

Chyluria and chylothorax after posterior selective fusion for adolescent idiopathic scoliosis

Alexander A. Weening¹ · Bernadette Schurink^{2,3} · Jelle P. Ruurda³ · Richard van Hillegersberg³ · Ronald L. A. W. Bleys² · Moyo C. Kruijt¹

Received: 3 November 2016 / Revised: 7 March 2017 / Accepted: 23 March 2017 / Published online: 5 May 2017
© Springer-Verlag Berlin Heidelberg 2017

Abstract



Purpose To describe and discuss the diagnostic and treatment complexity of lymphatic system complications after scoliosis surgery.

Methods Surgery for adolescent idiopathic scoliosis is very commonly performed with posterior pedicle screw instrumentation. Complications of the anteriorly based lymphatic system are, therefore, rare. We present a case with complications related to the lymphatic system, which have not been reported before after this type of surgery.

Results After standard Th3 to Th12 posterior spinal reduction and fusion of a moderate thoracic curve, chyluria and a chylothorax developed in an adolescent girl. This appeared to be caused by an obstruction of the thoracic duct. Thorax drainage and finally thoracoscopic intervention prevented further pulmonal impairment. The exact cause could not be identified and the persistent lymph drainage problems had to be treated with a medium chain triglyceride diet.

Conclusion With this report, we aim to create awareness of the lymphatic system in general and the possibility of severe complications, even after a posterior only approach of the vertebral column.

Keywords Scoliosis · Surgery · Chyluria · Chylothorax · Thoracic duct

Case presentation

An 18-year-old female was surgically treated for a 65°, right thoracic scoliosis. Her medical history was otherwise confined to an incidentally found and angiographically confirmed arteriovenous malformation (AVM) at the level of the eighth thoracic (Th) vertebra. Posterior spinal fusion Th3 to Th12 went uneventful (Fig. 1a, b). Two days after surgery, the urine catheter was obstructed by heavy flocculent urine with a milky aspect. Diagnostic workup could not confirm a nephrotic syndrome, but a high urine concentration of triglycerides (20.73 mmol/L) was found, characteristic for chyluria. Subsequent lymphoscintigraphy showed exceptional lymph drainage from both legs via the inguinal nodes and a thin retroperitoneal system, directly to the left kidney and bladder. No tracer entered the thoracic duct. It was assumed that lymph drainage was blocked

✉ Alexander A. Weening
a.a.weening@hotmail.com

¹ Department of Orthopedic Surgery, University Medical Centre Utrecht, Heidelberglaan 100, 3584 CX Utrecht, The Netherlands

² Department of Anatomy, University Medical Centre Utrecht, Utrecht, The Netherlands

³ Department of Surgical Oncology, University Medical Centre Utrecht, Utrecht, The Netherlands

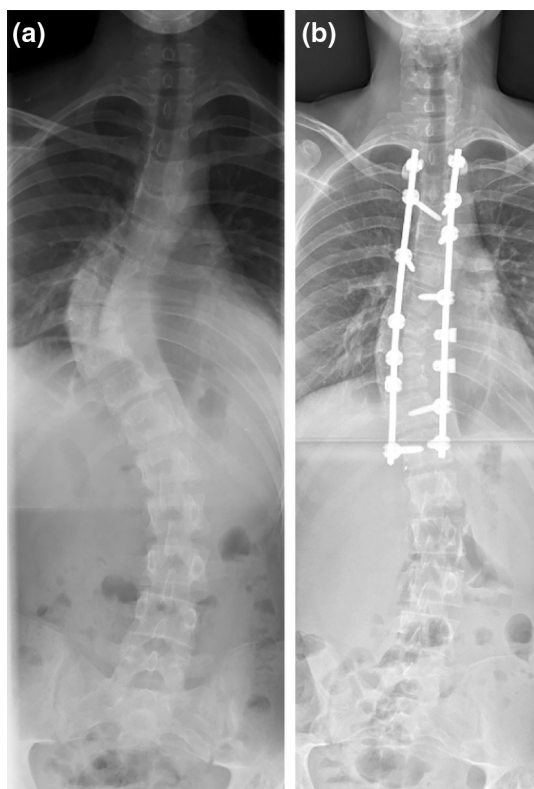


Fig. 1 **a** 65°, right thoracic scoliosis. **b** Posterior spinal fusion Th3 to Th12

proximally. An MCT diet was started and patient was followed-up in the outpatient clinic. A CT scan done here revealed an unexpected left-sided pleural effusion and a 2 mm anterior protrusion of the left Th5 pedicle screw. Upon drainage, the effusion appeared to be chylous, with a triglyceride level of 35.63 mmol/L. Daily, 1–2 L of chyle continued to leak from the chest drain and immediately after drainage the chyluria ceased. A subsequent ultrasound-guided intranodal lymphangiogram showed lymph drainage towards the cisterna chyli at the level of Th12, dilated intercostal branches and diffuse collateral leakage in the left hemithorax. Again, the thoracic duct could not be visualized (Fig. 3a, b).

Diagnostic imaging section

See Figs. 1, 2, 3.

Historical review of the condition, epidemiology, diagnosis, pathology, and differential diagnosis

The lymphatic system carries approximately 3 L of lymph every day, which contains high amounts of essential triglycerides, cholesterol, proteins, and T-lymphocytes [1].



Fig. 2 CT scan, transverse section—left Th5 pedicle screw, with a 2 mm anterior protrusion and a left-sided pleural effusion (after drainage confirmed to be chylous)

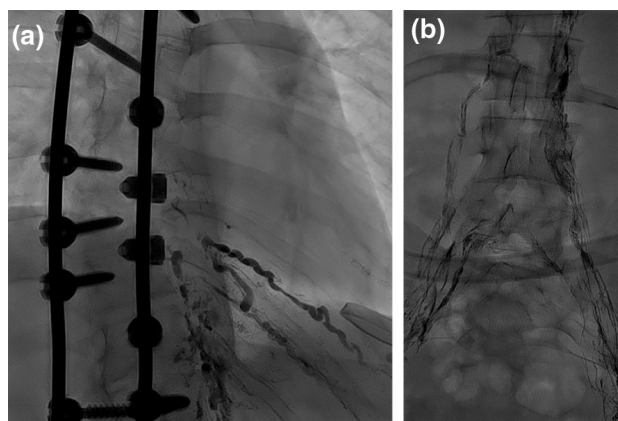


Fig. 3 **a** There is diffuse left collateral drainage and leakage in the thorax. Filling of the thoracic duct could not be visualized. **b** Lymphangiogram shows lymph drainage to the cisterna chyli

The thoracic duct is the main terminal lymph vessel, collecting lymph from the entire body except for the right upper quadrant [2]. It terminates in the venous system at or near the junction of the left internal jugular and subclavian veins, where a valve system prevents venous backflow [3]. Conditions affecting this system can be life-threatening as they may result in severe nutritional depletion and an impaired immune system [3, 4]. Damage to the lymphatic system usually results in obstruction and/or leakage of chyle and can manifest in different ways such as: (severe) lymphedema, so-called Elephantiasis, chylous ascites, chyluria, and chylothorax. In this report, we describe the two latter manifestations after scoliosis surgery.

Chyluria is the discharge of milky urine due to a lymphatic system abnormality. Causes can be either tropical or nontropical [5]. The former mainly occurs in Asia and is the result of a parasitic infection with *Wuchereria Bancrofti* that causes lymph vessel damage and obliteration via multiple mechanisms [6, 7]. Subsequent backflow causes distal lymphatics to dilate and lymphatico-urinary fistulas, most commonly located at the renal fornices, are formed

[6, 8–10]. Nontropical causes are rare and include congenital lymphatic malformations or obstruction of the lymphatic system, which can be either by (malignant) processes or iatrogenic [11].

Chylothorax has multiple causes that can be classified as medical or surgical [12]. The leading medical cause of chylothorax is a malignant process. Most commonly, this includes lymphomas or bronchogenic carcinoma [12]. Obstruction of or infiltration into the thoracic duct can increase intraluminal pressure, which leads to retrograde lymphatic flow and formation of a dilated collateral system. These vessels may rupture causing leakage of chyle into the pleural cavity [13, 14]. Other medical causes are trauma or spontaneous chylothorax, attributable to a sudden increase in duct pressure, such as coughing or vomiting [12].

Diagnostic workup of both conditions starts with evaluation of the milky fluid for triglyceride levels. Hereby, chyluria can be differentiated from nephrotic proteinuria, pyuria, and crystalluria, and screening for filariasis is important to exclude an infection with *Bancrofti* as a possible cause [15]. Empyema or cholesterol pleural effusion can mimic the white chylous liquid present in chylothorax [16]. Early diagnosis can be difficult, as not all effusions have the classic milky appearance; this is present in only 50% of patients with chylothorax [12].

Radiologic workup includes CT, which can provide clues for diagnosis (e.g., infiltrating tumors, malpositioned surgical implants), and nuclear imaging such as lymphangiography or lymphoscintigraphy when lymphatic (congenital) abnormalities or change in normal chyle flow (obstruction, ruptures) are suspected [11, 17].

Rationale for treatment and evidence-based literature

Based on the CT scan showing protrusion of the screw, lymphangiograms that showed no filling of the thoracic duct, and absence of obvious external congenital abnormalities, such as large lymphangiomas or hemangiomas, we hypothesized that in our patient, an obstruction in the thoracic duct should be the leading cause. Whether this was due to the (minimal) protrusion of the Th5 screw, the 50% correction of the curve and/or the presence of the AVM around Th8 could not be determined. As the chylothorax did not resolve with conservative measures including total parenteral nutrition (TPN) for 4 weeks, we exchanged the prominent screw for a shorter one. However, this did not solve the problem. In addition, the lung did not fully expand, even with permanent suction on the chest tube. Therefore, after another 3 week trial of TPN with

somatostatine at a final dosage of 13.2 mg/day, we decided to explore the left pleural cavity.

Conservative treatment is the first line of treatment for a chylothorax. Chest tube drainage and suction will obliterate the pleural space and by decreasing the chyle load a defect can heal [18]. This can also be accomplished with an MCT diet or total parenteral nutrition. Medium chain triglycerides are absorbed directly into the portal system and hereby diminishing the load on the lymphatic system [16].

Surgical management is advised when conservative treatment fails, if an imminent nutritional deterioration exists, or if the lung re-expansion is severely hindered by the formation of a fibrous membrane [3]. Surgical treatment should be directed at the underlying cause: infiltration and obstruction should be treated and/or ongoing leaks should be identified, sealed, or ligated at a more upstream level. Pleurodesis or pleurectomy is reserved as a salvage procedure for patients in whom the leak or the thoracic duct cannot be identified [19]. Pleuro-peritoneal shunting is described but has a non-structural character with a manual pumping system and a relative high percentage (10%) of occlusion [20]. When pulmonary expansion is compromised, a decortication and mobilization of the lung can be performed through a VATS, an open approach or a robot-assisted intervention as in this case [21]. Minimally invasive treatment options for chylothorax include thoracic duct embolization or needle interruption [14, 22].

The optimal timing of surgery or duration of conservative treatment is under debate. It is important to keep in mind that lymphatic defects can heal spontaneously; however, early surgery could prevent patient deterioration as may occur during prolonged conservative treatment. Decision making on when to intervene is based on the nutritional state of the patient and leakage rates [12, 16, 21].

In their review, Misthos et al. advocate early intervention in patients with leakage rates exceeding 1000 mL/24 h during the first seven postoperative days, and intervention after 1 week if output levels do not drop below 250 mL/day [21]. However, cut-off values for leakage rates vary, with other authors opting for surgical intervention when drains produce >1.5 L/day, >1 L/day for 5 days, or when leakage persists 2 weeks after surgery [12, 23].

The treatment regime for chyluria is similar, with initial conservative management followed by surgical treatment in patients with severe refractory chyluria. Severity is judged based on a number of factors, such as persistence of symptoms despite dietary measures and a history of chylous cloths causing colics and urinary retention [24]. Surgical options comprise sclerotherapy or renal pedicle lymphatic disconnection among others [11].

Procedure (surgery, intervention)

After 2 months of non-successful conservative treatment, we performed a left pleural cavity exploration with the aid of the daVinci Intuitive robot. After a fat-challenge, obvious leakage of several large collaterals along the tenth rib and near the AVM at Th8 was identified and these collaterals could be ligated successfully (Fig. 4). A decortication was performed, and because of the successful ligation, no formal pleurodesis was done.

Procedure imaging section

See Fig. 4.

Outcome, follow-up

After surgery, the patient was kept on TPN for another 2 weeks and continued an MCT diet for a period of 3.5 months to minimize chylous load. During this period, she experienced no macroscopic chyluria, no respiratory problems, and no signs of weight loss. Considering this stable clinical situation, a normal diet was tried again but unfortunately the chyluria re-appeared.

Surgical damage to the thoracic duct is not uncommon. During thoracoscopic esophagectomy, the thoracic duct is often ligated to reduce the incidence of postoperative chylothorax [25]. Lymph normally finds its way towards intercostal lymph vessels and pre-existent dilated collateral vessels or enters the venous circulation via lymphaticovenous anastomoses [13].

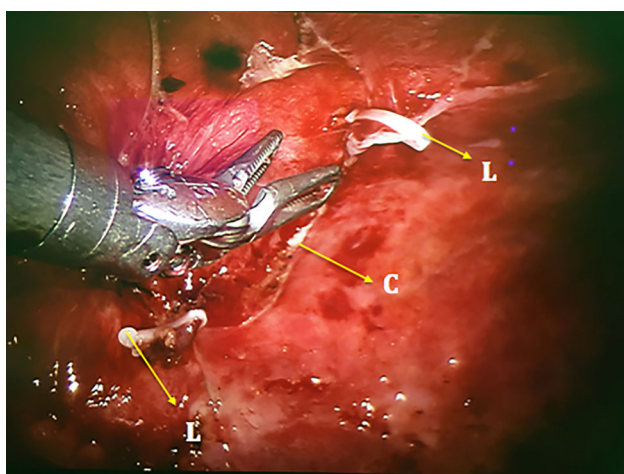


Fig. 4 Left-sided robotic thoracoscopic exploration. Several collaterals along the tenth rib and near the AVM at Th8 could be identified and ligated. *C* collateral, *L* ligature

Why this physiological phenomenon did not occur in our patient remains obscure. In terms of occlusion, there should not be much of a difference between an elective ligation and iatrogenic compression of the thoracic duct, other than the level of obstruction. Therefore, we hypothesize that the level of obstruction may be a key feature in our case. Where upper-GE surgeons clip the thoracic duct just above the level of the diaphragm (Th12), the level of obstruction in our case may have been much higher. With a higher obstruction, at Th8 where the AVM was found and where the apex of the curve was situated or even higher at Th5 where the screw was prominent, the amount of collaterals connected to the duct above the obstruction which can provide a functional bypass, decreases [26]. Consequently, the resistance will be higher, causing backflow (chyluria) or rupture of the collateral vessels (chylothorax).

Another hypothesis is that the patient indeed suffered from a lymphatic anomaly, where the AVM may have been a first indication. Both the vascular and the lymphatic system have a common embryologic origin; lymphatic vessels are capillary offshoots from the endothelium of veins and may follow a metameric distribution [27, 28]. Mixed combinations of both vascular and lymphatic malformations are rare but a few cases are described [28–30]. These reports further suggest that a chronic elevation in venous pressure due to an AVM causes hyperplasia of lymphatic ducts and the formation of collateral vessels. These vessels are fragile and changes in anatomy (e.g., during surgery) can easily cause rupturing of the distended lymphatic system [28].

The re-appearance of chyluria in our patient when eating normally implies that the collateral system is still insufficient and we are searching for a balance between moderate chyluria and an acceptable diet. Since she is in good shape, we see no other option than to wait for the collateral system to further adapt.

Compliance with ethical standards

Conflict of interest All authors declare that they have no conflict of interest.

Source of funding This paper and research have not been funded.

Informed consent Informed consent has been obtained from the case study, and therefore, participant included in this study.

Human participants and/or animals All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

- Phang KL, Bowman M, Phillips A, Windsor J (2014) Review of thoracic duct anatomical variations and clinical implications. *Clin Anat* 27(4):637–644
- Skandalakis J, Skandalakis L, Skandalakis P (2007) Anatomy of the lymphatics. *Surg Oncol Clin N Am* 16(1):1–16
- Talwar A, Lee HJ (2008) A contemporary review of chylothorax. *Indian J Chest Dis Allied Sci* 50:343–351
- Dumont AE, Mayer DJ, Mulholland JH, Jenner WE (1964) The suppression of immunologic activity by diversion of thoracic duct lymph. *Ann Surg* 160(3):373–382
- Sharma S, Hemal AK (2009) Chyluria—an overview. *Int J Nephrol Urol* 1:14–26
- Gulati S, Gupta N, Singh NP, Batra S, Garg S, Beniwal P et al (2007) Chyluria with proteinuria or filarial nephropathy? An enigma. *Parasitol Int* 56(3):251–254
- De Almeida AB, Freedman DO (1999) Epidemiology and immunopathology of bancroftian filariasis. *Microbes Infect* 1(12):1015–1022
- Maged A (1967) Renal chyluria. *Br J Urol* 39(5):555–559
- Diamond E, Schapira HE (1985) Chyluria—a review of the literature. *Urology* 26(5):427–431
- Akisada M, Tani S (1968) Filarial chyluria in Japan lymphography, etiology, and treatment in 30 cases! *Radiology* 90(2):311–317
- Panchal VJ, Chen R, Ghahremani GG (2012) Non-tropical chyluria: CT diagnosis. *Abdom Imaging* 37(3):494–500
- McGrath EE, Blades Z, Anderson PB (2010) Chylothorax: aetiology, diagnosis and therapeutic options. *Respir Med* 104(1):1–8
- Lameer C (1966) The development of chylothorax in obstruction of the thoracic duct. *Ned Tijdschr Geneesk* 110(34):1493–1495
- Chen E, Itkin M (2011) Thoracic duct embolization for chylous leaks. *Semin Intervent Radiol* 28(1):63–74
- Cheng JT, Mohan S, Nasr SH, D'Agati VD (2006) Chyluria presenting as milky urine and nephrotic-range proteinuria. *Kidney Int* 70(8):1518–1522
- Pillay TG, Singh B (2016) A review of traumatic chylothorax. *Injury* 47:545–550
- Nair SK, Petko M, Hayward MP (2007) Aetiology and management of chylothorax in adults. *Eur J Cardiothorac Surg* 32(2):362–369
- Chinnock B (2003) Chylothorax: case report and review of literature. *J Emerg Med* 24(3):259–262
- Adler RH, Levinsky L (1978) Persistent chylothorax. Treatment by talc pleurodesis. *J Thorac Cardiovasc Surg* 76(6):859–864
- Rheuban KS, Kron IL, Carpenter MA, Gutgesell HP, Rodgers BM (1992) Pleuroperitoneal shunts for refractory chylothorax after operation for congenital heart disease. *Ann Thorac Surg* 53(1):85–87
- Misthos P, Kanakis MA, Lioulas AG (2012) Chylothorax complicating thoracic surgery: conservative or early surgical management? *Updates Surg* 64(1):5–11
- Itkin M, Kucharczuk JC, Kwak A, Trerotola SO, Kaiser LR (2010) Nonoperative thoracic duct embolization for traumatic thoracic duct leak: experience in 109 patients. *J Thorac Cardiovasc Surg* 139(3):584–589
- Selle JG, Snyder WH, Schreiber JT (1973) Chylothorax: indications for surgery. *Ann Surg* 177(2):245–249
- Singh I, Dargan P, Sharma N (2004) Chyluria—a clinical and diagnostic step ladder algorithm with review of literature. *Indian J Urol* 20(2):79–85
- Linden PA, Swanson SJ (2005) Esophageal resection and replacement. In: Sellke F, del Nido P, Swanson S (eds) *Sabiston & spencer surgery of the chest*, 7th edn. Elsevier Saunders, Philadelphia, pp 627–651
- Schulman A, Fataar S, Dalrymple R, Tidbury I (1978) The lymphographic anatomy of chylothorax. *Br J Radiol* 51(606):420–427
- Jain RK, Padera TP (2003) Development. Lymphatics make the break 299(5604):209–210
- Kraus GE, Bucholz RD, Weber TR (1990) Spinal cord arteriovenous malformation with an associated lymphatic anomaly. *J Neurosurg* 73(5):768–773
- Djindjian R, Hurth M, Houdart R (1971) Spinal angiomas, segmentary vascular dysplasia and phacomatosis. *Rev Neurol (Paris)* 124(2):121–142
- Moore KL (1982) *The developing human: clinically oriented embryology*. WB Saunders, Philadelphia