

# A patient with an inspiratory stridor 25 years after lung surgery

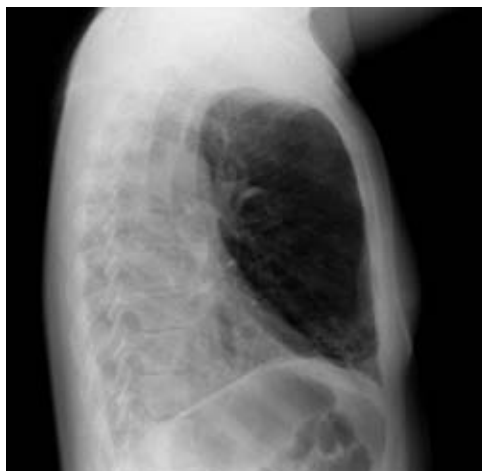
## Case history

A 32-year-old female regularly attended the outpatient clinic for pulmonology with recurrent pulmonary infections. Her medical history revealed an operation on the right lung 25 years previously for persistent tuberculosis in that lung. The nature of the operation was unknown at clinical admission. Besides this, asthma had been diagnosed 2 years previously. In a regular visit, the patient complained of progressive exertional dyspnoea, coughing and sputum expectoration. Chest auscultation revealed stridor. Chest radiography is shown in figure 1



## Task 1

How would you interpret the chest radiograph?



**Figure 1**  
Chest radiographs.

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**Answer 1**

The chest radiograph shows shifting of the mediastinum, trachea and heart towards the posterior part of the right hemithorax. Moreover, there is a scoliosis to the right. The abrupt ending of the right main bronchus is consistent with a right-sided pneumonectomy.

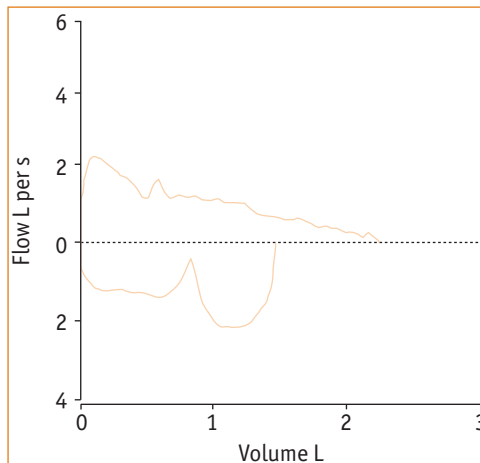
In requested documentation, it was found that a pneumonectomy of the right lung had been carried out 25 years previously.

Because of the symptoms of the patient, spirometry was performed (table 1; figure 2).

**Table 1 Spirometry data**

	Predicted L	Actual L	Actual/predicted %
VC	3.31	2.33	70.4
FEV <sub>1</sub>	2.84	1.30	45.7
FEV <sub>1</sub> /VC %	83.02	55.70	

VC: vital capacity; FEV<sub>1</sub>: forced expiratory volume in one second.



**Figure 2**  
Flow-volume curve.

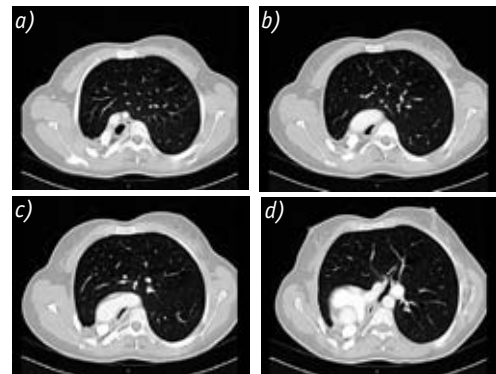
**Task 2**

What do the spirometry data and flow-volume curve suggest?

**Answer 2**

Vital capacity (VC) and forced expiratory volume in one second (FEV<sub>1</sub>) are decreased modestly and severely, respectively. The resultant FEV<sub>1</sub>/VC ratio is also decreased suggesting an expiratory obstruction. Furthermore, the inspiratory flow increases normally but a prominent notch is apparent halfway through inspiration. At this point, inspiratory flow is decreased almost to zero.

Compared with earlier spirometry of the same patient, in which a small notch was visible in the inspiratory part of the flow-volume curve, the inspiratory flow limitation had worsened. Diffusion capacity, which was 93% predicted 1 year previously, appeared to be unaffected. An obstruction of the upper airways was suspected; therefore, chest computed tomography (CT) was carried out (figure 3).



**Figure 3**  
CT scan of the thorax at the time of presentation.

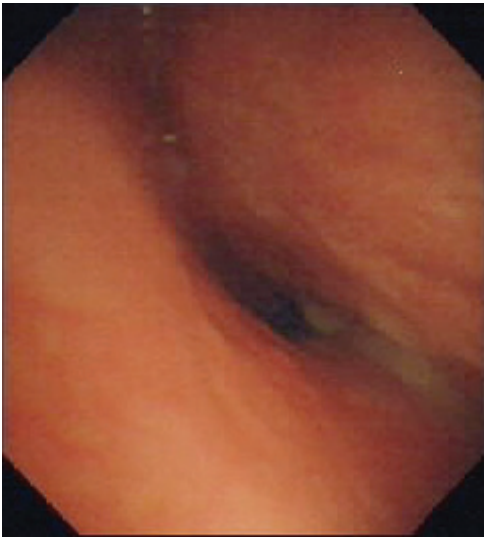
**Task 3**

Describe the CT scans.

**Answer 3**

The left lung has expanded and there is a complete herniation of the left lung to the right hemithorax. The mediastinum has shifted dorsally in the right hemithorax with anticlockwise rotation of the heart and great vessels. The heart is situated against the right posterolateral chest wall. The trachea is situated right paravertebral and is extremely narrow at the level of the aortic arch (figure 3c). Additionally, the remaining left main bronchus is narrowed (figure 3d), because the trachea and the left main bronchus are compressed by the aortic arch anteriorly, and by the oesophagus and thoracic spine posteriorly.

Bronchoscopy revealed near-total collapse of the trachea, consistent with severe tracheo-bronchomalacia in the distal end of the trachea and left main bronchus. No intraluminal obstructions or malignancies were found.



**Figure 4**  
Bronchoscopic view of the left main bronchus. Note the extreme narrowing.

**Task 4**

What is your diagnosis?

**Task 5**

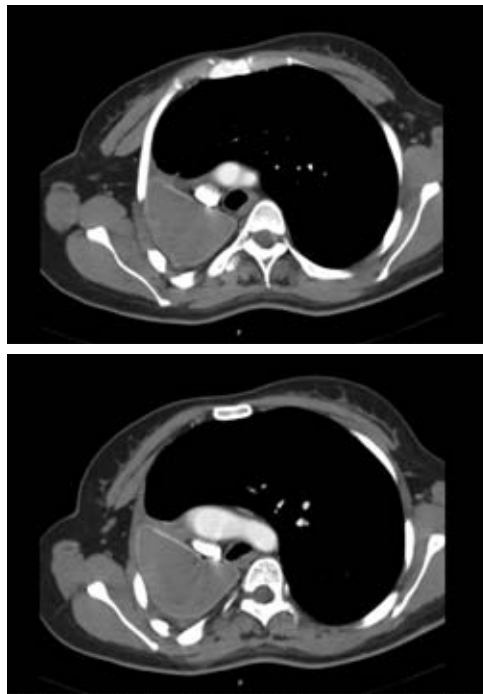
What would be your treatment?

**Answer 4**

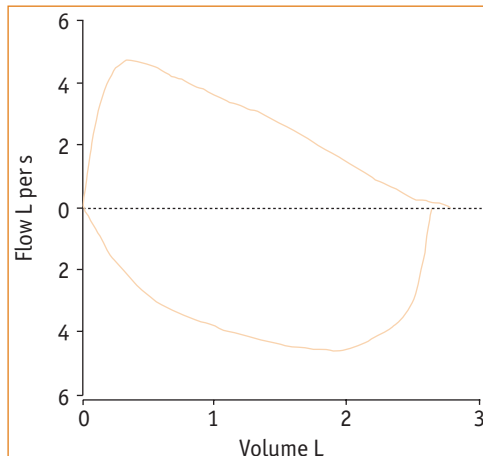
Postpneumonectomy syndrome.

**Answer 5**

The patient was treated with a rightsided thoracotomy and repositioning of the mediastinum with implantation of two silicone prostheses of 380 g and 170 g. These were covered with vicryl and fixed at the spine. Postoperative recovery passed without any complications and the patient's FEV1 increased to 1.67 L, with a normalised expiratory pattern. One year after surgery, FEV1 was 2.19 L (77% pred) with a completely normalised inspiratory flow (figures 5 and 6).



**Figure 5**  
CT scan of the thorax after surgery.



**Figure 6**  
Flow-volume curve one year after surgery.

## Discussion

Postpneumonectomy syndrome (PPS) is a rare complication after a pneumonectomy, with an estimated incidence of one case per 640 pneumonectomies [1]. Originally, it was described exclusively in patients with a right pneumonectomy [2]. Although PPS is seen mostly after right-sided pneumonectomy, it has now also been described in patients after left pneumonectomy, in cases of a rightsided aortic arch and even in cases with a normal left-sided aortic arch [3, 4].

PPS has been described in both children and adults [5] with varying times of presentation, ranging from 1 month to 20 years after pneumonectomy [4]. Our patient presented 25 years after pneumonectomy. The syndrome is characterised by an extreme rotation of the mediastinal structures (the heart, great vessels and trachea) into the pneumonectomy side. The rotation is caused by hyperinflation of the remaining lung, in combination with the negative pressure of the pneumonectomy space. After right pneumonectomy, the mediastinum will rotate anticlockwise with posterior displacement. The opposite takes place with left pneumonectomy, the mediastinum rotating clockwise. As a result of this displacement, stretching and narrowing of the main bronchus occurs, in turn causing compression of the bronchus between the left pulmonary artery and aorta in right PPS, while in left PPS the bronchus is compressed between the right pulmonary artery and the thoracic spine. The most common presentation of PPS is progressive exertional dyspnoea [5]. Other symptoms include stridor and recurrent respiratory infections. Additionally, bronchomalacia often develops, making treatment extremely difficult [4]. Furthermore, after pneumonectomy there is a risk of musculoskeletal deformities, particularly scoliosis [6].

Currently, PPS is corrected by intrathoracic prosthesis implantation using silicone, saline breast implants or expandable implants [7].

This case exhibits three unique features. First, the patient experienced not only an expiratory flow limitation but also an inspiratory flow limitation secondary to her tracheobronchomalacia. Secondly, extreme displacement of the heart and large vessels has occurred due to the complete filling of the thorax by the left lung. Thirdly, PPS came to light 25 years after surgery.

The incidence of tracheobronchomalacia as a complication of pneumonectomy is not known. However, the mechanism is thought to be secondary to prolonged compression of the cartilage rings between a great vessel and the

vertebral body, or the result of the interposition of the trachea between the pulmonary artery and aorta [4]. The cartilage rings become thinner and softer, with a loss of elasticity in the myoelastic elements, which no longer support the cartilage adequately [1, 8]. In healthy individuals, intrathoracic and intraluminal pressure differences cause dilation of the trachea with inspiration and narrowing with expiration. In malacia, the airway is willing to collapse when the extraluminal pressure exceeds the intraluminal pressure. This is more susceptible during coughing or forced expiration. In spirometry, a decreased FEV1 is seen with rapid declining expiratory flow, air trapping and elevated airway resistance.

Besides a decreased expiratory flow, this patient showed severe inspiratory impairment. Inspiratory collapse is less common in PPS and is likely to be a result of pressure on the trachea from the abnormally situated aortic arch and thoracic spine, in combination with the tracheomalacia and the increasing negative intraluminal pressure due to inspiratory flow (Venturi effect). There was an evident choke-point halfway through inspiration, with almost zero airflow. Thereafter, the intraluminal pressure increases and the trachea opens again [9, 10, 11].

Other patients have been described who experienced bronchial/tracheal narrowing with increased air turbulence as increased work of breathing and lower exercise capacity [1, 12].

The complete filling of the thoracic cavity with the remaining lung is a phenomenon which could be explained because the patient's pneumonectomy was performed at the age of 8 years. Exercise and lung function studies in patients who have undergone pneumonectomy during infancy demonstrate that normal physical endurance is common in adulthood [13], with spirometry results almost the equal to those predicted for healthy subjects [14].

Whether these findings must be explained by simple distension of the lung or pulmonary growth remains unclear. The initial process of adaptation to the available space may be purely mechanical, by recruitment and distension at the cost of the diminished reserve capacity [14]. After this initial mechanism, growth of the lung becomes more important.

The first report of growth of the remaining lung after pneumonectomy in an animal was published in 1892 [15]. Thereafter, many animal experiments [14, 16, 17] and observations in humans [13] have shown that the potential for compensatory lung growth diminishes with age,

although it is not known until what age compensatory lung growth can occur. The observed near-normal diffusion capacity in this patient suggests not only that the lung had expanded but also that growth/increase of lung tissue had occurred.

In summary, this was a patient with abnormal spirometry and chest radiography after right

sided pneumectomy in her youth. This case shows that PPS can occur even after a prolonged period, and it should therefore be considered in patients with previous lung resection and respiratory complaints. Furthermore, we demonstrate resolution of the dynamic inspiratory obstruction after rearrangement of the mediastinal structures.

#### References

1. Jansen JP, Brutel de la Rivière A, Alting MP, Westermann CJ, Bergstein PG, Duurkens VAM. Postpneumonectomy syndrome in adulthood: Surgical correction using an expandable prosthesis. *Chest* 1992; 101: 1167–1170.
2. Shepard JO, Grillo HC, McCloud TC, Dedrick CG, Spizarny DL. Right pneumonectomy syndrome: radiologic findings and CT correlation. *Radiology* 1986; 161: 661–664.
3. Shamji FM, Deslauriers J, Daniel TM, Matzinger FR, Mehran RJ, Todd TRJ. Postpneumonectomy syndrome with an ipsilateral aortic arch after left pneumonectomy. *Ann Thorac Surg* 1996; 62: 1627–1631.
4. Grillo HC, Shepard JO, Mathisen DJ, Kanarek DJ. Postpneumonectomy syndrome: diagnosis, management and results. *Ann Thorac Surg* 1992; 54: 638–651.
5. Bédard EL, Uy K, Keshavjee S. Postpneumonectomy syndrome: a spectrum of clinical presentations. *Ann Thorac Surg* 2007; 83: 1185–1188.
6. Westfelt JN, Nordwall A. Thoracotomy and scoliosis. *Spine* 1991; 16: 1124–1125.
7. Podevin G, Larroquet M, Camby C, Audry G, Plattner V, Heloury Y. Postpneumonectomy syndrome in children: advantages and long-term follow up of expandable prosthesis. *J Pediatr Surg* 2001; 36: 1425–1427.
8. Kelly RF, Hunter DW, Maddaus MA. Postpneumonectomy syndrome after left pneumonectomy. *Ann Thorac Surg* 2001; 71: 701–703.
9. Austin J, Ali T. Tracheomalacia and bronchomalacia in children: pathophysiology, assessment, treatment and anaesthesia management. *Ped Anaesthesia* 2003; 13: 3–11.
10. Carden KA, Boiselle PM, Waltz DA, Ernst A. Tracheomalacia and tracheobronchomalacia in children and adults. *Chest* 2005; 127: 984–1005.
11. Loring SH, O'Donnell CR, Feller-Kopman DJ, Ernst A. Central airway mechanics and flow limitation in acquired tracheobronchomalacia. *Chest* 2007; 131: 1118–1124.
12. Valji AM, Maziak DE, Shamji FM, Matzinger FR. Postpneumonectomy syndrome. Recognition and management. *Chest* 1998; 114: 1766–1769.
13. Werner HA, Pirie GE, Nadel HR, Fleisher AG, LeBlanc JG. Lung volumes, mechanics, and perfusion after pulmonary resection in infancy. *J Thorac Cardiovasc Surg* 1993; 105: 737–742.
14. Laros CD, Westermann CJJ. Dilatation, compensatory growth, or both after pneumonectomy during childhood and adolescence: a thirty-year follow-up study. *J Thorac Cardiovasc Surg* 1987; 93: 570–576.
15. Haasler F. Über compensatorische hypertrophie der lunge. [On compensatory hypertrophy of the lung.] *Virchows Arch Pathol Anat Physiol* 1892; 128: 527–536.
16. Dummill MS. The problem of lung growth. *Thorax* 1982; 37: 561–563.
17. Thurlbeck WM. Postnatal human lung growth. *Thorax* 1982; 37: 564–571.