

## Recurrent Idiopathic Chylothorax in Children: A Case Report

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### Abstract

### Case Report

Chylothorax is a rare disease (1-2% of pleural effusions), with a prevalence between 1/8600 and 1/15,000 births. It is characterized by the presence of chyle in the pleural cavity. Three categories of chylothorax are known: congenital, idiopathic or traumatic (usually postoperative) chylothorax. We report the observation of a 3-year-old child with idiopathic chylothorax revealed by respiratory symptoms, with pleural effusion and ipsilateral lung collapse on chest x-ray and CT. Cytology and chemical analysis of pleural fluid showed exudative fluid with a chylous appearance, high cell count (2800 cells/mm<sup>3</sup>), lymphocyte predominance (98%), and culture was sterile. Chylothorax is usually revealed by dyspnea, but also by nausea, vomiting, anorexia and/ or malnutrition. The diagnosis is suspected when the puncture brings back a milky liquid, confirmed by the presence of a triglyceride level greater than 1.2 mmol/ L and number of cells greater than 1000 cells/ mL, with lymphocyte predominance. Treatment of chylothorax can be conservative or surgical when conservative management fails and aims to stop permanent chyle leakage.

**Keywords:** Chylothorax; Pleural effusion; triglyceride; Medium-chain triglyceride diet; Total parenteral nutrition; Thoracic drainage; Pleuroperitoneal shunting; surgery clamping the breach.

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## INTRODUCTION

Chylothorax is a relatively rare cause of pleural effusion in children, accounting for 1-2% of pleural effusions. It is defined by the presence of chyle in the pleural cavity.

Three categories have been described: congenital chylothorax, which may be malformative or idiopathic [1], and traumatic chylothorax, especially post-operative.

Idiopathic chylothorax is very rare: 34 cases have been reported since 1960, of which only 13 are sufficiently detailed to allow a description of the main semiological features [2]. It occurs outside any malformative or traumatic context, and is reversible in the majority of cases.

In children, it is a potentially life-threatening disorder that may cause profound respiratory, nutritional (hypoalbuminemia), electrolyte (hyponatremia), and immunologic (lymphopenia, hypogammaglobulinemia, T-cell depletion) effects [2].

The diagnosis of chylothorax depends on analysis of the pleural fluid, with identification of chylomicrons or high triglyceride concentrations.

Patients with chylothorax may be treated by conservative means or surgery. Certain principles are common to both treatment options, including the treatment of the underlying cause, the reduction of chyle production, and the provision of adequate respiratory and nutritional support. Although case reports have been published, the optimal management of idiopathic chylothorax in children remains uncertain and may require a specific approach in children. Thus, the aim of the present study was to review the presentation, the different therapeutic approaches, and the outcome of idiopathic chylothorax in children [3].

## OBSERVATION

We report the case of a 3-year-old child. With a history of bilateral inguinal hernia operated on at the age of 2. Who had progressively developed dyspnea on exertion and then at rest, with marked weight stagnation.

On admission, the patient was afebrile, in respiratory distress with sustenance and polypnoea, and signs of malnutrition with muscle wasting.

Clinical examination revealed a right fluid effusion syndrome (abolished MV with dullness of the entire thoracic hemichamber).

Standard radiography showed an opaque right thoracic hemichamber, with left mediastinal discharge (Figure N° 1). The CT scan showed a large right pleural effusion pushing back the mediastinum, with almost total collapse of the right lung (Figure N° 2). Infectious workup negative.

A pleural puncture was performed with a lactescent appearance, then the liquid was sent for biochemical and cytobacteriological study:

WBC 2800 elements/ mm<sup>3</sup> predominantly lymphocytic 98%, without individualization of germs. A PCR study was also performed for *Mycobacterium tuberculosis* complex DNA, which was negative.

The pleural fluid was examined for malignant cells: inflammatory and lymphocytic cytology with reactive mesothelial hyperplasia.

The diagnosis of chylothorax was accepted, and the patient was put on a low-fat diet and endothoracic drainage with a jolly drain. The drain brought back 1700

cc of lactescent liquid over 4 days, then the drain stopped bringing back; clamped for 24 hours with a good control chest x-ray, the drain was removed (Figure N° 3).

1 month later, the child reconsulted with the same symptoms, but with greater respiratory distress and a more marked state of malnutrition. Thoracic drainage was performed with good clinical and radiological improvement, and the patient was discharged with the drain in place. 15 days later, the patient was readmitted with respiratory distress associated with abdominal distension.

Chest X-ray: recurrence of pleural effusion. Abdominal ultrasound showed abundant ascites.

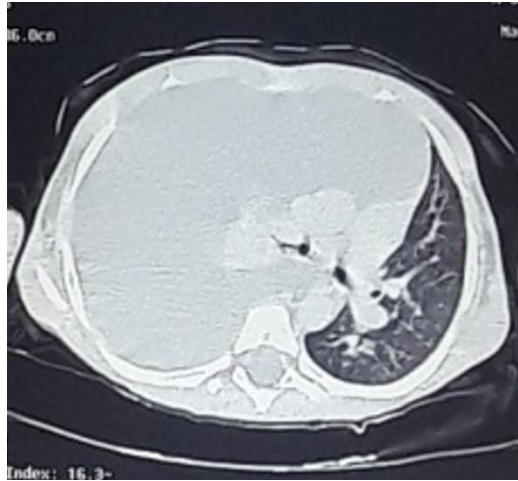
The patient underwent thoracic drainage, with placement of a peritoneal drain bringing back more chylous fluid.

In view of the recurrent chylothorax, the decision was taken to perform a thoracotomy; exploration of the thoracic duct showed the presence of a breach with chyle issuing through, clamping of the thoracic duct with 12 clips with application of spongicel and biological glue. The remainder of the exploration found a disinsertion of the diaphragm, which explains the passage of chyle into the abdominal cavity.

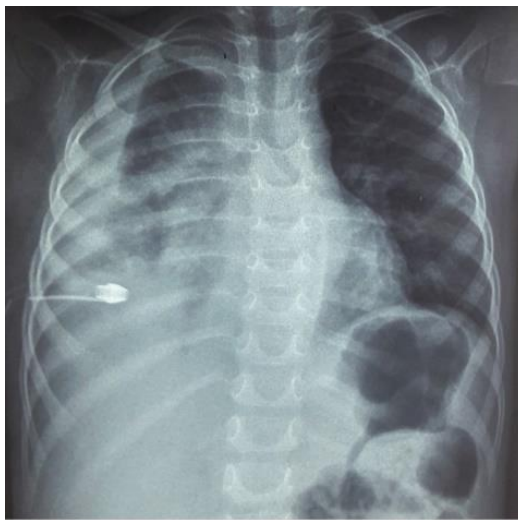
The clinical and radiological evolution was good, with a 3-year follow-up (Figure N°4).



**Figure N°1: chest x-ray revealing abundant right pleurisy with mass effect**



**Figure N°2:** Chest CT scan showing abundant right pleurisy with mass effect



**Figure N°3:** chest x-ray revealing improvement in pleurisy after drainage



**Figure N°4:** chest x-ray disappearance of the effusion after 3 years of surgical treatment

## DISCUSSION

The management of idiopathic chylothorax in children is still a matter of debate, Because of the low

number of cases reported in the literature. The aim of our study was to present our institutional experience of idiopathic chylothorax in children and describe our therapeutic strategie [3].

In this case - about a 3 years old child, with unilateral idiopathic chylothorax - the treatment with MCT diet, associated with TPN, was not able to stabilize the effusion. the conservative treatment failed, and had no place in our conduct, given that symptoms were intense from the onset of the disease. This even led to a spontaneous pleuroperitoneal shunt, because of the high pressure of the pleurisy. therefore, surgery was the only way to stabilize the effusion.

Chylothorax presents as a noninfectious pleural effusion causing nonspecific symptoms such as cough, asthenia or abdominal pain, and sometimes respiratory distress [4, 5].

Classically, pleural fluid analysis reveals a triglyceride level greater than 1.2 mmol/L and an absolute cell count greater than 1000 cells/mL, with a percentage of lymphocytes greater than 80% [4, 5]. Nutritional status and treatment status at the time of the collection of pleural fluid may modify the characteristics of the pleural fluid. It will be milky in the case of a fatty diet and light yellow if the patient has had no oral feeding. our patient presented with high levels of triglycerides, and a lymphocyte concentration greater than 2500/mm<sup>3</sup>, in the pleural fluid.

When the diagnosis of chylothorax is made and there is no apparent etiology, the recommended diagnostic procedures include a CT scan of the chest and abdomen to rule out a malignancy or tuberculosis and to detect associated abdominal lymphangiectasia, heart ultrasonography to look for pericardial effusion, and x-ray of the skeleton to eliminate the bone lysis characteristic of Gorham disease.

Biological assessment containing Serum electrolytes, serum albumin, and complete blood count with differential to look for lymphocyte depletion and hemostasis are not required for diagnosis, but are useful to determine the metabolic and nutritional status of the patient. Pulmonary function tests may show a restrictive ventilatory impairment and hypoxemia (6).

The optimal therapeutic strategy remains controversial, and it may be divided schematically into conservative treatment and surgical treatment. Conservative treatment includes the use of a low-fat diet supplemented with MCT or TPN (4,5).

In our case, an oral MCT-based diet was used initially, then the child was subjected to transient intestinal rest TPN, but without any improvement, contrary to a few studies carried out on chylothorax cases (5,7).

As such a diet has been reported to improve chylothorax [8, 9], TPN may be more effective during the initial period with earlier resolution of the condition [5, 9, 10]. After regression of chylothorax, a low-fat diet

without long-chain triglycerides is maintained to limit lymph flow.

However, such a diet may induce severe malnutrition from deficiency of energy, liposoluble vitamins and essential ( $\omega$ 3 and  $\omega$ 6) fatty acids [11]. Oral or intravenous supplementation of ADEK vitamins and essential fatty acids is also recommended.

Somatostatin can be added to the conservative treatment to improve chylous effusions in postoperative and neonatal chylothorax [12-14]. It reduces gastric, pancreatic, and intestinal secretions; decreases hepatic venous pressure; and reduces splanchnic blood flow.

For the case of significant respiratory distress, a drainage of the effusion by thoracocentesis or chest tube insertion is necessary; However, if the chyle reaccumulates, repeated thoracocentesis or continuous drainage may be associated with protein loss, lymphopenia, hypogammaglobulinemia, and abnormal lymphocyte function.

Surgery for cases that fail respond to initial conservative measures will be required in a minority of patients, and this is associated with a higher risk of complications [15].

Ideal timing for surgery is not clearly defined [4]. Some authors recommend surgery if effusion persists for longer than 2 weeks; others consider a pleural fluid production of greater than 100 mL per year of age in children [16]. Most authors, however, recommend an extended period of conservative management and do not proceed to surgical treatment unless there has been longer than 4 weeks of pleural effusion [4, 8, 17].

The different surgical procedures include chemical pleurodesis, pleuroperitoneal shunting and thoracic duct ligation or ligation of adjacent leaking lymphatics, which is difficult, mainly because of the uncertainty of the site of lymphatic involvement and the difficulty in surgical localization.

Pleurodesis is the method of choice for some authors [18]. Many different drugs have been used for chylothorax, such as fibrin glue, bleomycin, and talc [19–21], the choice of which depends less on scientific reports than on local experience [5].

Pleuroperitoneal shunting was first reported by Azizkhan *et al.*, in 1983 [22]. The principle of this technique is to reabsorb the chyle by the peritoneum. It is usually efficient and well tolerated [23, 24].

In our case, preference was given to ligation and closure of the thoracic duct breaches, as pleuroperitoneal shunting had already been performed spontaneously and had not improved the situation.

In the past, mortality owing to chylothorax has exceeded 50% [25]. Currently, the morbidity and mortality have decreased but remain significant [16]. There are no published data available regarding the outcome of idiopathic chylothorax in children;

In our patient, the chylothorax recurred several times, as we initially tried only conservative treatment and then drainage; but the evolution was miraculous after surgical treatment, the chylothorax disappeared, and the child regained weight.

## CONCLUSION

In practice, there is no universal consensus for the management of chylothorax, but everything depends on the child's initial clinical condition. Some authors advocate conservative management of idiopathic chylothorax in children, with a MCT-based regimen as first-line treatment and TPN as second-line treatment. somatostatin may be combined with TPN; when the chylous effusion has not stabilized after one month of TPN, surgery may be considered. In our experience, the amount of chylous production was enormous, with symptoms of respiratory distress that could not be tolerated, and surgery was the rule.

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