

ENDOSCOPIC PLACEMENT OF A PLEUROPERITONEAL SHUNT

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This paper describes a minimally invasive surgical technique to place a pleuroperitoneal shunt in a 15-year-old boy with primary lymphoedema. The patient had undergone multiple thoracenteses and chest tubes for a recurring left pleural effusion. In addition, he suffered from multiple abdominal lymphoceles and episodes of spontaneous bacterial peritonitis. This previously unreported minimally invasive technique to successfully place a pleuro-peritoneal shunt, allowed the patient to be discharged from hospital free of oxygen and external tubes. Information was obtained about the patient's abdomen which will guide future therapy.

Case report

During a three-year period, a 15-year-old boy with primary lymphoedema presented with recurrent left pleural effusions and persistent dyspnoea at the authors' hospital. He required two litres of oxygen continuously by nasal cannula to maintain his oxygen saturation above ninety-five per cent. He suffered two attacks of spontaneous bacterial peritonitis, one at age 13, and one at age 14, which resolved with antibiotic

therapy. Chest and abdominal computed tomography (CT) scans, performed at the time of the second attack of spontaneous bacterial peritonitis (age 14), revealed a left pleural effusion and multiple loculated lymphoceles (Figures 1 and 2). The patient underwent multiple thoracenteses, chest tubes and paracenteses for symptoms of dyspnoea and abdominal distension.

Varicosities may affect the superficial, deep, and perforating venous systems and may remain stable in size or gradually expand.

His past medical history was significant for the initial presentation of lymphoedema at age 12 (lymphoedema praecox), when he presented with a massively swollen left lower extremity. After suffering multiple episodes of cellulitis, progressive fibrosis and decreased mobility, he underwent a left lower extremity amputation. Following this, his abdominal and thoracic symptoms worsened, possibly because of increased accumulation of lymph in the proximal tissues and decreased absorption into the proximal lymphatics. Investigations also revealed chyluria and protein-losing enteropathy and he became cachectic. In order to experience less dyspnoea, he agreed to undergo placement of a pleuroperitoneal shunt.

Operative technique

After administration of general anaesthesia and antibiotics, the patient was placed in the right decubitus

position to better access the left pleural effusion and avoid the mediastinal structures. A 5mm port was inserted in the left posterior axillary line at the sixth intercostal space. Carbon dioxide was administered into the left chest cavity to a pressure of 9mmHg to compress the lung and create a space to visualise shunt placement. Chronic chylothorax was observed (Figure 3). The pleural portion of a pleuroperitoneal shunt (Denver Biomedical pleural effusion shunt, model 42-9000) was placed through a separate incision and guided to a satisfactory position by the 5mm, thirty-degree camera (Figure 4).

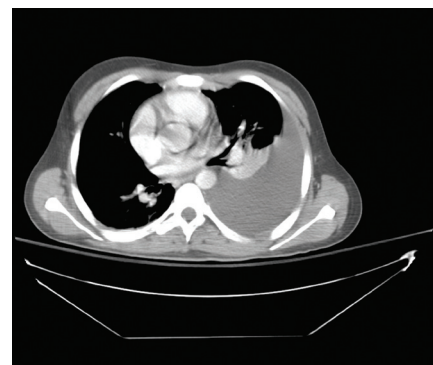


Figure 1. Computed tomography image of left pleural effusion (chronic chylothorax).



Figure 2. Computed tomography image of abdominal lymphoceles.

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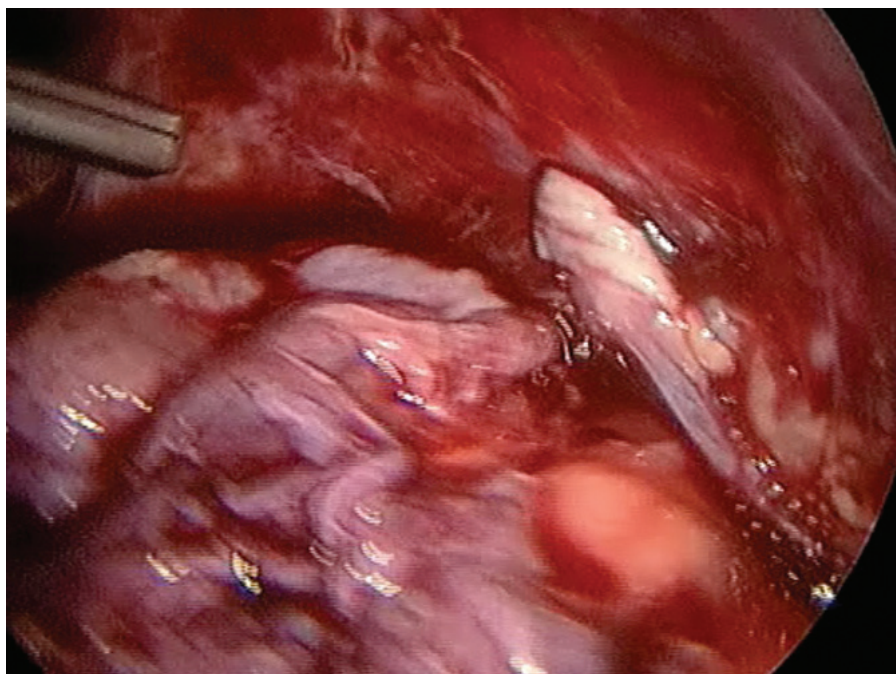


Figure 3. Thoracoscopic view of left pleural effusion (chronic chylothorax).

The shunt and pump chamber were tunnelled under the skin. The pump chamber contained a one-way valve and was sutured over a rib to provide a solid surface for compression. A 5mm incision was made in the left upper quadrant, a purse string suture was placed in the posterior rectus sheath and the 5mm port was inserted into the abdomen under direct vision. Pneumoperitoneum was obtained to a pressure of 13mmHg and the camera was inserted. Multiple adhesions and lymphoceles were observed in the abdominal cavity. With laparoscopic guidance, an area was dissected in which to place the abdominal portion of the shunt through a separate 5mm incision. The pump chamber was manually compressed and pleural fluid flowed into the abdomen.

Following the procedure, the patient experienced immediate relief of dyspnoea. He compressed his shunt for five minutes, four times daily, and this has been ongoing after discharge from hospital. He was discharged from hospital forty-eight hours after the procedure and has not required oxygen, hospital admission or thoracentesis for two years. His cachexia resolved.

Discussion

Recent molecular genetic studies have shown a correlation between abnormal lymphatic endothelial cell formation, lymphangiogenesis, lymphatic function and primary lymphoedema (Ji, 2008). Abnormalities of forkhead-related transcription factor (FOXO2), Ephrin B2, vascular endothelial growth factor receptor (VEGFR-3), antiangiogenic factor (angiopoietin-w, Prox-

I and podoplanin have proved to be important factors of the genetic cascade linked to dysfunction of the embryonic and postnatal development of the lymphatic system (Ji, 2008). A decrease in lymph transport leads to the clinical manifestations of primary lymphoedema (Ji, 2008).

Abnormalities of these factors result in a heterogeneous group of disorders and complex syndromes (Connell et al, 2010), whose pathogenesis in many cases has not been well delineated. Milroy's disease, congenital familial lymphoedema, was first described in 1892 (Milroy, 1892). Within the last decade it has been linked to an abnormal mutation of the vascular endothelial growth factor receptor (VEGFR-3) (Karkkainen et al, 2000).

Within the spectrum of lymphatic vascular disease, certain circumstances mandate specific interventions. Surgeons are frequently consulted to manage the manifestations of lymphoedema which include chylous ascites, chylothorax and lymphatic malformations. Patients may undergo multiple thoracenteses and paracenteses in order to control symptoms of dyspnoea and abdominal distension (Faul et al, 2000; Aalami

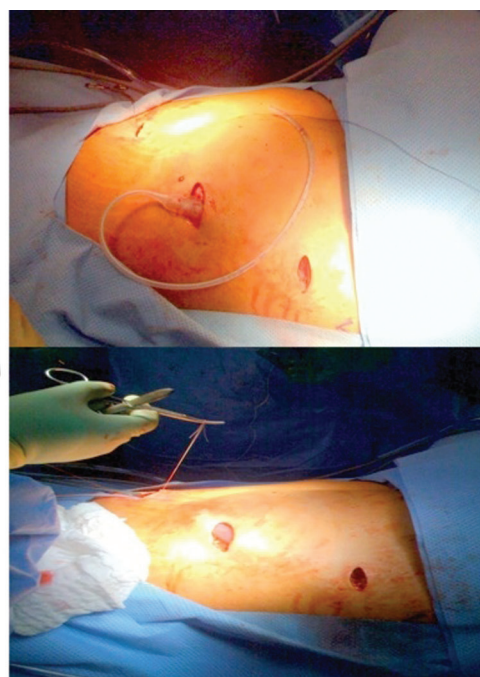
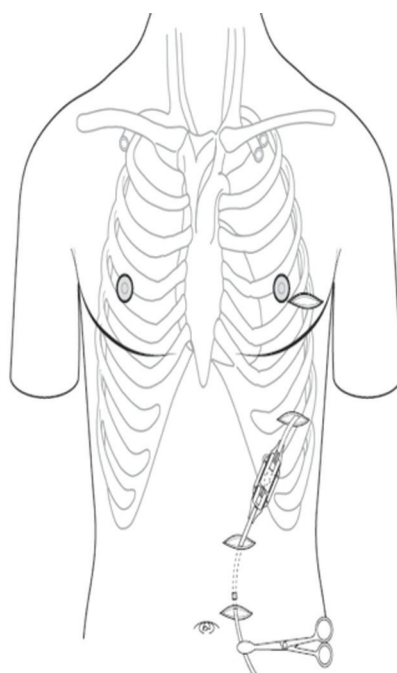


Figure 4. Incisions for pleuroperitoneal shunt placement.

et al, 2000). Since its introduction in 1982, the pleuroperitoneal shunt has been used with success by surgeons to control symptoms of pleural effusion and lymphoedema (Weese and Schouten, 1982). Engum et al (1999) reported the use of pleuroperitoneal shunts in the management of persistent chylothorax in seven infants (congenital lymphoedema), in which complete resolution of pleural effusions was seen in five.

To the authors' knowledge, this is the first report of adaptation of minimal access surgical techniques to manage the symptoms of dyspnoea in a patient with hereditary lymphoedema and a pleural effusion. The thoracoscope guided the positioning of the shunt and allowed adequate dissection to assure lung expansion. Multiple adhesions and lymphoceles were noted in the abdominal cavity. In the future, the authors will avoid the use of the veress needle and continue to access the abdomen directly.

In the authors' opinion, if the shunt malfunctions or becomes infected, the patient in this case may experience episodes of spontaneous bacterial peritonitis, ascites, pain and protein-losing enteropathy. Therefore, the patient and his family were instructed to present for outpatient follow-up annually and to go to the emergency room if experiencing pain, fever or dyspnoea.

Conclusion

Primary lymphoedema is a chronic disease, often associated with a mutation in a gene associated with the lymphangiogenesis process, which

leads to abnormal lymphangiogenesis in utero. A diffuse dysfunction of the lymphatic system occurs and presently, no technique is available to address this dysfunction at the molecular level (Ji, 2008, Ferrell et al, 2010). Currently, molecular models are undergoing investigation. Until such models are ready for human application, there will be a role for surgical intervention for symptoms of lymphoedema. In this case, the application of minimally invasive surgery to place a pleuroperitoneal shunt, allowed the physical, nutritional and psychosocial aspects of the illness to be addressed. **JL**

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Key points

- ▶▶ Primary lymphoedema is due to abnormal lymphangiogenesis and causes diffuse impairment of the lymphatic system.
- ▶▶ The genetic cascade linked to abnormal lymphangiogenesis is undergoing study, but no models are ready for human application.
- ▶▶ The cornerstones of therapy are conservative, but surgeons are frequently consulted to manage lymphoedema manifestations including chronic chylothorax and dyspnoea.
- ▶▶ Sporadic case reports have documented the use of pleuroperitoneal shunts for this symptom.
- ▶▶ This case reports the successful adaptation of minimal access surgical techniques to expand the use of pleuroperitoneal shunts in the setting of primary lymphoedema, chronic chylothorax and dyspnoea.