic studies in a safe and efficient manner adequate sedation and/or general anaesthesia is required. We describe a novel application of synchronous ventilation using a Carlens (Y) adaptor and the use of the bispectral index (BIS) for detection of cross-circulation in thoracopagus twins.

Case report

Fourteen-month-old (14.5 kg) thoracopagus conjoint twins were referred to our institution for cardiac evaluation and consideration for separation. TTE, TOE, ECG-g-MRI-A, and cardiac catheterization with angiography under general anaesthesia were performed. The twins were facing each other at an angle of 45°, conjoint at the chest, along the mid-sternal line, just below the clavicles to the upper abdomen. They were fairly nourished and although moderately cyanotic, were awake, alert and interactive. Twin A, the right-sided twin, had a hyperdynamic parasternal impulse, with a normal first heart sound and a single second heart sound. There was a systolic ejection murmur of grade 3/6 in both upper sternal borders, radiating widely to the entire precordium and posteriorly to both lung fields.

The pulses and perfusion of both twins were excellent, but there was marked cyanosis and clubbing of their nail beds. No hepato-splenomegaly was detected on physical examination. Before operation arterial pressure, heart rate, ventilatory frequency and room air oxygen saturation were 118/57 mm Hg, 138 beats min⁻¹, 60 bpm and 75% respectively in twin A and 103/69 mm Hg, 131 beats min⁻¹, 60 bpm, and 73% respectively in twin B.

All anaesthetic equipment were duplicated and i.v. accesses and drugs were colour labelled for easier identification. Standard monitoring (as per ASA guidelines) and BIS monitors were applied. Anaesthesia was induced in the operating room with sevoflurane in oxygen by facemask, first in twin...
A, while twin B was breathing oxygen. Two minutes later, twin A lost his eyelash reflex (BIS=45). Two minutes after loss of the eyelash reflex (BIS=45) in twin A, twin B became anaesthetized (BIS=47), and i.v. access was obtained in each twin without patient reaction. At this point, while end-tidal sevoflurane was 4.5% and the BIS values were 45 in twin A and 46 in twin B, propofol 7 mg was administered to twin A. Because of the fact that the babies were facing each other, twin B was lifted and tilted, so that twin A was optimally positioned for laryngoscopy. Intubation of the trachea was performed easily with an tracheal tube (ETT) size 4.5. The procedure was repeated for twin B. Muscle relaxation was achieved with rocuronium 7 mg to each twin. Because of the fact that the babies were facing each other, twin B was lifted and tilted, so that twin A was optimally positioned for laryngoscopy. Intubation of the trachea was performed easily with an tracheal tube (ETT) size 4.5. The procedure was repeated for twin B. Muscle relaxation was achieved with rocuronium 7 mg to each twin. The lungs in the twins were ventilated with two separate ventilators (Narkomed 6400, Dräger Medical, Inc., Telford, PA) with the same set up (tidal volume 70 ml, ventilatory frequency 28 bpm and preset pressure of 16 cm H₂O), but the end-tidal carbon dioxide values were different (4.8 kPa in twin A and 3.0 kPa in twin B). Considering the fact that the twins had a non-stimulating procedure and that their blood pressures were maintained at the lower range of the normal, the anaesthesia was maintained at a lighter level (BIS 65–70).

One hour after induction of anaesthesia, while twin A was fully relaxed, twin B showed signs of muscular activity; twin B required repeated doses of rocuronium of 3, 5, and 5 mg respectively over the next 15 min to achieve relaxation. A considerable cross-circulation from twin B to A was suspected. To confirm this, we administered 20 mg of propofol to twin B when the BIS value was 70 in both twins. Two minutes later the BIS value in twin A decreased to 45 while it remained 68–70 in twin B. After another 10 min the BIS in twin A returned to 70 (Fig. 1).

In order to provide synchronous ventilation in the MRI suite and to simplify ventilation during transports, a Carlens (Y) adaptor with Opti-Port™ right angle connector from a Broncho Cath™ set (Mallinckrodt Medical, Athlone, Ireland) was connected to the tracheal tubes through two straight Gas Sampling Connectors (Fig. 2). Despite the fact that only one ventilator was utilized (tidal volume 140 ml, ventilatory frequency 30 bpm, preset pressure 18 cm H₂O and FIO₂ 0.3), significant difference between the end-tidal carbon dioxide values was maintained (Fig. 3).

In the MRI suite, two MRI compatible monitors (Magnitude™, Invivo Monitoring System, Orlando, FL, USA) were utilized. However, only one ECG set was used as it was essential that MRI firing be consistently coupled to the ‘R’ waves of the ECG. The patients were ventilated with one Narkomed MRI-2 Anesthesia System (Dräger Medical Inc., Telford, PA, USA). The difference in the end-tidal carbon dioxide of the twins was still present. During the scan, repeated breath-holding periods (<50 s) were required but no arterial desaturation occurred.

After MRI and during cardiac catheterization (4 h procedure), one ventilator and two monitors were used. The end-tidal carbon dioxide values finally equalized to a mean of 4.8 kPa.

At the conclusion of the catheterization the twins were transported to the PICU and their tracheas were uneventfully extubated the following day.

The cardiac findings were as in the following: Twin A demonstrated levocardia, with tricuspid atresia and a rudimentary right ventricle, with double-outlet left ventricle, d-malposed great arteries and subpulmonary stenosis. Twin B demonstrated dextrocardia with atrial situs inversus, double-outlet right ventricle and pulmonary atresia. There was contiguity of the pulmonary venous atria of both twins, and contiguity of the dominant left-sided ventricle of twin A, to the right-sided ventricle of twin B, with these ventricles sharing a common AV valve apparatus (Fig. 4). This anatomy was consistent with Type C thoracopagus, wherein the twins share an atrium and a ventricle. This type is unfortunately not amenable to successful separation without sacrificing one or the other sibling. 4

Discussion

Some aspects of the anaesthetic considerations for cardiac MRI in paediatric patients and thoracopagus conjoint twins have recently been emphasized. 12
During ECG-g-MRI-A, morphological, qualitative and quantitative assessment of haemodynamic flow and volumes is possible. Intravascular and other advanced MR techniques, such as functional MRI, improve interventional and therapeutic management of children with congenital heart diseases (CHDs). 5

Odegard and colleagues 6 reported successful general anaesthesia with low incidence of complications in children with CHD, undergoing cardiac MR. Additionally, general anaesthesia had minimal adverse events when compared with sedation which has been associated with hypoxia in high-risk children scheduled for MRI and CT scan. 7 General anaesthesia may be preferable because of the long duration of these studies and the need for prolonged breath-holding periods during image acquisition. 6

In conjoint twins, synchronous ventilation is necessary to improve quality and decrease the time of the study. 8 We decided to use the Carlens (Y) adaptor to achieve synchronous ventilation. Interestingly, the twins had different values of end-tidal carbon dioxide during the first hour and during the MRI scan (4–6 h), which equalized towards the end of anaesthesia. We believe that one reason for this disparity could be the differences in the compliance and resistance of lungs between the twins that were further enhanced by different blood levels of rocuronium attributable to cross-circulation. Furthermore, twin B had pulmonary atresia, and received oxygenated blood from twin A, through the contiguity of their pulmonary venous atria and right and left ventricles, respectively. Only two very hypoplastic pulmonary veins were identified in twin B by Doppler interrogation. Although no imaging studies were done to evaluate for lung parenchymal hypoplasia, it is not unreasonable to speculate that this was present in twin B, given the much diminished pulmonary blood flow her lungs received. This would result in relatively more profound hypoxia in twin B, which may explain the marked differences in the end-tidal carbon dioxide of both twins, despite the fact that they were receiving similar ventilation patterns both initially, when two separate ventilators were utilized, and even later on, when the ventilation was synchronous.

Pharmacokinetics and pharmacodynamics are inconsistent in various types of twins. Usually there is more cross-circulation in the thoracopagus and craniopagus twins 9 than in other types, and therefore one can expect altered and unpredictable drug responses. 10 Estimation of circulatory mixing is useful to help calculate drug dosage and fluid replacement during surgery. 11 Drugs administered to one twin may have unexpected effects on the other,
These studies are time consuming and expensive. Recommended i.v. doses of anaesthetic agents for the combined body weight of the twins are usually halved and then divided into two equal doses to be administered to each twin. Reduced incremental doses are titrated against response and help minimize the dangers of compounding drug effects in one twin. Because the twins had contiguity at both the atrial and ventricular levels, this phenomenon was demonstrated during induction when inhaled sevoflurane to twin A affected both. This effect was not unexpected given the cardiac anatomy, as twin B had pulmonary atresia and was dependent on twin A for pulmonary circulation. Cross-circulation was also evident when twin B, but not twin A, required repeated doses of rocuronium to achieve muscle relaxation and when propofol administered to twin B resulted in a rapid decrease of the BIS score in twin A. It is highly probable that the effects of general anaesthesia enhanced inter-atrial shunting between their pulmonary venous atria, perhaps by decreasing the systemic vascular resistance in twin A more readily, so that the drug effects were present in twin B only when twin A presumably already had markedly higher blood levels. This illustrates the importance of determining the correct i.v. drug dosages by utilizing a combined body weight, which is usually halved and then divided into two equal doses that are then administered to each twin. Reduced incremental doses are titrated against response to help minimize the dangers of compounding drug effects in one twin. We used this technique in the anaesthetic management of our patients. Contrary to the bolus doses the fluid therapy or dose of continuously administered drugs that act by a constant plasma level, such as antibiotics, should be oriented at the total volume of distribution, that is, both twins.

Clamping and separation of vascular shunts and cross-circulation early during separation surgery prevent hypovolemic shock from ‘stealing’ of blood through the shared vessels. The blood pressure values of the twins can be different and arterial blood tends to shunt from the twin with higher blood pressure to the other with a lower pressure. This phenomenon may explain the enhanced effects of i.v. drugs in twin A, even when they were administered to twin B. Increasing the concentration of anaesthetics to lower the blood pressure in one twin can cause an overdose of anaesthetic in the other twin.

Isotope and contrast studies with technetium-99m sulphur colloid, radiolabelled albumin or red blood cells, and inhaled oxygen may be required to identify the extent and speed to which blood will cross from one twin to the other. These studies are time consuming and expensive.

Toyoshima and colleagues injected a bolus of indigo carmine in a pair of thoracoomphalopagus twins, and observed the appearance of the pigment in the urine of the other twin. They also provoked an intentional bleeding for 3 min in one of the twins; subsequently, the haemoglobin concentration of the two babies was the same. This demonstrated that acute haemorrhage from one twin resulted in a rapid and equal decrease in haemoglobin in both babies. This would also explain why both our patients were polycythemic, despite the fact that only twin B had pulmonary atresia, and twin A’s heart defect did not cause as profound cyanosis as twin B’s.

The BIS monitoring uses processed EEG that measures the hypnotic effects of anaesthetics and sedatives on the brain. Results from recent paediatric studies demonstrate that BIS provides useful clinical information. We used the BIS as a measure of evaluating the cross-circulation in our patients. During induction, while twin A was receiving sevoflurane by mask and twin B only oxygen, both became anaesthetized clinically (loss of eyelash reflex) and by the BIS score (45 and 47 respectively). Consequently, i.v. catheters were placed without patients’ reaction. The lack of effect of an appropriate dose of rocuronium (3 mg) raised the suspicion of cross-circulation, which was confirmed by a 63% reduction in the BIS score in twin A after administering propofol to twin B. A total of 13 mg (70% increase) of rocuronium was needed to paralyse twin B. The BIS monitor allowed for quick identification of cross-circulation. However, if surgery for separation is planned, careful angiographic or radioisotopic imaging of the cross-circulation is necessary for estimation of the cardiac output percentage which is exchanged, as one of the twins might be dependent on the other’s circulation for survival. It should also be recognized that the degree of cross-circulation is dynamic, highly dependent on both twins’ relative systemic vascular resistance.

In conclusion, we have described the anaesthetic management of a pair of thoracopagus twins for cardiac evaluation. Synchronous ventilation of the twins was achieved through a Carlens (Y) adaptor during both the procedures and transport. The complex logistical implications are obvious. We have also described the first use of BIS monitor for detection of cross-circulation in conjoint twins.

Acknowledgements
The authors would like to thank Aspect Medical Systems Inc., for their support and for providing the data recorder for the two BIS monitors, and Mrs Marcia Williams for kindly helping with the illustration.

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