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Case report

Bilateral chylothorax: A late complication of Kawashima procedure despite normal pulmonary pressures

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ABSTRACT

Introduction: Chylothorax, a relatively rare congenital heart disease early postoperative complication, is occurring more frequently due to complexity of cardiac surgeries.

Presentation of case: We present a 9-month-old boy who had hypoplastic left heart (HLH) syndrome with interrupted inferior vena cava (IVC) and bilateral superior vena cava (SVC) palliated with left sided modified Blalock-Taussig (MBT) shunt during neonatal period and second stage palliation with left sided bidirectional glen (BDG) procedure and right sided Kawashima procedure develop bilateral chylothorax two weeks after discharge.

Discussion: This is the first reported case in the literature of a patient who developed chylothorax with relatively low Fontan systemic venous pressures after a Kawashima procedure. Clinically important chylothorax may be a marker of poor long-term outcomes, demonstrating an inability to handle overwhelming lymphatic congestion.

Conclusion: Early diagnosis of chylothorax in complex cardiac surgeries may permit successful conservative management.

1. Introduction

Chylothorax after cardiac surgery is a known complication and its incidence is around 2.8% [1], 9.2% in high-risk patients [2]. Chylothorax is an early postoperative complication with median time to diagnosis of 6 days [3]. Common etiologies are injury to thoracic duct, high central venous pressure, or presence of thrombus in Superior vena cava (SVC) system [1–3]. Our patient developed chylothorax following left sided bidirectional glen and right Kawashima procedure with multiple interesting features. Chylothorax happened late in the postoperative period after patient was discharged home from the hospital. Effusions were large and occurred in the absence of high venous pressures or thrombus in the SVC system and were resolved with conservative management. This work has been reported in line with the SCARE 2020 criteria [9].

2. Presentation of case

Our patient is a 9-month-old boy with complex cyanotic congenital heart disease including hypoplastic left heart (HLH), double outlet right ventricle (DORV), interrupted inferior vena cava (IVC), bilateral

superior vena cava (SVC), mitral valve atresia and pulmonary stenosis. Patient has confirmed diagnosis of heterotaxy syndrome with left atrial isomerism, an interrupted inferior vena cava with azygos continuation. Family history and medication history is noncontributory. At 10 days of age, patient had palliation with left sided modified Blalock-Taussig (MBT) shunt with 3.5 mm Gore-Tex graft. At 8 months of age patient had cardiac catheterization to evaluate for bidirectional glen (BDG) procedure. The patient's hemodynamics data and cardiac anatomy are shown in Fig. 1.

One week after cardiac catheterization, he underwent second stage palliative cardiac surgery of takedown of left MBT shunt, right sided Kawashima procedure, left sided BDG, right pulmonary arterioplasty and atrial septectomy. Cardiopulmonary bypass time was 174 min with aortic cross clamp time of 106 min. His Glen pressures during postoperative periods were 9 to 10 mm Hg. Patient was discharged home on postoperative day 5. Three days after cardiac surgery in outpatient setting, patient was asymptomatic and SpO₂ on room air was 92%.

Fifteen days after cardiac surgery, patient developed tachypnea, cyanosis, SpO₂ of 75%. Chest X-ray showed bilateral plural effusions with right side predominance and worsening of pulmonary opacification (Fig. 2). On arrival in PICU, oxygen saturation was 80% with high-flow

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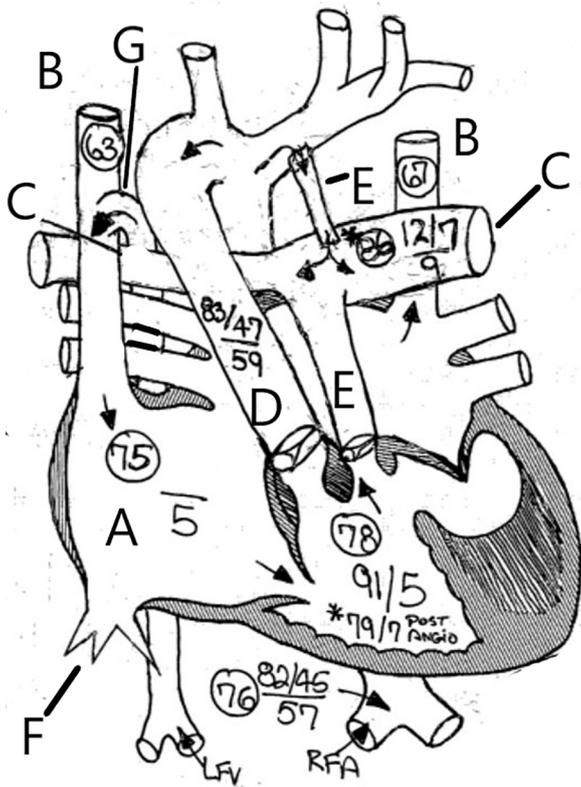


Fig. 1. Cardiac diagram showing anatomy, oxygen saturation and pressures. Numbers indicate systolic/diastolic and mean pressures in mm Hg. Circled numbers indicate oxygen saturations. A: Right atrium. B: Right and left superior vena cava (SVC). C: Right and left pulmonary arteries. D: Ascending aorta. E: Main pulmonary artery. F: Absent inferior vena cava (IVC). G: Vertical vein draining into right SVC.

nasal cannula at 10 lpm. Right side chest tube was placed and 500 ml of chylous pleural fluid was drained with significant improvement in respiratory symptoms. Pleural effusion analysis revealed elevated Triglyceride (TG) level (864 mg/dl), protein level (3 g/dl), LDH of 210, confirming chylothorax. Left sided chest tube was placed next day draining 110 ml of fluid and chest tube additionally drained 55 ml of fluid over next 24 h. The procedures were performed by the on-call intensivist. Internal jugular ultrasound ruled out evidence of clot. Patient was initially managed with low fat diet (Enfaport™, Mead Johnson nutrition, Evansville, IN. USA), diuretics, empiric sildenafil with partial improvement then maintained on total parenteral nutrition and octreotide infusion for complete resolution.

A significant reduction in pleural output was noted. With continued improvement the chest drain was removed at day 10 which allowed for incremental reduction of octreotide therapy and establishment of a low-fat oral diet and close observation for rebound chylous accumulation. Patient at one week follow-up in outpatient setting by pediatric cardiologist showed no recurrence of pleural effusion. He was found to be stable with appropriate growth and oxygen saturations. At the time of this writing, he continued to have a gastrostomy tube but had begun supplementing tube feeds with oral feeds which he tolerated well.

3. Discussion

The Bidirectional Glenn (BDG) procedure, which creates an anastomosis between the superior vena cava (SVC) and ipsilateral pulmonary artery (PA), is the second stage of repair in Fontan completion indicated for patients with anatomic or functional single ventricle. The Kawashima procedure is similar to BDG but adapted for interrupted inferior

vena cava (IVC) anatomy. The Fontan, which is the final procedure in the series, connects the IVC to the PA. Patients with interrupted IVC will have a Fontan-like circulation after undergoing the bidirectional Glenn/Kawashima due to the influx of splanchnic venous blood from azygous circulation. Chylothorax is less likely to occur after BDG (2.2%) compared to post-Fontan (12%) due to relatively lower postoperative systemic venous pressure [4]. Reported complications from the Kawashima procedure in the literature include severe cyanosis secondary to anomalous systemic venous return or unrecognized congenital porto-systemic venous connections [10].

This case is unique in that it is the first documented case chylothorax after the Kawashima procedure despite the presence of relatively low Fontan systemic venous pressures.

The mechanisms of chylothorax after cardiac surgery can be due direct trauma to the lymphatic vessels, less commonly secondary to central venous hypertension after cavopulmonary connection [5]. Relatively good lymphatic blood flow is maintained at a systemic venous pressure <15 mm Hg [6] and an animal study also has shown preserved thoracic lymph flow at relatively high systemic venous pressures [7]. Lymphatic abnormalities are also common in children with single ventricle physiology with palliative surgeries [8]. This evidence indicates that development of chylothorax after cardiac surgery is multifactorial. It is difficult to predict which patients develop chylothorax after high-risk cardiac surgery [6] and early identification and treatment is crucial for good outcome [4].

Our patient developed chylothorax relatively late in the post operative course and effusions were bilateral. Patient recovery in the hospital with conservative management and no recurrence after few months of follow up indicate temporary cause for chylothorax, most likely excess burden on lymphatic drainage. Despite high volume of chylothorax (>15 ml/kg/day), child recovered without surgical intervention.

Patient perspective

N/A.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

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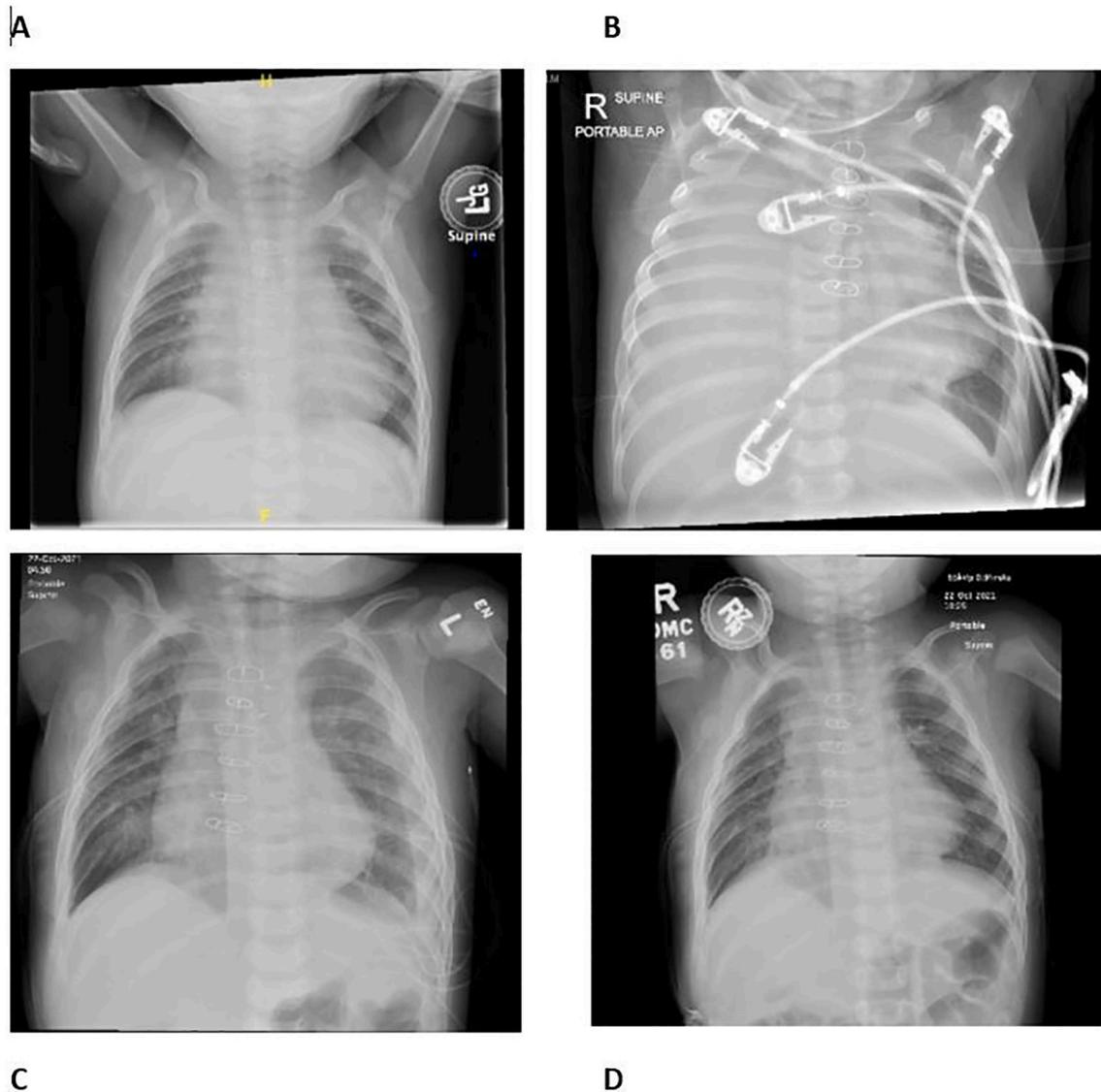


Fig. 2. Chest X-ray at various time periods of the disease process.

A: After cardiac surgery at the time of discharge. B: At the time of admission showing large right pleural effusion. C. After chest tube insertion on the right side. D. After left chest tube placement.

CRedit authorship contribution statement

JA was involved in the writing of the manuscript. RS and YT were involved in the editing/supervision of the manuscript.

Declaration of competing interest

The authors declare no conflicts of interests.

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