Introduction

We describe the successful separation of thoraco-omphalopagus conjoined twins at 42 weeks post-conceptual age. Though separation of conjoined twins has been described, both separation and survival are not invariably achieved. The challenges encountered in this case included the presence of cardiac anomalies that prevented delay of the separation until the recommended optimal age and size. Most separation surgeries are performed when twins are 6 months or older. Most separation surgeries are performed when twins are 6 months or older.

Clinical Aspects

The twins were diagnosed prenatally and delivered successfully at 35 weeks by caesarean section and weighed 3.7 kg combined. They were evaluated for separation by multiple imaging modalities, including echocardiogram, CT and MRI to delineate shared anatomical structures and circulation. One twin was diagnosed with Tetralogy of Fallot (twin B) and the other with ectopia cordis (twin A). In preparation for future separation, tissue expanders were placed when the twins were 3 weeks old under general anesthesia. During the first general anesthetic twin B had hypercyanotic spells and postoperative echocardiogram demonstrated more pronounced subpulmonary muscle bundles. A tentative separation was planned at the age of 6 months to allow for growth, and plans were made for emergent separation in anticipation of deterioration in clinical status. Separation could not be delayed as twin B (with Tetralogy of Fallot) became more symptomatic despite beta-blockade and supplemental oxygen. At 7 weeks of age and a combined weight of 6.2 kg, separation was undertaken.

Anesthetic management challenges included airway management secondary to facial proximity, hypercyanotic spells and restrictive lung disease leading to challenges with ventilation. Airway management was not problematic during prior encounters, it is possible that the tissue expanders reduced the flexibility that existed during previous intubations. To overcome the hypercyanotic spells, maintenance of intravascular volume was a priority. An infusion of esmolol was continued in twin B during the case. Phenylephrine was also used for a limited period at the beginning of the case. Anesthesia was maintained with a fentanyl infusion, minimal isoflurane and neuromuscular blockade. In addition, an intensive care unit ventilator was utilized intraoperatively for twin B to optimize ventilation which was challenging in both twins, particularly after skin closure. Primary skin closure after separation was successful despite only having tissue expanders in place for 4 weeks. Both twins were extubated within 2 weeks following separation and remained stable. Twin B has since had cardiac surgery for Tetralogy of Fallot.

Discussion

In order to have successful separation, conscientious planning and anatomic evaluation are essential. Medical advancements have allowed for improved preparation. Optimizing the disadvantaged twin with pharmacologic interventions remains a challenge in the presence of cross-circulation. Variables such as prematurity and cardiac anomalies only contribute to the difficulty of surviving the first months before and after separation. In our case, temporizing measures such as modified Blalock-Taussig shunt placement in the twin with Tetralogy of Fallot were considered. However, this still posed a tremendous risk in the event of possible shunt thrombosis and unstable physiology. After multi-specialty discussions, the risk of attempting separation earlier than most prior separation surgeries was found to be the best option in this case.

References