Fontan operation for the Cantrell syndrome using a clamshell incision

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Abstract

A median sternotomy could be difficult for a child with ectopia cordis and complex congenital cardiac anomalies. We report a patient with ectopia cordis, functionally single ventricle and bilateral superior vena cava, who underwent a staged Fontan procedure through a clamshell incision and the sternothoracotomy approach.

Keywords: Ectopia cordis • Cantrell syndrome • Functional single ventricle • Fontan operation • Bilateral superior vena cava

INTRODUCTION

Ectopia cordis is a rare congenital malformation. Previous reports indicate a poor prognosis for patients with ectopia cordis and congenital heart diseases. Although the more common thoracoabdominal ectopia cordis offers a better prognosis, there are a few reports of Fontan completion in ectopia cordis patients [1, 2]. The difficulties associated with the surgical approach to ectopia cordis patients are the factors inhibiting Fontan completion. We report an experience with a staged Fontan procedure in a patient with thoracoabdominal ectopia cordis and bilateral superior vena cava via the sternothoracotomy (clamshell incision) approach.

CASE REPORT

A 9-year old boy was born with thoracoabdominal ectopia cordis (Fig. 1A). Further workup included an echocardiogram demonstrating the transposition of the great arteries, ventricular septal defect, left ventricular outflow tract obstruction, hypoplastic right ventricle and bilateral superior vena cava. The patient’s heart protruded through the anterior diaphragmatic defect and was covered by very thin skin. The abdominal wall was only covered by an amniotic membrane. He was taken to the operating theatre for skin-flap coverage. At the age of 2 months, he underwent a modified Blalock-Taussig shunt procedure through a left thoracotomy. At 1 year of age, we chose a single ventricle pathway because of his hypoplastic right ventricle. He underwent bilateral bidirectional cavopulmonary anastomosis through sternothoracotomy (clamshell incision in the third intercostal space) without a dissection of adhesions between the heart and the skin because of his ectopia cordis. We performed cardiopulmonary bypass involving the insertion of a perfusion cannula into the ascending aorta and a venous drainage cannula into the right atrium. When both cavopulmonary anastomoses were completed, the superior vena cava was clamped in turn. Both cavopulmonary connections were placed as close together as possible. The postoperative course was uneventful, and the patient’s oxygen saturation was 85% at discharge from the hospital. At 5 years of age, he underwent thoracoabdominal wall reconstruction and umbilical plasty. At 9 years of age [weight 19 kg, height 114 cm], the patient presented with advanced hypoaxemia and dyspnoea on exertion; he was admitted to complete the Fontan operation. Until then, his parents had hesitated to risk a Fontan-type operation. Preoperative catheterization showed a mean pulmonary artery pressure of 8 mmHg, systemic arterial oxygen saturation of 83% and Nakata’s pulmonary arterial index of 156 mm²/m². A three-dimensional computed tomography image shows ectopia cordis before the Fontan operation (Fig. 2A and B). We performed the completion of the Fontan operation via resternothoracotomy (clamshell incision). Following the entrance to the right pleural cavity through the fourth intercostal space, we dissected and cut the sternum transversely at the height between the second intercostal spaces. The third and fourth right costal cartilages were cut (Fig. 2B). We conducted a standard cardiopulmonary bypass via the insertion of a perfusion cannula into the ascending aorta and a venous drainage cannula inserted into the right superior vena cava and the inferior vena cava. Myocardial protection was provided by crystalloid antegrade cardioplegia. The main pulmonary artery was divided and we closed the pulmonary valve by suture. An 18-mm expanded polytetrafluoroethylene graft, which was cut diagonally, was anastomosed to the right-to-main pulmonary artery in an end-to-side fashion. The inferior vena cava was anastomosed to the graft in an end-to-end fashion. We made a 3.5 mm fenestration in a side-to-side fashion. When the boy was weaned from the cardiopulmonary bypass, the central venous pressure was >20 mmHg. Transoesophageal echocardiography showed pulmonary venous obstruction due to the compression of his single pulmonary vein confluence by extracardiac conduit, which had not been diagnosed accurately. After cross-clamping the aorta and injecting cardioplegia solution, the right atrial opening showed a narrow pathway of a single pulmonary vein connection compressed by the conduit. We excised the wall of the route, cut its roof and augmented it with an expanded...
polytetrafluoroethylene patch. He was easily weaned from the cardiopulmonary bypass. The pressure of the pulmonary artery was 13 mmHg. He was taken to the intensive care unit, where he spent 5 days. On postoperative day 3, he was extubated, and on the following day, the chest tube was removed. His oxygen saturation was 95% at hospital discharge. A lung perfusion scan, in which the agent was injected from the lower extremity, showed 48% flow to the right lung and 52% to the left. Currently, 3 months after the surgery, the patient is in good health and had not been experiencing pain (Fig. 1B).

**COMMENT**

Ectopia cordis was present in 5.5–7.9 per 1 million live births. It is usually found in the presence of sternal defects, omphalocele, and ventral diaphragmatic hernia, pericardial defects, and congenital heart disease. These anomalies were reported by Cantrell et al. [3] as a pentalogy in 1958. Previous reports suggest a poor prognosis for patients with ectopia cordis and congenital heart disease. Thoracic ectopia cordis has been specifically associated with a poor prognosis. Although the more common thoracoabdominal ectopia cordis and lesser degrees of Cantrell’s pentalogy offer a better prognosis, there are a few reports of Fontan completion in ectopia cordis patients [1, 2]. It is challenging to perform midline sternotomy on ectopia cordis patients because of the adhesion between the skin and heart. In our opinion, the adhesion between the heart and skin in patients with ectopia cordis could make the operation field difficult. Some surgeons have reported the thoracotomy approach to perform cardiac surgery on ectopia cordis patients. The present patient had bilateral superior vena cava. It is difficult to deal with bilateral cavopulmonary anastomosis through thoracotomy. To overcome these problems, we adopted sternothoracotomy (clamshell incision) at the stage 2 operation which avoided the zigzag incision. This procedure revealed good surgical sight of the upper mediastinum structure, including the bilateral superior vena cave, ascending aorta, pulmonary artery and Blalock-Taussig shunt without dissection of the adhesion between the heart and the skin. However, we could deal with the clapshell incision using another intercostal space and expanded polytetrafluoroethylene sheet, which had been previously sheeted behind the sternum. A number of disadvantages of the clamshell incision have been described, including the ligation of both internal thoracic arteries and the potential for respiratory insufficiency [4]. Although this approach also causes more pain than median sternotomy, it is certainly better tolerated than posterolateral thoracotomy.
Conflict of interest: none declared.

REFERENCES


