Etiological factors and treatment of chylothorax in paediatric patients - a systematic review

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Abstract

Chylothorax is an accumulation of chyle in the pleural cavity. It is a relatively rare cause of pleural fluid triglyceride level greater than 110 mg/dl with a cholesterol level lower than 200 mg/dl confirms the diagnosis of chylothorax. Medical imaging are also necessary such as a non-invasive and easily accessible lung ultrasound. Symptoms of this disease are tachypnea, dyspnea, and in some cases dry cough. This review aims to summarize the current literature regarding chylothorax in children, analyze its possible etiologies and treatments. The causes of chylothorax are varied. It may appear after surgical interventions, traumas, infections and also be congenital. Iatrogenic factors are the most common cause of chylothorax in children with cardiothoracic surgeries. Management of chylothorax can be quite complex and highly variable, depending on patient’s condition and their response to the introduced treatment. Conservative treatment consisting of nutrition therapy, chest drain, and pharmacotherapy is typically a first-line of treatment. Diet modification consist in dietary...
supplements enriched with medium chain triglycerides (MCT) or starting the patient on a total parenteral nutrition (TPN). In most of the analyzed cases the conservative treatment alone proved sufficient in the management of chylothorax. In case of its failure, surgical treatment was a secondary therapy choice. One of the most common surgical procedures for pleural effusion is a thoracic duct ligation (TDL) or pleurodesis and both of these methods are highly effective therapy for chylothorax.

This review of the literature reveals a wide variety of causes and methods of treatment of chylothorax. There are no clear standards of management and the therapy is adjusted to the clinical condition of the patient.

Key words: chylothorax; paediatrics; thoracic duct ligation; low fat diet

1. Introduction

Chylothorax, defined as an accumulation of chyle in the pleural cavity, is a relatively rare cause of pleural effusion in children. The annual incidence of chylothorax is 14 cases per 100,000 children in Europe. Causes of chylothorax can be acquired or congenital. Lymphatic fluid relatively often results from damage to thoracic duct or a large lymphatic vessel. It is more common to accumulate on the right side of the chest. [1-3]

The diagnosis of pleural effusion begins with imaging tests. Ultrasound is the most commonly performed test because of its high-accessibility and non-invasiveness. However, an X-ray or a chest CT can also be ordered. In order to identify the detected fluid as chyle, some laboratory testing of the collected fluid is necessary. The pleural fluid triglyceride level greater than 110 mg/dl confirms the diagnosis of chylothorax. If the value is lower, detection of chylomicrons is necessary to confirm the diagnosis. Chylothorax must be distinguish from pseudochylothorax, which unlike chylothorax does not contain high concentrations of triglycerides or chylomicrons, but contains high concentrations of cholesterol [4].

During a patient interview, chylothorax manifests itself with tachypnea, dyspnea, chest pain and in some cases severe cough. The symptoms of fluid in the pleural cavity, such as diminished breath sounds or a decrease in the intensity of vocal fremitus can be detected during the physical exam [5]. Congenital chylothorax can manifest with general swelling, symptoms of respiratory failure or heart failure [4]. Occasionally, it can be detected during prenatal testing [5].

Chylothorax can lead to many, at times life-threatening, complications such as electrolyte imbalances (hyponatremia, hypoproteinemia, hypoalbuminemia), recurrent lower respiratory tract infections or clotting disorders. Malnutrition caused by chylothorax can lead to growth inhibition in children and favors development of opportunistic infections [4].

Considering the above symptoms and complications, which are often life-threatening for the children, it is important to have uniform treatment guidelines. This review aims to systematize the current knowledge about regarding chylothorax in children, as well as to analyze its possible etiologies and treatments.

2. Methods

Current literature was reviewed by searching for publications from the period between January 2015 and March 2021 using a search phrase “chylothorax AND children AND causes” and “chylothorax AND children AND treatment” in Pubmed and Scopus databases.

The inclusion criteria were: patient’s age <18, both sexes of patients and chylothorax as the main condition; types of articles: case studies, case series, retrospective studies, meta-analyses and prospective study. The exclusion criteria were as follows: “online ahead of print” articles, articles without full text access or available in a language other than Polish or English, as well
as articles where the presented data were not broken down by age (combined for both children and adults).

95 articles were included from among 418 reviewed articles. The included articles consisted of 44 case studies, 12 case series, 38 retrospective studies, and 1 prospective study. The collected clinical data dealt with the cases of 1075 children with chylothorax, and focused on its causes or treatment.

Statistical Analysis
The obtained data were analyzed in Statistica 13 Stasoft Software. The results are expressed as counts and percentages as well as the mean and standard deviation (SD) for categorical and continuous variables, respectively.

3. Results
Out of total 1075 patients, 280 (26%) didn’t specify the child sex. Among the remaining respondents, 512 (48%) were male. The patients' age ranged from 22.6 weeks of GA (gestational age) to 18 years, with an average age of 16 months. Among those studied, age of 301 patients is not mentioned. In 97% of children chylothorax was diagnosed postnatally. The mean gestational age at diagnosis was 31.7 weeks. Infants constituted the largest group of patients in which the first symptoms of chylothorax were observed (45%) [2-4], [6-106] Additional demographic information is shown in Table 1.

Table 1. Demographic-clinical data of patients with chylothorax [2-4], [6-106].

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Number of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children (total)</td>
<td>1075 (100%)</td>
</tr>
<tr>
<td>Diagnosis</td>
<td></td>
</tr>
<tr>
<td>Children with the prenatal diagnosis</td>
<td>28 (3%)</td>
</tr>
<tr>
<td>Children with the postnatal diagnosis</td>
<td>1047 (97%)</td>
</tr>
<tr>
<td>Age</td>
<td></td>
</tr>
<tr>
<td>Average age of children</td>
<td>16 months</td>
</tr>
<tr>
<td>Minimum and maximum ages at diagnosis</td>
<td>Min: 22.6 weeks of GA</td>
</tr>
<tr>
<td></td>
<td>Max: 18 years</td>
</tr>
<tr>
<td>Gestational age (GA) at diagnosis (mean, weeks)</td>
<td>31,77</td>
</tr>
<tr>
<td>The patient's age at onset of symptoms</td>
<td></td>
</tr>
<tr>
<td>prenatal</td>
<td>28 (3%)</td>
</tr>
<tr>
<td>&lt;=31 days</td>
<td>63 (6%)</td>
</tr>
<tr>
<td>&gt;=31 days- 12 months</td>
<td>487 (45%)</td>
</tr>
<tr>
<td>&gt;=12 months - &lt;7 years</td>
<td>165 (15%)</td>
</tr>
<tr>
<td>&gt;=7 - 18 years</td>
<td>31 (3%)</td>
</tr>
<tr>
<td>Unknown age</td>
<td>301 (28%)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Males</td>
<td>512 (48%)</td>
</tr>
<tr>
<td>Females</td>
<td>283 (26%)</td>
</tr>
<tr>
<td>Unknown sex</td>
<td>280 (26%)</td>
</tr>
</tbody>
</table>
4. Causes

Causes of chylothorax were known in 88 of the reviewed articles and referred to 1028 children, described based on the table 2.

Causes of chylothorax are extremely diverse. However, the analysis of the available literature shows that iatrogenic factors are the most common cause (70.8%). Among surgical procedures contributing to the development of chylothorax, cardiothoracic surgeries are most prevalent, with predominance of Fontan procedures and congenital heart defect corrective surgeries (61.2%) [2], [3], [6]-[30]. A significant group undergoing these procedures were infants with genetic conditions, such as Noonan syndrome, Down syndrome or Gorham disease. Patients with these genetic syndromes have multiple cardiovascular malformations, bone malformations and lymphatic abnormalities that make surgery difficult and contribute chylothorax. In this review of the literature, this problem appeared in 9 articles [12], [18], [24], [26], [31]-[35]. Chylothorax also occurred post-operatively in patients with congenital diaphragmatic hernia, tracheoesophageal fistula or after other thoracic operations (in 8% of the analyzed cases) [3], [15], [20], [27], [36]-[54]. 12,8% of children developed an effusion diaphragmatic hernia, tracheoesophageal fistula or after thoracic operations (8% of the analyzed cases) [3], [18], [24], [26], [31]-[35]. Nonetheless, the research shows that chylothorax certainly develops earlier in premature babies, patients with genetic conditions, such as Gorham-Stout disease or lymph flow disturbances caused by the damage to lymphatic vessels or anatomical defects. In our analysis, they accounted for 1% of the studied population [31], [68]-[70]. Rare, but important reasons for the development of the previously discussed effusion are: thrombosis (including thrombosis in the course of the nephritic syndrome), cancer (mainly cancers of the lymphatic system, such as: cystic hygroma, Hodgkin lymphoma, and non-Hodgkin lymphoma), intense vomiting and thoracic trauma (in 2,7% of the analyzed cases) [2], [27], [30], [37], [40], [68], [71]–[76]. In 12% of chylothorax cases, the cause could not be determined [3], [30], [34], [39], [77]–[81].

Table 2. The most common causes of chylothorax in children

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>Number of patients (n) total number - 1028</th>
<th>% of total number of patients</th>
<th>REFERENCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iatrogenic/ surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac/cardiothoracic surgery</td>
<td>629</td>
<td>61,2</td>
<td>[2], [3], [6], [7], [9]–[14], [16]–[23], [25]–[30], [59], [68], [96], [106]</td>
</tr>
<tr>
<td>Thoracic surgery</td>
<td>23</td>
<td>2,2</td>
<td>[20], [44]–[54], [68]</td>
</tr>
<tr>
<td>Congenital diaphragmatic hernia (CDH) repair</td>
<td>32</td>
<td>3,1</td>
<td>[3], [36]–[40]</td>
</tr>
<tr>
<td>CDH repair + ECMO treatment</td>
<td>20</td>
<td>2,0</td>
<td>[40]</td>
</tr>
<tr>
<td>Tracheostomy revision</td>
<td>1</td>
<td>0,1</td>
<td>[91]</td>
</tr>
<tr>
<td>Innominate vein occlusion/stenosis (due to central vein/</td>
<td>7</td>
<td>0,7</td>
<td>[92]</td>
</tr>
<tr>
<td>Condition</td>
<td>Incidence</td>
<td>Mortality</td>
<td>References</td>
</tr>
<tr>
<td>--------------------------------------------------------------------------</td>
<td>-----------</td>
<td>-----------</td>
<td>------------</td>
</tr>
<tr>
<td>Oesophageal atresia/tracheoesophageal fistula</td>
<td>5</td>
<td>0,5</td>
<td>[27], [41]–[43], [68]</td>
</tr>
<tr>
<td>Plication of the diaphragm</td>
<td>1</td>
<td>0,1</td>
<td>[68]</td>
</tr>
<tr>
<td>Liver surgery</td>
<td>1</td>
<td>0,1</td>
<td>[3]</td>
</tr>
<tr>
<td>Anoplasty and fistula closure</td>
<td>1</td>
<td>0,1</td>
<td>[43]</td>
</tr>
<tr>
<td>Extracorporeal membrane oxygenation cannulation</td>
<td>1</td>
<td>0,1</td>
<td>[27]</td>
</tr>
<tr>
<td>Minimally invasive surgery of neuroblastic tumors</td>
<td>3</td>
<td>0,3</td>
<td>[98]</td>
</tr>
<tr>
<td>Surgery of peripheral nerve tumors</td>
<td>1</td>
<td>0,1</td>
<td>[99]</td>
</tr>
<tr>
<td>Puncture of the internal jugular or subclavian vein-injury to the thoracic duct</td>
<td>1</td>
<td>0,1</td>
<td>[102]</td>
</tr>
<tr>
<td>Intercostal drainage of pneumothorax</td>
<td>2</td>
<td>0,2</td>
<td>[39]</td>
</tr>
<tr>
<td><strong>Congenital</strong></td>
<td><strong>110</strong></td>
<td><strong>10,7</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Central lymphatic flow disorder</strong></td>
<td>22</td>
<td>2,1</td>
<td>[32], [59], [60]</td>
</tr>
<tr>
<td><strong>Other</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trombosis of SVC</td>
<td>9</td>
<td>0,8</td>
<td>[37], [40], [71]–[73]</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>3</td>
<td>0,3</td>
<td>[2], [73]</td>
</tr>
<tr>
<td>Nephrotic syndrome</td>
<td>2</td>
<td>0,2</td>
<td>[2], [30]</td>
</tr>
<tr>
<td>Generalized lymphatic anomaly (GLA)</td>
<td>6</td>
<td>0,6</td>
<td>[68]</td>
</tr>
<tr>
<td>Gorham- stout disease</td>
<td>2</td>
<td>0,2</td>
<td>[31], [69]</td>
</tr>
<tr>
<td>Diffuse lymphangiomatosis</td>
<td>2</td>
<td>0,2</td>
<td>[68], [70]</td>
</tr>
<tr>
<td>Malignant lymphoma</td>
<td>4</td>
<td>0,3</td>
<td>[30], [73]</td>
</tr>
<tr>
<td>Lymphangioma</td>
<td>1</td>
<td>0,1</td>
<td>[30]</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>3</td>
<td>0,3</td>
<td>[68], [88]</td>
</tr>
<tr>
<td>Esophageal atresia</td>
<td>1</td>
<td>0,1</td>
<td>[37]</td>
</tr>
<tr>
<td>After chylopericardium and unsuccessful thoracic</td>
<td>1</td>
<td>0,1</td>
<td>[97]</td>
</tr>
</tbody>
</table>
5. Conservative treatment

Methods of conservative treatment were presented in 77 articles and referred to 531 children described in the chart 1 [2-3], [7], [11-14], [17], [19-20], [22-23], [25-29], [31-33], [35-45], [49], [51-52], [54-58], [60-64], [66], [70- 81], [84-88], [90-93], [95-103]. The vital part of the effective treatment of chylothorax is the identification of its cause, and the subsequent implementation of the targeted treatment. The analysis of the available literature shows that identifying the etiology of chylothorax is often unsuccessful (12.0 %). However, measures to reduce plural effusion are taken independently of the causal treatment by means of surgery or conservative methods [3], [5].

Most commonly the treatment of chylothorax begins with non-invasive methods, which involve diet modification and pharmacotherapy. The main goal of the nutrition therapy is the reduction in lymph production in order to limit its flow through the thoracic duct, and consequently minimize venous leakage. It is possible to achieve such a result by introducing dietary supplements containing medium chain triglycerides (MCT) or starting the patient on a total parenteral nutrition (TPN) [5], [82], [83]. As much as 33.7 % of children from the reviewed literature was treated with the first of the abovementioned methods [2], [17], [19], [23], [25], [33], [40], [43], [52], [56], [72], [77], [79], [81], [84]-[89]. It is preferred mainly in patients with normal circulatory and respiratory function [5]. MCT diet consists of triglycerides with saturated fatty acid chains of 8 to 12 carbons in length, which are absorbed directly from the portal vein, avoiding lymphatic drainage. Providing an easily accessible energy source is a clear advantage of this method. However, MCT can lead to essential long-chain fatty acid deficiency, as well as changes in glucose metabolism. Moreover, MCT metabolism is carnitine-dependent, thus such diet can be potentially dangerous for premature babies, who do not have sufficient ability for carnitine biosynthesis. This can result in partial oxidation of MCT, and subsequently an increased risk of metabolic acidosis [83]. In case of unsuccessful treatment with MCT or in hemodynamically unstable patients with substantial pleural effusion, a prompt implementation of TNP is preferred. TPN, has been used as a first line dietary modification treatment in 35.2% of patients from the reviewed literature [7], [11], [12], [27], [37], [41], [45], [55], [64], [66], [71], [73], [74], [78], [90]-[93]. TPN allows for a faster reduction in pleural effusion [83]. This method is not without its risks either. They include mucosal atrophy
of the digestive tract, a decrease in hormone secretion in the digestive tract, and liver dysfunction [94]. Less common methods of nutritional treatment include low-fat diet (2.6% patients [32, 38, 44, 49, 54, 57, 61, 92, 95, 96]), completely fat-free diet (5.8% [13, 14, 20, 46, 51, 73, 75, 76, 97, 98]) or in infants – milk with modified fat content (7.5% [3, 23, 84]).

At the same time, authors of many publications do not provide specific diet modifications (10.9%) [22, 26, 29, 31, 42, 60, 66, 70, 73, 80, 86, 99, 100].

The lack of clear guidelines regarding diet modifications poses a problem in developing appropriate treatment plans. Dietary modifications are often used interchangeably or synergistically in case of a failure of one of the treatment methods. One of the more common dietary modifications is implementation of first MCT, and then TPN as a second line of treatment (19.6%) [2, 19, 25, 56, 79, 84]. Occasionally, the authors did not precisely indicate which treatment – MCT or TNP was prescribed [28, 35, 39, 43, 62, 63, 101–103].

In case when the nutritional modification fails, pharmacological treatment using somatostatin and its synthetic analogue – octreotide, is implemented. Both substances act via decreasing the mesenteric and portal venous flow, as well as inhibiting the secretion of gastroenteropancreatic peptides and hormones. This, in turn, leads to a decrease in intestinal motility, reduced nutrient absorption from the intestines, and reduced chyle flow through the lymphatic vessels, in addition to a reduction in chyle formation. Research confirms both their effectiveness and safety, even in newborns [5]. Treatment with somatostatin or octreotide has been implemented in more than half of the patients from the reviewed literature, in addition to dietary modifications (56.3%) and it achieved a desired therapeutic effect in 63.2% of them. [2-3, 11-12, 17, 19-20, 25–28, 31–33, 37, 39, 41–43, 45, 52, 54–57, 62–64, 66, 70-71, 73, 77–80, 84, 86-87, 91-92, 95-96, 98, 103] Due to certain differences in pharmacokinetic properties and route of administration, octreotide is a more popular choice (79.9% of patients) than somatostatin (10.4%) [3,11-12, 17, 19-20, 25–27, 31-33, 37, 39, 41–43, 45, 52, 54–57, 60, 62–64, 66, 70-71, 73, 77-80, 84, 86-87, 91-92, 95-96, 98, 103]. It can be administered both intravenously and subcutaneously, while somatostatin can be administered only intravenously. Additionally, the synthetic analogue has a stronger effect and a longer half-life [5]. Still, some authors do not indicate which pharmaceutical has been administered (9.7%) [2, 28].

In many cases, chylothorax was over-complicated by other clinical issues. In such circumstances, a specific treatment was administered in order to limit the risk of further chylothorax progression and deterioration of patient’s clinical status. Sildenafil, which is a phosphodiesterase type 5 (PDE5) inhibitor, increases the concentration of cGMP in the cells of the smooth muscles of the pulmonary vessels, leading to pulmonary vasodilation, which prevents the development of pulmonary hypertension [17, 26, 55]. In the event of chyle infection in the pleural cavity, antibiotic therapy is initiated [17, 41, 66, 75]. Intravenous steroids are also frequently administered in order to reduce inflammation [19, 25, 32, 62, 72, 79, 80, 84, 87, 103]. Antithrombotic prophylaxis, involving heparin – mainly low molecular weight heparin, enoxaparin [71, 72, 95]. Sirolimus is a less commonly used medication [31, 57, 60, 61]. It is used in case of chylothorax accompanied by lymphangioleiomyomatosis. By inhibiting the mTOR pathway, sildenafil prevents deterioration of lung function in patients with this condition.

One of the elements of the conservative treatment for chylothorax is thoracentesis or chest drain, which allows for a quick removal of fluid from the pleural cavity, but most importantly it allows for a confirmation of the pleural effusion diagnosis. Chest drain is continued in case of a severe chylothorax accompanied by respiratory dysfunction and difficulties with lung expansion. In any case, chest drainage results in a significant depletion in the electrolyte, fluid,
protein, fat, immunoglobulin and lymphocyte content. Such losses are particularly dangerous for newborns, as they significantly disrupt the immune system, and thus promote infection [5]. Despite its many hazards, chest drain is used in a large number of patients (76.5 %) with an average duration of 20.9 days. Even though in many cases conservative treatment is effective and produces the desired outcomes (70.4%), in some patients it is not sufficient (29.6%) and another intervention, e.g. surgical, is necessary.

Chart 1. Conservative treatment of patients with chylothorax.

MCT- medium chain triglycerides, TPN- total parenteral nutrition

6. Surgical treatment
Methods of surgical treatment were presented in 40 articles and referred to 287 children described in the chart 2 [3], [7], [9-10], [12], [17], [19-20], [26-29], [31-32], [35], [37], [41-42], [50], [52], [54], [56-60], [62], [64], [66], [69-71], [78], [80], [87], [91-93], [95-97]. Surgical treatment needs to be considered if the conservative treatment has failed. Unfortunately, a uniform standard for such treatment has not yet been devised in case of chylothorax in children.

One of the basic surgical procedures for pleural effusion is a thoracic duct ligation (TDL). The site for TDL is typically selected between thoracic vertebrae eight and twelve, just above the aortic hiatus. TDL can be performed in two ways, but a thoracoscopic access provides a safe and effective treatment, while avoiding problems related to a major surgical procedure, such as an open thoracotomy [105]. This procedure was the most common one among the reviewed publications, used in 44.25% of the patients, in 65.35% it proved effective and did not require any further interventions [3], [9-10], [12] [19], [26–28], [35], [50], [52], [57], [66], [69], [80], [91-92]. However, in 22.05% of patients treated with TDL, pleurodesis was performed additionally [26], [27], [35], [92]. Pleurodesis involves injection of an obliterating agent into the pleural cavity, producing inflammation. The fibrotic tissue forming between the visceral and parietal pleurae eventually prevents fluid accumulation in the closed pleural space [5]. Pleurodesis was used as a stand-alone therapeutic procedure in 21.25% of patients and 90.16% of them were cured using this method [3], [17], [37], [41], [42], [52], [56], [58], [78], [93].
Among the less common procedures performed together with TDL are: embolization using fibrin glue, coils or lipiodol, diaphragmatic fenestration, thermal ablation of the pleura, angioplasty with stent placement, balloon angioplasty, catheter-directed thrombolysis, and pleuroperitoneal shunting (27.56%) [26–28], [69].

Another common, minimally invasive procedure is a thoracic duct embolization (TDE). It is a 3-stage procedure consisting of: lymphangiography, percutaneous thoracic duct cannulation, and TD embolization proximally to the leak. Lymphangiography allows to locate the site of the chyle leak, in addition to visualizing the anatomical variant of TD configuration [5]. According to this literature review, TDE has been performed in 23% of patients [3], [7], [20], [26], [27], [32], [54], [59], [96], [97]. This procedure is particularly effective in post-operative, post-traumatic chylothorax, hence its consideration as a first-line treatment is recommended [26]. Among the reviewed publications, polyglycolic acid sheets and fibrin glue were the most common agents used for TDE (33.33%) [27], [54]. Less popular agents were: lipiodol (27.27%) glue (15.15%) coils with glue (10.60%) and in 13.64% of cases, an agent used for TDE was not specified [3], [7], [20], [26], [32], [59-60], [96-97].

This literature review has shown a high efficacy of surgical treatment (87.11%). Only 8.01% of patients were resistant to the initial treatment and required additional medical interventions, and in 4.88% of patients, the treatments for chylothorax were unsuccessful, resulting in patient’s death.

Chart 2. Surgical treatment of patients with chylothorax

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>TDL</td>
<td>44.25%</td>
</tr>
<tr>
<td>TDE</td>
<td>23%</td>
</tr>
<tr>
<td>Pleurodesis</td>
<td>21.25%</td>
</tr>
<tr>
<td>Other</td>
<td>11.5%</td>
</tr>
</tbody>
</table>

TDL- thoracic duct ligation, TDE- thoracic duct embolization

7. Conclusions

The analysis of the gathered literature allows to conclude that iatrogenic factors are the most common cause of chylothorax in children, with cardiothoracic surgeries constituting a large portion. Congenital chylothorax, which can arise during fetal development, is also relatively common.

Pleural effusion most commonly results from infection, heart failure or cancer. Chylothorax most often occurs as a result of a chest trauma or due to a rupture of the thoracic duct during a cardiothoracic surgery. Chylothorax can also be diagnosed during fetal development or in newborns, in which case it is known as congenital chylothorax. Children with genetic conditions, such as trisomy 21, Noonan syndrome or Turner syndrome are more prone to chylothorax due to a disrupted lymph flow, resulting from an impaired development of the lymphatic vessels or their hypoplasia.

Chylothorax therapy typically begins with conservative treatment consisting of nutrition therapy, chest drain, and pharmacotherapy. Diet modification often consists in an
implementation of dietary supplements containing MCT as a first-line of treatment. If this treatment fails or if a child cannot be fed enterally, TPN is introduced.

Chest drain is a standard procedure in case of chylothorax diagnosis, even though it poses a risk of side effects.

Aside from the abovementioned interventions, pharmacotherapy has been implemented in the treatment of more than half of the patients. Among the analyzed cases, octreotide was chosen much more frequently than somatostatin, because of its high efficacy and proven safety in children. Additionally, intravenous glucocorticoids or antibiotics were implemented in order to prevent complications or minimize their possible effects.

In most of the analyzed cases the conservative treatment alone proved sufficient in the management of chylothorax. In case of its failure, surgical treatment was a secondary therapy choice. TDL was the most common surgical procedure that produced the desired outcome in most children, while pleurodesis proved highly successful both as an adjuvant therapy and a stand-alone treatment for chylothorax.

This review demonstrates that the management of chylothorax can be quite complex and highly variable, depending on patient’s condition and their response to the previous treatments. Highly effective and uniform treatment plans are difficult to devise, so all therapies should primarily target the original cause of chylothorax.

This literature review regarding chylothorax is based solely on retrospective and case studies, which is its main limitation. Moreover, in some instances, the authors do not provide detailed information regarding children’s age or comorbidities, and group the used pharmaceuticals and treatment methods together, making the therapies harder to compare. It is necessary to carry out the prospective studies in order to determine the effectiveness of the individual treatment methods.

References


