



Incidence and Etiology of Chylothorax after Congenital Heart Surgery in Children

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Abstract

Background: Chylothorax is a rare but serious postoperative condition with a high rate of morbidity and may lead to the mortality of children undergoing congenital heart disease (CHD) surgery. This study evaluated the specific surgical procedures associated with the higher risk of postoperative chylothorax.

Methods: We assessed 435 cases undergoing CHD surgery between April 2003 and May 2006. We detected postoperative chylothorax in 6 patients. The diagnosis of chylothorax was established based on the presence of an odorless fluid with the characteristic milky appearance of the fluid (except when the patients were fasting in the immediate postoperative period), a triglyceride level greater than 110 mg/dL or between 50 and 110 mg/dL with a pleural fluid white cell count greater than 1000, and more than 80% lymphocytes on differential when the pleural fluid was not chylous.

Results: Over a 37-month period, 435 (mean age = 51.6 months; 232 males) patients underwent various types of surgical procedures for CHD; 6 patients developed chylothorax after the Fontan operation; one patient died due to severe chylothorax; 3 patients were managed by nutritional modifications, diuretics, and thoracocentesis; and 2 patients required thoracic duct ligation. The Fisher exact test analysis showed a significant association between the Fontan operation and postoperative chylothorax (p value < 0.0001).

Conclusion: Our study showed a significant association between the Fontan surgery and chylothorax.

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Introduction

Several situations are known to be associated with chylothorax; none more so important than pediatric cardiac surgery.¹ A recent increase in the incidence of postoperative chylothorax, from the previously reported 1% or less²⁻⁵

to 2.5% to 4.7%,^{3, 6, 7} is well documented. Damage to the thoracic duct, disruption of accessory lymphatics, and increased pressure in the systemic vein exceeding that in the thoracic duct have been proposed as the possible causes of chylothorax after surgery for congenital heart disease (CHD).^{2, 3, 8}

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Postoperative chylothorax is a particularly serious complication and has the potential to cause significant morbidity and mortality.⁹ Chylothorax results in prolonged ventilator dependence, increased length of hospital stay, malnutrition, nosocomial infection, and death.^{6, 10-15} Patients with chylothorax usually remain asymptomatic until a large volume of chylous accumulates.^{5, 16} Chylous contains vitally important materials, and its accumulation over an extended period of time accounts for nutritional depletion, fluid and electrolyte loss, hypolipemia, and lymphocytopenia of T cells, which can contribute to immunodeficiency.^{5, 17}

Accordingly, awareness of the conditions associated with chylothorax helps to identify and manage this potentially hazardous condition earlier and to prevent its serious morbidities and mortality. Most of the studies published so far being case reports or small-sized trials, we sought to evaluate a large group of patients undergoing pediatric cardiac surgery in terms of postoperative chylothorax incidence and to determine the specific surgical procedures associated with the higher risk of postoperative chylothorax.

Methods

This study included all patients with CHDs who underwent cardiac surgery for the correction of their anomaly between April 2003 and May 2006. The parents of all the patients signed written informed consent. The institutional Ethics Committee approved the protocol of this study.

The patients' CHDs were diagnosed by physical examination, chest X-ray (CXR), and echocardiography. After the type of CHD had been diagnosed, decision for surgery in each patient was made by a team consisting of a cardiac surgeon and a pediatric cardiologist.

Postoperatively, pleural effusion was sought for by physical examination and CXR. The diagnosis of chylothorax was established based on the presence of an odorless fluid with the characteristic milky appearance (except when the patients were fasting in the immediate postoperative period) of the fluid, a triglyceride level > 110 mg/dl or between 50 and 110 mg/dL with a pleural fluid white cell count > 1000 and > 80% lymphocytes on differential.

Data collection was performed by reviewing case files, and demographic, clinical, radiographic, and laboratory investigations were recorded.

Our institutional management strategy following the diagnosis of chylothorax was to commence conservative management with nutritional modifications (such as high-protein and high-medium chain triglycerides diet), followed by surgical interventions for non-responders.

The data are expressed as frequencies and mean \pm standard deviation (SD). The characteristics were compared between the patients with and without chylothorax using the χ^2 and Fisher exact tests. A p value < 0.05 was considered statistically significant. For the statistical analyses, the statistical software SPSS (version 16.0) for Windows (SPSS Inc., Chicago, IL) was used.

Table 1. Incidence of chylothorax after various types of pediatric heart surgery

CHD type	Surgery type	N (female)	Mean Age (range)	Down Syn.	PE	CTX	Death
TOF	TC	97 (42)	54.88 m (5 m – 29 y)	1	5	0	1
AV canal	TC	16 (9)	44.65 m (7 m – 13 y)	2	0	0	0
VSD	TC	58 (23)	65.1 m (3 m – 24 y)	0	1	0	0
ASD	TC	17 (11)	116.1 m (18 m – 45 y)	0	0	0	0
TGA	ASO	28 (6)	3.3 m (0.3 m -36 m)	0	2	0	9
Unknown	Senning	7 (3)	19.5 m (1.8 m – 60 m)	0	0	0	1
PDA	TC	14 (10)	23.2 m (7 m – 98 m)	1	0	0	0
CoA	TC	12 (6)	17.5 m (0.9 m – 84 m)	0	0	0	0
Ebstein Anomaly	TC	6 (2)	78.0 m (5 y- 9 y)	0	0	0	0
TAPVR	TC	4 (1)	79.5 m (1 m – 25 y)	0	0	0	0
AS	AVR	5 (3)	74.5 m (0.6 m – 16 y)	0	0	0	0
Unknown	MBT shunt	61 (27)	44.9 m (0.2 m – 25 y)	1	1	0	3
Single Ventricle	Fontan	50 (20)	123.1 m (30 m – 33 y)	0	6	6	1
Unknown	Glenn	46 (24)	6.95 y (1 y – 39 y)	0	4	1	0
Unknown	RV to PA shunt	12 (5)	49.8 m (11 m – 96 m)	2	0	0	1
TA Type I	TC	1 (1)	3.0 y	0	0	0	0
AP window	TC	1 (1)	3.5 y	0	0	0	0

CHD, Congenital heart disease; PE, Pleural effusion; CTX, Chylothorax; TOF, Tetralogy of Fallot; TC, Total correction; AV canal, Atrioventricular canal; VSD, Ventricular septal defect; ASD, Atrial septal defect; TGA, Transposition of the great vessels; ASO, Arterial switch operation; PDA, Patent ductus arteriosus; CoA, Coarctation of the aorta; TAPVR, Total anomalous pulmonary venous return; AS, Aortic stenosis; AVR, Aortic valve repair; MBT shunt, Modified Blalock-Taussig shunt; RV, Right ventricle; PA, Pulmonary artery; TA Type I, Tricuspid atresia type 1; AP window, Aortopulmonary window



Table 2. Characteristics of the patients with postoperative chylothorax

Case number	Age	Sex	Dx	Previous surgery	Current surgery	Management of CTX
1	5 y	F	TGA + VSD + PH	4 y ago: PA banding 2 y ago: Bd. Glenn	Fontan	TDL after failure of low fat regimen
2	9 y	F	PS + TA	2 y ago: RtMBT	Fontan	TDL after failure of low fat regimen and chest tube
3	8 y	F	TA	2 y ago: RtMBT shunt	Fontan	Chest tube, low fat regimen, and diuretics
4	5 y	M	PA + TGA + MA + SI totalis	4 y ago: RtMBT shunt 2 y ago: Glenn	Fontan	Chest tube, low fat regimen, and diuretics
5	3 y	F	TA	1 y ago: AP shunt	Fontan	Chest tube, low fat regimen but died due to sever CTX
6	10 y	F	TA + PS	9 y ago: LtMBT	Fontan	Chest tube, low fat regimen, and diuretics

Dx, Diagnosis; CTX, Chylothorax TGA, Transposition of the great arteries; VSD, Ventricular septal defect; PH, Pulmonary hypertension; PA, Pulmonary atresia; Bd. Glenn, Bidirectional Glenn; TDL, Thoracic duct ligation; PS, Pulmonary stenosis; TA, Tricuspid atresia; RtMBT shunt, Right main Blalock-Taussig shunt; MA, Mitral atresia; SI, Situs inversus; AP, Pulmonary atresia

Results

Over 37 months, 435 (mean age = 51.6 months; 232 males) patients underwent various types of surgical procedures for CHD. Sixteen patients were excluded due to early postoperative mortality, and 403 patients continued their follow-up. According to the surgical procedures, the patients were divided into 17 groups. The types of congenital anomalies and surgical procedures, mean age, incidence rates of pleural effusion (4.5%) and chylothorax (1.43%), and death rate (3.8%) are summarized in Table 1.

Following cardiac surgery, 19 (4.53%) and 6 (1.43%) patients revealed pleural effusion and chylothorax, respectively. We found one death among the chylothorax cases. The underlying anomaly of the patients who showed postoperative chylothorax was single ventricle anomaly, which was corrected by the Glenn and Fontan procedures. The Fisher exact test analysis showed a significant association between the Fontan operation and postoperative chylothorax (p value < 0.0001). The characteristics of the patients who showed postoperative chylothorax are summarized in Table 2.

Discussion

This study enabled us to determine the incidence of chylothorax, which was 1.43% following pediatric heart surgery. All patients who revealed the important complication, chylothorax, had undergone the Fontan and Glenn surgery.

Chylous pleural effusion, or chylothorax, is an early postoperative complication.^{1, 18, 19} Although an infrequent complication after the repair of CHDs, chylothorax can

result in significant morbidity.²⁰ The postoperative leakage of the lymphatic fluid into the pleural space may result from the surgical disruption of the thoracic duct or one of its main tributaries, or increased pressure within the intrathoracic lymph system.

Longer time to diagnosis is correlated with increased drainage duration; therefore, morbidity associated with lipid and protein losses, immunosuppression, long-term chest tube and intravenous access, and hospitalization will increase subsequently.⁶ Additionally, treatment modalities are more effective if they are initiated earlier since in a late application of octreotide therapy for patients with relatively higher daily drainage of chyle no considerable improvement has been demonstrated.⁶ Taken together, it is understood that being aware of the risk factors of postoperative chylothorax is needed for an earlier diagnosis of this potentially hazardous complication, providing the patients with earlier management and obviating the need for more invasive therapies such as thoracic duct ligation.

As was mentioned earlier, the incidence of chylothoraces in our study was 1.43%, which was higher than 0.85% reported by Chan et al.²¹ while less than the incidence rates of 2.5%,³ 3.8%,⁶ and 4.7%⁷ reported by others.^{3, 22} In comparison to older studies, reporting 1% from 1979 to 1987 by Allen and co-workers⁶ and 1.1% from 1961 to 1969 by Higgins and associates,⁸ the incidence of chylothorax showed an increase in recent years, reflecting the increased complexity of surgery and possibly the earlier reintroduction of feeding after surgery.^{6, 7}

Because the thoracic duct is thin-walled and may be colorless, it may be difficult to identify. Consequently, it is susceptible to inadvertent injury during investigative or surgical procedures in the posterior mediastinum. The laceration of the thoracic duct during an accident or thoracic

surgery results in lymph escaping into thoracic cavity as well as pleural space, producing chylothorax. Although all types of intrathoracic manipulations may cause chylothorax, several congenital malformations have been reported to be more vulnerable to this complication.^{3, 4, 6} All procedures predisposing to increased systemic venous pressure have the risk of postoperative chylothorax.²¹ In particular, the bidirectional cavopulmonary shunt operation, Fontan-type procedures, and right ventricular dysfunction after the repair of Tetralogy of Fallot have been previously reported to increase the potential of postoperative chylothorax.^{3, 6} Our finding in this study, which demonstrated a significant association between the Fontan surgery and postoperative chylothorax, further corroborated the previous studies.

In addition to venous system manipulation, other types of closed heart procedures performed in the vicinity of the thoracic duct such as systemic-to-pulmonary arterial shunt insertion, repair of aortic coarctation, and ligation of the arterial duct, likewise predispose to the development of chylothorax as is evidenced in this and previous studies.^{3, 5}

In the present study, the only type of surgical procedure which showed a significant association with postoperative chylothorax was Fontan. This finding is in accordance with previous studies.⁶

During cavopulmonary anastomosis in the Fontan procedure, both systemic venous hypertension and resultant back-up of pressure into the thoracic duct on one hand, and mechanical injury to the thoracic duct during surgical manipulation on the other hand lead to impairment in chylous drainage and increased chyle loss.

The general consensus for the initial management of postoperative chylothorax in children is probably chest tube drainage and nutritional support.⁷ Conservative management for several weeks appears justified as the resolution of chylothorax has been reported in up to 77% of patients after giving either medium chain triglycerides or total parenteral nutrition for up to 45 days with an average of about 12 days.^{7, 18, 23} Previous studies have suggested that the persistence of chyle output for more than 3 weeks¹⁸ and lesions associated with elevated systemic venous pressure^{3, 6} are risk factors for the failure of conservative management. Given that the Fontan procedure carries a higher risk of prolonged chylous pleural drainage,^{6, 21} it may necessitate earlier and more aggressive therapy for these patients. Our results in the current study are encouraging for further studies to evaluate whether earlier therapeutic interventions can reduce the duration of chylous drainage after Fontan and increase the efficacy of conservative management, which may eventually decrease hospitalization duration, improve prognosis, and prevent more invasive procedures like thoracic duct ligation.

Conclusion

The Fontan operation is associated with higher risk of postoperative chylothorax and necessitates the awareness of surgeons for earlier diagnosis followed by proper management.

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