

[NM-179] Anesthetic Management for Separation of Thoraco-omphalopagus Conjoined Twins Complicated by Tetralogy of Fallot

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Objective

To describe the successful separation of thoraco-omphalopagus conjoined twins at 42 weeks post-conceptual age. Though separation of conjoined twins has been described, separation and survival continue to be a challenge. 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 at least 6 months old or more.

Clinical Aspects

The thoraco-omphalopagus twins were diagnosed prenatally and delivered successfully at 35 weeks by caesarean section and weighed 3.7kg combined. They were evaluated for separation with perinatal echocardiogram, CT-MRI was utilized to evaluate 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 it was noted that twin B was symptomatic, developing right ventricular outflow obstruction. Repeat echocardiogram of twin B showed a more pronounced subpulmonary muscle. A tentative separation was planned at the age of 6 months to allow for growth. Additionally, plans were made in anticipation of possible deterioration in clinical status. Separation could not be delayed as the twin with Tetralogy of Fallot become more symptomatic requiring beta-blockade and supplemental oxygen. Each twin weighed 3.1 kg at the time of separation surgery. Anesthetic management challenges included airway management and ventilation. Primary skin closure after separation was successful despite only having tissue expanders in place for 4 weeks. Both twins were extubated a week following the separation and are stable. This continues to be a precarious period in their recovery course.

Discussion

In order to have successful separation, conscientious planning and anatomic evaluation are essential. Medical advancement has 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 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. After multi-specialty discussions the risk of attempting separation earlier than most prior separation surgeries was found to be the ideal option in this case.

References

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