

Successful Surgical Separation of Thoraco-Omphalopagus Symmetrical Conjoined Twins in Iran: Two Case Reports

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Received: 19 January 2019

Revised: 06 April 2019

Accepted: 28 April 2019

What's Known

- The principle of conjoined twin separation depends on physiological and anatomical factors. Each twin has specific abnormalities that should be fully identified before planning the surgery.
- A comprehensive preoperative investigation of the anatomy of each twin is necessary to predict reconstruction outcomes.

What's New

- The first successful separation of two pairs of thoraco-omphalopagus conjoined twins in Iran is described.
- It is important to identify critical steps of the surgical separation and make contingency plans for possible partial liver donation and cross-circulation between twins.

Abstract

Conjoined twins are derived from the division of a single fertilized ovum; a phenomenon accompanied with multiple congenital anomalies. Such twins are identical, of the same sex, and more likely to be female. Most twins die during the embryonic period, and only 18% survive longer than 24 hours. There are complex anomalies in thoraco-omphalopagus twins that makes them unlikely to live long enough to undergo separation. Treatment of this uncommon condition presents both surgical and anesthetic challenges. The management of rare anomalies is difficult even for skilled surgeons. Therefore, it is logical to use the knowledge gained from previous experiences.

We herein present the first successful surgical separation of two pairs of thoraco-omphalopagus conjoined twins at the Pediatric Surgery Center of Namazi Hospital (Shiraz, Iran). In both cases, the neonates had separate hearts and common pericardium. Contrast-enhanced computed tomography of two sets of twins showed fusion of sternum, pericardium, diaphragm, and left lobe of liver. Critical steps of the surgical separation were identified and contingency plans were made for possible partial liver donation and cross-circulation between twins. The separation procedure and reconstruction of the abdominal wall were successfully performed. Both pairs of twins, now 6- and 8-year-old, are healthy and have normal growth and are under follow-up.

Please cite this article as: Bahador A, Nikeghbalain S, Foroughi M, Beizavi Z, Sahmeddini MA, Eghbal MH, Vafaei H, Malekhosseini SA, Bahador M. Successful Surgical Separation of Thoraco-Omphalopagus Symmetrical Conjoined Twins in Iran: Two Case Reports. *Iran J Med Sci.* 2020;45(2):143-147. doi: 10.30476/ijms.2019.81060.

Keywords • Twins, conjoined • Congenital abnormalities • Reconstructive surgical procedures

Introduction

Conjoined twins, a congenital anomaly, develop due to the failure of anatomical separation of two identical fetuses; with an incidence of approximately 1 in 50,000 live births. These twins are identical, of the same sex, and more likely to be female. Most twins die during the embryonic period and only 18% survive longer than 24 hours.¹ Conjoined twins are classified according to the most prominent site of union, namely ventral union, dorsal union, and lateral union.² There are complex anomalies such as cardiac abnormalities in thoraco-omphalopagus twins that makes them unlikely to live long enough to undergo separation.¹ Although conjoined twins can survive without separation, surgical

separation is nowadays the main treatment, as it provides the opportunity for normal and long-term function.³ The first successful surgical separation of conjoined twins was performed by König in 1689. Since then, more than 100 cases have been reported with one or both twins surviving.⁴ Treatment of this uncommon condition presents both surgical and anesthetic challenges. Moreover, the management of rare anomalies is difficult even for skilled surgeons. It is therefore important to learn from the knowledge and experiences gained from previous surgeries.

We herein present the first successful surgical separation of two pairs of thoraco-omphalopagus conjoined twins. In both cases, the twins were male with common pericardium and liver. The separation procedure and reconstruction of the abdominal wall were successfully performed at the Pediatric Surgery Center of NemazeeNamazi Hospital affiliated to Shiraz University of Medical Sciences, Shiraz, Iran.

Case Presentation

Case I: First Pair of Twins

In March 2011, an ultrasound examination of a 26-year-old woman in week 13 of pregnancy showed symmetrical conjoined twins. She had no history of twins or congenital anomalies. The twins were male, oriented face to face, and attached together from the mid-sternum down to the umbilicus (figure 1). The mother was under the observation of a gynecologist and delivered the neonates via cesarean section. Apgar scores of the twins were 5 and 6 at 1 minute, 9 and 10 at 5 minutes after birth. Their total weight was 4,700 grams. Postnatal echocardiography revealed that the neonates had separate hearts and common pericardium. Color Doppler ultrasonography and contrast upper gastrointestinal (GI) examinations performed 4 days after birth showed a separate upper GI tract. Contrast-enhanced computed tomography (CT) performed 5 months after birth showed the fusion of sternum, pericardium, diaphragm, and the left lobes of the liver (figure 2). Surgical separation of the twins was performed when they were 5 month old.

Case II: Second Pair of Twins

In January 2013, an ultrasound examination of a 31-year-old woman in week 16 of pregnancy showed thoraco-omphalopagus symmetrical conjoined twins. Her pregnancy was uneventful. The twins were delivered via cesarean section, and their Apgar scores were 7 and 8 at 1 minute, 10 and 10 at 5 minutes after birth. Their total weight was 5,400 grams and the



Figure 1: The twins attached together from the mid-sternum down to the umbilicus.

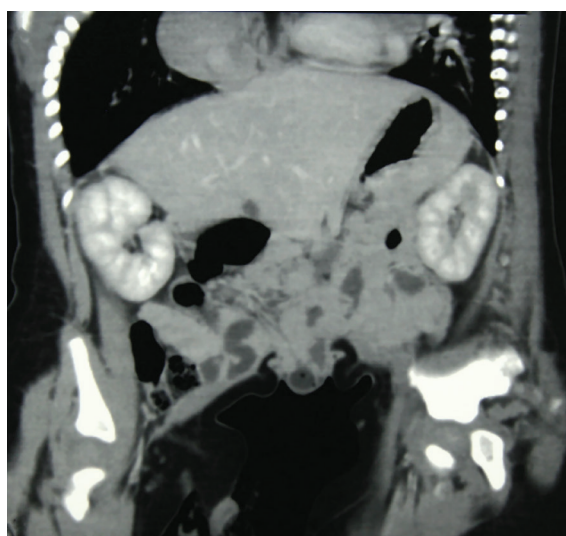


Figure 2: Abdominal computed tomography revealed common pericardium and shared a liver.

twins shared the bottom of the sternum down to the umbilicus (figures 3a and 3b). Postnatal echocardiography revealed that the neonates had separate hearts and common pericardium. Abdominal ultrasonography showed that the liver was shared over the full length of the lateral sector of the left lobes. The other organs were unremarkable. The contrast GI examination showed a separate upper intestinal tract. An enhanced contrast CT-scan performed 4 months after birth confirmed the ultrasonography and upper GI results. Surgical separation of the twins was performed when they were 4 month old.

Surgical Separation

Preoperative laboratory tests (complete blood count, blood gas, liver function test, and renal function test) of the two pairs of twins were performed and were all within the normal range. An experienced multidisciplinary team composed of a pediatric surgeon, gynecologist,

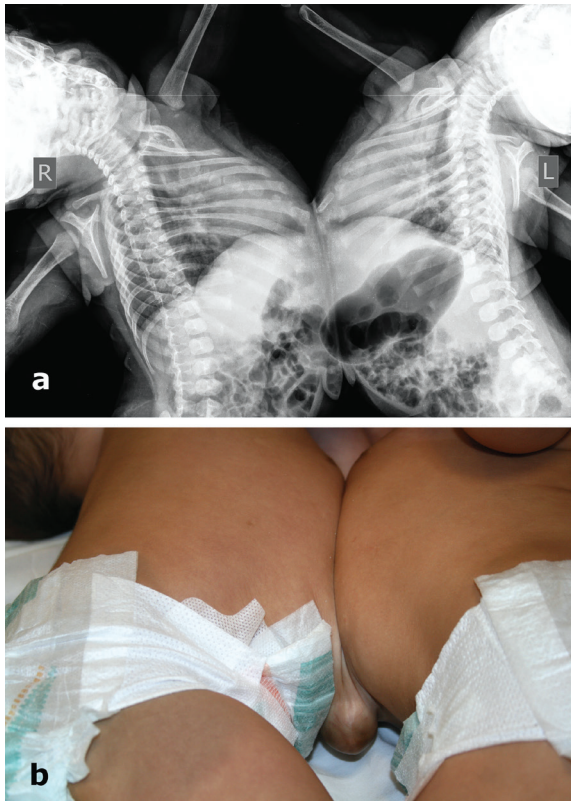


Figure 3: a) Radiography revealed thoraco-omphalopagus conjoined twins. b) The twins shared the bottom of the sternum down to the umbilicus.

pediatrician, and two anesthesiologists managed the separation procedure.

The twins were anesthetized and lateral position intubation in both twins was performed by two expert anesthesiologists under video laryngoscopy. After preparation, at first, a longitudinal incision was made over the abdominal segment of the bridge and extended down to the symphysis pubis. Exploration revealed that the GI tract was separate and only the left lobe of the liver was fused along

the entire length. After ligation of common teres ligament, an umbilical tape was passed under the left lobes while piercing the midway of the triangular ligament. Liver dissection using the cavitron ultrasonic surgical aspirator (CUSA), argon beam, and bipolar electrocautery was performed. There were no significant changes in the central venous pressure (CVP), blood pressure, and hemodynamics of the twins during liver separation. After separating the liver and following the second exploration, the incision was extended upward and the attached parts of the sternum, pericardium, and diaphragm were separated. The incision was gradually extended posteriorly and downward until the twins were separated. One of the twins was transferred to another operating room. The lower chest and abdominal wall of both twins were closed with a dual mesh. The entire operation time was approximately 210 minutes. The twins were transferred to a neonatal intensive care unit, where they were ventilated for 3 and 4 days, respectively. On day 4, enteral feeding was started and increased to the maximum tolerated level. After 1 week, the mesh was covered with skin. The twins were discharged after 10 and 13 days, respectively.

Separation of the second pair of twins was performed as described for the first pair. The entire operation time was 185 minutes. The neonates were ventilated for 4 days, after which enteral feeding commenced and increased to the full volume. Skin closure was performed 1 week after the initial surgery. The twins were discharged 10 days after separation.

Preoperative CT angiography evaluation of the shared liver vasculature was not performed. An intraoperative evaluation indicated that the liver vasculature and biliary systems were totally separated in both pairs and liver transplantation



Figure 4: Both pairs of twins are healthy and have normal growth.

was not required. Both pairs of twins were admitted 4 months later to remove the mesh for the reconstruction of the abdominal wall and umbilicus, and closure of the fascia and skin. This procedure was performed uneventfully. Both pairs of twins, now 6- and 8-year-old, are healthy and have normal growth and are under follow-up (figure 4). Written informed consent was obtained from the parents of both pairs of twins for the publication of the present case reports.

Discussion

Conjoined twins are regarded to be monozygotic, monochorionic twins of the same sex with an identical chromosomal pattern typically associated with specific anomalies. One of the most common types of conjoined twins is thoraco-omphalopagus, comprising 75% of such cases.⁵ Surgical separation of these twins has been always a challenge and may require novel techniques. We have described two pairs of thoraco-omphalopagus twins with common pericardium and liver. These were the first successful surgical separations of conjoined twins in Iran.

Thoraco-omphalopagus twins have a high incidence of cardiovascular anomalies with different types of cardiac fusion; 90% share a common pericardial sac.⁶ The severity of cardiac malformation seems to be the critical prognostic factor. Prenatal diagnosis permits a systematic approach to obstetric management and optimal fetal care. It also helps to plan urgent or elective separation. However, the outcome of urgent separations is poor with up to 50% mortality rate.⁷ If prenatal echocardiography shows cardiac fusion in the early period of gestation, it is reasonable to consider termination of the pregnancy. In our cases, prenatal diagnosis was made at 13 and 16 weeks of gestation and only revealed common pericardium in both pairs of twins.

In addition to in-utero evaluation, conjoined twins require early and detailed investigation of their cardiopulmonary, GI, genitourinary, and skeletal systems. Hence, ultrasonography, color Doppler, echocardiography, and contrast GI studies should be performed. These non-invasive investigations can define the roadmap to determine the possibility and the optimal time for separation.⁸

The timing of elective separation is a controversial issue. It seems that the first week after birth will determine the survival potential of each twin. During this period, there is no need for invasive surgery in stable twins. Due to the potential risks of invasive surgery (hospitalization, general anesthesia, and infection), the “watch

and wait” approach is more logical. O’Neil and colleagues reported a survival rate of 50% in twins separated during the neonatal period. However, the survival rate increased to 90% after the age of 4 months since the twins were older, more stable, and could better tolerate interventions.⁹ In our cases, we separated the twins at the respective ages of 4 and 5 months after birth, because they were hemodynamically stable. Since it is feasible at this age, partial liver transplantation was considered.

When the heart is not fused, major problems related to the alimentary tract, chest, and abdominal wall are to be expected. The most shared segments of the GI tract are duodenum and jejunum, which can be revealed by a contrast GI study. In the case of a fused liver, an angiography is rarely necessary, since a CT-scan and colored Doppler ultrasonography can reveal the anatomic details.¹⁰ Al-Rabeeh and colleagues proposed four classifications based on the extent to which the hepatobiliary system was shared; ranging from a totally separated biliary system to complex sharing of the entire extrahepatic biliary tree.¹¹ Fallon and Olutoye emphasized that preoperative determination of sufficient liver mass with adequate drainage for each twin was necessary, and intraoperative ultrasonography might be beneficial to localize structures.¹²

The surgical team should be ready for unexpected conditions. If liver separation is not possible, and there is a need for liver transplantation, mothers should be evaluated and prepared for partial liver donation in order to perform living-related liver transplantation. In our cases, the intraoperative evaluation revealed a fully separated biliary tree and vascular system. Hence, there was no need for liver transplantation. Despite potential cross-circulation between twins, we did not observe any significant changes in their CVP, blood pressure, and hemodynamics during liver separation. This was possibly due to different vessel sizes and the low degree of cross-circulation.

Skin closure is a challenge and should be carefully considered prior to the separation procedure. To cover the chest and abdominal wall, various methods (skin graft, local skin flap, tissue expander, free vascularized flap, and mesh) have been used. Tannuri and colleagues used mesh instead of a tissue expander, because data from the literature had shown complications due to the use of a tissue expander in as many as 57.2% of twins. They also stated that skin graft was unnecessary, because of spontaneous epithelialization that occurs following adequate nutritional support.¹

In the present study, we used mesh to cover the abdominal organs after separation. Skin closure was performed one week later, followed by an abdominal wall reconstruction 4 months after separation. Seemingly, the surgeon's preference and size of the defect are the most important factors in selecting the best method of reconstruction.

Conclusion

Separation of conjoined twins is one of the most challenging surgical operations. It requires extensive perioperative investigations and multidisciplinary collaborations for optimal separation and reconstructive outcomes.

Conflict of Interest: None declared.

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