

Technique Using a Titanium Plate to Reconstruct the Inferior Sternal Cleft in Pentalogy of Cantrell: Lessons From Four Patients

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Abstract

Background

By reviewing the outcomes of four patients, we summarize our experience with the strategy of using a titanium plate to reconstruct the inferior sternal cleft in pentalogy of Cantrell (POC).

Methods

This was a retrospective analysis of 4 patients who visited our department between January 2000 and June 2020 concurrent with POC. All four patients underwent an operation, as well as cardiac ultrasound and thoracoabdominal computed tomography (CT). Cardiac malformations achieved satisfactory correction according to echocardiographs. A titanium plate was used to repair the sternal and supraumbilical abdominal defects. The hospital course, operative data, and outpatient records were reviewed.

Results

All 4 patients had partial sternal clefts, and 4 patients underwent a single-stage operation. All 4 cases of ectopia cordis were eventually repositioned. The defect in the sternum and supraumbilical abdomen was repaired using a titanium plate. One patient with double-outlet right ventricle (DORV) developed low cardiac output syndrome and pulmonary infection, and symptomatic treatment was administered until discharge. The mean follow-up time ranged from 9 months to 10 years. No patient developed pectus excavatum, and there were no cases of retrosternal seroma or pneumothorax. The titanium plate was migratory in the second patient and was dislodged at another hospital 3.5 years postoperation, and a fibreboard was formed in the area where sternal cleft; the beating heart could not be observed outside the thoracoabdominal or thoracic wall. In the first patient, the titanium plate was torn with a small fissure at 2 years postoperation, but the fissure was not enlarged during follow-up. In the other two patients, the appearance of both the abdominal wall and lower sternum and cardiac function were good. The New York Heart Association function class was I in all four patients.

Conclusion

The use of a titanium plate to construct the neosternum can yield a satisfactory exterior appearance of the thorax with a partial sternal cleft, but long-term outcomes need to be examined further.

Introduction

Pentalogy of Cantrell (POC) is a rare disease that occurs in 5 per million live births and was first described in 1958 by Cantrell et al. Despite the rarity of POC, most reports of POC are case reports, and only approximately 185 cases have been reported around the world; the largest series of patients was 22, followed by a series of 8^{1,2}. POC is characterized by the presence of five major malformations: midline supraumbilical abdominal wall defect, lower sternal defect, diaphragmatic pericardial defect, anterior

diaphragmatic defect and various congenital abnormalities of the heart². Because of the multiformity of its clinical manifestations and severity of malformations, survival rates are as low as 37%³ and even 27%¹ according to some reports. Many surgeons are experientially deficient and easily misdiagnose this disease because of its extremely low incidence; to date, most reported cases in the literature have been sporadic. Repair of the inferior sternal cleft has been a hot topic and has attracted much attention, but there has been no consensus of opinions. The biggest challenge is the choice of an appropriate material to repair sternal clefts, and we previously reported on our method⁴. Here, we summarize our experience in successfully treating 4 patients with this complex congenital anomaly and describe the long-term outcomes of reconstruction of the inferior sternal cleft with a titanium plate⁴.

1. Patients And Methods

1.1. Patients

The Ethics Committee of the Children's Hospital of Chongqing Medical University approved the study protocol. The parents of the patients provided written informed consent.

From January 2000 and June 2020, we enrolled 4 patients in this study: 3 males and 1 females. All were full-term infants. Their weights on admission ranged from 3.4 kg to 7.5 kg, and their ages ranged from 3 months to 6 months. The first patient showed ventricular septal defect (VSD) and atrial septal defect (ASD); the second patient, who had patent ductus arteriosus (PDA) and ASD, also presented with VSD; the third and fourth patients presented with double-outlet right ventricle (DORV) (subaortic VSD) with pulmonary stenosis (PS); the fourth patient also showed pulmonary valve stenosis, left pulmonary artery stenosis, and persistent left superior vena cava, with collateral circulation; All the patients had partial sternal clefts, and ectopia cordis was incomplete with stable haemodynamics (additional patient characteristics are shown in Table 1). The respiratory and circulatory systems of the 4 patients were all stable. In all 4 patients, ectopia cordis was not obvious (incomplete ectopia cordis) and was covered with loose skin, and only the cardiac apex protruded outside the chest. In these patients, the beating heart could be observed outside the thoracoabdominal or thoracic wall.

Table 1
Anatomical Characteristics of the 6 Patients with Pentalogy of Cantrell

Patient (No.)	Sex (M/F)	Age at operation	Cardiac malformations	Other accompanying complications	Postoperative complications
1	F	4 months	ASD, VSD, PDA	None	The titanium plate chapped
2	M	3 months	ASD, VSD	None	The titanium plate migrated
3	M	6 months	DORV, PS, VSD	None	None
4	F	6 months	DORV, PS, VSD	None	Low cardiac output syndrome

DORV: double-outlet right ventricle; PS: pulmonary stenosis; VSD: ventricular septal defect; ASD: atrial septal defect; PDA: patent ductus arteriosus; COA: coarctation of aorta.

1.2. Methods

The incision was made through median sternotomy from the angulus sterni to the lower end of the abdominal defect. Then, the hypodermis was separated from the heart. Care was taken during this step not to injure the coronary arteries or diverticulum. The pericardium and diaphragm were explored first to ascertain the presence of a severe diaphragmatic hernia. The intracardiac anomaly was corrected with standard cardiopulmonary bypass. The heart was pushed back into the thoracic cavity, and the pericardium was closed with an expanded Gore-Tex membrane to protect the heart from titanium. One titanium plate (Autocam Medical, Kentwood, MI, USA) was anchored to the sternum and ribs using 3 – 0 PDS-II or medical silk interrupted sutures (Ethicon, Somerville, NJ, USA) to repair the sternum and chest wall defect⁴ monitoring for changes in the heart rate, blood pressure and rhythm.

2. Results

Sternal clefts can be classified into two major forms: partial and complete⁵. All 4 patients had partial sternal clefts; patients 1 and 2 had simple ASD and/or VSD, and both were treated with conventional cardiac surgery. Patients 3 and 4 had DORV (subaortic VSD), mild PS and a mild right ventricular outflow tract; these two patients were treated with DORV correction and dredging of the right ventricular outflow tract. In patient 4, reconstruction of the left pulmonary artery was performed. All four patients underwent a single-stage operation, and in all of these patients, the heart was replaced into the thoracic cavity without causing significant haemodynamic changes. A titanium plate was used to repair the defect in the sternal cleft and abdominal wall. In patient 4, low cardiac output syndrome occurred due to the complex cardiac malformation after medical treatment and discharge, but there were no serious postoperative complications. The follow-up period ranged from 9 months to 10 years. No patient developed pectus excavatum, and there were no cases of retrosternal seroma or pneumothorax. In all four patients, the appearance of both the abdominal wall and lower sternum and cardiac function were good. The New

York Heart Association function class was I in the patients who underwent an operation. Although the titanium plate was migratory in the second patient and was dislodged at another hospital 3.5 years postoperation, and a fibreboard was formed in the area of the sternal cleft, the beating heart could not be observed outside the thoracoabdominal or thoracic wall. In the first patient, the titanium plate was torn with a small fissure at 2 years postoperation (Figure A), but the fissure was not enlarged, and the exterior appearance of the thorax was normal. In the other two patients, the exterior appearance of the thorax was normal, and cardiac function was good.

3. Discussion

Pentalogy of Cantrell (POC) is a rare disease, and all of the patients exhibit chest wall malformations, a defect in the lower part of the sternum. It may result in severe paradoxical movement of mediastinal structures and respiration in infants, which can impair oxygenation and result in CO₂ retention, acidosis, cyanosis, and/or dyspnoea⁶. It can also result in respiratory infections and blunt or piercing trauma to the heart⁵. Cardiovascular function may be impaired because of the reduction in thoracic volume.

Sternal clefts are malformations caused by failure in the fusion of sternal elements. The first sternal cleft reported was in 1740⁷. The aetiology of sternal cleft deformity is unknown; however, studies on ventral body development in mice indicate impairment in Hoxb gene expression as a possible factor. Preoperative assessment should include an echocardiogram, chest radiograph, and computerized tomography (CT) scan with three-dimensional reconstruction illustrating the extent of the bony lesion.

The greatest challenge in repairing these anomalies is the construction of the neosternum; additionally, the sternal cleft may predispose patients to blunt or piercing trauma to the heart. Although many methods for constructing the neosternum have been proposed, none are ideal. The question is what should be used to repair the sternal cleft: autologous tissues or prosthetic materials? Some reports have suggested⁸ certain principles for reconstruction of inferior sternal clefts, which include the following: 1) rigid protection of the heart without compression, 2) preferable use of autologous tissue, 3) dynamic reconstruction of the thoracic cage, 4) uncompromised growth, and 5) minimal donor site morbidity.

Autologous tissues are ideal and the first choice, as accepted by most surgeons^{8,9}. This method has the advantages of reducing the risk of infection and allowing synchronous growth with the body. There have been several excellent reviews describing experience with this approach, such as those by Kim CW¹⁰ and Sabiston¹¹, who reported an excellent operative effect. However, these techniques are greatly limited by the size of the defect and corresponding autologous tissues that can be obtained, especially in neonates⁸. Additionally, these therapies all have the common disadvantages of increased surgical trauma and bleeding, and donor site morbidities such as retrosternal seroma and pneumothorax can occur; in addition, they are technically more difficult to perform. In these approaches, ribs and costal cartilage are the most common materials used as autologous tissues to construct the neosternum, but this can cause serious consequences. Research has shown^{5,9} that intraoperative complications occur at

a rate of 4% due to pericardial or pleural tears during sternal dissection and that postoperative complications, such as retrosternal seroma and pneumothorax, occur at a rate of 17%. Pectus excavatum is another postoperative complication that occurs in 25% (1/4) of patients.

Many prosthetic materials have been used for sternal cleft reconstruction, such as silicone elastomers and titanium plates^{4, 10, 12}. We reported on our previous experience in 2016⁴. In our case, we used a titanium plate to repair the sternal cleft and achieved a good appearance. First, a mould of the abdomen-thorax was made according to exact data from three-dimensional chest CT reconstruction. Second, the size of the titanium plate was demarcated on the mould as the defect of the sternum and supraumbilical abdomen. Then, we arched the titanium plate(Figure B) in order to provide more room in the thoracic cavity for the heart, which sometimes resulted in multistage operations because ectopia cordis needed to be avoided. In addition, with the use of a titanium plate, much damage was avoided, i.e., the ribs and costal cartilage remained intact, and the pectorals were not dissociated. Furthermore, this approach reduced the difficulty of repairing the thoracoabdominal wall by minimizing the tension of the sutures because the rectus abdominis could be sutured to the lower end of the titanium plate. The merits of titanium plates include stiffness, histocompatibility, resistance to infection and low interference with CT and magnetic resonance imaging^{13, 14}. Based on our experience, this method is suitable for patients in whom ectopia cordis is not severe and the heart can eventually be repositioned and for patients in whom the area of the sternal cleft is large but ectopia cordis is not severe. While this method is also controversial, the lack of synchronous growth with the body is problematic. The string will be broken, which might be an underlying reason for migration, which is also a serious postoperative complication. In some cases, as in ours, the titanium plate may even need to be removed. In addition, because of lacking synchronous growth, the titanium plate would be torn when fixed with steel wire. Other complications, such as psychological problems, are persistent drawbacks, although these deficiencies did not occur in our patients.

At the early stage of our long-term follow-up, no complications occurred, and a good appearance was achieved in all four patients(Figure C and D). We believe this approach is an excellent scheme that can be used to construct the neosternum in the patients with partial sternal clefts. However, with a prolonged follow-up time, complications such as migration and avulsion occurred, and we believe that some improvements should be made to perfect this approach. We analysed the process of this operation and concluded that the most important reason why migration and avulsion occurred was the method of how the titanium plate fixed, as we ignored the lack of synchronous growth with the body of the titanium plate. In the first and second cases, we sutured the titanium plates to the ribs in order to fix the titanium plate firmly, and we presume that with the growth of the rib, the titanium plates were torn or the sutures were broken, and then migration or avulsion occurred. In the rest of the cases, we fixed the titanium plate to the muscle directly, and no migration or avulsion occurred.

4. Conclusion

POC is a rare disease. Reconstruction of the sternal cleft is key for favourable long-term outcomes, but how to reconstruct the sternal cleft is controversial, and there is no ideal method. We reported our experience with the treatment of sternal cleft reconstruction by using of a titanium plate, which yielded a satisfactory exterior appearance of the thorax, but long-term outcomes need to be examined further.

The List Of Abbreviations

POC: pentalogy of Cantrell; CT: computed tomography ; DORV: double-outlet right ventricle; PS: pulmonary stenosis; VSD: ventricular septal defect; ASD: atrial septal defect; PDA: patent ductus arteriosus; COA: coarctation of aorta.

Declarations

Ethics approval and consent to participate: This research received ethical approval and consent. It did not require the approval of an ethics committee.

Consent for publication: All patient information contained in this manuscript has been published with the consent of their parents.

Availability of data and materials: The data and materials in the manuscript are available, and the original data for the relevant results are owned by myself; I can be contacted if needed.

Competing interests: There are no conflicts of interest or legal economic disputes in this study.

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Authors' contributions: Linyun Xi, the first author, designed the work, acquired, analyzed, and interpreted the data, drafted the work and substantively revised it. Chun Wu, the second author, substantively revised it. Zhengxia Pan, the third author, provided some suggestions for this work. Ming Xiang, the corresponding author, designed the work, drafted the work, substantively revised it, and approved the submitted version.

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Figures

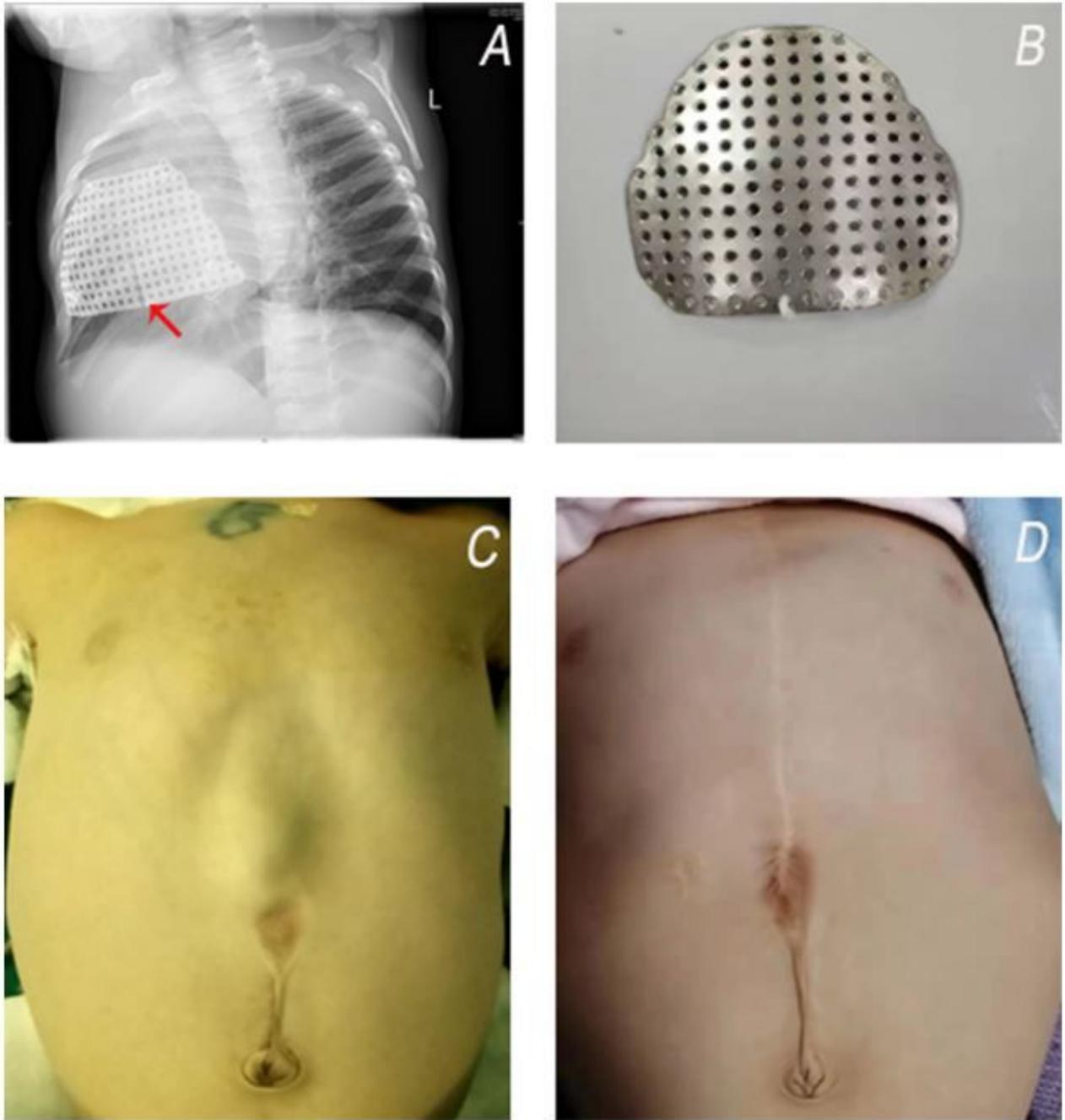


Figure 1

A: The titanium plate was torn with a small fissure (red, filled, arrow); B: a titanium plate was tailored; C: Preoperative view; D: Postoperative view.