

**Table 1** Main characteristics of previous described cases associating Poland syndrome and pneumothorax. All patients were male.

Case number	Age	Hemi thorax*	Musculoskeletal anomalies <sup>†</sup>	Pneumothorax <sup>‡</sup>	Apical bullous alterations	Ref.
1	16	Left	None	Contralateral <sup>°</sup>	Bilateral	(5)
2	27	Right	Brachydactyly <sup>§</sup> of the ipsilateral hand	Ipsilateral <sup>°</sup>	Bilateral	(5)
3	22	Right	Ipsilateral superior micromelia <sup>#</sup> with shoulder and elbow joint mobility restriction	Ipsilateral	None	(8)
4	19	Right	None	Ipsilateral	Right	(9)
5	21	Right	None	Ipsilateral	Right	(9)

\* Location of pectoralis muscle deformity; † Additional musculoskeletal deformities; ‡ Location in relation to the pectoralis muscle deformity; § Shortening of fingers; # Limb shortening; ° Recurrent pneumothorax.

fore this population may benefit from greater clinical surveillance.

The possible hypothesized association between these two clinical entities is dependent on the reporting and analysis of more cases. Also, longitudinal epidemiological studies may be useful.

## Disclosure

The authors report no financial support, off-label or investigational drug use.

## Conflicts of interest

The authors have no conflict of interest to declare.

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## Late biliobronchial fistula

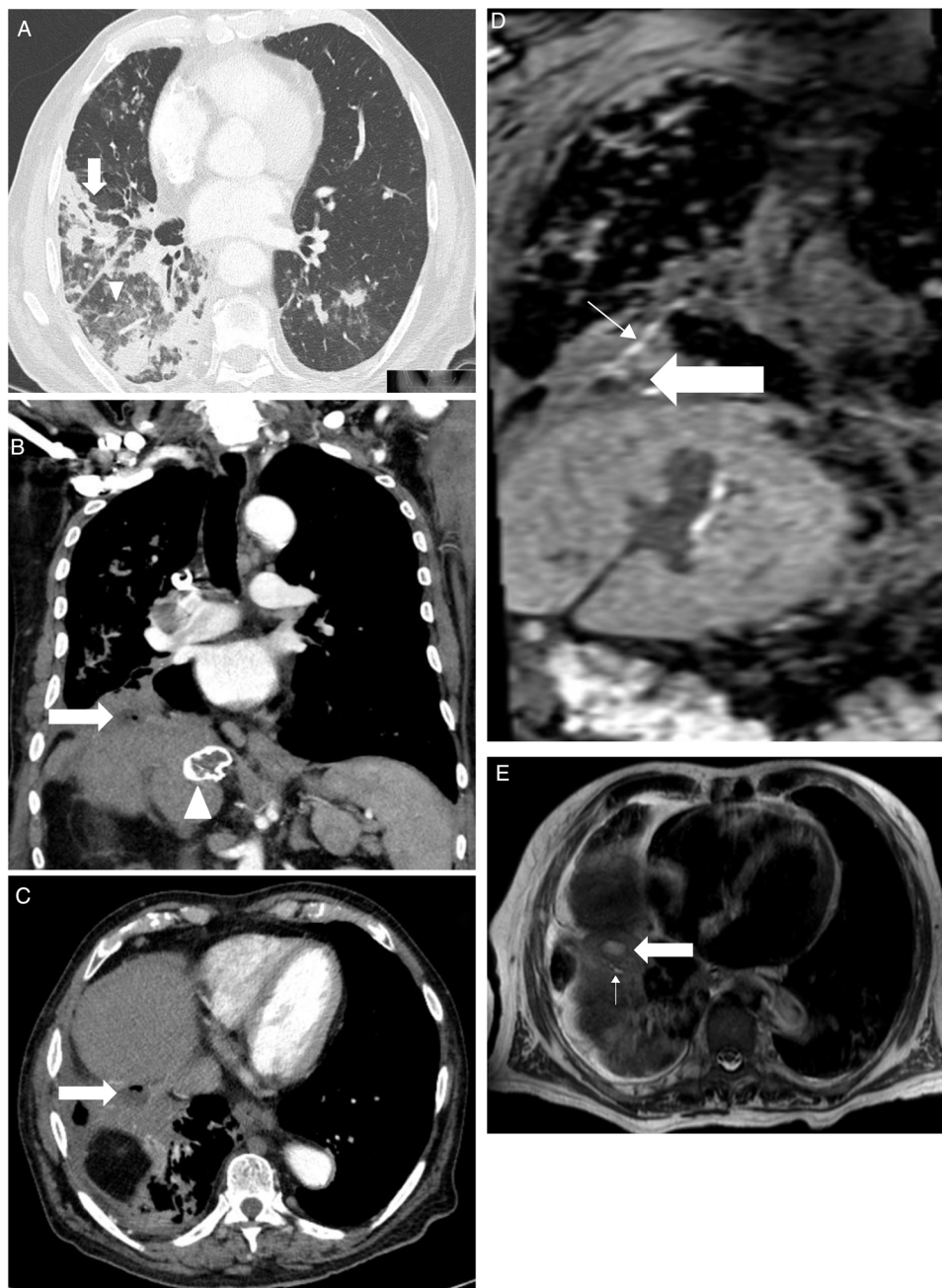


Biliobronchial fistula (BBF) is a rare condition characterized by communication between the bile duct and the bronchial tree.<sup>1</sup> The first case was described by Peacock's in 1850 in a 20-year-old woman with hepatic echinococcosis.<sup>2</sup> Diagnosis

is mainly clinical, with radiological or endoscopic support,<sup>3</sup> and therapeutic options range from conservative to invasive with highly variable results.<sup>4</sup> We present the case of a patient admitted to our service with this unusual disorder.

An 88-year-old male was hospitalized with fever, shortness of breath and increased expectoration in the previous month. Fourteen years earlier he had had a partial hepatectomy for a biliary fistula secondary to the removal of a hydatid cyst, with elevation of the right hemidiaphragm as a result of the surgery, and residual lesions to the ipsilateral lung base. He was diagnosed with bronchiectasis with

*Abbreviations:* BBF, bronchobiliary fistula; CT, computerized tomography.



**Figure 1** A: Axial cut computerized tomography. Pulmonary infiltrates patched with airborne bronchogram (thick arrow) and areas of tarnished glass predominantly right lower lobe (arrowhead).

Figure 1B: Coronal cut computerized tomography. A small abscess is shown in the diaphragmatic region (thick arrow) and calcified hydatid cyst (arrowhead).

Figure 1C: Axial cut computerized tomography. Abscess (thick arrow) between the posterior hepatic rim and the lower right lobe, with a small air bubble inside.

Figure 1D: Coronal cut of hepatic magnetic resonance imaging in hepato-specific phase with bile duct contrast excretion at the same level as Figure 1B. Abscess (thick arrow) and fistulous path (thin arrow) are shown.

Figure 1E: Axial cut of hepatic magnetic resonance imaging in hepato-specific phase with bile duct contrast excretion at the same level as Figure 1C. Fistulous tract (thin arrow) and abscess (thick arrow) leading to the anterobasal segment of the lower right lobe.

chronic bronchial infection by *Pseudomonas aeruginosa* and treated with inhaled colistine. In the days prior arriving to the Emergency Department, he had been treated with tobramycin and intravenous amikacin (home hospitalization). On physical examination, he was tachypneic, afebrile, had bilious expectoration, and crackles and hoarseness could be heard throughout the right hemithorax. Blood tests showed leukocytosis ( $15.110/\text{mm}^3$ ) with 84% of segmented and 7% of stems; creatinine 1.18 mg/dL; Na 129 mmol/L; GGT 83 UI/L; FAL 303 UI/L; total proteins 5.7g/dL; C-reactive protein 24.2 mg/dL and procalcitonin 1.32 ng/mL; PaO<sub>2</sub> 56 mm Hg. Bronchofibroscopy showed, from the larynx and throughout the right bronchial tree, abundant yellowish bilious-looking fluid, compatible with bilioptisis, with an inflammatory mucosa of the lower right bronchus lobe without endobronchial lesions. In CT we observed pulmonary infiltrates patched with airborne bronchogram, areas of tarnished glass predominating in the right lower lobe (Figure 1A), a peridiaphragmatic cystic lesion at the base of the right lung with a small air bubble and post-operative liver changes with peripheral calcification in the caudate lobe suggesting a healing hydatid cyst (Figures 1B and 1C). Liver ultrasound showed right hepatectomy with hypertrophy of the left hepatic lobe without biliary lithiasis. Liver MRI and cholangio-MRI scans showed a fistulous tract and subdiaphragmatic abscesses (Figures 1D and 1E). A plastic biliary stent was placed by endoscopic retrograde cholangiography. We also performed a sphincterotomy for biliary decompression. At the same time, the subdiaphragmatic collections in which *Pseudomonas aeruginosa* was cultivated were drained. During the course of antibiotic treatment (ceftazidime and tobramycin), *Staphylococcus haemolyticus* bacteremia occurred, forcing the addition of vancomycin. The patient worsened progressively with renal function deterioration, dying a few days later.

Biliobronchial fistula is a rare condition for which diagnosis is established from the presence of bilioptisis and is confirmed by techniques such as bronchofibroscopy or various imaging studies. It may be congenital or secondary to trauma (most common, including bile duct surgery), liver disease (hydatid cyst and amoebic liver abscess), or bile duct obstruction. A frequent complication in chronic stages, as in our case, is the presence of bronchiectasis in the pulmonary segment involved.<sup>5</sup> Occasionally, there may be recurrent pneumonia and ipsilateral pleural effusion, with secondary sepsis, probably caused by chemical pneumonitis that produces bile in the bronchial mucosa. Imaging studies used to confirm the diagnosis (CT scan, ultrasound, hepatic nuclear magnetic resonance, nuclear magnetic resonance, percutaneous cholangiography, and endoscopic retrograde cholangiopancreatography) must demonstrate the fistulous tract.

There is no general agreement on how to deal with BBFs. Ong et al used subcutaneous octreotide and succeeded in reducing bilious cough in a patient and speeding up the closure of the fistula, but its use is limited and is not effective if there is underlying infection, neoplasia or obstruction.<sup>6</sup> The definitive treatment is surgical fistulectomy with soft tissue reconstruction, but due to its significant morbidity and mortality and the frequent re-operations it leads to, more conservative interventions have also been proposed aimed

at reducing pressure in the biliary tract, such as endoscopic retrograde biliary drainage or percutaneous transhepatic biliary drainage. Depending on the size and location of the BBF, another conservative option would be to try to close the fistula through bronchofibroscopy. The sealing material used should achieve an inflammatory reaction of the mucous membrane that causes the permanent closure of the fistula. There is a wide range of synthetic substances and biological derivatives that can be applied through the flexible bronchoscope.<sup>7</sup>

What can be learned from this case is that faced with a patient with a history of a traumatic process in the hepatobiliary tract (even if it is old), bilious sputum and bronchiectasis in the right lung, this condition should be suspected.

### Author's Contributions

RA, MET, AMdeA and LV were responsible for the conception and design of the study, and wrote and edited the manuscript. All authors read and approved the final manuscript.

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### Conflicts Of Interest

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