



## Case report

## Pleural cerebrospinal fluid shunting causing trapped lung: A respiratory physician's approach to management and prevention

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## ABSTRACT

Cerebrospinal fluid (CSF) shunting into the pleural space can cause complications such as long-standing pleural effusions and trapped lung. These complications can be difficult to manage due to the propensity of effusions to recur, and the irreversible nature of trapped lung. This report describes the case of a woman with a pleural CSF shunt who developed chronic pleural effusions and trapped lung over two years, following a 24-year period without any respiratory shunt complications. Management options for this patient included thoracentesis, lung decortication, insertion of an indwelling pleural catheter, and shunt revision. Advocating for pleural shunt revision when symptomatic or increasingly large pleural effusions occur may prevent the development of trapped lung.

## 1. Case presentation

In January 2016, a 40-year-old woman presented with a one week history of progressive dyspnea following thoracentesis two weeks previous. She had a history of myelomeningocele, marked scoliosis, and hydrocephalus secondary to a Chiari II malformation. Her hydrocephalus had been managed with an intrathecal to pleural shunt at the age of 14. This patient had a good functional status with no shortness of breath until two years prior to this presentation. Over these two years, she developed increasing dyspnea on exertion and recurrent increasingly large left-sided pleural effusions which were believed to be complications from her shunt. During this period, five therapeutic thoracenteses were performed. These procedures initially led to significant improvement in the patient's dyspnea for a few months following each intervention. However subsequent thoracenteses provided decreasing benefit, with the fifth only providing one week of relief. The patient was referred to our pleural disease clinic following this presentation.

When she presented to us, the patient had modified Medical Research Council (mMRC) class III dyspnea. She was not taking any medications. She had no constitutional symptoms such as fever, malaise, night sweats, or weight loss. She is a lifelong non-smoker with no history of workplace or environmental exposures that we could identify. Family history is non-contributory.

Her physical exam demonstrated marked scoliosis, absent air entry

to the left lung, and apex beat displacement to the right. Oxygen saturation was 96% on room air. She was tachycardic with a heart rate of 110 bpm and her blood pressure was 160/110 mmHg. No pulsus paradoxus was present.

Pulmonary investigations included multiple chest radiographs and chest computed tomography (CT). Previous radiographs showed increasingly large pleural effusions, evidence of non-expanding lung and hydropneumothorax following thoracenteses (Fig. 1). When she came to us, thoracentesis was performed again as the patient was symptomatic. Afterwards, the CT scan showed a completely collapsed left lung, left-sided pleural effusion and pneumothorax, and right-sided mediastinum shift (Fig. 2).

Two weeks later, the patient presented to the emergency room with increasing dyspnea and a persistent hydropneumothorax. A chest tube was inserted and placed on suction for four days leading to minimal lung expansion. Upon its removal, a chest radiograph demonstrated a residual hydropneumothorax. The fluid analysis exhibited a transudative process with no signs of infection or malignancy. Based on the chronicity of the effusion, post-drainage radiology results, and the fluid characteristics, the patient was diagnosed with trapped lung. Placement of a tunnelled indwelling pleural catheter was considered but ultimately not performed due to fears that, although rare, an infection in the pleural space could spread to her spinal canal. She was referred to her previous neurosurgeon who revised her shunt and inserted an intrathecal to peritoneal shunt.

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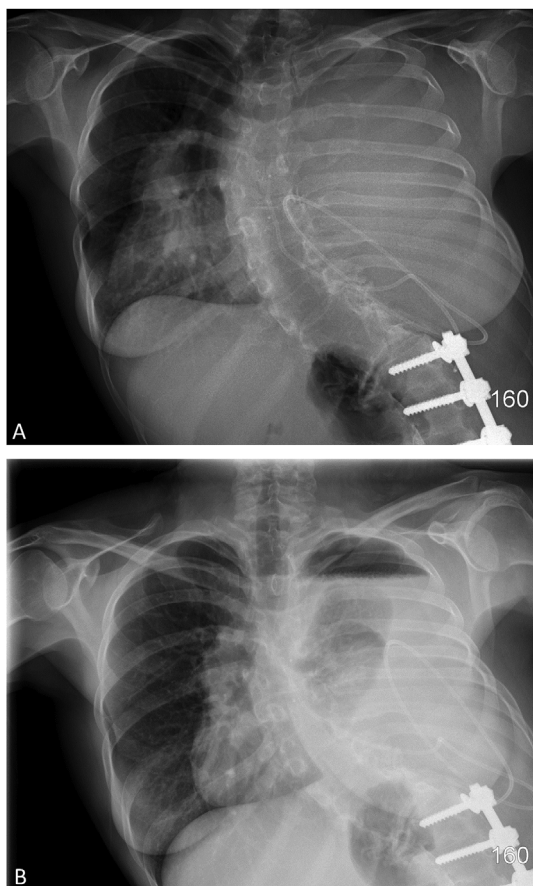


Fig. 1. (A) Chest radiograph taken before thoracentesis demonstrating a large left-sided pleural effusion. (B) Post-thoracentesis chest radiograph demonstrating an air-fluid level over the left lung apex, compatible with hydro-pneumothorax, and minimal left lung expansion.



Fig. 2. (A) Post-thoracentesis computed tomography scan showing left hydro-pneumothorax, left lung collapse, and rightwards mediastinum shift.

Following these interventions, the patient reports mMRC class I dyspnea. However this is of little concern to her as she performed a minimal amount of strenuous activity at her healthy baseline, prior to her respiratory complaints. She feels dyspnea does not limit her daily functioning at present. Her current chest radiograph displays a

persistent hydro-pneumothorax. Visceral pleural decortication or insertion of a chronic indwelling pleural catheter was offered but, as the patient is now feeling well, she was not interested in pursuing these interventions.

## 2. Discussion

Shunting of cerebrospinal fluid (CSF) is a commonly used technique to manage hydrocephalus. These shunts typically originate in the ventricles and terminate in the peritoneal or pleural cavity. Ventriculoperitoneal (VP) shunts are generally preferred due to their relatively easy revision and fewer, less serious complications [1]. If the peritoneum is an unsuitable shunting location due to infection, thrombosis, adhesion or ascites, ventriculopleural (VPL) shunts are a suitable alternative. [2–9]. Unfortunately, recurrent symptomatic pleural effusions, particularly in infants [3,10], are a well-documented complication of this shunt [3,6,11]. This likely occurs due to plugging of parietal pleural stoma by CSF constituents [12]. Since the absorptive capacity of the parietal pleura stoma is normally 28 times the amount of fluid it produces [13], the additional volume of fluid shunted into the pleural space should not cause effusion otherwise [9,10]. Another significant complication that can arise from a VPL shunt is non-expandable lung (or fibrothorax). This can occur due to chronic irritation from a long-standing pleural effusion leading to severe fibrosis of the visceral pleura. Two cases of VPL shunting leading to this complication have been reported. The first case, described in 1992, affected a 3-year-old child [10], and the first case involving an adult was reported in 2008 [14]. Both of these patients went on to have their shunts revised to VP shunts and had surgical decortication performed to alleviate dyspnea.

Non-expandable lung is a complication that occurs when the lung cannot expand to reach the thoracic wall, preventing the contact of the parietal and visceral pleurae [15]. The lung's expansion is restricted by a fibrous peel on the visceral pleural membrane which develops due to pleural disease. Non-expandable lung can be further be categorised into two separate, but related, diagnoses: lung entrapment and trapped lung [16]. Lung entrapment occurs secondary to active pleural disease such as malignancy, hemothorax, or parapneumonic effusion [15,17]. Its treatment should focus on management of the underlying cause of pleural irritation [16]. Without adequate management, the disease can progress to trapped lung and the peel will be permanent [18]. In trapped lung, this thick mature peel continues to cover the visceral pleura even though the causative inflammation has resolved, resulting in a unilateral chronic effusion [15]. If fluid removal is performed, air will temporarily take the place of any removed fluid and a negative pressure environment will be created (pneumothorax ex vacuo). This environment will promote rapid fluid reaccumulation and the pleural effusion will return [19]. While the effusion in trapped lung can cause symptoms such as dyspnea, draining of this effusion often causes pain [16] making this problematic to adequately manage for a symptomatic patient.

For trapped lung, the choice of treatment modality depends on the patient's symptoms. If they are asymptomatic, observation is warranted to ensure the effusion is not worsening [18]. If the patient has dyspnea and fluid removal through thoracentesis has been found to improve this, placement of a chronic indwelling catheter can be considered for symptom alleviation [20]. Though this procedure will not immediately lead to significant lung expansion, the relief of pressure on the diaphragm allows for more efficient inspiratory muscle function, relieving dyspnea [21,22]. Recurrent drainage attempts through an indwelling catheter may, theoretically, expand the trapped lung over time although this has not been documented. If a more definitive approach is indicated, surgical decortication is the only curative treatment [18]. This procedure involves the removal of the fibrous peel covering the lung hoping to cause permanent lung expansion [23].

When considering decortication, the chronicity of trapped lung is an important factor to consider. Much of the literature regarding

decortication concerns the management of empyema. It has been noted that an increased duration of trapped lung leads to poorer outcomes, including more frequent complications [24–27], increased post-operative pain, lengthier hospital stays [25], and increased 30-day mortality [27]. Additionally, a study by Stefani et al. [26] found that about 70% of patients who had decortication delayed by only 21–30 days had worse outcomes compared to earlier decortication [26]. This being said, even patients with long-standing trapped lung have been found to have improvement in lung function [25] and dyspnea [28] following decortication. Therefore surgical decortication is still recommended regardless of trapped lung chronicity.

Though it is an uncommonly performed procedure, it is important to realise VPL shunts can lead to long-standing effusions and the development of trapped lung, leaving patients with permanent symptoms. In addition, the duration of an effusion likely negatively impacts chances for lung expansion. For these reasons, we recommend that respiratory physicians advocate for the early revision of VPL shunts following either a symptomatic effusion or dramatic increase in size of a patient's effusion, regardless of an extended history of shunt patency.

### 3. Conclusion

The present article described a case of a woman with a pleural CSF shunt, recurrent pleural effusions and trapped lung. Due to the challenges in managing these conditions, a preventative approach should be considered. Respiratory physicians should advocate for pleural shunt revision following the development of associated pleural effusions, in order to prevent the development of trapped lung.

Informed consent was obtained from the patient in regards to the submission of this report.

### Conflicts of interest

The authors have no conflicts of interest to declare.

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