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# Postpneumonectomy syndrome in a newborn after esophageal atresia repair

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ABSTRACT

*INTRODUCTION:* Postpneumonectomy syndrome (PPS) is an ominous complication, caused by mediastinal shift following massive lung resection.
*PRESENTATION OF THE CASE:* A neonate with oesophageal atresia and tracheo-oesophageal fistula developed acute respiratory distress shortly after surgery, despite mechanical ventilation. The patient was found to have an associated oesophageal right lung that collapsed after oesophageal atresia repair and a left pulmonary artery sling causing left main bronchus stenosis mimicking a postpneumonectomy syndrome.
*DISCUSSION:* We will describe the diagnostic work-up and the therapeutic measures used both in the acute phase and as definitive treatment in this challenging case.
*CONCLUSIONS:* Neonatologists and paediatric surgeons should be aware of this rare association that may cause acute life threatening and worsening of patient's clinical status. Prompt realignment of the mediastinum in the normal position is critical to obtain rapid improvement of the patient's clinical conditions.


## 1. Introduction

Postpneumonectomy syndrome (PPS) is an ominous complication, with prevalence in children ranging from 1 in 6 [1] to 1 in 5 pneumonectomies [2]. It is caused by mediastinal shift following massive lung resection that results in compression and stretching of the great vessels and airways. It usually occurs after right pneumonectomy [2], performed for infections, congenital malformations, or metastatic tumours. We report on a patient with oesophageal atresia (OA) who developed a PPS following OA repair. The case represented an authentic diagnostic and therapeutic challenge.

## 2. Case presentation

A 1-day-old girl was referred to our department for the suspect of OA. She was born by caesarean section at 37 weeks of gestation after an uneventful pregnancy. Family history was unremarkable. Her birth weight was 2.1 kg and the Apgar score was 7/7 at 1/5 min. On admission she was well, with normal oxygen saturation during spontaneous breathing. Chest X-rays showed the nasogastric tube folding at the level of the first thoracic vertebra and a distended stomach (Fig. 1a), suggesting OA with distal tracheo-oesophageal fistula (TOF). Echocardiography demonstrated normal left sided aortic arch, a large patent ductus arteriosus and hypoplastic right pulmonary artery. Screening for other associated anomalies was negative. Preoperative fiberoptic tracheoscopy was performed to exclude the presence of a tracheo-oesophageal cleft, and to measure the oesophageal gap. The tip of the bronchoscope was placed at the level of the fistula and a contrasted naso-gastric tube was inserted in the upper pouch. The distance between the two was measured, which was 1.5 cm [3]. She underwent uneventful surgical ligation and section of the TOF and primary oesophageal anastomosis. On post-operative day one, she had progressive clinical worsening with marked hypoxia despite mechanical ventilation. Chest X-rays showed total collapse of the right lung and right mediastinal shift (Fig. 1b). A fiberoptic tracheobronchoscopy performed by an experienced otolaryngologist revealed a long malacic trachea, extending to the left main bronchus. The fistula was closed and the right main bronchus was absent. This finding was missed during the first bronchoscopy. The suspect of a right oesophageal bronchus associated with the OA was confirmed by oesophagogram showing the origin of the right main bronchus from the oesophagus, caudal to the patent anastomosis (Fig. 1c).

The right lung collapse caused massive mediastinal shift and rotation, clinically mimicking a PPS. A high resolution CT scan with 3D images reconstruction showed a collapsed hypoplastic right lung with a single, small, right pulmonary artery and an associ-

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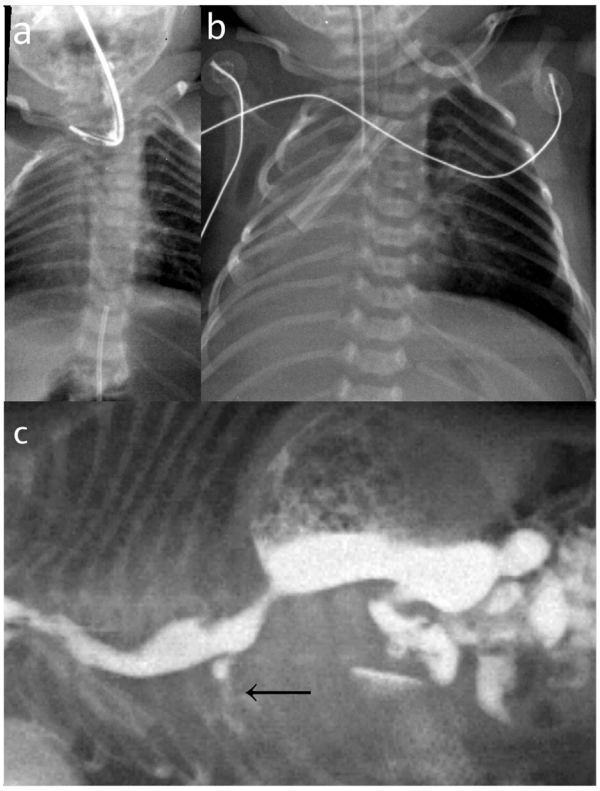


**Fig. 1.** (a) Chest X-rays on admission with nasogastric tube folding in the upper pouch and distended stomach, suggesting an oesophageal atresia with distal tracheo-oesophageal fistula. Note, the right lung is inflated. (b) Post-operative chest X-rays. The right lung is collapsed and the mediastinum is markedly shifted towards the right hemithorax. (c) Oesophagogram with the patient lying on the right side. The right main bronchus (arrow) originates from the right side of the oesophagus, caudal to the anastomosis.

ated left pulmonary artery sling (Fig. 2a), not visible during the pre-operative echocardiography focused to the side of the aortic arch. To push the mediastinum in the correct position, a 12 CH Foley catheter was inserted in the right hemithorax and sterile normal saline (50 ml) was infused (Fig. 2b–d), and serial saline infusions were performed in the next days. In the occasion of surgical correction of the left pulmonary artery sling (day 22 of life) a tissue expander was inserted in the right hemithorax to stabilize the mediastinum. On post-operative day 25, a right pneumonectomy was performed and the tissue expander was replaced with a larger one. Post-operative course was uneventful; she was extubated on post-operative day 2, and discharged home on post-operative day 24. She is alive and well at 24 months of follow-up with moderate thoracic asymmetry and good respiratory function.

## 3. Discussion

We present a case of acute PPS following OA repair in a patient with associated right oesophageal lung.

### 3.1. Communicating bronchopulmonary foregut malformations

The association of OA with an abnormal bronchial implantation on the oesophagus is extremely rare and falls in the so-called communicating bronchopulmonary foregut malformations (CBPFM). In our series of 256 patients with OA/TOF consecutively treated from 1995 we found only one such patient and Gou et al. report one CBPFM in 143 patients with OA/TOF [4]. Gerle et al. first used the term "bronchopulmonary foregut malformations" in 1968 to describe a spectrum of congenital malformations including pulmonary sequestration with or without connections to the alimentary tract [5]. Srikanth et al. later suggested the term CBPFM to describe a segment of lung tissue connected to the foregut, and introduced a classification [6]. The first group of this classification is represented by sequestered lung tissue associated with OA/TOF and connected to the distal oesophagus.

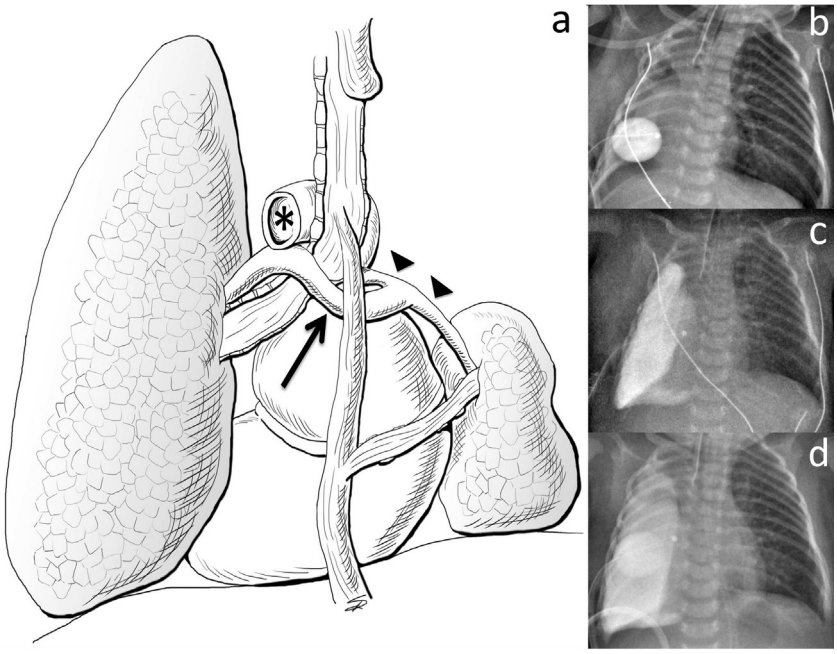


**Fig. 2.** (a) Artist rendition of the patient's preoperative anatomy (posterior view). Oesophageal atresia with tracheo-oesophageal fistula. The right main bronchus originates from the lower oesophagus. The left pulmonary artery (arrow) sling originates from the right pulmonary artery (arrow heads), and courses posterolateral to the left main bronchus and reaches the left hylum, thereby causing left main bronchus stenosis together with the aortic arch (*). (b) A 12 CH foley catheter was inserted in the right hemithorax. (c) An iatrogenic hydrothorax was progressively induced with normal saline and contrast medium. (d) Note, the mediastinum gradually regaining its normal position at the end of the procedure.

### 3.2. Clinical presentation

In our patient, respiratory conditions deteriorated shortly after surgery. Surgical repair of EA suddenly impeded the ventilation to the lung fed by the oesophageal bronchus, thereby leading to total right lung collapse and contralateral single lung ventilation. The pathogenetic cascade is confirmed by the only reported case that did not have surgical ligation of the fistula after detection of the anomaly [7]. That patient's respiratory conditions remained stable because the fistula allowed airflow to the right lung [7]. In our patient, the limitation of the preoperative endoscopic study to exclude a tracheo-oesophageal cleft and to define the oesophageal gap misled our intraoperative management and delayed the diagnosis of CBPFM. A detailed tracheobronchoscopy would probably have allowed the diagnosis of CBPFM before OA repair, as suggested by the subsequent tracheobronchoscopy that showed the absence of the right main bronchus. In any case, in the event of acute worsening of clinical conditions during OA/TOF repair or shortly thereafter, with unilateral total lung collapse, the presence of an associated CBPFM should always be born in mind and actively investigated. Oesophagogram is the gold-standard diagnostic test as it shows the aberrant origin of the oesophageal bronchus. The study should be performed in lateral decubitus as the bronchus originates from the side of the oesophagus, usually the right one. Oesophagoscopy should be avoided in the acute setting as it can cause disruption of the fresh anastomosis. In our patient, clinical manifestations mimicked PPS. PPS is a dreaded complication of pneumonectomy. In neonates and infants, lung resection is generally well tolerated, with good long-term outcomes. However, there are anatomic characteristics specific to children that are thought to predispose them to specific post-operative complications. Mediastinal structures are more elastic when compared with adults and their cartilage is softer. Moreover, during childhood, lungs are more compliant due to the higher content of elastic tissue. This relative flexibility of the mediastinal structures could lead to contralateral pulmonary distension after a pneumonectomy. Mediastinal shift and rotation causes compression and stretching of the great vessels and airways that in association with overdistension of the remaining lung can result in critical worsening of pulmonary function leading to major respiratory failure, the so called PPS. This condition is most frequently encountered in children than in adults for the anatomical characteristics mentioned above. PPS can occur almost immediately after pneumonectomy but is more common in the progressive and delayed form. In our patient, OA/TOF repair was followed by acute complete right lung collapse, simulating a right pneumonectomy, with massive right-sided mediastinal shift, clinically mimicking a PPS. The left pulmonary artery sling that caused left main bronchus stenosis further worsened the clinical picture. As a consequence, both air and blood flow in the remaining left lung was acutely reduced by the mediastinal shift. When such a clinical picture occurs in patients with group 1 CBPFM, the term pseudo-PPS seems more appropriate as they do not undergo lung resection surgery.

### 3.3. Treatment

Classically, treatment of CBPFM is to resect the aberrant lung tissue with lobectomy or pneumonectomy and mediastinal repositioning using rigid prostheses (e.g. breast prostheses) or tissue expanders in the empty hemithorax [8,9]. Reimplantation of the anomalous bronchus is the ideal management but was attempted in only one patient with CBPFM associated with OA, which was successful [10]. This option should be considered in patients with normal pulmonary vasculature and without lung tissue damage due to aspiration or prolonged consolidation. However, in some cases PPS may be life-threatening, and its treatment requires emergency repositioning of the mediastinum in axis. In our patient, we performed an iatrogenic hydrothorax by infusing sterile normal saline in the right hemithorax, which was followed by immediate improvement of her clinical conditions. This was a simple, rapid, and effective method to regain clinical stability. Serial infusions were performed to maintain the mediastinum in axis, which allowed the patient to sustain clinical stability and to undergo cardiac surgery. In our patient, bronchial reconstruction was not possible as definitive treatment because the right bronchus was too short, not allowing a good anastomosis with the trachea, and the lung remained collapsed too long to allow its re-expansion. Stable mediastinal repositioning was obtained with an expandable prosthesis placed in the right hemithorax.

## 4. Conclusions

In patients with OA, the development of post-operative severe respiratory distress and unilateral lung collapse must lead to the suspect of an associated CBPFM. Neonatologists and paediatric surgeons should be aware of this rare association that may cause acute life threatening worsening of patient's clinical conditions. In the case of PPS development, prompt recognition of the syndrome and prompt realignment of the mediastinum is critical. Infusion of sterile normal saline in the affected hemithorax is a simple, fast and effective method that allows pushing the mediastinum in the correct position, thereby leading to dramatic improvement of the clinical conditions.

## Conflicts of interest

The authors have no potential source of conflict of interest.

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## Ethical approval

Present report complies with the guidelines for human studies and animal welfare regulations. The Ethics Committee of our Hospital has approved the report. No animal experiments were conducted.

## Consent

Although no identifying details are reported, parents of the patient have given their informed consent.

## Author contribution

C. Iacusso: data collection, writing the paper.P. Bagolan: data collection and interpretation, paper revision.S. Bottero: data interpretation.A. Conforti: data collection.F. Morini: data interpretation, paper writing and revision.

## Guarantor

F. Morini.

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