Chylothorax after surgery for congenital heart disease in children

Simone Rolim Fernandes Fontes Pedra, Carlos Augusto Cardoso Pedra, Valmir Fernandes Fontes, Maria Virginia Tavares Santana, Paulo Paredes Paulista

Departments of Pediatric Cardiology and Cardiovascular Thoracic Surgery, “Dante Pazzanese” Institute of Cardiology, São Paulo, Brazil

Abstract We reviewed retrospectively the clinical and surgical data from 9 children with postoperative chylothorax secondary to cardiac surgery for congenital heart disease seen over a 3 year period. Mean age was 26 months and 6 patients were female. The procedures performed were 2 Blalock-Taussig shunts, 6 Bidirectional Glenn anastomoses and 1 modified Fontan procedure. Diagnosis of chylothorax was made by classical means. The overall incidence of chylothorax was 0.8%, with the relative incidences being 2.5% for Blalock-Taussig, 2.7% for Fontan, and 11.7% for the Glenn procedures. All patients were initially treated conservatively with chest tube drainage and hypercaloric diet supplemented with medium chain triglycerides. Parenteral nutrition was needed in 2 patients, one because of sepsis and the other because of progressive malnourishment. High output fistulas and long periods of drainage were noted after cavo-pulmonary procedures. Up to 12% weight loss was observed in 6 patients, one patient was successfully treated of pneumonia and another died of Pseudomonas aeruginosa sepsis. Two patients required chemical pleurodesis because of unabated drainage, with the others being successfully treated conservatively. Chylothorax is a common complication after bidirectional Glenn procedures, and is associated with considerable morbidity and mortality. Earlier operative intervention may be required in these patients. Pleurodesis is a simple, safe, and effective procedure to stop chylous flow.

Key words: Chylothorax; postoperative; congenital heart disease; management

CHYLOTHORAX IS AN UNCOMMON AND serious complication following any thoracic procedure which involves manipulation in the area of the thoracic duct.1-4 It is estimated to occur with an incidence of 0.25 to 0.5% after surgery for congenital heart disease.1,2 Its management has been controversial. Variable results have been obtained using different methods of treatment. Furthermore, studies in children are rare, and numbers are low. Conservative treatment includes chest drainage or repeated thoracocentesis together with an elemental diet and/or parenteral nutrition.3,4 The optimal timing of operative intervention has not been fully established, and several surgical approaches have been proposed, including early reoperation to ligate the fistula or the thoracic duct,12,13 chemical pleurodesis,14-16 placement of pleuroperitoneal shunts,17-20 application of fibrin glue to putative sites of leaks21 and thoracoscopic ligation of the fistula.22 In this study, we report our experience in the management of this difficult problem, with emphasis on the modes of therapy and the morbidity and mortality with which it is associated.

Materials and methods

From January 1991 to December 1994, nine patients developed chylothorax following
Table 1 Patients, cardiac diagnosis and type of operation

<table>
<thead>
<tr>
<th>Case</th>
<th>Age*</th>
<th>Cardiac Diagnosis</th>
<th>Type of operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>22</td>
<td>PA</td>
<td>BT - Left</td>
</tr>
<tr>
<td>2</td>
<td>01</td>
<td>DIV + PS</td>
<td>BT - Left</td>
</tr>
<tr>
<td>3</td>
<td>72</td>
<td>DORV</td>
<td>Fenestrated TCPC</td>
</tr>
<tr>
<td>4</td>
<td>02</td>
<td>TA</td>
<td>Bi - Glenn</td>
</tr>
<tr>
<td>5</td>
<td>22</td>
<td>DORV</td>
<td>Bi - Glenn</td>
</tr>
<tr>
<td>6</td>
<td>50</td>
<td>DIV + PS</td>
<td>Bi - Glenn</td>
</tr>
<tr>
<td>7</td>
<td>32</td>
<td>DIV + PS</td>
<td>Bi - Glenn</td>
</tr>
<tr>
<td>8</td>
<td>14</td>
<td>DIV + PS</td>
<td>Bi - Glenn</td>
</tr>
<tr>
<td>9</td>
<td>14</td>
<td>TA</td>
<td>Bi Caval Glenn</td>
</tr>
</tbody>
</table>

*M: male; P: Pulmonary atresia; PS: Pulmonary stenosis; DIV: double inlet ventricle; DORV: Double outlet right ventricle; TA: Tricuspid atresia; TCPC: Total cavo pulmonary connection.

Table 2 Chylothorax features

<table>
<thead>
<tr>
<th>Case</th>
<th>Interval after operation</th>
<th>Duration of drainage *</th>
<th>Volume of Drainage (+)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2º PO</td>
<td>05</td>
<td>3,6</td>
<td>Successful CM</td>
</tr>
<tr>
<td>2</td>
<td>1º PO</td>
<td>03</td>
<td>3,5</td>
<td>Successful CM</td>
</tr>
<tr>
<td>3</td>
<td>19º PO</td>
<td>18</td>
<td>7,8</td>
<td>Successful CM</td>
</tr>
<tr>
<td>4</td>
<td>18º PO</td>
<td>22</td>
<td>4,4</td>
<td>DW 50% Pleurodesis</td>
</tr>
<tr>
<td>5</td>
<td>19º PO</td>
<td>24</td>
<td>19</td>
<td>Death</td>
</tr>
<tr>
<td>6</td>
<td>14º PO</td>
<td>50</td>
<td>7,2</td>
<td>TALC Pleurodesis</td>
</tr>
<tr>
<td>7</td>
<td>26º PO</td>
<td>25</td>
<td>5,8</td>
<td>Successful CM</td>
</tr>
<tr>
<td>8</td>
<td>23º PO</td>
<td>09</td>
<td>6,3</td>
<td>Successful CM</td>
</tr>
<tr>
<td>9</td>
<td>33º PO</td>
<td>06</td>
<td>13</td>
<td>Successful CM</td>
</tr>
</tbody>
</table>

* Days; (+): ml/Kg/Day; PO: Post operative day; CM: Conservative management.

operations for treatment of congenital cardiovascular disease at “Dante Pazzanese” Institute of Cardiology. The patients ranged in age from 40 days to 6 years (mean: 26 months) and 6 were female. Table 1 shows details of each patient, age at time of surgery, the cardiac diagnosis and the type of operation performed.

The diagnosis of chylothorax was made based on the following characteristics of the pleural effusion: milky appearance, odorless, a protein content greater than one half that of the plasma, triglyceride content greater than 500 mg/dl and positive Sudan III test.5,9

After the diagnosis, all patients were treated conservatively with chest tube drainage and a low fat, hyperproteic and hypercaloric diet supplemented with medium chain tryglicerides. Parenteral nutrition was undertaken if there was deterioration of general condition or a gastrointestinal problem. Those patients submitted to cavo-pulmonary procedures (Bidirectional Glenn anastomoses and the Fontan operation) were also treated with diuretics, inotropes and vasodilators in order to relieve systemic venous pressure overload. The decision to stop the conservative management was made after discussion with the surgical team, although copious chylous flow that persisted longer than 3 weeks was accepted as an indication for surgery.

Results

The 9 cases of chylothorax occurred in a cohort of 1137 operations for correction of congenital heart disease, an overall incidence of 0.8%. The relative incidences were 2.5% for Blalock-Taussig shunts, 2.7% for Fontan operations, and 11.7% for the bidirectional cavo-pulmonary anastomoses.

Table 2 indicates details for each case, the features of the chylothorax, and the final
outcome. The first two cases occurred after Blalock-Taussig shunts, and were diagnosed shortly after the operation. They involved low output fistulas and short periods of drainage. The remaining cases followed cavo-pulmonary procedures, and were diagnosed later than two postoperative weeks. They had high output fistulas and long periods of drainage. All pleural effusions occurred on the left side. In our fifth patient the effusion progressed to involve the right side.

All 9 patients were initially treated in a conservative manner. In the fifth and the sixth patients parenteral nutrition was required. This was because of sepsis in the fifth and because of deterioration of general condition and malnourishment in the sixth. Two patients needed surgical treatment because of copious chylous flow that had persisted for more than 21 days. Both patients were successfully treated by chemical pleurodesis. In the fourth patient hypertonic glucose (DW 50%) was instilled directly through the chest tube, while the sixth patient needed an open thoracotomy, with instillation of talc suspension in the pleural space.

Despite of our efforts to maintain a reasonable nutritional status, 4 patients showed low levels of seric albumin, and 6 had a weight loss of between 4 and 12%. Pneumonia was diagnosed and successfully treated in the sixth patient, but the fifth patient developed Pseudomonas aeruginosa sepsis, and died on the 24th day of drainage.

After a mean follow up of one year and a half, all surviving patients were free of recurrences, including those treated with pleurodesis.

Discussion

After cardiac surgery, chylothorax is classically described as a complication of extra pericardial operations, especially those involving mobilization of the aortic arch, including ligation of the arterial duct, repair of vascular rings and coarctation of aorta, and Blalock-Taussig shunts. In these procedures, the thoracic duct (or its tributaries) is directed injured during the process of dissection. Because there is a great variability in the anatomic distribution of the lymphatic channels (with some passing through the thymic tissue), chylothorax can also, but infrequently, follow intrapericardial procedures. Systemic venous hypertension due to increased right-sided cardiac pressure is another cause of chylous leakage to the pleural space. The elevated pressure is transmitted to the lymphatic system, retards the flow through the thoracic duct, and promotes dilation of the lymphatic vessels with eventual rupture to the pleural space. Since systemic venous hypertension is the rule in caval-pulmonary procedures (bidirectional Glenn anastomoses and Fontan operations), chylothorax is not uncommonly found following these procedures. In keeping with this observation, we found an incidence of postoperative chylothorax in our patients submitted to the bidirectional Glenn anastomoses, higher than for our Blalock-Taussig procedures. Mustard and Senning operations, particularly when associated with superior caval venous obstruction, are other causes of chylothorax due to similar mechanisms.

As highlighted by others, we found a clear difference in the clinical picture between traumatic chylothorax and chylothorax secondary to systemic venous hypertension. The latter had a longer interval between operation and diagnosis, with a higher volume and a longer period of chylous drainage. This phenomenon can be explained in several ways. When there is systemic venous hypertension, more time is required for dilatation and rupture of the lymphatic vessels in the mediastinum and pleural space. Thus, the interval between the operation and diagnosis is longer. Systemic venous hypertension also increases lymphatic pressure, stimulating lymph production and impairing its drainage. This sustained pressure overload is also responsible for higher volumes and longer periods of drainage. It is unlikely that the longer duration observed in our patients was related to the type of treatment, as we followed the same protocol of management in all.

Opinions differ regarding the respective advantages of using a diet rich in medium chain triglycerides, or total parental nutrition, in the treatment of this condition. It is known that medium chain triglycerides are absorbed directly into the portal system, bypassing the intestinal lymphatics and thus reducing thoracic lymphatic flow. This has led to their use in nutritional support of patients with chylothorax. Others, however, have demonstrated that either medium chain triglycerides, or even water alone, can increase chylous flow. Some, therefore, advocate enteric rest and total parenteral nutrition, although this approach has not been shown to reduce the duration of chylous drainage. In our protocol for conservative treatment, we avoided the use of parenteral nutrition as a first line of therapy because of the risks associated with this therapeutic modality. Patients with chylothorax after cardiac surgery are generally prone to infections because they are immunosuppressed due
to the malnutrition associated with the basic cardiac disease. This is exacerbated by the intense postoperative catabolic state and the huge loss of proteins and immunoglobulins through the fistula. If one employs enteric rest with total parenteral nutrition, it is obligatory to insert a central venous catheter to provide adequate calorie support. As it will remain in place for a considerable period of time, this can greatly increase the risk of infection, specially bacteremia or sepsis related to the catheter, as observed by others. Moreover, the catheter itself is thrombogenic and can eventually lead to caval venous obstruction, worsening substantially the final outcome. Other arguments against the sole use of total parental nutrition are the recent evidences that one of the major functions of the gastrointestinal tract is immunological. Bowel rest results in gastrointestinal mucosal atrophy which compromises the integrity of the mucosal barrier and enhances exposure to endogenous gut bacteria and/or toxins. In our opinion, parenteral nutrition should be employed only in cases when it is impossible to use the gastrointestinal tract, or when there is an obvious and progressive weight loss despite the use of an hyperproteic, hypercaloric diet supplemented with medium chain triglycerides. Even then, if it is possible, we try to avoid a central venous line, favoring a peripheral approach.

Administration of albumin or colloidal solutions may be imperative if albumin is low in the serum and there is low oncotic pressure. Patients with systemic venous hypertension should also receive inotropic support associated with diuretics and vasodilators in order to relieve systemic venous pressure overload.

As the spontaneous closure of a thoracic lymph fistula is unpredictable, timing of operative intervention has not been fully established. Most authors recommend an extended period of conservative treatment, but the crucial decision is: how long to continue with the conservative management? Copious chylous flow that lasts for more than 2–3 weeks, or more than 100 ml per day per year of age for 5 days, or more than 100 ml/kg/day, and severe nutritional, metabolic or immunological disturbances are generally accepted as guidelines to stop the conservative management. Operation is also indicated when the lung is “trapped” and requires decortication, usually as a result of delayed diagnosis. Recently, Bond and co-workers identified risk factors associated with failure of non-operative management. They showed that patients with elevated central venous pressure, or superior caval venous thrombosis, were not likely to improve with conservative therapy. They recommended prompt identification of these risk factors in order to consider earlier operative intervention. On the other hand, Nguyen and colleagues didn’t notice this correlation between central venous hypertension and the need for earlier intervention, even using the same initial approach, with total parental nutrition as the sole conservative treatment.

Since its original description by Lampson and co-workers in 1948, the ligation of the thoracic duct (or one of its tributaries) substantially improved the morbidity and mortality in patients with chylothorax. Although this technique has been reported to be successful in 90% of cases, it requires a new and extensive thoracotomy and is also associated with technical difficulties. Because of this, new surgical approaches were developed. The use of pleuroperitoneal shunts in the treatment of refractory chylothorax in neonates was introduced by Azizkhan and colleagues in 1983. In 1989, the same group published a larger series, and noted that chylous flow was successfully treated in 92% of cases. The new method provided retention of the nutritional and immunological elements of the chyle. Thus, they recommended early intervention after only 5 days of drainage. Nevertheless, the procedure failed to relieve chylothorax secondary to caval venous thrombosis. Alternative forms of therapy were advocated in this circumstance. Although the Fontan operation is associated with marked elevation of both superior and inferior caval venous pressures, Sade and coworkers successfully employed a pleuroperitoneal shunt for persistent pleural drainage after such procedure. This simple technique is a reasonable alternative in experienced centers. Recently, successful thorascopic identification of the site of chylous leak and its ligation has been described in adults. It is also an attractive option of treatment due to its less invasive nature.

The idea of treating chylothorax by promoting chemical pleurodesis is not new, having been introduced in the 1940’s. Since then, several substances have been shown to produce a sterile inflammation of the pleura culminating with adhesions between the visceral and parietal layers. In this way the leak of chyle can be sealed off, maintaining full expansion of the lung. In our study, the two patients managed in this manner achieved rapid and effective closure of their fistulas, with no recurrences in the medium
term. The procedure itself is technically simple, safe, less invasive than ligation of the fistula or the thoracic duct and, probably, at least as effective.

Although the limitation of our study is its small number of patients, our experience has shown that chylothorax is a common complication following bidirectional Glenn procedures. Conservative management was successful in the majority of the patients, although it was associated with considerable morbidity and mortality. Recognizing that elevated central venous pressure is a risk factor for failure of nonoperative management, patients with cavo-pulmonary type of anastomoses should be considered for earlier surgical intervention in order to avoid serious complications from chylous losses. Pleurodesis is technically simple and can be safely and effectively employed to achieve this goal.

Acknowledgments

We thank Dr. Paulo Zelinsky who encourage us to report this experience, and Sandra Regina Ferreira Mourão for her secretarial assistance in preparing this paper.

References

33. Frazer AC. Differentiation in the absorption of olive oil and oleic acid in the rat. J Physiol (Lond) 1943; 102: 308-312.


