# CHAPTER 55
# CHYLOTHORAX

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## Introduction

Chylothorax is a rare entity and is defined as an effusion of lymph in the pleural cavity. The chyle may have its origin in the thorax or in the abdomen or in both. Leakage usually occurs from the thoracic duct or one of its main tributaries.

## Demographics

There are no known racial, gender, age, or geographical variations to chylothorax. This is due to its aetiology. However, it is known to occur in up to 4% of patients after cardiothoracic surgery.

## Pathophysiology

The thoracic duct develops from outgrowths of the jugular lymphatic sacs and the cisterna chyli. During embryonic life, bilateral thoracic lymphatic channels are present, each attached in the neck to the corresponding jugular sac. As development progresses, the upper third of the right duct and the lower two-thirds of the left duct involute and close. The wide variation in the final anatomic structure of the main ductal system attests to the multiple communications of the small vessels comprising the lymphatic system. The thoracic duct originates in the abdomen at the cisterna chyli located over the second lumbar vertebra. The duct extends into the thorax through the aortic hiatus and then passes upward into the posterior mediastinum on the right before shifting toward the left at the level of the fifth thoracic vertebra. It then ascends posterior to the aortic arch and into the posterior neck to the junction of the subclavian and internal jugular veins.

The chyle contained in the thoracic duct conveys approximately three-fourths of the ingested fat from the intestine to the systemic circulation. The fat content of chyle varies from 0.4 to 4.0 g/dl. The large fat molecules absorbed from the intestinal lacteals flow through the cisterna chyli and superiorly through the thoracic duct. The total protein content of thoracic duct lymph is also high. The thoracic duct also carries white blood cells, primarily lymphocytes (T cells)—approximately 2,000 to 20,000 cells per milliliter. When chyle leaks through a thoracic duct fistula, considerable fat and lymphocytes may be lost. Eosinophils are also present in a higher proportion than in circulating blood. The chyle appears to have a bacteriostatic property, which accounts for the rare occurrence of infection complicating chylothorax.

## Aetiology

Effusion of chylous fluid into the thorax may occur spontaneously in newborns and has usually been attributed to congenital abnormalities of the thoracic ducts or trauma from delivery. The occurrence of chylothorax in most cases cannot be related to the type of labor or delivery, and lymphatic effusions may be discovered prenatally.

Chylothorax in older children is rarely spontaneous and occurs almost invariably after trauma or cardiothoracic surgery; however, some patients with thoracic lymphangioma may present in this older age group. Operative injury may be in part a result of anatomic variations of the thoracic duct. Neoplasms, particularly lymphomas and neuroblastomas, have occasionally been noted to cause obstruction of the thoracic duct. Lymphangiomatosis or diffuse lymphangiectasia may produce chylous effusion in the pleural space and peritoneal cavity. Extensive bouts of coughing have been reported to cause rupture of the thoracic duct, which is particularly vulnerable when full following a fatty meal. Other causes include mediastinal inflammation, subclavian vein or superior vena caval thrombosis, and misplaced central venous catheters (Table 55.1).

*Table 55.1: Causes of chylothorax.*

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|---|
| **Lymphatic malformation (nontrauma)** |
| Thoracic duct atresia/aplasia/hypoplasia/dysplasia |
| Lymphangioma |
| Lymphangiomatosis |
| Intestinal lymphangiectasia (protein-losing enteropathy) |
| Fontan procedure |
| **Thoracic duct injury (trauma)** |
| Cardiothoracic operations |
| Oesophageal atresia |
| Diaphragmatic hernia |
| Penetrating trauma (stab or gunshot injury) |
| **Malignant** |
| Lymphoma |
| Kaposi sarcoma |
| Mediastinal teratoma |
| **Infectious** |
| Tuberculosis |
| Filariasis |
| Pneumonia |
| Pleuritis and empyema |
| **Idiopathic (associated with)** |
| Down syndrome |
| Noonan syndrome |
| Gorham's disease |
| Hydrops foetalis |
| Turner syndrome |
| Lymphoedema |
| **Transudative** |
| Cirrhosis of the liver |
| Fontan procedure |
| Heart failure |
| Nephritic syndrome |
| **Miscellaneous** |
| Sarcoidosis |
| Amyloidosis |

## Clinical Presentation

The accumulation of chyle in the pleural space from a thoracic duct leak may occur rapidly and produce pressure on other structures in the chest, causing acute respiratory distress, dyspnea, and cyanosis with tachypnea. In the foetus, a pleural effusion may be secondary to generalised hydrops, but a primary lymphatic effusion (idiopathic, secondary to subpleural lymphangiectasia, pulmonary sequestration, or associated with syndromes such as Down, Turner, and Noonan) can cause mediastinal shift and result in hydrops or lead to pulmonary hypoplasia. Postnatally, the effects of chylothorax and the prolonged loss of chyle may include malnutrition, hypoproteinaemia, fluid and electrolyte imbalance, metabolic acidosis, and immunodeficiency.

In a neonate, symptoms of respiratory embarrassment observed in combination with a pleural effusion strongly suggest chylothorax. Similar findings are noted in the traumatic postoperative chylothorax. In the older child, nutritional deficiency is a late manifestation of chyle depletion and occurs when dietary intake is insufficient to replace the thoracic duct fluid loss. Fever is not common.

## Diagnosis

Chest roentgenograms typically show massive fluid effusion in the ipsilateral chest with pulmonary compression and mediastinal shift. Bilateral effusions may also occur. Aspiration of the pleural effusion reveals a clear straw-colored fluid in the fasting patient, which becomes milky after feedings. Analysis of the chyle generally reveals a total fat content of more than 400 mg/dl and a protein content of more than 5 g/dl. In a foetus or a fasting neonate, the most useful and simple test is to perform a complete cell count and differential on the fluid; when lymphocytes exceed 80% or 90% of the white cells, a lymphatic effusion is confirmed. The differential can be compared to that obtained from the blood count, where lymphocytes rarely represent more than 70% of white blood cells.

Lymphangiography is useful for defining the site of chyle leakage or obstruction with penetrating trauma, spontaneous chylothorax, and lymphangiomatous malformation. However, in a nontraumatised patient, the site of lymphatic leakage is often difficult to localise. Lymphoscintigraphy may be an alternative to lymphangiography, as it is a faster and less traumatic procedure.

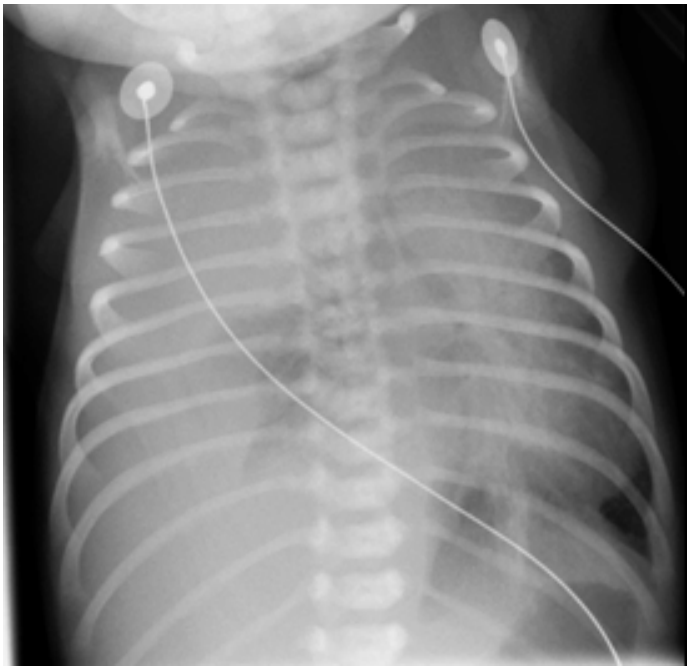

*Figure 55.1: Right-sided congenital chylothorax in a newborn.*

## Management

### Nonoperative Management

Thoracentesis may be sufficient to relieve spontaneous chylothorax in occasional infants; however, chest tube drainage will be necessary for the majority of patients. Further, tube drainage allows quantification of the daily chyle leak and promotes pulmonary re-expansion, which may enhance healing. Chylothorax in newborns usually ceases spontaneously. In some cases of congenital chylothorax, supportive mechanical ventilation may be necessary because of insufficient lung expansion, persistent foetal circulation, or lung hypoplasia. In cases of severe chylothorax leading to nonimmunologic hydrops foetalis, antenatal management by intrauterine thoracocentesis or pleuroperitoneal shunting should be considered in the absence of significant underlying malformations.

For postnatal chylothorax, since identifying the actual site of the fluid leak is difficult, surgery is often deferred for several weeks. Most cases of traumatic injury to the thoracic duct can be managed successfully by chest tube drainage and replacement of the protein and fat loss. Feeding restricted to medium- or short-chain triglycerides theoretically results in reduced lymph flow in the thoracic duct and may enhance spontaneous healing of a thoracic duct fistula. However, it has been shown that any enteral feeding, even with clear fluids, greatly increases thoracic duct flow. Therefore, the optimum management for chyle leak is chest tube drainage, withholding oral feedings, and providing total parenteral nutrition (TPN). Cultures of chylous fluid are rarely positive; therefore, providing long-term antibiotics during the full course of chest tube drainage is not considered necessary. In nonresolving chylothorax, subcutaneous injection of octerotide, a somatostatin analogue, at 10 $\mu$g/kd/day in 3 divided doses is reported to have excellent results in a number of case reports and should be tried prior to surgical intervention.

### Surgical Management

When chylothorax remains resistant despite prolonged chest tube drainage (2–3 weeks) and TPN, thoracotomy on the ipsilateral side may be necessary. The decision whether to continue with conservative management or to undertake surgical intervention should be based on the nature of the underlying disorder, the duration of the fistula, the daily volume of fluid drainage, and the severity of nutritional and/or immunologic depletion. Ingestion of cream before surgery may facilitate identification of the thoracic duct and the fistula. When identified, the draining lymphatic vessel should be suture ligated above and below the leak with reinforcement by a pleural or intercostal muscle flap. When a leak cannot be identified with certainty, or when multiple leaks originate from the mediastinum, ligation of all the tissues surrounding the aorta at the level of the hiatus provides the best results. Fibrin glue and argon-beam coagulation have also been used for ill-defined areas of leakage or incompletely resected lympangiomas.

Thoracoscopy may occasionally be used to avoid thoracotomy. The leak, if visualised, can be ligated, cauterised, or sealed with fibrin glue. If the leak cannot be identified, pleurodesis can be accomplished with talc or other sclerotic agents under direct vision through the thoracoscope, but this technique should probably be avoided in infancy due to the potential consequences on lung and chest wall growth. If there is concomitant chylopericardium, a pericardial window can be fashioned.

During any thoracotomy, if chyle leak is noted, the proximal and distal ends of the leaking duct should be ligated.

Pleuroperitoneal shunts have been reserved for refractory chylothorax. A Denver double-valve shunt system is the type most commonly employed; it is totally implanted and allows the patient or parent to pump the valve to achieve decompression of the pleural fluid into the abdominal cavity where it is reabsorbed.

## Prognosis

Prognosis depends largely on the aetiology of the chylothorax. A mortality rate of 12.8% among paediatric patients with a nontraumatic chylothorax has been reported. This rate may be reduced by appropriate support with TPN and timely intervention.

## Evidence-Based Research

Table 55.2 presents a systematic review of using someatostatin or octreotide as a treatment option for childhood chylothorax.

*Table 55.2: Evidence-based research.*

| | |
|---|---|
| **Title** | Somatostatin or octreotide as treatment options for chylothorax in young children: a systematic review. |
| **Authors** | Roehr CC, Jung A, Proquitte H, Blankenstein O, Hammer H, Lakhoo K, Wauer RR. |
| **Institution** | Department of Neonatology, Charité Campus Mitte, Universitätsmedizin Berlin, Berlin, Germany; John Radcliffe Hospital, Department of Paediatric Surgery, Oxford, UK. |
| **Reference** | Intensive Care Med. 2006; 32(5):650–657. Epub 2006 Mar 11. |
| **Problem** | Chylothorax is a rare but life-threatening condition in children. To date, there is no commonly accepted treatment protocol. Somatostatin and octreotide have recently been used for treating chylothorax in children |
| **Intervention** | Summarisation of the evidence on the efficacy and safety of somatostatin and octreotide in treating young children with chylothorax |
| **Comparison/ control (quality of evidence)** | Design: Systematic review: literature search (Cochrane Library, EMBASE and PubMed databases) and literature hand search of peer reviewed articles on the use of somatostatin and octreotide in childhood chylothorax. Patients: Thirty-five children treated for primary or secondary chylothorax (10/somatostatin, 25/octreotide) were found. |
| **Outcome/effect** | Ten of the 35 children had been given somatostatin, as intravenous (IV) infusion at a median dose of 204 μg/kg per day, for a median duration of 9.5 days. The remaining 25 children had received octreotide, either as an IV infusion at a median dose of 68 μg/kg per day over a median 7 days, or subcutaneous injection at a median dose of 40 μg/kg per day and a median duration of 17 days. Side effects such as cutaneous flush, nausea, loose stools, transient hypothyroidism, elevated liver function tests and strangulation-ileus (in a child with asplenia syndrome) were reported for somatostatin; transient abdominal distention, temporary hyperglycaemia and necrotising enterocolitis (in a child with aortic coarctation) for octreotide. |
| **Historical significance/ comments** | A positive treatment effect was evident for both somatostatin and octreotide in the majority of reports. Minor side effects have been reported; however, caution should be exercised in patients with an increased risk of vascular compromise to avoid serious side effects. Systematic clinical research is needed to establish treatment efficacy and to develop a safe treatment protocol. |

## Key Summary Points

1. Chylothorax may be congenital or traumatic, most commonly postoperative.
2. Diagnosis is by means of pleural tap analysis showing more than 80% lymphocytes on the differential count.
3. Optimum treatment includes chest tube drainage, nothing by mouth and nutritional support with total parenteral nutrition (TPN). Feeding restricted to medium chain triglycerides may be tried in the absence of TPN, and is often used once the leak has subsided with the patient on TPN.
4. A somatostatin analogue may be tried before surgical intervention.
5. Surgery is reserved for the refractory chylothorax with either direct ligation of the leak where feasible, ligation of the duct and all periaortic tissues at the aortic hiatus, or utilisation of a pleuroperitoneal shunt.

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