How should chylothorax be managed?

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SUMMARY  The management and complications of chylothorax occurring beyond the neonatal period were reviewed retrospectively. Records from 15 patients treated between 1976 and 1986 were analysed; a combination of thoracocentesis, chest drain insertion, and dietary modification were successful in abolishing chyle leakage in 10 cases. One child died from complications of cardiac surgery rather than from the chylothorax, and surgical intervention was necessary in the remaining four patients and included pleurectomy in three and thoracic duct ligation in the fourth. Lymphopenia, hypoalbuminaemia, hyponatraemia, and weight loss were the most common complications of conservative management and tended to occur in those patients with the longest duration of drainage. Postoperative recovery after pleurectomy and thoracic duct ligation was uneventful. We conclude that conservative management of chylothorax will be successful in most cases. Complications of such a policy are fairly common but rarely serious.

Chylothorax after the neonatal period may be traumatic or non-traumatic in origin. Traumatic chylothorax is most commonly associated with surgery for congenital heart disease or in the posterior mediastinum. Other causes may be accidental and trivial, such as coughing; rarely, it may be part of the range of child abuse. Neoplasm, subclavian vein thrombosis, mediastinal lymphangiectasia, and parasitic infection are examples of non-traumatic aetiology.

Chylothorax in childhood is a potentially life threatening condition yet sufficiently uncommon to ensure that few centres have much experience in its management. Although many authors recommend a conservative approach, some have advocated consideration of early surgical intervention in cases of traumatic chylothorax. There is therefore some debate about the benefits of surgical as opposed to dietary treatment. We have generally employed the dietary approach, in the form of thoracocentesis, chest drain insertion, and dietary modification, with surgery if this fails. We reviewed our last 10 years’ experience of chylothorax to establish the effectiveness of this policy and the risks inherent in both medical and surgical treatment.

Patients and methods

Fifteen patients treated for chylothorax between 1976 and 1986 were identified by means of a hospital diagnostic coding index. Patient records were analysed to determine mode of presentation, underlying condition, duration and volume of chyle leak, details of management employed, incidence of complications, and outcome.

Results

The aetiology, management, and outcome for each case are summarised in the Table.

Aetiology. The median age at presentation was 20 months (range 3 months to 9 years). Twelve children had undergone thoracotomy before development of chylothorax, 10 for congenital heart defects. One child had a left lower lobectomy and another resection of an intrathoracic Ewing’s sarcoma, followed in both by chyle leakage. Ligation of a persistent ductus arteriosus was the single most common antecedent of chylothorax, which complicates 1% of all such operations in our hospital.

Three patients presented without previous surgery. In one of these an upper respiratory tract infection associated with cough seemed to be the precipitating factor and no underlying cause was ever found. The other two patients had abnormalities of the lymphatic system; one had a pleural lymphangioma diagnosed at pleurectomy and the other was a child with Klippel-Trenaunay-Weber syndrome in whom chylothorax followed an attempt to reduce the size of his lymphoedematous leg with occlusive bandages.
Diagnosis. The three children with chylothorax unrelated to surgery presented with a history of increasing shortness of breath over several days. Six of the 12 patients who underwent thoracotomy were diagnosed before they became symptomatic, four because of continuing fluid loss from chest drains inserted at operation and two when pleural effusions were seen on routine postoperative x ray films. Chylothorax in six children who had undergone thoracic surgery presented with increasing dyspnoea from two to as many as 60 days postoperatively (median 16 days).

In 12 patients the pleural fluid had a classical milky appearance. In three others, before enteral feeding had been restarted after surgery, pleural fluid was serous in nature. The extent to which pleural fluid was subjected to laboratory analysis varied from patient to patient. Diagnosis of chylothorax was made on the basis of a number of factors: clinical history, milky appearance of the fluid, continued sterile effusions, chylomicrons visible on light microscopy, 20–50 leucocytes per high power field, with 90% or more being lymphocytes, and fluid and protein content approaching that of plasma. Protein concentration measured in four patients varied from 24 to 55 g/l. All but one effusion was left sided. In addition to chylothorax, one patient developed chylopericardium, which required surgical drainage.

Management. All the children were initially managed conservatively from the time of diagnosis. Initial thoracentesis was always followed by reaccumulation of chyle that necessitated chest drain insertion. Dietary modification was employed in all patients. Twelve were given a low fat high protein diet supplemented with medium chain triglycerides (MCT). Typically, this would consist of vegetables, lean meat or white fish, and bread without butter or margarine. Fried foods other than potato chips cooked in MCT oil were excluded as were cakes and biscuits. Milk was provided as skimmed milk supplemented with carbohydrate (glucose polymer) and MCT oil. Three children received an elemental diet as their only form of nutrition. Chyle drained from the chest was generally replaced with an equivalent volume of intravenous plasma.

Chyle production. We examined the maximum volume of chyle drained in relation to body weight in any 24 hour period. Three patients were receiving parenteral fluids only postoperatively, yet still drained a mean volume of 42 ml/kg/day over two days. In one patient we opted to substitute total

### Table: Aetiology and outcome in 15 cases of chylothorax

<table>
<thead>
<tr>
<th>Age (years/months)</th>
<th>Sex</th>
<th>Precipitating event</th>
<th>Maximum chyle loss in 24 hours (ml/kg body weight)</th>
<th>Total No of days of drainage</th>
<th>Complications</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>50</td>
<td>F</td>
<td>PDA ligation</td>
<td>4</td>
<td>26</td>
<td>—</td>
<td>Resolution, pleurectomy</td>
</tr>
<tr>
<td>0/3</td>
<td>F</td>
<td>PDA ligation</td>
<td>12</td>
<td>12</td>
<td>Low sodium</td>
<td>Resolution</td>
</tr>
<tr>
<td>1/6</td>
<td>F</td>
<td>PDA ligation</td>
<td>5</td>
<td>10</td>
<td>Lymphopenia</td>
<td>Resolution</td>
</tr>
<tr>
<td>0/7</td>
<td>F</td>
<td>PDA ligation</td>
<td>32</td>
<td>8</td>
<td>Lymphopenia</td>
<td>Resolution</td>
</tr>
<tr>
<td>1/8</td>
<td>F</td>
<td>Cavo-pulmonary shunt</td>
<td>9</td>
<td>64</td>
<td>Low sodium, low albumin, weight loss</td>
<td>Resolution</td>
</tr>
<tr>
<td>1/9</td>
<td>M</td>
<td>Cavo-pulmonary shunt</td>
<td>8</td>
<td>15</td>
<td>Low sodium</td>
<td>Resolution</td>
</tr>
<tr>
<td>0/10</td>
<td>M</td>
<td>Cavo-pulmonary shunt</td>
<td>10</td>
<td>7</td>
<td>—</td>
<td>Resolution</td>
</tr>
<tr>
<td>6/0</td>
<td>F</td>
<td>VSD repair</td>
<td>90</td>
<td>11</td>
<td>Lymphopenia</td>
<td>Death</td>
</tr>
<tr>
<td>0/11</td>
<td>F</td>
<td>VSD repair, pulmonary artery band</td>
<td>33</td>
<td>7</td>
<td>Lymphopenia, weight loss</td>
<td>Resolution</td>
</tr>
<tr>
<td>1/4</td>
<td>F</td>
<td>Left Blalock-Taussig operation</td>
<td>86</td>
<td>8</td>
<td>—</td>
<td>Resolution</td>
</tr>
<tr>
<td>0/10</td>
<td>F</td>
<td>Left lower lobectomy</td>
<td>12</td>
<td>26</td>
<td>Lymphopenia</td>
<td>Resolution</td>
</tr>
<tr>
<td>9/0</td>
<td>F</td>
<td>Ewing’s sarcoma</td>
<td>17</td>
<td>7</td>
<td>—</td>
<td>Thoracic duct ligation</td>
</tr>
<tr>
<td>6/0</td>
<td>F</td>
<td>Pleural lymphangioma</td>
<td>39</td>
<td>30</td>
<td>Low albumin, weight loss</td>
<td>Resolution, reaccumulation, pleurectomy</td>
</tr>
<tr>
<td>5/0</td>
<td>M</td>
<td>Klippel-Trenaunay-Weber syndrome</td>
<td>24</td>
<td>29</td>
<td>Weight loss</td>
<td>Resolution, reaccumulation, pleurectomy</td>
</tr>
<tr>
<td>1/9</td>
<td>F</td>
<td>Cough</td>
<td>76</td>
<td>53</td>
<td>Lymphopenia</td>
<td>Resolution</td>
</tr>
</tbody>
</table>

M=male; F=female; PDA=persistent ductus arteriosus; VSD=ventricular septal defect.
parenteral nutrition for an MCT milk feed that was producing chyle flow around 12 ml/kg/day but found no reduction in the rate of leak over the next 48 hours until we reverted to a low fat enteral diet. The median maximal 24 hour chyle drainage in patients on our standard low fat enteral diet was 17 ml/kg.

Outcome. The median duration of chyle leak was 12 days (range four to 64 days) and complications tended to occur in those patients who drained the most chyle (Table). These included loss of more than 8% body weight, hypoproteinaemia, hyponatraemia, and peripheral blood lymphocyte counts that fell two standard deviations below the mean for age.5

Discussion

Chyle in the pleural space may compress intrathoracic structures and embarrass respiration. The first line of management is therefore removal of fluid by needle aspiration followed by insertion of an intercostal chest drain should reaccumulation occur. The appearance of the fluid is classically milky but, as three of our cases showed, may be serous if enteral feeding that includes fat is not being given. A protein content similar to the plasma protein concentration and a preponderance of lymphocytes are further evidence that the fluid is chyle. A lipophilic green dye (coal tar dye, Drug and Cosmetic Green no 6) given with a high fat meal will colour chyle, making it distinguishable from other body fluids and may be used as a diagnostic test where doubt persists.6

As enteral fat will greatly increase thoracic duct lymph flow7 most conservative regimens involve a low fat intake. This may be supplemented with medium chain triglycerides (MCT), which are absorbed directly into the portal venous system without forming chylomicrons.8 Some care should be exercised when using diets high in MCT; essential fatty acid (linoleic acid) deficiency may follow unless linoleic acid, present only in long chain triglycerides, provides at least 4% of energy.

We have tried a number of dietary strategies that involve low fat diet, including supplementation with MCT or parenteral Intralipid (KabiVitrum). We have also used elemental diets that contain most fat as MCT. In our experience, it is not possible to identify any single dietary manoeuvre that is superior in reducing chyle flow. Although total parenteral nutrition might seem to be the logical way to reduce chyle formation, our limited experience suggests that it is not immediately effective. Total parenteral nutrition does, however, provide an alternative method for maintaining adequate nutritional intake and has been used successfully in the management of thoracic diet fistulas by some authors.9

The long term complications of conservative management include the secondary effects of chyle loss—namely, nutritional depletion through loss of protein and electrolytes and immune deficiency through loss of immunoglobulins and lymphocytes.10 Despite careful dietary management and the replacement of chest drain losses, one third of our patients lost more than 8% of their body weight or became hypoproteinaemic. One 18 month old baby developed measles while in hospital, despite having no direct contact with an infected child on the ward at the same time, but otherwise there was no clear evidence that patients became immunocompromised.

If severe, the occurrence of such nutritional complications during the course of conservative management is an argument for surgical intervention. If chyle leakage seemed undiminished after three weeks of dietary management, or when reaccumulation of chyle followed relaxation of dietary fat restriction, surgery was considered. When diet seemed to be steadily reducing chyle leak we were prepared to continue with our medical management for longer than three weeks. Nine of our patients with chylothorax after thoracotomy and the patient with non-traumatic chylothorax all responded to conservative treatment, although in three this took more than three weeks (four, eight, and nine weeks, respectively). We would not agree, therefore, with Selle et al that early surgical intervention should be considered in traumatic chylothorax in children.2 Both patients with underlying lymphatic disorders and one with chylothorax after persistent ductus arteriosus ligation initially responded to dietary treatment but reaccumulated fluid when dietary fat was increased. As the actual point of leakage was impossible to define pleurectomy rather than thoracic duct ligation was successfully employed to prevent further accumulation of chyle within the pleural space. In the case of the child with an intrathoracic sarcoma, chyle leak was noted at the initial tumour resection. As duct damage seemed to be the most likely cause at that time, ligation was performed after only one week of failed conservative management.

We conclude that conservative management in the form of chest drainage and low fat diet will lead to successful resolution of most chylothoraces in children. When such a policy seems to be steadily reducing chyle leak it may be pursued for weeks or months with the likelihood of success, provided that the nutritional state of the child is well maintained. The risks of nutritional impairment, electrolyte

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imbalance, and secondary immune deficiency together with the cost of prolonged admission to hospital must be weighed against the risks inherent in further surgery. Failure of conservative manage-
tment to reduce chyle leak after three weeks or reaccumulation of chylothorax after relaxation of restricted enteral fat intake are indications for surgical intervention. Evidence of underlying lymphatic disorder is probably an indication for earlier rather than later surgical management. If a localised thoracic duct injury is not identified pleurectomy is the surgical procedure of choice.

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