

Perioperative management of two week old male mono-mono thoraco-omphalpagus dicephalus conjoined twins for repair of omphalocele and congenital diaphragmatic hernia

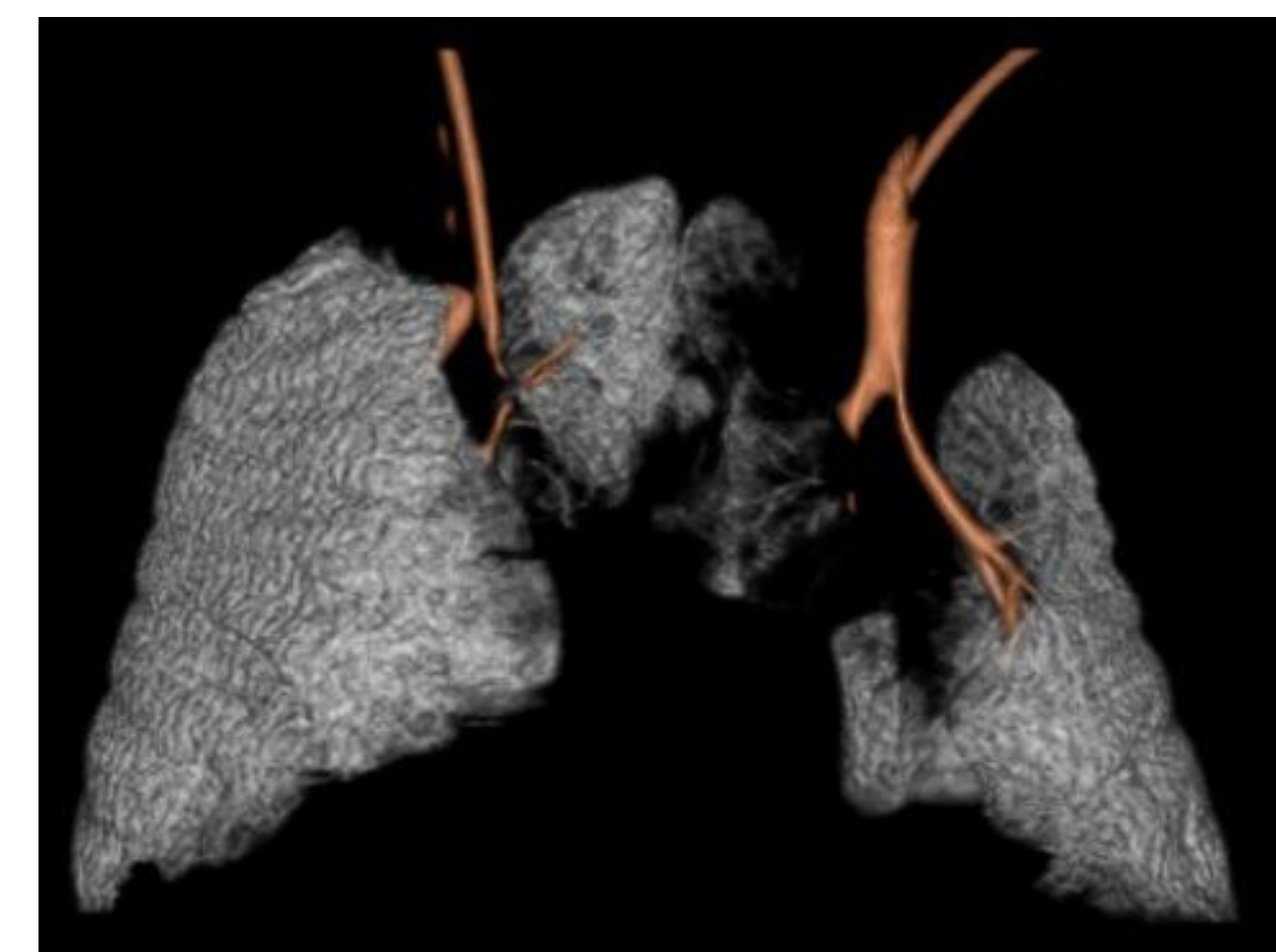


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Introduction

After a successful Cesarean-section, a 5 Kg pair of mono-mono thoraco-omphalpagus dicephalus conjoined twins were born at 36 2/7 weeks gestation. The twins share a descending aorta and IVC. Twin A's cardiac ECHO showed congenital heart disease with AV canal, double outlet right ventricle, and pulmonary stenosis with balanced circulation and a history of SVT, stable on propranolol at the time of the operation. Both twins have respiratory failure requiring intubation at birth, congenital diaphragmatic hernia with Twin A's stomach in medial portion of shared thorax, and omphalocele with obstruction. Twin B exhibited worsening pulmonary hypertension with increased oxygenation requirements in the 24 hours prior to the operation.

Note: all information and pictures are with written permission from the mother of the twins for educational purposes.



Preparation

One day prior to the planned procedure, all members of the multidisciplinary team including nursing, anesthesiology, and general surgery met in the designated operating room to discuss the plan for layout of the theatre, control of traffic, transport to operating room, blood loss, monitoring, positioning, color-coding for each twin (Twin A was green, Twin B was pink), and the action plan for potential complications. Given the nature of the patients, two anesthesiology teams were involved, with an attending and fellow comprising each anesthetic team.

The concern of balancing the respiratory requirements of each twin with significant circulatory mixing was also discussed in detail during preparation for the procedure. This required the use of two anesthesia ventilators with full monitor set ups. The patients do not have any shared components of the respiratory tract. See picture for MRI reconstruction of pulmonary systems. This necessitated the use of an operating room that had the capacity for the double set up; in this case a room that was designed to accommodate perfusion for cardiopulmonary bypass. The picture immediately adjacent shows the room set up with color coding in place.

Both patients were already intubated and collectively had a left saphenous PICC and right cephalic PICC. The decision was to get one peripheral IV on each upper extremity to allow medications and volume to be given directly into each patient's circulation to minimize mixing prior to reaching cerebral circulation. Additionally, a right upper extremity arterial line was planned to allow for continuous monitoring of qualitative cardiac output for Twin A.



Captions

Upper Left: The twins upon completion of the operation with color differentiation used to distinguish all monitors, lines, and endotracheal tubes

Lower Left: Two anesthesia ventilators and monitor set ups were required for this case; extensive color differentiation was used throughout

Upper Right: CTA reconstruction of the skeleton and circulation (AP view)

Lower Right: CTA reconstruction of the airways and lungs (AP view)

Case

On the morning of the surgery, all four members of the anesthesiology team transported the infants from the NICU to the operating room using Mapleson circuits attached to the endotracheal tubes following administration of fentanyl and vecuronium. The TPN, lipid, and midazolam infusions were continued throughout the perioperative encounter. Upon arrival, general anesthesia using a high narcotic technique was used for intra-operative management. A balance of crystalloids, 5% albumin, and packed red blood cells were used for fluid resuscitation. Defibrillator pads were placed over Twin A's thorax should he become unstable with SVT or other rhythm.

Two ventilators were used with significantly different settings, catered by each anesthetic team to the need of the respective twin being cared for, with much communication being required given different FiO₂ settings and significant mixing of circulations between the twins. Twin A required an FiO₂ of 21% throughout the procedure to allow for a balance between pulmonary and systemic circulations. Twin B required higher FiO₂, up to 60%, to compensate for pulmonary hypertension during the case. Repair of the diaphragmatic hernia with plication and omphalocele with placement of a percutaneous gastric tube in each twin's stomach proceeded without complications. Successful management of conjoined twins procedures relies on close communication, cooperation, and extensive preparation for the planned procedure.

Discussion

The incidence of conjoined twins is estimated to be approximately 1 in 50,000 pregnancies with only around 40% resulting in a live birth for a true incidence of around 1 in 200,000 live births¹. One of the most comprehensive discussions on the subject is that of Spencer in theoretical and analytical embryology of conjoined twins in which he defines 8 classifications for conjoined twins^{2,3}. This designates the most prominent site of conjunction along with the suffix -pagus. In this case this was the thorax and abdomen.

The most important consideration for conjoined twins is understanding the anatomy, especially shared circulation, in understanding if separation is possible⁴. In this case, with so much shared circulation and organs of the abdomen, separation is not possible. The possibility of an emergency separation during this procedure was discussed, but deemed not an option given the amount of time required for separation. In cases of conjoined twins that are not already intubated, the induction and airway management can pose one of the largest challenges, especially if the patients are different planes (facing each other for example) which complicates the ability to ventilate one while manipulating the airway of the other. Fortunately, this was not an issue for this case. Careful communication amongst all teams is critical during all cases of conjoined twins.

References

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