

Congenital Absence of Pericardium in Babies with Patent Ductus Arteriosus

I O Azhar, MBBS, R Anas, MS

Cardiothoracic Surgery Unit, Hospital Raja Perempuan Zainab II, 15586, Kota Bharu, Kelantan, Malaysia

SUMMARY

We report a case of two babies with absence of pericardium and patent ductus arteriosus (PDA). The absence of pericardium was found coincidentally during PDA ligation. The PDA was successfully ligated but the pericardium was not reconstructed. Postoperatively, the agenesis of the pericardium did not interfere with cardiac function.

KEY WORDS:

Pericardium, congenital absence of pericardium, patent ductus arteriosus

INTRODUCTION

Congenital absence of pericardium is a rare developmental defect that results from faulty partitioning of the pleuropericardic cavity during the 5th week of development. It is an unusual defect with less than 500 cases reported in the literature with estimated incidence of 1 in 14,000 based on autopsies. The incidence of PDA has been reported to be approximately 1 in 2,000 births, and is inversely related to the maturity of the infant. In this publication, we present two clinical cases of combination of these defects.

CASE REPORTS

Case 1

This term baby girl was born via emergency lower segment caesarian section for fetal tachycardia in chorioamnionitis mother. Her birth weight was 2650 grams. However, she was noted to have signs of respiratory distress and was admitted to NICU. She was then intubated at 10 minutes of life. Chest X-ray was done and noted to have left sided congenital diaphragmatic hernia (CDH).

After stabilization of her condition, she was referred for surgical intervention of diaphragmatic hernia. Laparotomy and reduction of left sided CDH was done at day eighth of life. Intraoperatively, there was a large posterolateral (Bochdalek) defect 5 x 4cm at left diaphragm. Anterior lip of left diaphragm poorly developed while posterior lip is well developed. Postoperatively, she represented with manifestation of cardiac failure. Echocardiogram was performed and revealed large PDA measured 4.1mm with bidirectional shunt and atrial septal defect (ASD) secundum measured 3.8mm with left to right shunts. Absence of pericardium was not detected.

Subsequently, she underwent PDA ligation. Intraoperatively, noted PDA measured 4mm in diameter, vertical position, altered anatomy of the heart and surrounding structures. There was no pericardial tissue. The left lung well developed, and not hypoplastic. The identified PDA was clipped and no attempt was made to repair or reconstruct the pericardium. Postoperatively, she was stable and clinically improved.

Case 2

A 4-month-old Malay boy was born premature at 29 weeks of gestation. His birth weight was 1200 grams. He was admitted into NICU since birth due to respiratory distress. He is a known case of ventilator acquired pneumonia with left lung bullae. He had history of prolonged ventilation since birth. Chest x-rays revealed bullae involving about two third of his left lung with collapsed consolidation of his right lung. CT Thorax suggestive of bullae over apicoposterior of left lung. Echocardiogram was done and showed PDA measured 3.1mm with left to right shunts and small fenestrated ASD measured 2mm with left to right shunts, and again the pericardial agenesis was missed.

Left thoracotomy with left upper lobe cystectomy and decortication was done, with PDA clipping. Intraoperatively, left upper lobe was small, had partially been dissolved by the infective process, and displaced inferiorly by "lung cyst". Cyst is lined by thick cortex occupying the upper half of the left side of the chest. The remnant left lobe "cocooned" by the thick cortex. It was more like an abscess cavity with thick pus within. PDA was hidden below a very thick cortex, measuring about 3mm in diameter. The left sided pericardial tissue noted partially absence. The identified PDA was clipped and no attempt was made to repair or reconstruct the pericardium. Postoperatively, he was improved and able to be extubated.

DISCUSSION

Congenital absence of the pericardium is rare, with less than 500 cases reported in the literature. The majority of reports are based on postmortem examinations¹, with fewer cases are coincidental findings that occurred during surgery³. In our patients, the absence of pericardium was found incidentally during PDA clipping.

Congenital agenesis of pericardium can be divided into three groups – left sided, right sided and bilateral defects. Left sided absence of pericardium is the most common defects, while

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*Corresponding Author: Azhar Ibrahim Omar, Cardiothoracic Surgery Unit, Hospital Raja Perempuan Zainab II, 15586, Kota Bharu, Kelantan, Malaysia
Email: azharibrahimomar@gmail.com*

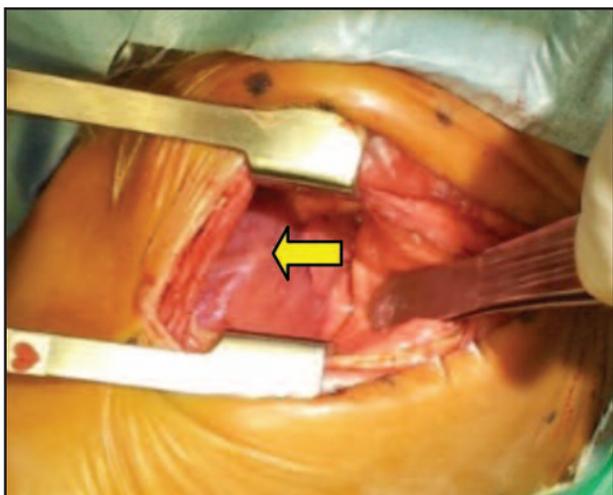


Fig. 1 : Complete absence of pericardium (arrow).

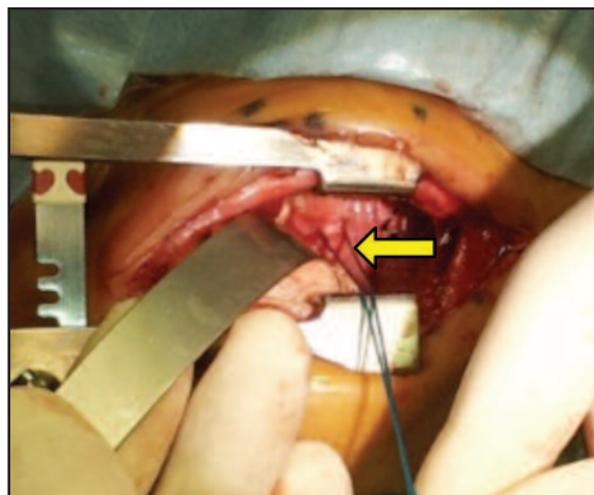


Fig. 2 : Patent ductus arteriosus is ligated (arrow).

right sided and bilateral defects are both rare. Each of these forms can be subdivided into complete or partial. Complete absence of the left pericardium is the most common defect accounting for 76% of all pericardial defects⁴. Our cases, the first baby was found to have complete absence of left pericardium, and the second baby have partial absence of left pericardium. Patients with partial defect of the pericardium are more at risk of morbidity than those with complete defects, as herniation of the heart through a partial defect may occur resulting in strangulation of the heart and death. Complete defects usually have a benign prognosis and require surgical intervention only in the presence of complications or debilitating symptoms. However, partial defects may lead more often to mechanical complications requiring surgical intervention.

Previous reports have shown that congenital absence of pericardium is commonly associated with congenital heart disease, including ventricular and atrial septal defect, PDA, valvular defects, and tetralogy of Fallot³. Our patients, she was found to have PDA and ASD secundum, while the boy has PDA with small fenestrated ASD.

CDH occur relatively frequent with about 30-40% of these patients have associated malformations. Most commonly are cardiac defects, but also neural tube defects, skeletal, urinary tract, and gastrointestinal abnormalities, which have a major influence on outcome and survival². Our first case, the patient presented with CDH with associated cardiac defects. Agenesis of the pericardium and anterior diaphragmatic hernia are also features of Cantrell's pentalogy, combined with abdominal wall defect, agenesis of the lower sternum, and cardiac malformation. In our first patient, she presented with cardiac malformations, and the diaphragmatic defect was in the posterolateral position. Our second case, he had no other congenital anomalies despite of lung cyst and abscess. The absence of pericardium and PDA in this baby is likely associated with prematurity.

In our cases, both of babies were found to have absence of pericardium incidentally intraoperatively and cannot be picked up by echocardiography. Most patients are asymptomatic. The most common symptom of complete absence of pericardium is vague chest pain and dyspnea. If absence of pericardium is suspected in patients present with these symptoms, various modalities of investigation can be performed. These include chest radiography, echocardiography, cardiac computed tomography (CT) scan and cardiac magnetic resonance imaging (MRI). MRI findings of complete pericardial defect were presented by Gutierrez et al. in 1985⁵. They predicted that MRI would become the preferred technique for the definitive diagnosis of absence pericardium due to its capability to image in any plane. MRI coronal sections through the heart demonstrated actual absence of pericardium. There was leftward displacement of the heart, a bulging pulmonary artery and herniation of lung between the base of the heart and the left diaphragm⁵.

The decision to proceed with surgical procedure was made on the basis of debilitating symptoms and herniation of the heart. Surgical procedures employed for patients with congenital absence of pericardium include left atrial appendectomy, division of adhesions, pericardiectomy, extension of the defect, or pericardioplasty. For patients with debilitating symptoms, pericardioplasty is recommended⁴. Immobilization of the heart with pericardial reconstruction will lead to symptomatic improvement.

CONCLUSION

We conclude that patients born with congenital diaphragmatic hernia and patent ductus arteriosus should routinely undergo vigilant investigation to exclude absence of the pericardium and other cardiac anomalies, which tend to be associated with this congenital defect. Absence of the pericardium can be diagnosed using echocardiography, CT scanning and magnetic resonance imaging⁴. However, in certain condition it tends to be under diagnosed and was only found during cardiothoracic surgery.

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