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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.

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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 intracardiac 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-spleenomegaly 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. 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 H_2O), 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 H_2O and $F_{I_{O_2}}$ 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.

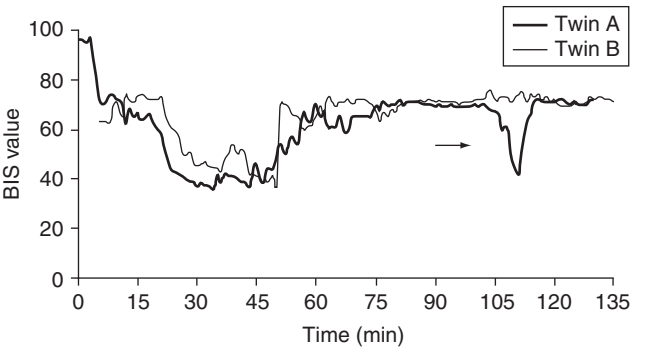

Fig. 1 BIS values during induction of anaesthesia and cardiac evaluation studies. The arrow indicates administration of propofol bolus (see text for details).

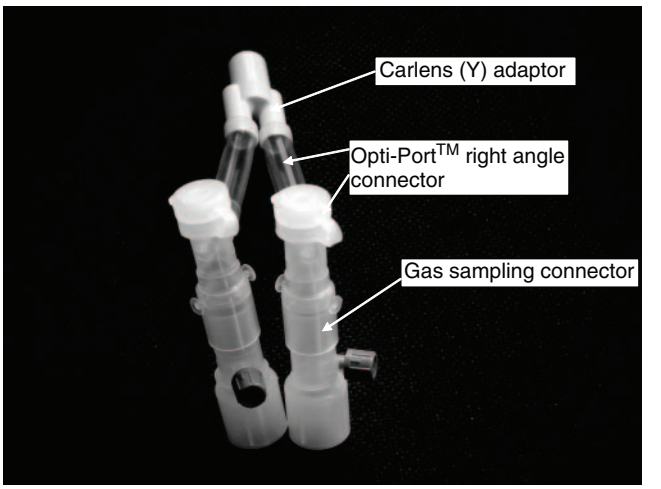

Fig. 2 Carlens (Y) adaptor with Opti-Port™ right angle connector used for ventilation.

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, dmalposed 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.[1 2]