Case report

Right atrial isomerism with infracardiac total anomalous pulmonary venous connection complicated by hiatal hernia

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Abstract

A 31-day-old girl with right atrial isomerism, a single atrium, a single right ventricle, pulmonary atresia, patent ductus arteriosus, coarctation of the pulmonary artery, infracardiac total anomalous pulmonary venous connection, and a large hiatal hernia was referred to our institution. We successfully performed a first-stage open palliation comprising the creation of a systemic-to-pulmonary arterial shunt, pulmonary artery angioplasty, and correction of the total anomalous pulmonary venous connection. During cardiopulmonary bypass, the hiatal hernia was also repaired through the median sternotomy itself without another laparotomy. The postoperative course was uneventful, and we avoided the potential complications of pulmonary venous obstruction and enteral feeding. She underwent a bidirectional Glenn operation and Fontan completion at 5 and 19 months of age, respectively.

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1. Introduction

Right atrial isomerism, resulting from a lateralization defect, can present as bilateral right-sidenedness and carries a poor prognosis because of the associated complex congenital cardiac malformations. Besides cardiac lesions, defective lateralization also leads to abnormalities in the gastrointestinal system including malrotation of gut and hiatal hernia [1]. Patients with small hiatal hernias are sometimes asymptomatic and surgical intervention is therefore not necessary in all cases [2]; however, large hiatal hernias cause gastroesophageal reflux, leading to poor feeding and vomiting. Furthermore, the hernial sac may exert space-occupying effects on the surrounding tissues and cause recurrent pneumonia, atelectasis, and compression of the atrium and pulmonary vein (PV) [1,3].

We encountered the case of an infant who had right atrial isomerism with a large hiatal hernia, and semi-urgent surgical repair was performed because of pulmonary venous obstruction (PVO) and infracardiac total anomalous pulmonary venous connection (TAPVC). Owing to concern regarding possible pulmonary venous compression by the large hiatal hernia and anticipation of a complicated postoperative course, we performed a first-stage open palliation with concomitant hiatal hernia repair through the same median sternotomy.

2. Case report

A 31-day-old girl (height, 48.5 cm and weight, 2.2 kg) was referred to our institution for surgical intervention. Echocardiography demonstrated right atrial isomerism, dextrocardia, a single atrium, a common atrioventricular valve (CAV), a single right ventricle, pulmonary atresia, coarctation of the pulmonary artery (PA), a right aortic arch, patent ductus arteriosus, infracardiac TAPVC with PVO, and bilateral superior vena cava. The ductus arteriosus was maintained by continuous lipo-prostaglandin E1 infusion. A chest X-ray and multidetector-row computed tomography (Fig. 1) revealed a large hiatal hernia (sliding type) and a transverse symmetrical liver. (Spontaneous closure of the ductus venosus was progressing slowly, and the patient suffered from PVO. Furthermore, she experienced feeding difficulties due to the hiatal hernia. However, at the institution where she was previously admitted, she was considered inoperable due to multiple risk factors, and she was only medically treated with digoxin, diuretics, sedative drug, and lipo-prostaglandin E1 infusion.)

On the day after admission, she underwent a semi-urgent surgery comprising (1) correction of TAPVC with anastomosis of the PV confluence to the posterior aspect of the common atrium and ligation of the vertical vein (conducted with low flow perfusion and moderate hypothermia), (2) PA angioplasty with total resection of the ductal tissue and direct end-to-end anastomosis using her own PA wall, and (3) creation of a systemic-to-pulmonary (SP) shunt between the left
innominate and left pulmonary arteries using a 3.0 mm ePTFE graft.

After cardiac repair, a pediatric surgery team attempted surgical correction of the hiatal hernia through the same median sternotomy with the heart still beating and before cardiopulmonary bypass (CPB) weaning. The hernial sac in the left pleural space was approached through a pleurotomy. Resection of the hernial sac and plication of the esophageal hiatus were performed without laparotomy. The duration of CPB and aortic cross-clamping was 287 and 49 min, respectively. The sternum was left open during the immediate postoperative period, with a delayed sternal closure on postoperative day 6.

We initiated enteral feeding on postoperative day 7 and were able to continue without any feeding difficulty. Postoperative gastroesophageal reflux or recurrence of hiatal hernia.

Postoperative cardiac catheterization showed equal bilateral PA growth, no anastomotic or branching PVO, and moderate CAVV regurgitation (revealed by echocardiography performed just before discharge and progressed gradually thereafter). She underwent a second-stage palliation consisting of a bilateral bidirectional Glenn operation, CAVV plasty, and an SP shunt takedown at 5 months of age and a Fontan completion with an extracardiac conduit at 19 months of age.

3. Discussion

In patients with right atrial isomerism and TAPVC, early and late mortality remain high [4,5]. Previous reports have identified infracardiac and mixed TAPVC, preoperative PVO, pulmonary atresia, and the need for an SP shunt surgery as significant risk factors of mortality in such cases. Our case had multiple risk factors for high mortality, thus requiring special attention.

Some literatures have previously reported hiatal hernia in patients with right atrial isomerism [1] and it has been estimated that the overall mortality among cases with right atrial isomerism with and without hiatal hernia is similar. However, large hiatal hernias cause feeding difficulties, including gastroesophageal reflux, vomiting, gastrointestinal bleeding, and exert space-occupying effects on the surrounding tissues leading to recurrent pneumonia, atelectasis, and compression of the atrium and PV [1,3]. Furthermore, the optimal timing of hiatal hernia repair in patients with single ventricle physiology is controversial.

Although PVO has not been reported after the repair of infracardiac TAPVC caused by the hernial sac, compression of the vertical vein of the infracardiac TAPVC has been reported previously [1]. In our case, the confluence of the PV and vertical vein was made to course along and curve toward the right side by the large hernial sac (Fig. 1); hence, there was a concern that the hernial sac could compress and deform the PV confluence if the hiatal hernia was left unrepaired.

Although a transabdominal approach is generally the first choice for hiatal hernia repair, a transthoracic approach facilitates dissection of a sufficient length of the esophagus and tension-free repair and provides better exposure [6]. Further, if the midline skin incision was extended and laparotomy was performed in this case, the transverse liver might have proved to be an obstacle while approaching the esophagus and stomach (Fig. 1); hence, we selected the transthoracic approach through the median sternotomy and approached the hernial sac during CPB without an additional laparotomy.

To our knowledge, such a concomitant surgery has not been reported previously. The advantages of this concomitant surgery are as follows. (1) Only one midline skin incision is needed. (2) Compression of the common atrium and PV by the hernial sac that occurs after infracardiac TAPVC repair can be avoided. (3) Enteral feeding can be initiated in the early postoperative period. However, this procedure also has
some disadvantages: (1) this approach and hiatal hernia repair through the same median sternotomy needs CPB support for cardiac decompression; therefore, it may be more invasive. (In this case, we considered hiatal hernia repair without CPB assist to be more invasive, regardless of the total CPB time.) (2) The operative field is farther and deeper from the median sternotomy than from a posterolateral thoracotomy. (3) To perform this concomitant surgery, the cardiac apex and the hernial sac must be contralateral; if these are ipsilateral, the approach during CPB may be difficult.

In conclusion, we successfully performed a first-stage open palliation comprising correction of the infracardiac TAPVC, PA angioplasty, and creation of an SP shunt with concomitant hiatal hernia repair through the same median sternotomy and thereby avoided the potential complications of PVO and enteral feeding.

References