Chylothorax

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55.1 Introduction

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

55.2 Demographics

There are no known racial, gender, age, or geographical variation to chylothorax. This is due to its aetiology [1]. Chylothorax is a common cause of a pleural effusion to result in respiratory compromise in a neonate. Yet the most common cause of a chylothorax is postcardiothoracic surgery [2] in children, with an increasing incidence reported of up to 6.6%. Risk factors in this group include subclavian vein thrombosis or damage to the thoracic duct.

Congenital chylothorax may occur in 1: 6000–10,000 live births and is typically associated with either lymphatic malformations or in association with chromosomal anomalies, such as Noonan syndrome or trisomy 21. There is a slight male preponderance and right-sided chylothoracies are more common in the congenital variant.

55.3 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 [3].

The chyle contained in the thoracic duct conveys approximately three quarters 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. Total protein content of thoracic duct lymph is also high. When chyle leaks through a thoracic duct fistula, considerable fat and lymphocytes may be lost. The thoracic duct also carries white blood cells, primarily lymphocytes (T cells)—approximately 2000–20,000 cells per milliliter. Eosinophils are also present in higher proportion than in circulating blood. Chyle appears to have a bacteriostatic property, which accounts for the rare occurrence of infection complicating chylothorax [4].

55.4 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 [1].

Chylothorax in older children is rarely spontaneous and occurs almost invariably after trauma or cardiothoracic surgery; [2] 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 venacaval thrombosis, and misplaced central venous catheters (► Box 55.1).

Box 55.1 Causes of Chylothorax

- *Lymphatic malformation (nontrauma)*
  - Thoracic duct atresia/aplasia/hypoplasia/dysplasia
  - Lymphangioma
  - Lymphangiomatosis
  - Intestinal lymphangiectasia (protein losing enteropathy)
- *Thoracic duct injury (trauma)*
  - Cardio-thoracic operations
  - Esophageal atresia
  - Diaphragmatic hernia
  - Penetrating trauma (stab or gun-shot injury)
- *Malignant*

- Lymphoma
- Kaposi sarcoma
- Mediatinal teratoma

- *Infectious*
  - Tuberculosis
  - Filariasis
  - Pneumonia
  - Pleuritis and empyema
- *Idiopathic (associated with)*
  - Down syndrome
  - Noonan syndrome
  - Hydrops fetalis
  - Turner syndrome
  - Lymphedema
- *Miscellaneous*
  - Sarcoidosis
  - Amylodiosis
- *Transudative*
  - Cirrhosis of the liver
  - Heart failure
  - Nephritic syndrome

55.5 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 fetus, a pleural effusion may be secondary to generalized hydrops, but a primary lymphatic effusion (idiopathic, secondary to subpleural lymphangiectasia, pulmonary sequestration or associated with syndromes such as Down, Turner, and Noonan syndromes) 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, hypoproteinemia, 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.

55.6 Diagnosis

Chest roentgenograms typically show massive fluid effusion in the ipsilateral chest with pulmonary compression and mediastinal shift (Fig. 55.1). Bilateral effusions may also occur. Aspiration of the pleural effusion

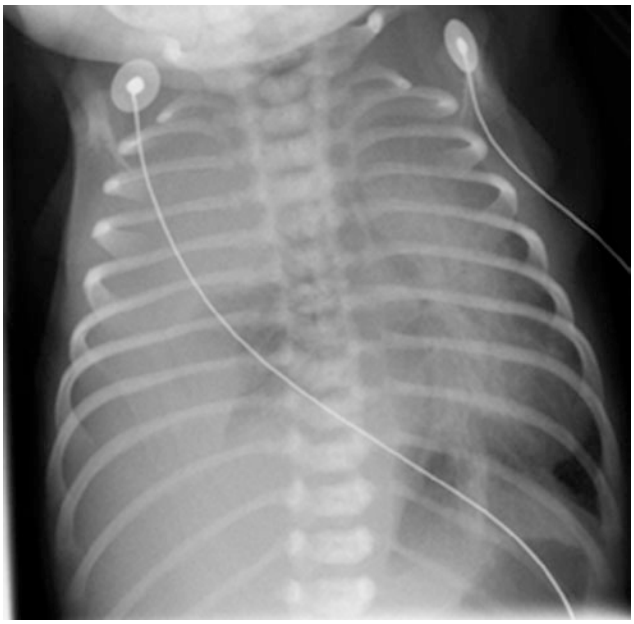

Fig. 55.1 Right-sided congenital chylothorax in a newborn

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, a protein content of more than 5 g/dl, and a ratio of pleural fluid to serum cholesterol less than 1, and the presence of chylomicrons. In a fetus 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. If chyle leak is noted at operation, the proximal and distal ends of the leaking duct should be ligated [5, 6].

Lymphangiography is useful for defining the site of chyle leak or obstruction with penetrating trauma, spontaneous chylothorax, and lymphangiomatous malformation. However, in a nontraumatized patient, the site of lymphatic leakage is often difficult to localize. Lymphoscintigraphy [7] may be an alternative to lymphangiography as it is a faster and less traumatic procedure.

55.7 Management

55.7.1 Conservative

Thoracentesis may be sufficient to relieve spontaneous chylothorax in occasional infants; however, chest tube drainage will be necessary for the majority. Further, tube drainage allows quantification of the daily chyle leak and promotes pulmonary reexpansion, which may enhance

healing. Chylothorax in newborns [8] usually ceases spontaneously. In some cases of congenital chylothorax, supportive mechanical ventilation may be necessary because of insufficient lung expansion, persistent fetal circulation, or lung hypoplasia. In cases of severe chylothorax leading to nonimmunologic hydrops fetalis, antenatal management by intra-uterine thoracocentesis can be considered [9]. 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 result 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) [10]. Cultures of chylous fluid are rarely positive; providing long-term antibiotics during the full course of chest tube drainage is not considered necessary. In nonresolving chylothorax subcutaneous injection of octerotide [11], a somatostatin analogue, at 10 μg/kd/day in three divided doses is reported to have excellent result in a number of case reports and should be tried prior to surgical intervention. There has been demonstrated through numerous studies that conservative management is successful in >80% of cases.

55.8 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 [12]. The leak, if visualized, can be ligated, cauterized, 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 because of the consequences on lung and chest wall growth. If there is concomitant chylopericardium, a pericardial window can be fashioned.

Pleural peritoneal shunts have been reserved for refractory chylothorax [13–15]. 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.

55.9 Prognosis

Prognosis largely depends on the aetiology of the chylothorax. Mortality rate of 12.8% among pediatric patients with a nontraumatic chylothorax has been reported.

55.10 Conclusion

Chylothorax may present as hydrops fetalis or mild respiratory distress. Although the majority of these lymph leaks resolve spontaneously, long-standing chylothorax leads to both nutritional and immunological deficiencies. Conservative medical therapy remains the mainstay of treatment, with surgical intervention required in refractory cases.

Title	Chylothorax in Infants and Children
Authors	James Tutor
Institution	Department of Pediatric Pulmoanry Medicine, University of Tennessee Health Science Center, Memphis, Tennessee
Reference	Pediatrics. 2014;133(4):722–33
Problem	Review of the literature pertaining to the causes, diagnosis, management, and outcomes of congenital and traumatic chylothorax in children
Intervention	Literature review of the current methods of diagnosis and management of chylothorax
Comparision/control	Literature review
Outcome/Effect	Good review paper that is currently the most up-to-date with published views on the management of chylothorax
Significance	Comprehensive, up-to-date review of the management of the pediatric chylothorax

Key Points

1. Chylothorax may be congenital or traumatic.
2. Diagnosis is by means of pleural tap analysis.
3. Optimum treatment includes chest tube drainage, nil by mouth and nutritional support with TPN. Feeding restricted to medium chain triglycerides may be tried in the absence of TPN.
4. Somatostatin analogue may be tried before surgical intervention.
5. Surgery is reserved for the refractory chylothorax with either ligation of the duct where feasible or utilization of a pleural peritoneal shunt.

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Further Reading

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