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CME Review

Chylothorax: Diagnosis and Management in Children

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EDUCATIONAL AIMS

THE READER WILL FEEL CONFIDENT TO:

- Discuss the anatomy and physiology of the lymphatic system and its relevance when assessing a patient with chylothorax.
- Describe a practical approach for diagnosing chylous effusion in children.
- List the common causes of chylothorax
- Discuss and illustrate the different imaging techniques for the evaluation of chylothorax.
- Explain the principles of treatment and describe the treatment modalities of chylothorax in children.

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SUMMARY

Chylothorax is the accumulation of chyle in the pleural space, as a result of damage to the thoracic duct. Chyle is milky fluid enriched with fat secreted from the intestinal cells and lymphatic fluid. Chylothorax in children, is most commonly seen as a complication of cardiothoracic surgery but may occur in newborns or conditions associated with abnormal lymphatics. The diagnosis is based on biochemical analysis of the pleural fluid, which contains chylomicrons, high levels of triglycerides and lymphocytes. Investigations to outline the lymphatic channels can prove helpful in some cases. Initial treatment consists of drainage, dietary modifications, total parenteral nutrition and time for the thoracic duct to heal. Somatostatin and its analogue octreotide may be useful in some cases. Surgery should be considered for patients who fail these initial steps, or in whom complications such as electrolyte and fluid imbalance, malnutrition or immunodeficiency persist. Surgical intervention may be attempted thoracoscopically with repair or ligation of the thoracic duct.

INTRODUCTION

Chylothorax is a relatively rare cause of pleural effusion in children. It is defined by the accumulation of chyle in the pleural space that occurs as a result of damage to the thoracic duct by rupture, laceration, tear or compression.[1–4] Chyle is a milky coloured fluid enriched with emulsified fat (chylomicrons) absorbed by the intestinal cells and transported by lymphatic channels that converge to become the thoracic duct and pass into the circulation. The diagnosis of chylothorax depends on analysis of the pleural fluid, with identification of chylomicrons or high triglyceride concentrations. The incidence of chylothorax in children is unknown; however, it is predominantly seen as a complication following cardiothoracic surgery and occasionally in newborns. Other causes include congenital malformations of the pulmonary or thoracic lymphatic system, major neck surgery, superior vena cava obstruction, pleural or mediastinal malignancies and dysmorphic syndromes such as Turner's or Noonan's Syndrome. Chylothorax is a potentially life-threatening disorder that can lead to serious metabolic, immunologic and nutritional complications. Initial management is the same regardless of the cause of chylothorax. Low fat diet, total parenteral nutrition (TPN) and increasingly, surgical procedures have been described for management.

The aim of this article is to outline the causes of chylothorax in children, present a paradigm for investigations and describe the various management options.

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ANATOMY OF THE THORACIC LYMPHATIC SYSTEM

Knowledge of the anatomy of the thoracic lymphatic system is important in the assessment and management of a patient with chylothorax. The thoracic duct is formed by coalescing intestinal and lumbar lymphatics at the level of the cisterna chyli, a triangular dilatation anterior to the body of the second lumbar vertebra, behind the aorta. It enters the thorax through the aortic hiatus of the diaphragm between thoracic vertebrae 10 to 12 (T10 to T12). In the thorax, it ascends through the posterior mediastinum anterior to the vertebra on the right side, behind the oesophagus and pericardium (it is separated from the pericardium by a recess of the right pleural cavity). At the level of T5 to T6, the thoracic duct crosses to the left side to enter the superior mediastinum, ascending behind the aortic arch. The duct then exits the thorax through the superior thoracic aperture passing into the neck where it forms an arch that is anterior to the scalenus muscle, a few centimetres above the left clavicle (level of C7). After turning laterally, it enters the circulation at the junction of the left internal jugular and left subclavian veins.

Anatomic variations of the thoracic duct are common. Duplication, triplication or other anatomical variations are present in nearly 35% to 50% of the population.[5–8] There is also an extensive lymphatic network of collaterals. This richness of collaterals is such that the thoracic duct can be ligated at any point during its thoracic or cervical course without stasis.[7]

The thoracic duct drains lymph from the lower limbs, abdomen, intestinal chyle and left thorax, head, neck and upper limb. The lymphatic flow from the right side of the head, neck and thorax, right upper limb, right side of the heart and the convex surface of liver, is drained by the right lymphatic duct and does not contain chyle.

The clinical relevance of knowing the thoracic duct anatomy is to ascertain the level of disruption and possible aetiologies. Rupture of the thoracic duct between the diaphragm and T5 usually produces a right-sided chylothorax, and above T5, a left-sided chylothorax.[7,9] When bilateral chylothoraces occur, damage of the duct is usually located where it passes the mid-line at the level of T5 or part of a more diffuse lymphatic condition.[10] Rupture of the lymphatics in the abdomen causes a chylous ascitis, which is a leakage of chyle into the peritoneal cavity. This condition will not be reviewed in this paper.

The thoracic duct has numerous valves forcing chyle flow proximally. Flow is dependent on the inflow of food (especially fat) and fluid into the intestine. The forward flow is helped by intermittent compression of the cisterna chyli during breathing, and by increase intra-abdominal pressure on inspiration.[7]

PHYSIOLOGY

The lymphatic system has three primary functions. Firstly, it transports the lipids and lipid-soluble vitamins (A, D, E and K) absorbed by the lacteals, lymphatic capillaries in the gastro-intestinal tract (small intestine) to the systemic circulation. Secondly, it collects the excess of fluid from the interstitial spaces along with extravasated proteins that cannot be absorbed directly into the blood capillaries and returns them to the circulation. Thirdly, it constitutes an essential part of the immune system, in particular, returning lymphocytes to the circulation.

The thoracic duct is the main vessel for the transport of chyle and other nutrients from the intestine to the circulation. It normally transports between 1.5 to 2.5 L daily; however it can transport up to 4L of chyle per day in an otherwise healthy adult. Chyle is a non-inflammatory, alkaline and bacteriostatic fluid that is composed mainly of fat, cholesterol, electrolytes, proteins, glucose and abundant lymphocytes (Table 1). Almost all the chyle is delivered from the intestinal lacteal system.[11] Thoracic duct flow varies depending on the diet, medications, intestinal function and physical activity. Flow through the thoracic duct can increase by two to ten fold for 2 to 3 hours after ingestion of fat and by 20% after drinking water. At the same time, other factors that increase the interstitial fluid pressure will also increase the lymph flow.

Knowledge of lipid metabolism is required to understand the relationship between diet, fat content and thoracic duct flow. Normal dietary fat consists mainly of triglycerides with different fatty acid chain lengths, and each are metabolized differently. Medium-chain triglycerides (MCT) contain 6 to 12 carbon fatty acids, and are absorbed from the gastrointestinal tract directly to the portal circulation and transported to the liver for metabolism.[12–14] Long-chain triglycerides (LCT) contain >12 carbon fatty acids, constitute up to 95% of the total triglycerides in the diet and are water-insoluble. Therefore, they require transformation in the form of lipoproteins (chylomicrons) to be transported in the blood.[15,16] Chylomicrons are macromolecules rich in LCT, cholesterol, phospholipids and proteins. They are extruded from the intestinal mucosal cells into lacteals, and from there to the lymphatic system through the thoracic duct into the venous system. This explains its milky appearance, and allows a simple bedside test for chylothorax. During fasting, chyle is usually clear owing to a low fat content and low protein concentration.

DIAGNOSIS

Chylothorax should be suspected when there is extensive pleural effusion occurring in a neonate, after cardiothoracic or mediastinal surgery, in patients with a mediastinal mass or in the presence of major lymphatic malformations (see Fig. 1). The diagnosis of chylothorax is confirmed by examination of the pleural fluid (see Table 1). Thoracocentesis is a simple and safe method of obtaining fluid that will reveal the presence of chyle.

The pleural fluid from a chylothorax is typically milky and does not clot. However, it may not be milky in a fasting patient or in postoperative patients with reduced dietary fat intake.[3] A milky or turbid appearance of the fluid may also be seen with an empyema or when there is a chronic pleural effusion with high content of cholesterol but no triglycerides or chylomicrons (pseudochylothorax). Therefore, chylothorax must be diagnosed by biochemical analysis of the fluid. The key finding is the presence of chylomicrons, which can be seen after staining with Sudan III.[17,18] This stain requires special cytological preparation of the pleural fluid that may not be freely available. Triglyceride concentration of

Table 1
Characteristics and biochemistry of chyle

Component/Feature	
pH	7.4 – 7.8
Colour	Milky (clear in starvation)
Sterile	Yes
Bacteriostatic	Yes
Total fat	0.4 – 6 g/dl
Cholesterol	65 – 220 mg/dl
Triglycerides	> 1.1 mmol/L (>110 mg/dl).
Total Protein	2 – 6 g/dl
Albumin	1.2 – 4.1g/dl
Globulin	1.1 – 3.1 g/dl
Electrolytes	Similar to plasma
Glucose	2.7 – 11 mmol/L
Cellularity	
Absolute cell count	> 1,000 cell/L
Lymphocytes	> 80%
Erythrocytes	50 – 600/mm^3
Chylomicrons	Yes

Adapted from Straaten et al. (1993)[20], Buttiker et al. (1999)[3] and Agrawal et al. (2008)[19].

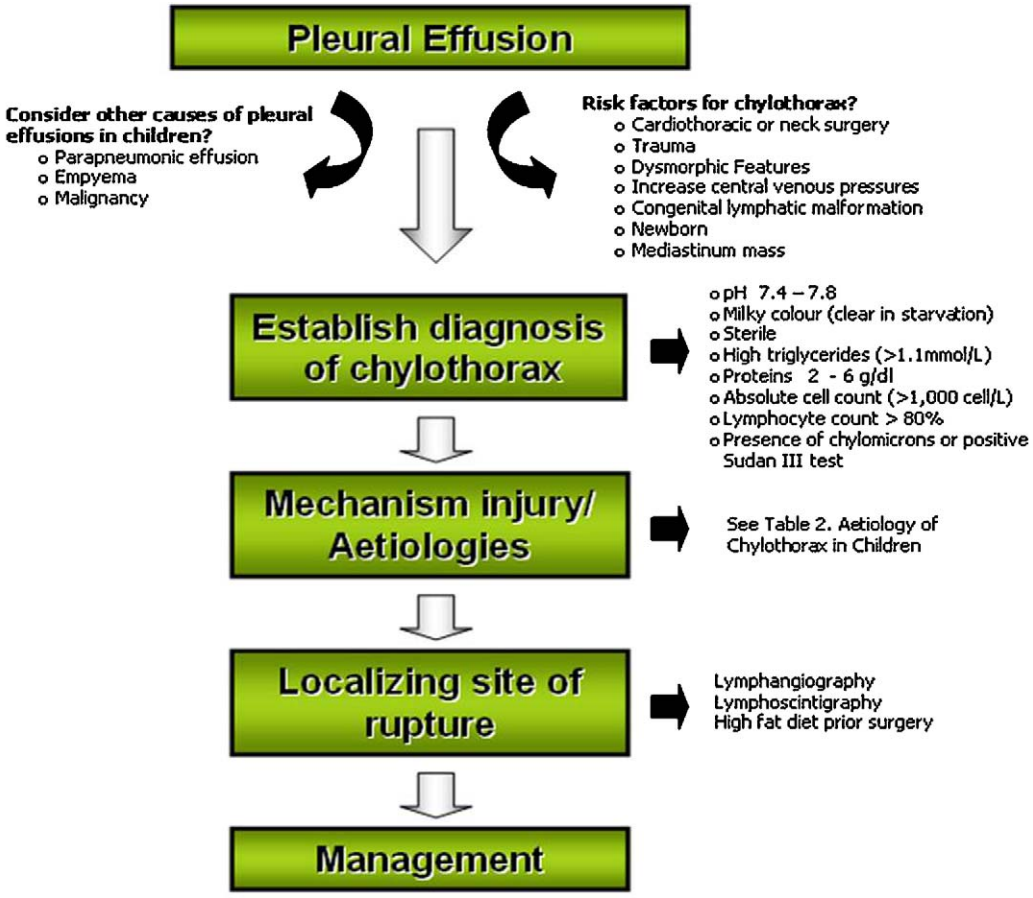

Figure 1. Algorithm for the Assessment and Management of Chylothorax in Children.

the fluid is a simpler method but only positive if the concentration is above 1.1 mmol/L (concentrations between 0.56 and 1.1 mmol/L are equivocal).[3,17–19] If there is doubt about the diagnosis, administration of a high fat meal by mouth or via a nasogastric tube will result in a dramatic change in the colour, triglyceride and chylomicron content of the pleural effusion, confirming the presence of chyle leak.

Other characteristics of the pleural fluid from chyle leak are the abundance of T lymphocytes, along with products that are usually transported by lymphatics such as proteins (immunoglobulins, clotting factors), vitamins, electrolytes and other products of digestion. Buttiker and colleagues (1999)[3], analysed 39 children with chylothorax, in 36 (92%) the total cell count was >1000 cell/μL and >90% of these were lymphocytes.[3] Lymphocyte count of the pleural fluid could be a useful marker of chylothorax when the diagnosis is uncertain.

AETIOLOGY OF CHYLOTHORAX IN CHILDREN

Leakage of chyle into the pleural space occurs as a result of damage to the thoracic duct. The aetiology may vary according to the age of the child, mechanism of injury (recent surgery, trauma, high central venous pressure) or result from congenital abnormalities of the lymphatics that may or may not be part of an associated condition (eg Down's or Noonan's syndrome). We have divided the causes of chylothorax in children into five categories: congenital, traumatic, high central venous pressure, malignancy or miscellaneous. (see Table 2).

Chylothorax is the most common form of pleural effusion in the first few days of life.[20] This may occur as an unexpected finding in an otherwise healthy baby or be secondary to abnormalities of lymph vessels as found in congenital lymphangiectasia, pulmonary lymphangiectasia, or associated with syndromes such as Turner's, Down's and Noonan's Syndrome.[21–24] It may also result from various congenital defects of the thoracic duct such as absence or atresia.[16,25] Congenital chylothorax is a common manifestation of non-immune hydrops fetalis. Chylothorax associated with hydrops is believed to occur as a result of an abnormal development of the lymphatic vessels. Nevertheless, congenital chylothorax can cause hydrops by impairing venous return and/or loss of protein into the pleural space leading to generalized hypoproteinemia and generalized oedema.[23,26,27] The cause of neonatal chylothorax in the absence of identified lymphatic abnormality has never been clearly explained. While it is often considered in the category of congenital chylothorax, the mechanism is thought to be either traumatic, with rupture of the thoracic duct by hyperextension of the spinal column or secondary to increased systemic venous pressure during birth, especially in complicated deliveries.[28,29]

Congenital abnormalities of the lymphatics do not always present in the newborn period. Pulmonary lymphangiomatosis and lymphangiectasia are the two major abnormalities of lymphatics associated with chylothorax.[24,30] Pulmonary lymphangiomatosis is a focal proliferation of well differentiated lymphatic tissue, frequently associated with lymphatic abnormalities in other organs. The majority of lymphangiomas present in the first 2 years of life, however they may not be recognised until adulthood. Lymphagiomas that appear in head, neck and axial skeleton can extend into the mediastinum. Approximately 1% of all lymphangiomas are confined to the chest. Thoracic lymphangiomas tend to present after a period of latency. In pulmonary lymphangiectasia,

Table 2
Aetiology of Chylothorax in Children

Aetiology
A. Congenital
Congenital lymphatic malformations
Lymphangiomatosis
Lymphangiectasia
Atresia of the thoracic duct
Associated with various syndromes
Down Syndrome
Noonan Syndrome
Turner Syndrome
Hydrops fetalis
B. Traumatic
Surgical
Cervical
Excision of lymph nodes
Thoracic
Surgery for congenital heart diseases
Surgery for mediastinal tumours
Surgery for congenital lung malformations
Others
Invasive diagnostic and therapeutic procedures
Subclavian vein catheterization
Non-iatrogenic trauma
Hyperextension or stretching of chest wall or thoracic spine
Forceful cough or vomiting
Child birth
C. High central venous pressure
Thrombosis of the superior vena cava or subclavian vein
Post-Fontan surgery
D. Malignancies
Lymphoma
Teratoma
Sarcoma
Neuroblastoma
E. Miscellaneous
Benign tumours
Tuberculosis/Histoplasmosis
Sarcoidosis
Transdiaphragmatic movement of chylous ascites

the lungs show diffuse dilatation of the interlobular and subpleural lymphatics. Primary lymphangiectasia presents in neonates and is often fatal. Secondary lymphangiectasia can result from conditions associated with abnormal lymph drainage and/or increase lymph production. Cases associated with pulmonary venous obstruction or congenital heart defects present early in life. Diagnosis can be made by lymphangiography, computerised tomography or magnetic resonance imaging, however, confirmation usually requires lung biopsy.[30,31] Other cases of lymphatic abnormalities such as lymphangiomatosis and lymphatic dysplasia syndrome are almost exclusively seen in adults.

Traumatic chylothorax results from damage to the thoracic duct by rupture or laceration. Postoperative chylothorax has been described after almost any surgical procedure performed in the chest.[32] In children, the most common setting is following cardiothoracic surgery, but can occur after scoliosis or neck surgery.[33] In children, the reported incidence of chylothorax after cardiothoracic surgery is between 0.85% and 6.6%.[2,33–35] Other traumatic causes of chylothorax include laceration of the thoracic duct during catheterization of the subclavian vein or by direct trauma as a result of penetrating chest trauma.[36,37]

Non-iatrogenic traumatic chylothorax has been described in children following sudden hyperextension of the spine, severe coughing and vomiting. Non-accidental injury, however, may be a cause of traumatic thoracic duct rupture that can masquerade a 'spontaneous' chylothorax in young children.[38–40] An additional cause of traumatic chylothorax involves the forces involved in the mechanism of birth.

Increased venous pressure in the superior vena cava or subclavian vein secondary to venous thrombosis or obstruction due to surgical procedures may cause rupture of the duct and/or its collaterals due to excessive venous pressure.[2,41–44] High central venous pressure secondary to a Fontan procedure has been reported to result in protein-losing enteropathy, intestinal lymphangiectasia and chylothorax, independently of the risk of thoracic duct laceration.[43,45]

Malignancies, while one of the most common causes of chylothorax in adults they are a less prevalent cause in children. Extrinsic compression of the thoracic duct or direct invasion of the thoracic duct may cause rupture and leakage of chyle. Lymphomas are the most common type of tumour associated with chylothorax (60 – 70% of cases) however; any mediastinal tumour (eg teratoma and neuroblastoma) has the potential to cause chylothorax.[1,46–48]

In children, the miscellaneous group includes those with benign tumours, sarcoidosis, and chronic infections such as tuberculosis and histoplasmosis.[49–51] Chylothorax has been reported in the setting of chylous ascitis and transdiaphragmatic passage of chyle from the peritoneal cavity.[52] In adults, chylothorax has been associated with cardiac failure, lymphangioleiomyomatosis (LAM), yellow nail syndrome, mediastinal radiation therapy, hypothyroidism and pancreatitis.[15]

CLINICAL MANIFESTATIONS

The clinical presentation of chylothorax results from the accumulation of pleural fluid. Initially patients can be asymptomatic, however dyspnoea, cough and chest discomfort develop with time. The severity of symptoms depends on the size of the effusion. Rapid accumulation of large volume of fluid can lead to adverse haemodynamic complications with significant cardiorespiratory difficulties. Patients with non-traumatic chylothorax usually present with chest discomfort and dyspnoea on exertion, or as incidental finding on a chest radiograph. The onset of symptoms is usually gradual; however if there is a significant leakage of chyle, the child can present with significant respiratory distress. A history of recent surgery or trauma may be relevant. Physical examination should include recognition of different risk factors for chylothorax such as dysmorphic features, superior vena cava obstruction, or lymphatic malformations elsewhere.

Congenital chylothorax presenting antenatally can act as a space-occupying lesion and cause restriction of normal development of the foetal lungs; therefore, causing some degree of lung hypoplasia. At birth it usually presents with respiratory distress in the first few days of life associated with bilateral or unilateral dullness to percussion and poor air entry.[53]

Amongst patients with chronic chylothorax associated with pulmonary lymphatic malformations, muscle wasting, weight loss and other signs of malnutrition can be present.[30]

INVESTIGATIONS

A chest radiograph will usually be performed to identify pleural fluid. This will assess the size and location of the effusion. Use of lateral decubitus radiograph or ultrasound can determine whether there is free fluid in the pleural space (simple, non-clotting effusion) or organised, as seen in empyemas.[54] Once the diagnosis of chylothorax is made by pleural fluid analysis, if there is no obvious cause (for example cardiac surgery or trauma), investigations should be performed to outline lymphatic vessels, identify the site of chyle leakage and finally to ascertain the cause of chylothorax (see Fig. 1). Imaging studies such as computerised tomography (CT) scans, lymphangiography and lymphoscintigraphy can be helpful.

Lymphangiography and lymphoscintigraphy are two specific lymphatic imaging modalities. Both require the administration of a contrast agent into the lymphatic system. There are several routes to achieve this: introduction through interstitial (intradermal or subcutaneous) administration, direct administration into a cannulated lymphatic vessel, or intravenous injection.[55,56] Lymphangiography implies the direct administration of an iodinated contrast agent into a cannulated lymph vessel. A simultaneous chest radiograph or CT will delineate the lymphatic anatomy. It is widely used in adults, as it is the best study to delineate the lymphatic anatomy and is very useful defining the site of chyle leak or obstruction. The sensitivity for detecting thoracic duct leaks may be relatively low because it may be difficult to visualize the entire length of the thoracic duct due to poor mixing of oily contrast medium and chyle. However, successful identification rates of up to 81% have been reported in adults.[57] There are significant limitations for its use in children, in particular the technical skills to cannulate lymph vessels and pain. Lymphangiography carries several complications such as infection, respiratory distress, and damage to lymphatics.

Lymphoscintigraphy is a nuclear imaging technique that utilizes radionuclides as contrast agents. Filtered 99m-technetium is the most common radionuclide used in United States. Usually, intradermal or subcutaneous injections are administered. Lymphoscintigraphy may be an alternative to lymphangiography as it is a faster and less traumatic procedure.[58] It has been used in children as an easy and non-invasive study with no irradiation [9,59] {See Illustrative case}.

CT scan may be required to image the mediastinum, especially if non-traumatic chylothorax is suspected. CT has been performed after lymphangiography detecting even small amounts of contrast material in the pleural space. Magnetic Resonance Imaging (MRI) may be better in some instances for imaging the mediastinum. MR lymphography involves interstitial or intravenous injection of gadolinium-based contrast agents. Interstitial MRI lymphography has been used in adults with good delineation of lymphatic vessels.[60,61]

Direct visualization of the chyle leak point is sometimes required. In recent times video-assisted thoracoscopic surgery (VATS) has replaced open thoracotomy. Biopsies of any suspect area should be taken during this procedure including lung biopsy if indicated (usually for lymphatic malformations).

Other complementary studies should be performed when there is clinical suspicion of a specific underlying condition. For example, if child abuse is suspected, careful physical examination with radiographic studies should be performed. If malignancy is suspected, tumour markers, bone marrow aspiration and CT scan of chest and abdomen will be indicated.

MANAGEMENT

Principles of Treatment

The approach to management of chylothorax is the same regardless of the cause of the chylothorax (See Table 3). No treatments have been subject to randomized controlled trials. Most of our knowledge of the management of chylothorax in children comes from small case series.[2,3,34,62] The six principles of the management of chylothorax in children are outlined in Fig. 2.

Initial Drainage

The initial step in all cases should be aspiration of the pleural fluid. This first thoracocentesis is usually for diagnostic purposes, however if the size of the effusion compromises respiration, and/or if the collection is likely to reoccur, then a chest tube should be inserted for continuous drainage of the pleural space.

Table 3
Treatment of Chylothorax

A. Non-operative Management
Thoracocentesis (single or multiple)
Continuous drainage (intercostal tube insertion)
Dietary Modifications
Fat-free diet
Medium-chain triglyceride diet
Total parenteral nutrition
Somatostatin and analogues (Octreotide)
Pleurodesis (chemical or radiation)
B. Surgical treatment
Ligation of the thoracic duct or mass ligation
Thoracotomy
VATS
Chest tube or thoracoscopic pleurodesis
Pleuroperitoneal shunts

Quantification of drainage is useful to determine clinical improvement and also to guide the clinician with regard to fluid imbalance. Some centres have adopted a therapeutic approach with daily drainage as a guide for clinical improvement or failure (<10 ml/kg/day improvement, >10 ml/kg/day failure, after 4 weeks of nonsurgical management).[2]

Dietary Modifications

The aim of chylothorax management is to reduce the flow of chyle through the thoracic duct while waiting for spontaneous healing. This is usually managed by fat-free diet with the addition of MCT. MCT consists of triglycerides with saturated fatty acids of 8 to 12 carbon chain length that are absorbed directly into the portal venous system bypassing lymphatic drainage. A more aggressive option is complete enteric rest using with total parenteral nutrition (TPN).[33]

Somatostatin and synthetic analogues (Octreotide)

Somatostatin is an endogenous hormone with a wide range of actions that include the gastrointestinal tract. Octreotide is a synthetic, long-acting somatostatin analogue.[35,63–65] Somatostatin and octreotide are the only pharmacologic agents that have been used successfully in the management of chylothorax in children.[35,65,66] There is, however, no consensus regarding the timing of introduction of these agents. The mechanism of action of somatostatin and octreotide in treating chylothorax is unclear. The

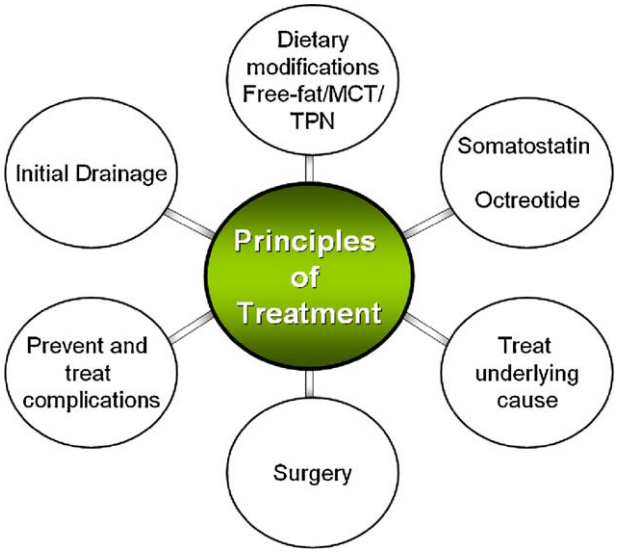

Figure 2. Principles of treatment of chylothorax in children.

most likely explanation is reduction of intestinal blood flow by vasoconstriction of the splanchnic circulation with reduction of lymphatic fluid production.[66,67] They also decrease gastrointestinal motility, decrease the volume of gastric, pancreatic and biliary secretions, which in turn decreases lymphatic flow in the thoracic duct.[15,63,65] In dogs, octreotide decreases the absorption of triglycerides, and this may be a relevant mechanism of action in humans.[68] Octreotide has the advantage over somatostatin of a longer half-life, greater potency and the option of subcutaneous administration.[63,65] The timing of introduction and duration of treatment in children is unknown. Some authors argue that the use of octreotide earlier in the clinical course may reduce fluid and electrolyte complications and may allow an earlier removal of the intercostal tubes; however, the level of evidence upon which this recommendation is made is poor.

There are several modes of administration and doses of somatostatin or octreotide to consider. Somatostatin and octreotide can be given as a continuous intravenous infusion or as an intravenous bolus, given twice daily. Octreotide can be also given subcutaneously. Roehr and colleagues (2006)[65] published a systematic review on the use of somatostatin or octreotide as a treatment for chylothorax in children. A total of thirty-five children treated for chylothorax were reviewed, ten children were given somatostatin and the remaining 25 children received octreotide. Somatostatin was given as an intravenous infusion at a median dose of 204 μg/kg/day (range 10 – 288 μg/kg/day). Octreotide was mainly used as an intravenous infusion at a median dose of 68 μg/kg/day (range 7.2 - 240 μg/kg/day). When given subcutaneously the median dose was 40 μg/kg/day (range 2 – 68 μg/kg/day). The median duration of treatment was 9.5 days for somatostatin compared to 7 days for intravenous octreotide or 17 days for subcutaneous octreotide.[65] Other authors have suggested starting with 0.5 μg/kg/hr of octreotide as an infusion with gradually increasing the dose up to 10 or 12 μg/kg//hr.[63–65,68] Reduction of lymphatic flow rate is usually evident within 3 or 6 days after initiation of treatment.[63,65,69]

Both somatostatin and octreotide are considered safe with few side-effects. Side effects include hyperglycaemia, hypothyroidism, cramps, nausea, diarrhoea, renal impairment, necrotizing enterocolitis and liver dysfunction.[63,65]

Other therapies have been described in single case reports in adults and include nitric oxide, high positive-end expiratory pressure ventilation and etilefrine. Etilefrine, is a sympathomimetic drug that has been used in a small number of adults for the management of postoperative chylothorax with no side effects. It causes systemic smooth muscle contraction and is thought to decrease chyle flow by constriction of the thoracic duct.[70]

The response to medical therapies for the treatment of chylothorax (dietary modifications and/or adjunctive medications) may take many weeks. Most series performed in children recommend up to 2 – 4 weeks until surgical procedures are considered. Non-operative management of chylothorax in children is successful in more than 80% of reported cases, including those patients with chylothorax following cardiothoracic surgery.[1–3,33–35,62]

Surgery

Surgery should be considered when medical management of chylothorax has failed to reduce chyle flow and allow healing of the duct. There is no consensus on the timing of surgery. Most authors advocate three to four weeks of medical therapy;[2,3] nevertheless, a case for earlier surgery could be made when if there is a well identified site of chyle leak and high flow that precludes spontaneous healing.[9] Successful surgery may also shorten hospitalization and reduce the risks of malnutrition, and immunosuppression.

There are numerous surgical approaches described for thoracic duct ligation, although comparisons are difficult given the many aetiologies and variable use of concomitant pleurodesis (surgical or chemical). If the site of rupture can be identified, for example by lymphangiography, direct surgical ligation of the thoracic duct represents the most definitive treatment.[6] Recently, video-assisted thoracoscopic surgical (VATS) approach has been recommended as it has a lower rate of complications and better cost-effectiveness.[71] Regardless of the surgical approach, visualization of the site of chyle leakage can be difficult. Intraoperative manoeuvres such as injection of 1% Evans blue dye in the thigh or a 200 ml mixture of milk and cream given to the patient a few hours before operation can help visualise the duct.[9,72,73] When the thoracic duct or site of leak is not identifiable, a mass ligation of the thoracic duct and its surrounding tissue can be done between the aorta, azygos vein and oesophagus, adjacent to the vertebral body.[74]

One of the largest series of chylothorax in children reported 51 children with postoperative chylothorax at a median age of 11 months (range 4 days - 19.6 years). Chylothorax developed at a median of 9 days after operation (range, 0 to 24 days). Twenty-one of them were identified before octreotide was available. All but one responded to MCT, while the remaining received complete enteric rest with TPN. Two patients (10%) required surgical intervention at two to four weeks. The remaining 30 were identified after the use of octreotide had been reported; 12 (40%) resolved completely with MCT diet alone, 17 (56%) received octreotide in addition to dietary management. and one required surgery before octreotide was commenced. Complete resolution of chylothorax was seen in 14 out of 17 patients (82%) treated with octreotide at 15.3 ± 5.5 days after starting octreotide with no side effects from the octreotide. The remaining three required surgical intervention.[34]

Another approach to management of chylothorax has been obliteration of the pleural space, either chemically (tetracycline, talc or povidone-iodine) or surgically.[75] Pleurodesis is commonly performed with the assistance of VATS although the sclerosing agent can be instilled through the chest tube. This treatment has been employed effectively in several case reports when MCT diet and octreotide failed and direct thoracic duct surgery was not attempted.[76]

Pleuroperitoneal shunts provide another way of draining chyle from the pleural space and have the advantage of not losing the chyle. The shunts are a one-way subcutaneous connection between the pleura and the peritoneum. It has been considered a safe and effective treatment for persistent chylothorax in infants, although, it requires daily pumping.[77,78] To our knowledge, there is nothing published outside infancy to guide the use of this therapy amongst children and adolescents with refractory chylothorax.

Other approaches have included fluoroscopically guided percutaneous transabdominal embolization of the thoracic duct with platinum coils.[79] In cases of severe chylothorax leading to non-immunologic hydrops fetalis, antenatal management by intra-uterine thoracocentesis, or by insertion of a pleuro-amniotic shunt can be considered.[20] Pleurodesis OK-452 may prevent pulmonary hypoplasia, thus improving respiratory function at birth.[80] Radiotherapy has been used in the context of patients with complex lymphatic malformations and secondary chylothorax.[81]

Prevention and treatment of the complications of chylothorax

Some of the complications of chylothorax include malnutrition, hyponatremia, fluid imbalance, respiratory distress, increased risk of thrombosis, and secondary immunodeficiency. None have been extensively studied [82,83]. Immunodeficiency in patients with lymphopenia and hypogammaglobulinemia in chylothorax has been suggested; however not well documented.

In regards to hypogammaglobulinemia, Orange et al (2003),[84] published a series of eight children who had acute post-traumatic

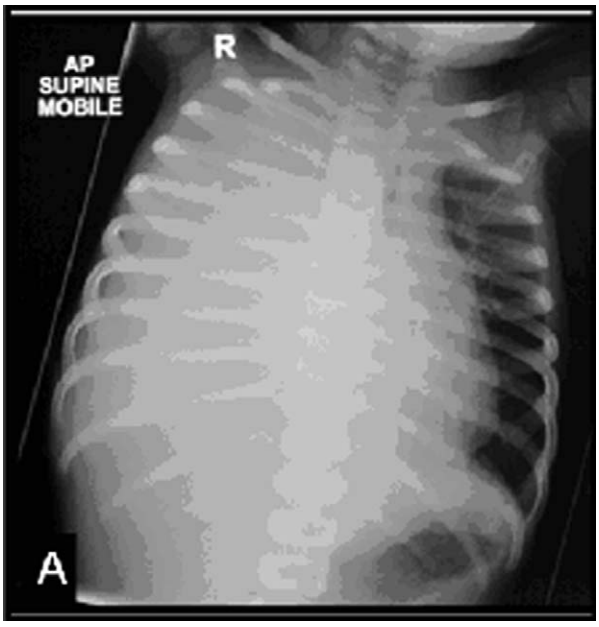

Figure 3A. Chest x-ray done on day of admission showing a complete opacification of the right hemi-thorax with mediastinal shift to the left. **Figure reproduced with permission from the Med J Aust.**[9]

chylothorax (all but one from cardiothoracic surgery) resulting in lymphopenia, hypogammaglobulinemia and other immunologic abnormalities. All patients received intravenous immunoglobulin (IVIG) to maintain IgG within the normal range. Six out of eight children (75%) had serious infections before administration of IVIG, compared to four out of eight (50%) during the period of IVIG. There was preservation of protective levels of tetanus-specific antibodies and no serious infections attributable to hypogammaglobylinemia or cellular immunodeficiency in this group. Although this was not a controlled study, no clinical immunodeficiency was associated with chylothorax and IVIG replacement was not shown to be clinically beneficial.[84]

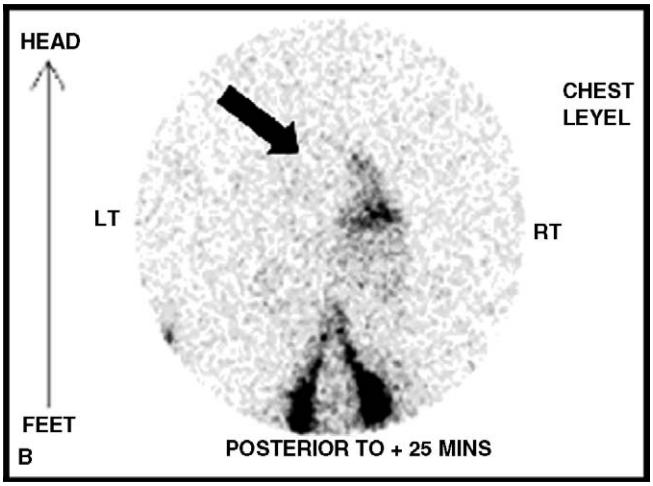

Figure 3B. Lymphoscintigraphy. Following the intradermal administration of microcolloid (containing radionuclide) simultaneously into the web spaces between the second and third toes of both feet, there was rapid passage of the radionuclide delineating lymphatic channels of both lower limbs. Twenty-five minutes after instillation of microcolloid, there was a rapid leakage of lymphatic fluid into the right hemithorax (at the level of the arrow) suggesting a significant rupture of the thoracic duct. There was no leakage into the left hemithorax, therefore, the site of rupture was suspected be localized below T5 (right hemithorax). **Figure reproduced with permission from the Med J Aust.**[9]

CONCLUSION

Chylothorax is a rare cause of pleural effusion in children, particularly beyond the neonatal period. The diagnosis is made by analysis of the pleural fluid with chylomicrons, triglycerides and lymphocytes present. In the absence of obvious risk factors such as surgery or recognised lymphatic abnormalities, the aetiology of chylothorax can be challenging to determine. Understanding the anatomy and physiology of the thoracic duct is vital for assessment and management. Initial treatment consists of drainage, dietary modifications and other medical therapies to diminish chyle flow, thus allowing the duct to heal spontaneously. Somatostatin and octreotide may be useful in the management of chylothorax in some cases. Failure of these measures associated with the presence of complications such as infection, malnutrition, fluid imbalance and prolonged hospitalization should result in early surgical intervention. In rare cases more complex interventions such as pleurodesis may be required. The prognosis of chylothorax in children depends on the aetiology of disruption of the thoracic duct and the associated abnormalities may influence the outcome.

FUTURE DIRECTIONS

- Determine factors that will predict success of any specific treatment early in the course of the disease.
- Prospective randomised controlled studies comparing different therapeutic approaches are needed.
- Determine the ideal dosage regimen, initiation time and administration route for somatostatin and octreotide.
- Improvement in imaging techniques for the visualization of the lymphatic vessels. These techniques should be simple and non-invasive, therefore feasible to use in children.

PRACTICE POINTS

- Chylothorax is a rare cause of pleural effusion in childhood which can be difficult to diagnose and manage.
- The diagnosis of chylothorax is made by identification of chylomicrons or high triglyceride concentrations in the pleural fluid.
- Diagnostic techniques such as lymphangiography or lymphoscintigraphy are useful for defining the site of chyle leak.
- Initial treatment of chylothorax is the same regardless of the cause: Drainage, nutritional support and measures to diminish chyle flow.
- More than 80% of children with chylothorax will respond to non-operative treatment in less than 4 weeks.
- Prognosis depends on the underlying aetiology.

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?CME SECTION

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Educational questions

Answer true or false to the following questions:

1. With regards to the anatomy of the thoracic duct:
   a. It is formed by coalescing intestinal and lumbar lymphatics at the level of the 5th thoracic vertebra.
   b. It enters the thorax through the aortic hiatus of the diaphragm and ascends through the posterior mediastinum on the left side of the thorax.
   c. Anatomic variations are infrequent.
   d. It drains lymph from the lower limbs, abdomen, intestinal chyle and left thorax, head and upper limb, the remaining will be drained by the right lymphatic duct.
   e. Rupture of the thoracic duct between diaphragm and T5 will produce a left-sided chylothorax.
2. Regarding the diagnosis of chylothorax in children:
   a. The diagnosis should be suspected in patients with persistent pleural effusion following cardiothoracic or mediastinal surgery.
   b. Diagnosis can be made by the presence of a milky or turbid appearance of the fluid, biochemical analysis of the fluid is not required.
   c. The presence of chylomicrons will confirm the diagnosis of chylothorax.
   d. Triglyceride concentration of the fluid is positive if the concentration is above 1.1 mmol/L.
   e. Another characteristic of the pleural fluid in chylothorax is the abundance of eosinophils.
3. The following are causes of chylothorax in children:
   a. Congenital abnormalities of the lymphatic vessels such as pulmonary lymphangiomatosis and lymphangiectasia.
   b. Syndromes such as Down's, Turner's and Noonan's Syndrome.
   c. Cardiothoracic surgery
   d. Malignancies such as Lymphoma or Teratoma
   e. Any condition causing high central venous pressure
4. With regards to investigations available for chylothorax in children:
   a. Investigations should be performed in all cases to delineate the lymphatic vessels and hopefully identify the site of chyle leakage.
   b. Lymphangiography is the easiest and safest study to perform in children with chylothorax.
   c. Lymphoscintigraphy is a nuclear imaging technique that is used in children as an alternative to lymphangiography as it is faster and less traumatic.
   d. CT scan should be performed in those cases with non-traumatic chylothorax to image the mediastinum.
   e. Direct visualization of the site of rupture can be done by VATS.
5. Regarding various treatment options for chylothorax in children:
   a. The approach and initial management of chylothorax in children is different in all cases and will depend on the underlying condition.
   b. The main aim of dietary modifications in the management of chylothorax is to improve nutrition.
   c. Somatostatin and octreotide have been found to be useful and relatively safe medications in the management of chylothorax, although its mechanism of action in treating chylothorax is still unclear.
   d. Early surgical approach is recommended in most cases of chylothorax in children given the poor response to medical management.
   e. Ligation of the thoracic duct is the most common surgical approach for management of chylothorax in children, but this procedure is usually recommended after 2 – 4 weeks of medical therapy.

ILLUSTRATIVE CASE: CHYLOTHORAX IN A 2-YEAR-OLD CHILD

A 2-year-old girl presented with acute respiratory distress. Initial chest x-ray was performed (see Figure A) showing opacification of the right hemithorax. Investigations performed confirmed a chylothorax with no obvious cause. Forceful vomiting was considered the likely cause. Lymphocintigraphy (see Figure B) was a key investigation as it confirmed the presence of chyle leak between the diaphragm and T5 (right hemithorax). This investigation facilitated surgical ligation of the thoracic duct and surrounding tissue, and should be considered in children with chylothorax where the location of thoracic duct rupture is not known.