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MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

CASE REPORT: CLINICAL CASE

Isolated Chylopericardium in an Infant With Hypoplastic Left Heart Syndrome

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ABSTRACT

Isolated chylopericardium is an exceedingly rare complication in any age group. Review of the limited published case series shows the most common cause of isolated chylopericardium to be cardiac surgery. We present a case of isolated chylopericardium after a bidirectional Glenn procedure in an infant with hypoplastic left heart syndrome. (Level of Difficulty: Intermediate.) (J Am Coll Cardiol Case Rep 2020;2:2115–9) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 7-month-old female infant with a fetal diagnosis of hypoplastic left heart syndrome (HLHS) was born at full term to a 17-year-old primigravid mother. The child’s past surgical history was significant for a Norwood procedure with Sano shunt placement at 4 days of life and the need for balloon angioplasty of the Sano shunt, right pulmonary artery, and coarctation of aorta at 2 months of age for persistent desaturation and oxygen dependency. At 6 months of age, she underwent a bidirectional Glenn procedure in addition to an atrial septectomy and bilateral pulmonary arterioplasties (as indicated in Figure 1).

Her post-operative course was complicated by a parainfluenza virus infection requiring noninvasive positive pressure ventilation and inhaled nitrous oxide support. On post-operative day (POD) 4, a transthoracic echocardiogram (TTE) revealed a trivial pericardial effusion. She recovered gradually and was started on her home formula of completely hydrolyzed protein.

Given her persistent supplemental oxygen dependency and respiratory distress, a repeat TTE was done on POD 28, and it showed a moderate-size pericardial effusion extending circumferentially. On physical examination, she was in mild to moderate respiratory distress with tachypnea and equal air entry in bilateral lung fields. No gallop, muffled heart sounds, or friction rub were auscultated. She received...

LEARNING OBJECTIVES

- To recognize the need for a high degree of suspicion for CP following congenital heart surgery in children who have a persistent pericardial effusion and evidence of cardiopulmonary insufficiency.
- To consider early management of persistent post-operative CP after congenital heart surgery with surgical drainage for rapid resolution and decrease in length of stay.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors’ institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the JACC: Case Reports author instructions page.

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prednisolone, furosemide, and ibuprofen out of concern for post-pericardiotomy syndrome. The electrocardiogram was significant for only sinus tachycardia and right ventricular hypertrophy.

Her oxygen requirement persisted; thus a repeat TTE (Figure 2) was completed on POD 43 and showed a moderate pericardial effusion with fibrous stranding, located predominantly in the posterior (1.4 cm) and apical (9 mm) regions. As seen in Figure 3, computed tomography (CT) of the chest depicted an enhancing pericardial effusion suggesting lipid-rich content. Hounsfield units for this effusion were +6 to −6. Hounsfield units quantify radiodensity on CT, and these values did not align with expected values of, for example, blood (+13 to +75). At this point, the decision was made to drain the pericardial fluid with pericardial window creation. Figure 4 indicates the pericardial effusion on transesophageal echocardiogram before the pericardial window creation.

**PAST MEDICAL HISTORY**

Her past medical history included HLHS.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis included post-pericardiotomy syndrome, pericardial effusion, hemopericardium, chylopericardium (CP), and cardiac tamponade.
INVESTIGATIONS

Milky pink pericardial fluid was aspirated; studies demonstrated a positive chylomicron screen and a triglyceride level of 660 mg/dl. Total pericardial nucleated cells were 16,330/μl with the differential showing 100% lymphocytes. The concurrent serum triglyceride level was 72 mg/dl.

MANAGEMENT

Because of her prolonged hospitalization and persistent oxygen dependency, the decision was taken to drain the pericardial effusion surgically over a trial of a low-fat diet. As determined during the cardiothoracic surgery evaluation, the effusion was not accessible by a percutaneous route. She underwent pericardial window creation with drain placement that drained copious, milky, loculated fluid inferior to the heart. She was kept nil per os and supported with total parenteral nutrition soon after pericardial window creation. The drain was removed after 2 days. She was placed on a low-fat enteral formula subsequently.

DISCUSSION

An isolated CP is a rare occurrence in pediatrics. A literature review done by Yu et al. (1) looking at all case reports (adult and pediatric) of primary or idiopathic CP from January 1950 to December 2015 noted that idiopathic cases accounted for about 35% of all the reported cases. The most common cause of CP in pediatrics is a complication of congenital heart surgery. The incidence of isolated CP in children remains unknown apart from a few published reports from single-center studies. Reports published from single centers of CP following cardiac operations have noted an incidence of 0.12% to 0.22% (2,3), representing 0.0004% to 0.15% of the complications of cardiac surgery (4). Most cases were treated with pericardiocentesis or a low-fat or medium-chain triglyceride diet. Treatment of prolonged drainage with pericardial-peritoneal shunting has been suggested as well (5).

There is a double layer of lymphatic networks that drain pericardial surfaces. The anterior surface drains into pericardial fat and areolar tissue lymphatics, whereas the posterior surface drains into the lymphatics of the reflected mediastinal pleura (6). Any impairment of this drainage during surgery in this region can lead to the formation of a CP. Among intrathoracic tributaries of the thoracic duct, about 9% of them arise from the heart. Major tributaries are the right and left efferent lymphatic trunks (RET and...
The LET, respectively) (7). The LET arises from the left ventricle and ascends behind the main pulmonary artery to drain into the thoracic duct after joining the right paratracheal lymph nodes. The RET, conversely, arises from the right ventricle and travels between the ascending aorta and the main pulmonary artery, then left of the thymus gland to drain into the thoracic duct. Because of this anatomic course of RET, surgery manipulating major vessels has the potential to injure the RET inadvertently and contribute to the development of CP (7). Published reports also note post-operative leaks in thymic lymphatic tissue that are associated with CP (3). This observation also explains the higher incidence of CP in children compared with adults because children have thymic tissue rich in lymphatics.

There are 2 classes of clinical presentations described. Some patients present asymptptomatically wherein the intrapericardial drain begins to drain chylous fluid during the post-operative period. The second and more common presentation consists of signs of tamponade in the few weeks after surgery; initially, these cases can be more difficult to differentiate from post-pericardiotomy syndrome (2). The review by Yu et al. (1), looking at total of 104 adult cases, reported that in a majority (71%) of patients, the conservative approach was unsuccessful, and surgical interventions were needed, including pericardial window creation, thoracic duct ligation, thoracic duct embolization, and pericardioperitoneal shunt. We have not come across studies looking at a comparison of percutaneous versus surgical drainage of pericardial effusion, but a single-center retrospective study in adults showed no difference in mortality between the 2 techniques but a higher recurrence rate with percutaneous drainage (9).

Conservative management of CP includes ruling out other reversible causes (e.g., jugular venous thrombosis), draining the chylous fluid, limiting total fluid intake, and instituting a low-fat diet with medium-chain triglycerides. Patients with refractory cases may require complete cessation of an enteral diet and support with parenteral nutrition until the drainage resolves. Use of somatostatin analogue infusion, intended to limit splanchnic blood flow, is also recommended to decrease overall lymphatic flow. Patients with persistent high chylous output may benefit from surgical intervention in the form of a thoracic duct ligation or chemical or surgical pleurodesis. A review by Yu et al. (1), looking at total of 104 adult cases, reported that in a majority (71%) of patients, the conservative approach was unsuccessful, and surgical interventions were needed, including pericardial window creation, thoracic duct ligation, thoracic duct embolization, and pericardioperitoneal shunt. We have not come across studies looking at a comparison of percutaneous versus surgical drainage of pericardial effusion, but a single-center retrospective study in adults showed no difference in mortality between the 2 techniques but a higher recurrence rate with percutaneous drainage (9).

FOLLOW-UP

By 3 days after pericardial window surgery, she was transitioned to room air with no distress. She remained hemodynamically stable and was discharged home on POD 7. No reaccumulation of pericardial effusion was noted on outpatient follow-up.
visits with pediatric cardiology in her hometown 2 weeks after the discharge.

**CONCLUSIONS**

CP is an exceedingly rare occurrence in the pediatric patients, with a post-cardiac surgery complication representing the most common cause. It is even less common in children without any known syndromes or chromosomal abnormalities. We report this rare case where not only did CP develop in the patient after a bidirectional Glenn procedure, but also the diagnosis was suggested retrospectively on the basis of echogenicity of the fluid detected on imaging, including CT and TTE. CP is particularly dangerous because it can lead to tamponade physiology if left untreated. This rare condition is difficult to treat with conservative measures, as reported by Yu et al. (1) from their extensive review; therefore, early surgical interventions should be considered.

**AUTHOR DISCLOSURES**

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**REFERENCES**


**KEY WORDS** chylothorax, congenital heart defect, pericardial effusion