

Congenital bronchial atresia in adults: thoracoscopic resection

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Abstract: Congenital bronchial atresia (CBA) is a rare congenital malformation consisting in an interruption of a lobar or—more frequently—of a segmental bronchus. It leads to mucus impaction and hyperinflation of the obstructed lung segment. It causes infectious complications and, in the long term, destruction of the adjacent lung parenchyma. Thus, a surgical resection is usually indicated, even in asymptomatic patients.

Keywords: Congenital bronchial atresia (CBA); pulmonary resection; thoracoscopy

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Introduction

Bronchial atresia is a congenital malformation arising during the formation of bronchi. It consists in a partial defect of a bronchus—usually at a segmental level—leading to the distension of the corresponding pulmonary parenchyma. They belong to the group of congenital pulmonary airway malformations (CPAM), which is different from the group of bronchopulmonary sequestrations (BPS) (1). Both malformations can be associated. CPAM are most often diagnosed in the neonatal period or in the young child (2), and sometimes in the adult, as in the case of the patients of this series. Pneumothorax (3), infection (4) and, in the long term, degradation of the pulmonary parenchyma are the main complications (1). Surgery is indicated, especially now that—in most patients—it can be safely performed without opening the chest (5).

Patients and methods

Between 2007 and 2017, eight patients were operated on for a congenital bronchial atresia (CBA) in our department. All patients had a chest computed tomographic (CT) scan with

intravenous contrast and, more recently a 3-dimensional modelisation based on CT. Intraoperative and postoperative data (including demographic criteria, clinical symptoms, surgical procedure, and final histology) were recorded from this database which was approved from our Institutional Review Board (CEPAR 2013-002).

All eight patients were operated on by a full thoracoscopic approach with a fissure-based dissection of vascular elements. The technique has already been reported (6) and will not be described here.

Results

Among the eight patients, there were five males and three females with a median age of 29.5 years (range, 14 to 48 years). All patients were symptomatic with recurrent pneumoniae but one, in which CBA was incidentally discovered.

CT scan was in favor of CBA in all cases usually showing a cystic lesion within a distended parenchyma (*Figure 1*). 3D modelisation, based on injected CT, can clearly demonstrate the bronchial interruption and the pathological hyperinflated parenchyma (*Figure 2*). Five out of the

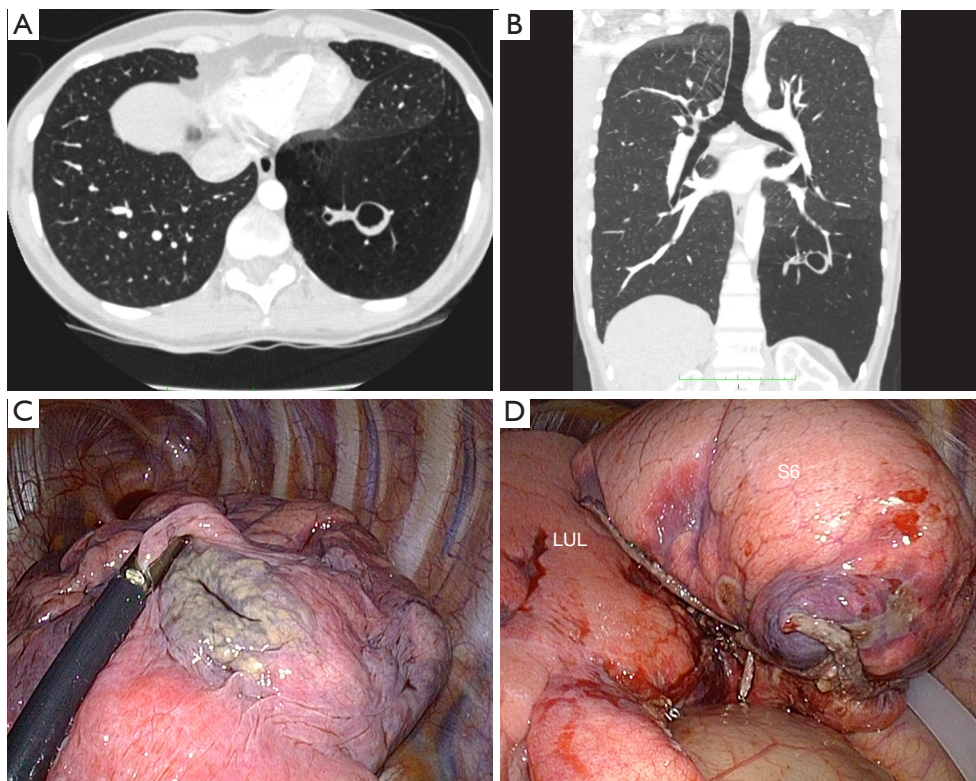


Figure 1 Congenital bronchial atresia of the left basilar segments. (A,B) CT-scan showing a cystic lesion with hypovascularisation; (C) thoracoscopic view before resection; (D) thoracoscopic view after basilar segmentectomy. LUL, left upper lobe.

pulmonary resections were a lobectomy and 3 an anatomic segmentectomy. No intraoperative complication occurred. Mean operation time was 204 min (range, 127–315 min), and mean blood loss was 89 mL (range, 0–200 mL). Five patients (62.5%) had minor postoperative complications: air leak (n=4) and pneumothorax (n=1) after chest tube removal, requiring drainage. Mean drainage duration was 5.5 days (range, 3–11 days). Mean hospital stay was 7.1 days (range, 4–14 days).

The diagnosis of CBA was confirmed on final pathological examination.

Discussion

Bronchial atresia is a rare congenital malformation related to a defect in vascularization during the development of the bronchial bud, resulting in a cul-de-sac termination of one or more bronchi. The adjacent pulmonary parenchyma develops normally and secretes mucus, which accumulates and forms mucoid impactions and/or bronchocele (1,2). Collateral ventilated alveoli

(Kohn's pores) provoke air retention and distension of the lung downstream of the atresia, which can cause recurrent infections and even mycobacterium infection (4) and, in the long term, destruction of the corresponding parenchyma. Pneumothorax can also reveal the malformation (3). Atresia is more common in segmental bronchi than in lobar ones. It can affect several territories (1,2). Although this abnormality is more often segmental than lobar (7), lobectomy is the most frequently reported resection since the remaining parenchyma is usually compressed by the abnormal segment, which is hyperinflated by trapping and by mucus accumulation (7,8). In a series of 29 bronchial atresias in children reported by Morikawa *et al.*, a segmentectomy was performed in only three cases (9). Other congenital malformations may be associated with bronchial atresia such as pectus excavatum (10). At histology, CBA is defined by an obstructed bronchus, which does not communicate with the rest of the bronchial tree, associated with bronchocele and surrounded by an over-inflated pulmonary parenchyma. Sometimes

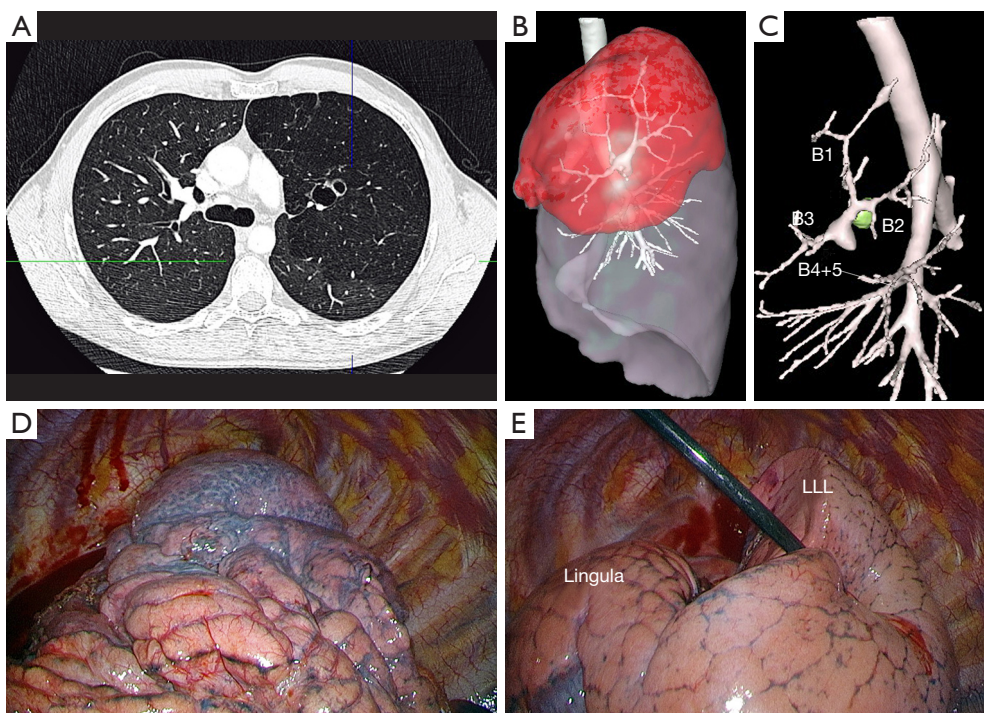


Figure 2 Congenital bronchial atresia of the left upper lobe involving segments 1, 2 and 3. (A) CT-scan showing a cystic lesion with hypovascularisation; (B) 3D-modelisation demonstrating the demarcation line between normal and pathological parenchyma; (C) 3D-modelisation showing atresia of the B1+2+3 bronchial trunk without communication with the left bronchial tree; (D) thoracoscopic view before resection; (E) thoracoscopic view after basilar segmentectomy. LLL, left lower lobe.

communication with the bronchial tree is preserved, but its light is occluded. Also, normal bronchoscopy does not eliminate the diagnosis of atresia. Thoracic CT demonstrates malformation by typically showing a cystic lesion within a distended parenchyma zone. 3D modelisation provides an additional help to the surgeon.

Bronchial atresia is most often diagnosed in the neonatal period or in childhood, with a radiological abnormality or repeated lung infections in the same territory, explaining why the largest series reported are pediatric (1,9). It is rarely discovered in adults, especially after 40 years (11). All our patients were operated by a full thoracoscopic approach (9). These patients are indeed young, with no comorbidity and with a good quality parenchyma. The limit between the healthy and pathological parenchyma facilitates thoracoscopic segmentectomy (12,13). In two patients of our series, the segment adjacent to the malformation was too pathological and a lobectomy seemed justified. In one of our patients with an atresia involving two different lobes, we limited the extent of the resection to a lobectomy by leaving in place a pathological segment in order to prevent

a re-expansion issue.

Conclusions

- (I) Surgical treatment of CBA seems justified, even in asymptomatic cases, to prevent recurrent infectious pulmonary complications and long-term lung alteration.
- (II) Parenchyma-sparing treatment should be discussed whenever possible.
- (III) A thoracoscopic approach should be used whenever possible.

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None.

Footnote

Conflicts of Interest: D Gossot is consultant for an instrument manufacturer (Delacroix Chevalier). The other authors

have no conflicts of interest to declare.

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