

Etiology and Management of Chylothorax Following Pediatric Heart Surgery

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ABSTRACT ***Background:*** **Chylothorax after congenital heart surgery (CHD) is a potentially challenging complication. The purpose of this study was to review our experience with the management of chylothorax following congenital heart surgery.** ***Methods:*** **Between September 1997 and August 2006, of 1341 pediatric patients undergoing correction of congenital heart disease in our institution, 18 (1.3%) developed chylothorax postoperatively. Surgical procedures included tetralogy of Fallot repair in 10 patients, ventricular septal defect closure (one), atrial septal defect with pulmonary stenosis repair (one), Fontan procedure (three), coarctation of the aorta repair (one), aortopulmonary shunt (one), and ligation of patent ductus arteriosus in one patient. All patients followed a therapeutic protocol including complete drainage of chyle collection and controlled nutrition. Somatostatin was used adjunctively in six (33.3%) patients. Surgical intervention was reserved for persistent lymph leak despite maximal therapy. Following resolution of chylothorax, a medium-chain triglyceride diet was implemented for six weeks.** ***Results:*** **There were no deaths. Fifteen patients (83.3%) responded to conservative therapy. Lymph leak ranged from 2.5 to 14.7 mL/kg per day for 8 to 42 days. Three patients with persistent drainage required thoracotomy with pleurodesis to achieve resolution, in two of which previously attempted chemical pleurodesis with doxycycline proved ineffective. Duration of lymph leak in this subgroup ranged from 15 to 47 days with 5.1 to 7.4 mL/kg per day output.** ***Conclusions:*** **Postoperative chylothorax is an infrequent complication of surgery for congenital heart disease and can occur even after median sternotomy in the absence of pathologically elevated venous pressure or Fontan circulation. Although hospitalization can be prolonged, conservative therapy is effective in most cases, while surgical pleurodesis proved successful in the refractory cases.** doi: 10.1111/j.1540-8191.2008.00781.x *(J Card Surg 2009;24:369-373)*

Postoperative chylothorax in childhood is a potentially serious complication of any thoracic surgical procedure and its management represents a serious challenge. In particular, chylothorax occurring after pediatric congenital heart surgery is an uncommon complication, occurring in 0.6% to 2% in reported series.[1,2] The intent of our study was to explore the etiology and present our experience with the management of this type of chylothorax.

MATERIALS AND METHODS

The hospital records of all children with a postoperative chylothorax following congenital heart surgery at our institution between September 1997 and August 2006 were reviewed. During this period, 1341 pediatric patients underwent surgery for congenital heart disease (CHD). Of these, 18 patients (1.35%), nine males and nine females, aged 5 to 68 (median 19.5) months old and with body weight 6 to 18.7 (median 9.2) kg, developed chylothorax 1 to 15 days postoperatively. Surgical procedures preceding the occurrence of lymph leak included tetralogy of Fallot (TOF) repair in 10 patients, ventricular septal defect (VSD) closure (one), atrial septal defect (ASD) closure with pulmonary stenosis (PS) correction (one), Fontan procedure (three), coarctation of aorta (CoA) repair (one), patent ductus arteriosus (PDA) ligation (one), and placement of a central aortopulmonary (A-P) shunt in a patient with complex CHD (one). Surgical access had been achieved via median sternotomy in 15 and left thoracotomy in three patients. Patient characteristics and procedures are listed in Table 1.

Chylothorax was suspected when persistent chest tube drainage occurred or a pleural effusion developed with the characteristic milky fluid. Microscopic and biochemical analyses of this fluid revealing a triglyceride (TG) level > 1.2 mmol/L and total cell count $> 1000/\mu$L, predominantly lymphocytes established the diagnosis.

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TABLE 1
Patient Characteristics and Procedures

Patients	Age (Month)	Diagnosis/ Operation	Duration of Leak (Days)	Amount of Leak (mL/kg/Day)	Somatostatin	Doxycyclin	Surgery	Comments
1	23	TOF	24	5.04	Yes	Yes	Yes	Reoperated day 30
2	13	TOF	8	3.86	No	No	No	
3	13	ASD, PS	47	7.39	Yes	Yes	Yes	Reoperated day 15 Noonan's syndrome
4	10	VSD	30	2.53	No	No	No	
5	9	TOF	15	5.28	Yes	No	No	
6	19	TOF	36	5.71	No	No	No	
7	31	PDA	25	8.71	Yes	No	Yes	Reoperated day 2
8	22	TOF	8	4.70	Yes	No	No	
9	5	CoA	2	5.67	No	No	No	
10	42	Fontan	10	3.92	No	No	No	
11	22	TOF	26	14.7	No	No	No	
12	28	Complex CHD/ A-P shunt	20	6.25	No	No	No	
13	50	Fontan	13	2.8	No	No	No	
14	15	TOF	38	9.1	Yes	No	No	
15	68	Fontan	42	6.05	No	No	No	
16	20	TOF	16	7.69	No	No	No	
17	15	TOF	16	4.1	No	No	No	
18	19	TOF	16	5.57	No	No	No	

All patients were treated conservatively, initially, with complete pleural drainage of the chyle collection and either enteral low-fat diet or, in most cases, total parenteral nutrition (TPN). A somatostatin analogue was used adjunctively in six patients (33.3%). Attempted chemical pleurodesis with doxycycline via chest tube was unsuccessful in two patients. Surgical intervention was reserved for persistent lymph leak despite maximum medical treatment and was necessary in three patients. All patients continued on medium-chain triglyceride (MCT) diet for a total of six weeks following resolution of chylothorax and on low-fat diet for another six weeks (Fig. 1).

RESULTS

There were no deaths. Fifteen patients (83.3%) responded to conservative therapy. The median duration of lymph leak in these patients was 16 days (range 8 to 42), and the median lymph leak 5.6 (range 2.5 to 14.7) mL/kg per day. In two out of three patents with persistent drainage, chemical pleurodesis with doxycyclin via chest tube proved ineffective. The median duration of lymph leak in this medically refractory subgroup was 24 (range 24 to 47) days with median output 7.4 (range 5 to 8.7) mL/kg per day. These patients underwent thoracotomy, lymphatic ligation, and abrasion pleurodesis resulting in postoperative resolution of chylothorax.

CONCLUSION

Injury to the main thoracic duct or its branches is possible after virtually any procedure performed near its route, typically during aortic procedures performed in the left hemithorax. However, most of our patients (83.3%) who developed chylothorax had undergone a sternotomy, and surgical manipulation remote from the course of the thoracic duct. The etiology of lymph leak in these patients is unclear but may involve injury to small lymphatic vessels around the ascending aorta, the superior vena cava, or the pulmonary artery trunk and its branches, which may require extensive dissection for cannulation or repair. Variations in lymphatic pathways and the presence of accessory or minor lymphatic channels in the mediastinum may render them vulnerable to inadvertent trauma during these procedures, resulting in chylous effusion without interference of the main thoracic duct. Chylopericardium has been reported as a rare complication after operations for congenital heart disease.[3] Isolated chylopericardium is difficult to diagnose and may be mistaken for postpericardiotomy syndrome. This raises the question whether the pleural effusions seen in our sternotomy cases are in fact pericardial collections draining through an opening in the pleura in one of the hemithoraces and presenting as chylothoraces. It should be mentioned that we routinely open both pleurae (drained separately to prevent postoperatively fluid collection). It is of interest that in two of our patients, the presentation of chylothorax in the right hemithorax coincided with the development of a mild pericardial effusion.

Chylothorax can also occur after obstruction in the superior vena cava without anatomic injury of lymph channels. Blalock et al. proved experimentally that occlusion of the superior vena cava (SVC) can produce chylothorax.[4,5] Therefore, certain cardiovascular procedures like the Fontan operation are more prone to this complication due to the expected increase in SVC pressure. However, no sign of increased central venous pressure was noted in our group of patients, and only three of the 40 patients (7.5%) who underwent a Fontan procedure during the same period developed postoperative chylothorax, this complication occurring despite a low Fontan pressure (~12 mmHg).

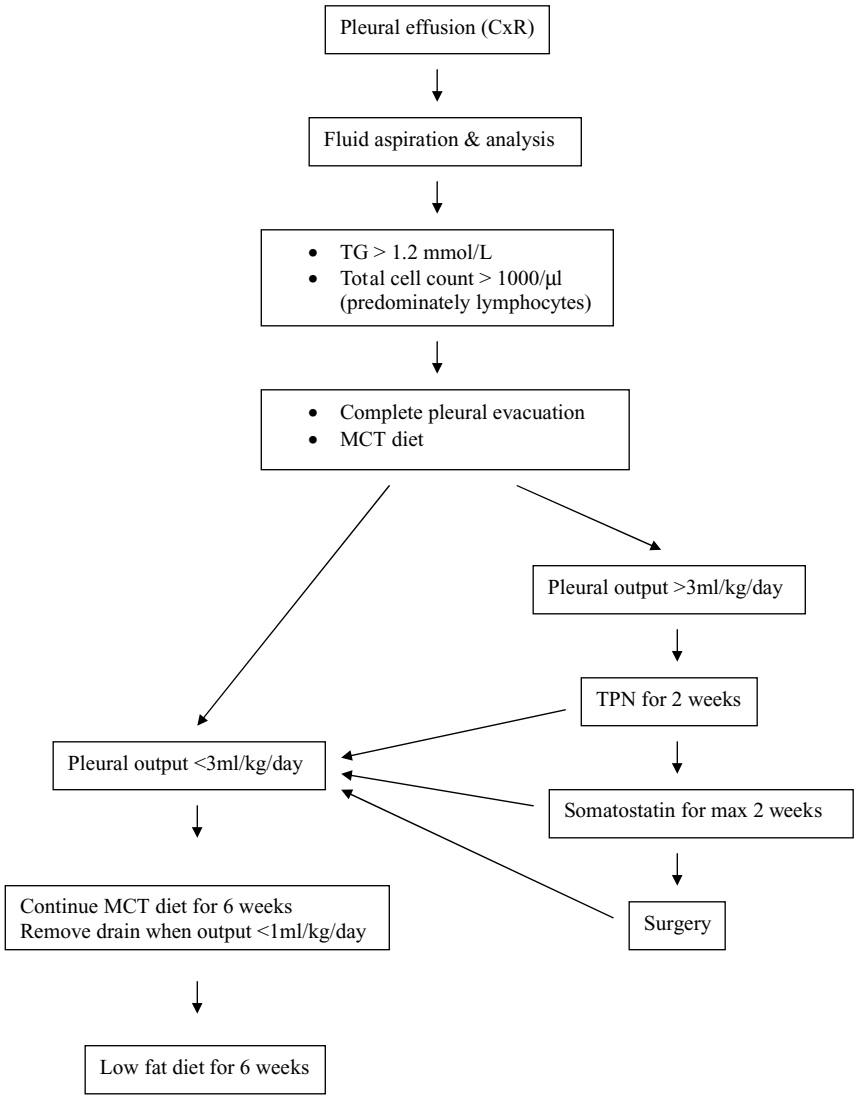

Figure 1. *Protocol for postoperative chylothorax treatment.*

Lymphatic dysplasia with spontaneous chylothorax is also a feature of Noonan and Turner syndromes.[6,7] One of our patients who developed chylothorax after ASD closure had Noonan's syndrome.

The morbidity of chylothorax is potentially severe, as large losses of fluids, proteins, lipids, and white blood cells can cause dehydration, nutritional deficiency, and immunologic dysfunction. Nonetheless, intrapleural infections are rare, possibly due to the bacteriostatic nature of the chylous fluid.[8] Accurate diagnosis and early treatment are important for achieving a favorable outcome and, therefore, the development of a pleural effusion after a thoracic procedure should always raise the suspicion of possible chylothorax. Diagnosis is based on examination of the effusion. Chyle is a lymphatic fluid enriched with fat absorbed by intestinal cells and transported via the thoracic duct into the circulation. The fluid has a milky appearance when chylomicrons are present, while during starvation it appears clear with a light yellow tint. Microscopic and biochemical analyses of the pleural effusion should show triglycerides > 1.2 mmol/L and cell count > 1000 cell/μL with a predominance of lymphocytes.[9,10] Because dietary fat in neonates and infants either consists mainly of triglycerides, or is metabolized to them during digestion, the presence of cholesterol cannot be expected. Therefore, it is best to measure triglyceride levels in a suspicious pleural fluid.[11]

Upon diagnosis, aggressive medical therapy should be commenced immediately.[12-14] Our therapeutic protocol (Fig. 1) involves an initial approach with complete evacuation of pleural effusion to obtain good lung expansion. In addition, no or low-fat enteral diet is established. It has been shown that long-chain fatty acids of 12 or more carbon atoms undergo a second esterification and enter the lymph as chylomicrons, whereas medium-chain fatty acids pass directly into the portal system coupled to albumin.[15] This has led to the adoption of a medium-chain triglyceride diet as nutritional support for patients with chylothorax. When oral

diet is not successful, oral intake is forbidden and total parenteral nutrition (TPN) infusion instituted. This conservative approach was effective in 15 (83.3%) of our patients, although a lengthy hospitalization was necessary. The time limit for conservative treatment is three to four weeks as most of our patients responded favorably to medical therapy within that time frame. Conservative treatment should not be thought to be free of complications. Parenteral nutrition itself may engender several problems related to infection, thrombosis, or cholestasis.[16] However, we have not encountered such problems in our small series of patients.

There is scarce clinical evidence that somatostatin may be helpful in affecting the rate and amount of thoracic chyle leak.[17] Somatostatin reduces gastric, pancreatic, and intestinal secretion.[18-20] The inhibition of serotonin and other gastrointestinal peptides also appears to reduce intestinal absorption and decrease hepatic venous pressure gradient and splachnic blood flow. This influence on the hemodynamics of splachnic circulation (increased splachnic arteriolar resistance) and intestinal motility (decreased gastrointestinal blood flow) may be reflected in a reduction of chyle output.[21] In six of our patients (33.3%), a somatostatin analogue was added as an adjunct to the initial conservative treatment and was administered as a continuous intravenous infusion in gradually increasing dosage for a maximum period of two weeks. In our limited experience, this appeared to be the maximum period of time during which the drug may have exhibited its effect. The latter was demonstrated by a gradual decrease in chest tube drainage once started, coming to a full stop after a maximum of 14 days in the two patients in whom this approach was successful. However, since only two (33.3%) of somatostatin-treated patients responded, and furthermore since reduction in chyle flow in those patients was gradual (and therefore possibly could have been achieved even without somatostatin), the effectiveness of this modality remains dubious.

Tetracycline has been used successfully for chemical pleurodesis in patients with chylothorax,[22] but this drug is no longer available as a solution. However, doxycyclin pleurodesis used in some of the cases as an alternative to tetracycline, had no effect whatsoever on the amount and/or the drainage rate.

If conservative therapy fails to control the leak, surgical intervention is advocated, although specific and precise criteria for abandoning conservative therapy are neither standardized nor generally accepted. It is accepted, however, that if, despite conservative therapy, lymphatic leakage persists beyond a few weeks or if massive fluid and nutritional losses present danger to the patient, precluding thus prolongation of medical treatment, operative intervention is indicated. Lampson reported the first successful ligation of the thoracic duct for chylothorax in 1947.[8] Operations for postoperative chylothorax include one or a combination of several possibilities including direct closure of leaking lymphatics, ligation of the main thoracic duct at the hiatus, and/or mechanical pleurodesis. Several disadvantages of the supradiaphragmatic thoracic duct ligation via right thoracotomy, including variations in ductal anatomy, often lead to failure of this approach, resulting in continuous chylous effusion with the associated morbidity.[2] Our preferred surgical strategy is to perform a thoracotomy on the affected side aiming to identify and control the specific lymphatics involved, if possible with the aid of preoperative cream administration. In addition, abrasion pleurodesis completes the procedure. This approach was successful in the three patients, one of who had a lymphatic leak associated with an operation around the descending thoracic aorta (PDA ligation).

Our data confirm that postoperative chylothorax following surgery for congenital heart surgery is rare and in some instances may be connected to chylopericardium. Nonoperative management can be successful in the majority of cases.[23] Somatostatin may play an adjunctive role in some patients, while doxycycline chemical pleurodesis seems entirely ineffective. Surgical intervention can be reserved for the minority of patients who fail to respond to medical treatment and it is successful. Overall appropriate therapy of this complication may be lengthy but can prevent significant morbidity and mortality.

REFERENCES

1. Le Coultre C, Oberhansli I, et al: Postoperative chylothorax in children: Differences between vascular and traumatic origin. J Pediatr Surg 1991;26:519-523.
2. Bond SJ, Guzzetta PC, Snyder ML, et al: Management of pediatric postoperative chylothorax. Ann Thorac Surg 1993;56:469-473.
3. Campbell RM, Benson LN, et al: Chylopericardium after cardiac operations in children. Ann Thorac Surg 2001;72:193-196.
4. Blalock A, Cunningham RS, Robinson CS: Experimental production of chylothorax by occlusion of the superior vena cava. Ann Surg 1936;104:359-364.
5. Szabo G, Magyar Z: Effect of increased systemic venous pressure on lymph pressure and flow. Am J Physiol 1967;212:1469-1974.
6. Goens MB, Campbell D, Wiggins JW: Spontaneous chylothorax in Noonan syndrome. Treatment with prednisone. Am J Dis Child 1992;146(12):1453-1456.
7. Bottner F, Sandmann C, Semik M, et al: Chylothorax after surgery for thoracic deformity in Noonan syndrome. Orthopedics 2005;28(1):71-73.
8. Lampson LS: Traumatic chylothorax. A review of the literature and report of a case treated by mediastinal ligation of the thoracic duct. J Thorac Surg 1948;17:778-781.
9. Staats BA, Ellefson RD, Budahn LL, et al: The lipoprotein profile of chylous and nonchylous pleural effusions. Mayo Clin Proc 1980;55:700-704.
10. Straaten van HL, Gerards LJ, Krediet TG: Chylothorax in the neonatal period. Eur J Pediatr 1993;152:2-5.
11. Büttiker V, Fanconi S, Burger R: Chylothorax in children—Guidelines for diagnosis and management. Chest 1999;116:682-687.
12. Robinson CL: The management of chylothorax. Ann Thorac Surg 1985;39:90-95.
13. Milsom JW, Kron IL, Rheuban KS, et al: Chylothorax: an assessment of current surgical management. J Thorac Cardiovasc Surg 1985;89:221-227.
14. Marts BC, Naunheim KS, Fiore AC, et al: Conservative versus surgical management of chylothorax. Am J Surg 1992;164:532-535.

15. Frazer AC: Differentiation in the absorption of olive oil and oleic acid in the rat. J Physiol (Lon) 1943;102:306-312.
16. Allen EM, van Heeckeren DW, Spector ML, et al: Management of nutritional and infectious complications of postoperative chylothorax in children. J Pediatr Surg 1991;26:1169-1173.
17. Cheung Y, Leung MP, Yip M: Octreotide for reatment of postoperative chylothorax. J Pediatr 2001;139:157-159.
18. Nakabayashi H, Sagara H, Usukura N, et al: Effect of somatostatin on the flow rate and triglyceride levels of thoracic duct lymph in normal and vagotomized dogs. Diabetes 1981;30:440-445.
19. Bac DJ, Van Hagen PM, Postema PTE, et al: Octreotide for protein-losing enteropathy with intestinal lymphangiectasia. Lancet 1995;345:1639.
20. Grosman I, Simon D: Potential gastrointestinal uses of somatostatin and its synthetic analogue octreotide. Am J Gastroenterol 1990;85:1061-1672.
21. Davis SN, Granner DK: Insulin, oral hypoglycemic agents, and the pharmacology of the endocrine pancreas. In Gilman AC, Hardman JG, Limbird LE, Molionoff PB, Ruddon RW (eds): Goodman and Gilman's The Pharmacological Basis of Therapeutics. 9th ed. Pergamon Press, New York, 1996, pp. 1512-1513.
22. Gil-Suay V, Martnez-Moragon E, de Diego A, et al: Chylothorax complicating a thoracic aortic aneurysm. Eur Respir J 1997;10:737-739
23. Jing-Ming Wu, Chih-Ta Yao, et al: Postoperative Chylothorax: Differences between patients who received median sternotomy or lateral thoracotomy for congenital heart disease. J Card Surg 2006;21:249-253.