Successful surgical repair of d-transposition of the great arteries in a separated conjoined twin

Gang Chen, Bing Jia, Wen-Bo Zhang
Shanghai, China

Background: **Conjoined twins are very rare examples of congenital malformations. In 75% of thoraco-omphalopagus conjoined twins, the intracardiac anatomy determines outcome and long-term survival.**

Methods: **We successfully separated one case of thoraco-omphalopagus conjoined twins, with one having d-transposition of the great arteries. After control of sepsis, the twin underwent an arterial switch and complete repair of her cardiac defect.**

Results: **She made a full recovery and was discharged from the hospital 20 days after surgery.**

Conclusion: **Careful planning, skillful surgical separation and cardiac surgery by a combined medical and surgical team is the key to save the twins in such a rare case.**

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Key words: **conjoined twins;**
sugery;
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Introduction

Conjoined twins are very rare examples of congenital malformations. The worldwide incidence is 1 in 50-200 000 live births with a female/male ratio of 1:3.[1] Thoraco-omphalopagus, the most common type, accounts for approximately 70% of cases,[2,3] resulting in cardiac and hepatic sharing, the extent of which determines the outcome and long-term survival after surgical separation.[4] We successfully separated one case of thoraco-omphalopagus conjoined twins with one having a complex congenital cardiac defect: d-transposition of the great arteries (d-TGA). After surgical separation, the twin with d-TGA underwent a successful arterial switch repair at 25 days of life. We describe here the treatment of this extremely rare condition.

Case report

Female thoraco-omphalopagus conjoined twins (combined weight, 5.895 kg) were born at 36 weeks of gestation by elective caesarean section. They were fused from the xyphoid to the umbilical region (total length, 9 cm) (Fig. 1). They had normal and independent heads and extremities. Twin B was found cyanotic with systemic oxygen saturation (SO_2) of 80%–83%. Echocardiography showed twin A had a patent ductus arteriosus (PDA), and twin B had d-TGA, intact ventricular septum and PDA. The aorta and pulmonary arteries of twin B were in the anterior-posterior plane and the pulmonary artery was 20% larger than the aorta, while there was commissural alignment of the pulmonary and aortic valve. The atrial septal defect was measured at 8 mm and considered adequate. MRI and

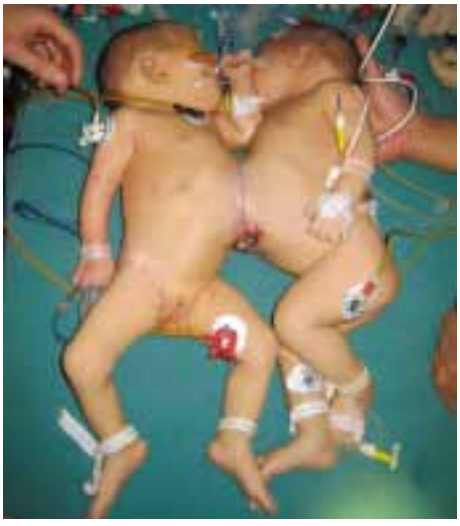

Fig. 1. Appearance of the thoraco-omphalopagus twins at age of 8 days.

Author Affiliations: Children's Heart Center, Children's Hospital of Fudan University, Shanghai 201102, China (Chen G, Jia B, Zhang WB)

Corresponding Author: Bing Jia, 399 Wanyuan Road, Shanghai 201102, China (Tel: +86-021-64931532; Fax: +86-021-64931517; Email: jiabing2012@hotmail.com)

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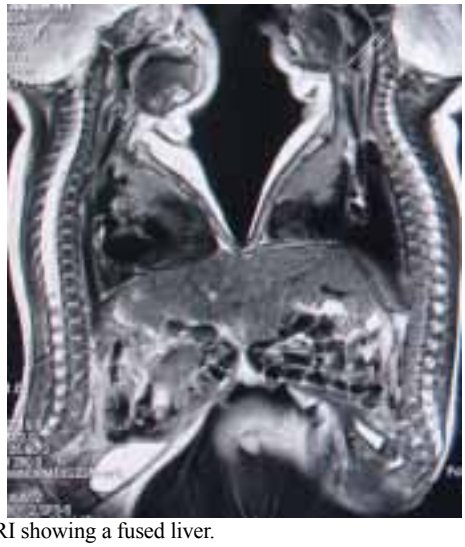

Fig. 2. MRI showing a fused liver.

CT of the chest and abdomen demonstrated a fused liver but a separate pancreaticobiliary system and gallbladder (Fig. 2). CT angiography confirmed the intracardiac anatomy in the twins.

To maintain the patency of the ductus arteriosus of twin B, prostaglandin E1 (PGE1) was administered via the intravenous route and titrated to keep SO_2 at 80%-85%. Twin A was on low-flow oxygen to stimulate the spontaneous closure of PDA.

On the eighth day of life, twin B developed fever, with abdominal distention, hematochezia and increased respiratory rate. A diagnosis of neonatal necrotizing enterocolitis (NEC) was made and confirmed by abdominal radiography. Urgent surgery including separation was considered as the only chance of survival for the conjoined twins.

After uneventful anesthesia of the twins, performed by two anesthetic teams, an incision was made from the xyphoid to the umbilical region. The liver was divided equally in the mid-section, and the twins were separated. The abdomen of twin A was closed by freeing the rectus abdominis muscle on both sides of the midline without tension. Twin B underwent an exploratory laparotomy, and thus an ascending perforation of the colon was found and treated with right hemicolectomy and ileostomy. Abdominal closure was difficult with tension, so retention stitches were used. The twin required a midline abdominal skin graft on day 10 due to an area of the incision failing to approximate.

After separation, twin A was returned to the Neonatal Intensive Care Unit. She was ventilated for 10 days and made an uncomplicated recovery. Twin B was transferred to the Cardiac Intensive Care Unit (CICU), ventilated and underwent systemic administration of antibiotics and PGE1 to maintain SO_2 at 80%-83%. She remained septic with fever. An abdominal drain was inserted on the second postoperative day to decompress the abdomen and was removed on day 10.

On the 25th day, the SO_2 of twin B dropped to 60%-63%. The infection was under control and her general status had improved markedly. We decided to undertake an arterial switch to correct the intracardiac defect. The baby was placed on cardiopulmonary bypass under deep hypothermia at 18°C and low-flow extracorporeal circulation. A complete repair was performed. The coronary anatomy was normal for d-TGA with the right and left ostia facing sinus and commissural alignment. Translocation of the coronary artery buttons was achieved without difficulty. The aortic re-anastomosis was completed without tension and the pulmonary artery was reconstructed with a pericardial patch. The cross-clamp time was 60 minutes. The baby was transferred to CICU with the sternum open, fully ventilated, and on low-dose inotropic support. The chest was closed on the second postoperative day. She was extubated on day 10 and discharged from the hospital 20 days after cardiac surgery.

Discussion

The indication and timing of separation of conjoined twins is directly related to thorough assessment of the shared anatomical area and vital organs. In many cases, separation is not possible because of shared organs, and over 75% of these babies will die in the first 3 months of life.[2,3] Even if surgical separation is deemed possible, early separation has been reported to carry a higher mortality.[1] Complete assessment of organ-sharing by CT and MRI, anatomical description of the liver and biliary tract and full cardiac assessment is essential for the planning of surgical separation.[5,6] Surgical separation is recommended at around 3 months of age, provided that enhanced nutrition, optimal physiology and preoperative planning by different medical teams are ensured.[7]

Twin B was found to be cyanotic and a diagnosis of d-TGA was made. Our initial plan was to proceed with separation at 10 days of life to allow for improved physiological stability and to perform intracardiac repair 10 days later. Unfortunately, twin B developed early NEC, which prompted urgent separation. The development of NEC in infants with congenital heart disease taking PGE1 is not rare.[8] The twins were followed up carefully, and NEC was diagnosed in time. The postoperative recovery of twin B was complicated by persistent abdominal distention and systemic signs of infection that necessitated abdominal drainage and prolonged use of antibiotics.

An arterial switch for d-TGA is recommended before 3 weeks of life (before left-ventricular (LV) wall thickness decreases). Following the experience of Zhang et al[9] in conjoined twins, we monitored the thickness of the wall and LV shape with echocardiography to ensure that the left ventricle remained volume- and pressure-loaded in preparation for the arterial switch. The atrial septal defect was measured 8 mm in diameter and we refrained from a balloon septostomy as it has been shown to help maintain the LV thickness. If the intra-abdominal infection had not been controlled by 3 weeks of age, we would have planned a staged procedure to prepare the left ventricle in the following weeks. Fortunately, complete cardiac repair was possible 15 days after separation.

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