# Chylothorax Following Bidirectional Cavopulmonary Shunt (Glenn Procedure) in an Infant with Complex Congenital Heart Disease: A Case Report.


## ABSTRACT

**Background:** Chylothorax or pseudochylothorax is a serious complication that can occur following pediatric cardiac surgery. We report the case of a 17-month-old infant with complex congenital heart disease of the single-ventricle type, who developed recurrent massive pleural effusion after a bidirectional cavopulmonary shunt (Glenn procedure). This case illustrates a complication that, while not uncommon, can prove challenging to manage.



**Keywords:** Chylothorax, Glenn Procedure, Bidirectional Cavopulmonary Shunt, Single Ventricle, Congenital Heart Disease, Pleural Effusion, Pediatric Cardiac Surgery.


## INTRODUCTION

The bidirectional cavopulmonary shunt (Glenn procedure) represents an intermediate surgical step in the staged palliation of single-ventricle congenital heart disease. It enables passive pulmonary circulation via the superior vena cava [1]. However, this non-pulsatile circulation predisposes patients to specific complications, including pleural effusions, chylothorax, and persistent hypoxemia. These complications are reported in approximately 30% of postoperative cases [2,3]. We report a case of chylothorax causing acute respiratory distress in an infant following a bidirectional cavopulmonary shunt.

## CASE REPORT

### 1. Patient demographics and medical history

We report the case of a 17-month-old male infant, the youngest of four siblings, born at term following a poorly monitored pregnancy, with no parental consanguinity. He had been followed since the age of one year for complex congenital heart disease comprising: single ventricle, mitral valve atresia, right-sided aorta, sub-valvular and valvular pulmonary stenosis, and an interatrial communication of 7 mm. One month prior to admission, the patient underwent a bidirectional cavopulmonary shunt (Glenn procedure). The immediate postoperative course was uneventful with good hemodynamic stability, and pulmonary arterial pressure rose from 10 mmHg to 15 mmHg.

**2. History of presenting illness**

The illness began 15 days before admission with the gradual onset of afebrile respiratory distress. Chest imaging revealed a large left-sided pleural effusion, which prompted a first thoracocentesis. The course was complicated by recurrence one week later, necessitating a second drainage procedure yielding approximately 600 mL of purulent-appearing fluid. Given the persistence of symptoms, the patient was referred to the Pediatric Intensive Care Unit (PICU) for further management.

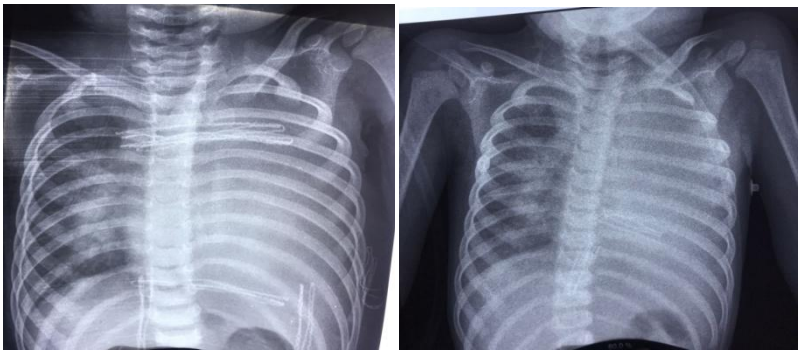
**3. Physical examination on PICU admission**

On admission, clinical examination revealed preserved consciousness (GCS 15/15), tachycardia at 154 bpm, tachypnea at 40 cycles/min, $SpO_2$ at 65% on supplemental oxygen, peripheral cyanosis, and signs of increased work of breathing (intercostal retractions, nasal flaring).

**4. Investigations**

Laboratory investigations are summarized in Table

1. Chest X-ray showed persistent left-sided pleural effusion.



2. Pleural fluid analysis (Table 2) confirmed the diagnosis.

**Table 1:** Laboratory Investigations on PICU Admission

| Parameter | Result |
|---|---|
| Hemoglobin | 11.7–13.7 g/dL |
| White blood cells | 11,980/mm³ |
| Platelets | 405,000/mm³ |
| CRP | 6.3 mg/L |
| $Na^+$ | 134 mmol/L |
| $K^+$ | 3.1–3.8 mmol/L |

**Table 2:** Pleural Fluid Biochemical Analysis

| Parameter | Result |
|---|---|
| Total proteins | 46 g/L (exudative) |
| Triglycerides | 2.68 mmol/L (≈ 237 mg/dL) — diagnostic threshold: ≥ 1.24 mmol/L |
| Cytobacteriological exam | Sterile |

A triglyceride level of 2.68 mmol/L (237 mg/dL), well above the diagnostic threshold of 1.24 mmol/L (110 mg/dL), confirmed the diagnosis of chylothorax. The high protein content was

consistent with an exudative effusion. Sterile cytobacteriological examination excluded superinfection.

**5. Management and outcome**

In the PICU, the patient received furosemide 1 mg/kg every 6 hours to reduce systemic venous pressure and lymphatic flow, aspirin 20 mg/kg/day for its antiplatelet and anti-inflammatory properties in the context of cavopulmonary circulation, and continuous chest tube drainage. The clinical course was favorable, with progressive respiratory improvement allowing transfer to a non-intensive care ward.

## DISCUSSION

**1. Epidemiology**

Chylothorax is one of the most feared postoperative complications in pediatric cardiac surgery. According to North American multicenter databases (PC4 and PHIS), the overall incidence ranges from 2.8% to 3.8% [1]. This incidence is significantly higher in neonates (6.9%), patients with single-ventricle physiology (6.9%), chromosomal anomalies (5.2%), and major non-cardiac anomalies (6.4%) [4]. A recent study using the US National Inpatient Sample (2016-2019) confirmed that cavopulmonary shunting (Glenn and Fontan) carries the highest postoperative chylothorax incidence among all cardiac surgeries [5]. In single-ventricle series, chylothorax complicates up to 17% of Glenn procedures, particularly in patients with pre-existing lymphatic abnormalities on MRI [3]. The presence of a systemic right ventricle has been identified as an independent risk factor, owing to ventricular dysfunction and elevated central venous pressure [6].

**2. Diagnosis**

Diagnosis rests on pleural fluid biochemistry. A triglyceride level >110 mg/dL (1.24 mmol/L) with cholesterol <200 mg/dL (5.18 mmol/L) is considered diagnostic,

corresponding to a 99% diagnostic probability [7]. In our case, triglycerides measured 2.68 mmol/L (237 mg/dL), well above threshold. The initially 'purulent' appearance of the fluid underscores the importance of systematic biochemical analysis in any recurrent post-Glenn effusion.

### 3. Management

Initial management follows a stepwise conservative approach, guided by the 2022 PC4 Chylothorax Work Group multicenter consensus algorithm [8]. Dietary modification with a medium-chain triglyceride (MCT) diet is the cornerstone; a two-week duration has been shown to reduce drainage duration without increasing recurrence risk [9]. Furosemide reduces systemic venous pressure and lymphatic flow. Chest tube drainage is indicated for large effusions with respiratory compromise. Octreotide should be introduced in case of treatment failure or recurrence [8,10]. In our case, the combination of chest drainage, furosemide, and aspirin resulted in a favorable outcome without requiring somatostatin analogues.

## CONCLUSION

This case clearly illustrates the pathophysiological and therapeutic complexity of post-Glenn chylothorax. Knowledge of its epidemiology (6.9% incidence in single-ventricle patients), its mechanisms (thoracic duct injury, venous hypertension, pre-existing lymphatic dysfunction), and the updated management algorithm (MCT diet, diuretics, octreotide in case of failure [8,10]) is essential for any clinician managing complex congenital heart disease. Systematic biochemical analysis of all pleural effusions following cavopulmonary surgery, application of a standardized stepwise protocol, and early identification of risk factors are the key determinants of a favorable outcome.

## DECLARATIONS

**Patient Consent:** Written informed parental consent was obtained for publication of this case report.

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