Management of Postoperative Chylothorax after Paediatric Cardiac Surgery

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Chylothorax refers to the accumulation of lymphatic fluid in the pleural space. Its presence in the neonatal and pediatric population has been associated with numerous conditions, mainly chest trauma, thoracic surgery, extracorporeal membrane oxygenation and primary or metastatic malignancy, particularly lymphoma.

chylothorax congenital heart disease breast milk

1. Introduction

1.1. Definition and Epidemiology

Chylothorax refers to the accumulation of lymphatic fluid in the pleural space. Its presence in the neonatal and pediatric population has been associated with numerous conditions, mainly chest trauma, thoracic surgery, extracorporeal membrane oxygenation and primary or metastatic malignancy, particularly lymphoma. In newborns rapidly increased venous pressure during delivery may lead to thoracic duct rupture. Less common causes include thrombosis of the duct, superior vena cava or subclavian vein, lymphangiomatosis, restrictive pulmonary diseases, tuberculosis, histoplasmosis and congenital anomalies of the lymphatic system [1][2].

Postoperative chylothorax is a known complication after pediatric cardiac surgery for congenital heart disease (CHD), and its incidence increased consistently over the last decades, a finding that could be reflecting the increased complexity of surgical procedures performed. It has been speculated though that augmented awareness of this complication has led to increased diagnosis rather than a true increase in incidence [3][4][5]. According to reports from the Pediatric Cardiac Critical Care Consortium (PC4) and Pediatric Health Information System (PHIS) databases [5][6], the overall incidence of chylothorax in pediatric patients following congenital heart surgery or heart transplantation ranges between 2.8–3.8%, with higher rates being observed in neonates (6.9%) and patients with single ventricle physiology (6.9%), chromosomal/genetic anomalies (5.2%) and major noncardiac anomalies (6.4%).

Thrombotic events related to upper extremity central venous catheters (CVC) (OR 10.63; 95% CI 4.24–26.64 when compared with no upper extremity CVC group) and neonatal population (OR 4.7; 95% CI 2.7–8.1 when compared with children > 1 year old) yielded the highest OR in a multivariate adjusted model evaluating risk factors for chylothorax development after cardiac surgery in children [6].

Several mechanisms are implicated in the development of postoperative chylothorax after CHD surgery, such as damage to the thoracic duct or collateral lymphatics during dissection or superior vena cava cannulation and increased systemic venous pressures associated with cavopulmonary anastomosis (Glenn shunt or Fontan operation) or right ventricular dysfunction. Further risk factors include the type and complexity of the surgical procedure performed as well as the duration of both the cardiopulmonary bypass and aortic cross-clamp [7][8].

With respect to the type of surgery, atriopulmonary and cavopulmonary anastomoses have been associated with the highest incidence of postoperative chylothorax (5.7%), followed by operations related to the complete correction of transposed great arteries (4.3%), the repair of aortic arch anomalies (3.7%), such as coarctation of the aorta and interrupted aortic arch and the complete repair of total anomalous pulmonary venous drainage (3.7%), while repair of atrial septal defect is associated with lower incidence of chylothorax (0.9%) [5].

2. Diagnosis of Postoperative Chylothorax

Diagnosis of chylothorax is usually based on pleural fluid analysis, which is typically characterized by a white blood cell count > 1000 cells/μL with >70–80% lymphocytes and, in enterally fed patients, a triglyceride concentration > 110 mg/dL. A pleural fluid triglyceride level greater than serum triglyceride level is also characteristic of chylous effusions. Chylous fluid is classically being reported as milky or turbid. Nevertheless, this can be misleading, especially in fasted patients.

Chylothorax in the postoperative period is usually suspected and thus should be checked for when chest tube drainage either becomes persistent or develops the characteristic milky appearance, especially after the patient has been fed, as well as when a similar appearing fluid is found after draining a newly discovered effusion [9][10][11][12]. Of note, researchers mention that there are no specified diagnostic criteria that need to be met before applying a therapeutic protocol for chylothorax management [2][3][4][13]. Qualitative methods for diagnosing chylothorax, such as the microscopic examination of fluid stained with Sudan III showing fat globules, have been described in the literature as well, adding further complexity and confusion with regard to chylothorax definition [14].

For diagnostic purposes, researchers advocate the use of an algorithm, such the one proposed by Skouras et al. [15], which is based on pleural fluid analysis and, if needed, lipoprotein electrophoresis. Pleural fluid triglyceride levels > 110 mg/dL or <50 mg/dL establish or exclude the diagnosis of chylothorax, respectively, for most instances [15], while the presence of chylomicrons in pleural fluid as demonstrated by lipoprotein electrophoresis, which is considered to be the gold standard in the diagnosis of chylothorax [16], clarifies the field in ambiguous cases, such as those with pleural fluid triglyceride levels 50–110 mg/dL or those involving fasted (or malnourished) patients. The proposed algorithm for the diagnosis of postoperative chylothorax is presented by Skouras et al [15].

3. Treatment Goals and Current Therapeutic Approaches in the Management of Postoperative Chylothorax in Children after Surgery for CHD

Chyle constitutes of cellular components with lymphocytic predominance, lipids, electrolytes, glucose, bicarbonate and proteins, such as albumin, clotting factors, complementary elements, antibodies and peptide hormones. Excessive and prolonged chylous losses can result in homeostatic disorders, protein energy malnutrition, clotting and immunological abnormalities, rendering these patients susceptible to significant risks, including, but not limited to, hypovolemia, hypoproteinemia, thrombi formation and systematic infections [2][3]. Chylothorax after pediatric cardiac surgery has been associated with an increased duration of postoperative mechanical ventilation, intensive care unit (ICU) and hospital lengths of stay, cost and mortality [5][6].

The main treatment goals for chylous effusions are to decrease and, eventually, cease the thoracic lymph flow and allow for the lymphatic vessels to heal or develop [2][3]. Conservative nutritional management strategies include either a combination of nil-per-os (NPO) and total parenteral nutrition (TPN), or enteral feeding with MCT diet, fat-modified breast milk or low-fat diet. While conventional fats are absorbed via lymphatic system and require chylomicron formation, medium-chain triglycerides (MCT) are absorbed by intestinal cells and directly transported into the portal system without chylomicron formation [17]. An MCT-based diet, from a pathophysiologic perspective, potentially could reduce lymphatic flow along the thoracic duct and, thus, minimize chyle output drainage.

Pharmacological agents, such as octreotide, can be added as adjunctive therapy to reduce the lymphatic flow. Additional medical management including, but not limited to, respiratory support, the optimization of cardiovascular performance, immunoglobulin and clotting factor supplementation and correction for electrolyte and bicarbonate deficiencies, is offered mainly on a case-by-case basis guided by institutional or expert opinions, as evidence-based treatment choices for post-operative chylothorax in children and older children are lacking [2][4].

Despite generally accepted medical and, under certain circumstances, interventional or surgical management for postoperative chylothorax in neonates and infants following cardiovascular surgery, there is a lack of consensus regarding both a definition that would lead to a change in patient management and the output classification of chylous effusions as low and high as well as the optimum care of these patient population [4]. Nutritional support management strategies vary depending on center and physician experience and/or preference.

While most studies suggest enteral nutrition with MCT for low drainage output chylothorax after cardiovascular surgery in children [3][4][14][18][19], significant variance of practice exists regarding nutritional management of high-output chylous effusions. Several authors suggest NPO with bowel rest and TPN initially [2][13][14][18], while others propose enteral feeding with MCT [3][4][19][20]. Of note, there is significant inconsistency among studies regarding the duration of enteral feeding in high-output states.

Most authors agree though that it is reasonable to switch from an MCT enteral diet to TPN for unresolved high output chylous drainage (i.e., >10 mL/kg/day), paying special attention to excluding raised central venous pressures or central venous thrombosis. Clinicians should not underestimate the potential complications related to TPN administration, such as venous thrombosis and sepsis, as well as the protective function of enteral feeding on the gut barrier through luminal nutrient provision [3][4][19].

Octreotide (or somatostatin) can be added as a pharmacologic adjunct in the management of young infants with postoperative chylothorax. Its use has been described in several published institutional protocols, with most authors suggesting its utilization as a second line treatment [2][3][13][21][22][23]. Mixed data regarding octreotide's effect on hospital length of stay and chylothorax duration have been reported in the literature, which could be attributed, at least partially, to the differences of local protocols, namely the time interval between the onset of chylothorax and the commencement of octreotide infusion [24]. Nevertheless, although the use of octreotide is mainly based on reports from children with congenital chylothorax, representing a different entity from postoperative chyle leak [4], it can be useful in reducing the volume of chylous effusions postoperatively after failure of conservative approach, with its use accompanied by an acceptable safety profile [22][25].

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