

Congenital Bronchopulmonary Foregut Malformation: Systematic review of the literature

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Abstract

Background : Congenital bronchopulmonary foregut malformation (CBPFM) is a rare congenital malformation involving both the digestive and respiratory system. Early diagnosis is difficult, and delayed recognition may result in considerable complications. The aim of the study was to identify and analyze the clinical characteristics and radiological features of CBPFMs. **Methods :** A systematic review was conducted in accordance with PRISMA guidelines. PubMed, Ovid database, EMBASE were searched for relevant publications to identify all published case-reports of CBPFM since 1992. Data about the demography, clinical presentation, pathology, imaging features, treatment and prognosis were collected. **Results :** Sixty-one cases were included in our study. Cases were aged from 1 day to 59 years with the majority aged 3 years or younger. The most common type was group III (37.7%), followed by group II (29.5%) group I (27.9%) and group IV (4.9%). The presentations included respiratory distress (32.8%), cough/choking following food intake and other presentations associated respiratory infection. Thirty-eight cases (62.3%) were diagnosed by upper gastrointestinal series (UGI). Misdiagnosis was common. Eight cases (13.1%) of the included cases died. **Conclusions :** Early recognition and extensive delineation of the anatomy of CBPFM are important to correct these anomalies successfully. UGI is the first choice to confirm the abnormal bronchus communicating with the esophagus. Resection of abnormal pulmonary tissue, lobe or even unilateral lung is preferred. Reconstruction procedures are feasible in selected patients.

Background

Communicating bronchopulmonary foregut malformation (CBPFM) is a rare congenital anomaly defined by a patent congenital communication between the esophagus or stomach and an isolated portion of the respiratory tract. Lesions including a lobar bronchus arising from the esophagus are described as an esophageal bronchus. If the main bronchus originates from the esophagus, it is termed esophageal lung¹. In 1992, based upon the occurrence of related defects and the level of the communication, Srikanth et al. reviewed 57 cases and proposed a system to classify CBPFM into four groups². Early diagnosis of CBPFM is difficult for the rarity of this entity and the nonspecific symptoms. We presented an infant with CBPFM and strictly reviewed the available literatures since Srikanth's classification proposed to describe the demography, clinical presentation, pathology, imaging features, treatment and prognosis of CBPFM.

Methods

Case

A 3.4kg female baby was born after an uneventful pregnancy. After born, she presented with mild respiratory distress and chest infection. A computed tomography (CT) scan was performed, which showed hypoplasia of the right lung and a dextrocardia. Antibiotics was administered, and the symptoms

alleviated. After that, the baby repeatedly presented with recurrent episodes of fever, cough, tachypnea and retraction and controlled by oral antibiotics. At 6 months old, she was admitted for persistent high fever (40°C) and dyspnea. CT scan showed hypoplasia of the right lung with multiple air bronchograms. The right mainstem bronchus originated from the distal esophagus and coursed to the right lung (Figure 1A). The upper gastrointestinal series (UGI) using iohexol (Omnipaque 350, GE Healthcare, Shanghai, China) as done to rule out the possibility of H-type tracheoesophageal fistula (TEF). It revealed filling of the right main bronchus and bronchial tree, which directly originated from the esophagus (Figure 1B). The bronchoscopy revealed a blind ended right bronchial stump and thus, the diagnosis of CBPFM (group II) was established. The echocardiography showed a small patent ductus arteriosus with good biventricular function and the left pulmonary artery arising distally from the right pulmonary artery.

Intra-operative findings revealed a heavily consolidated hypoplastic right lung with a single and thin right pulmonary artery and single right pulmonary vein. Diffuse abscesses were formed in the lung parenchyma (Figure 1C). The bronchus to the lung arose from the lower third of the esophagus. Right pneumonectomy with resection of esophageal bronchus was performed, and the abnormal opening in the esophagus was repaired. Postoperative recovery was uneventful, and the temperature returned to normal on the second day after surgery. A second UGI of the entire esophagus did not detect any leaking or fistula. The patient was discharged on postoperative day 7. She was doing well on follow-up 6 months later.

Figure 1 (A) CT scan showed hypoplasia of the right lung and the right mainstem bronchus originated from the distal esophagus. (B) The UGI revealed filling of the right main bronchus from the esophagus with opacifying the bronchial tree on the right side. (C) Diffuse abscesses were formed in the right lung parenchyma.

Systematic review

For reviewing of the literature, we conducted the systematic review following the recommendations of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement³. PubMed, OVID database and EMBASE were systematically searched for relevant articles published in English from January 1992 to August 2018. The search terms/syntax in PubMed was as follows: (bronchopulmonary foregut malformation OR "esophageal bronchus" OR "esophageal lung"). The titles and abstracts of all potentially relevant articles were read to determine their relevance. Full articles were also scrutinized if the title and abstract were unclear. Reference lists of identified articles were screened to ensure completeness of the search. All identified articles were independently assessed by two authors. Detailed data regarding patient characteristics, symptoms, initial diagnoses, diagnostic work-up, treatment and outcome were extracted into an electronic data sheet in a standardized manner.

The inclusion criteria were as follows: (I) Original studies describing individual cases of CBPFM in accordance with the definition proposed by Srikanth. (II) Studies describing adequate clinically relevant details. The exclusion criteria were as follows: (I) The malformation did not comply with the pattern of CBPFM. (II) Individual cases previously described in another study. Review articles and editorials were excluded. No statistical analysis was used for this review.

Figure 2. Preferred reporting items for systematic reviews and meta-analyses flowchart.

Results

One hundred and seventy-two papers were identified by searching the databases, one additional article was identified by manual searching. No reports were repetitive. Eighty-three reports were eligible for full-text screening. Of them, 38 papers matched our criteria and were included^{2, 4-40}. The selection study process is showed in the PRISMA flowchart in Figure 2. They included 60 cases of CBPFM. The 134 papers excluded were review articles, animal cases, published in a language other than English or malformation not meeting the diagnostic criteria depicted by Srikanth et al. Therefore, 61 cases were reviewed including our case (Table 1). The table in the appendix shows the details of the included cases.

Demographic characteristics

The gender of five cases was not reported, there were 26 males and 30 females, and male to female ratio was 0.87:1. The sex distribution among each CBPFM group was similar. The age was from 1 day to 59 years. A large proportion of these patients were aged 1 year or younger at presentation (n = 46) and 38 presented immediately after born. Seven adults have been described.

Classification and side

Among the included 61 cases, there were 13 cases in group IA, 4 cases in group IB, 18 cases in group II, 23 cases in group III and 3 cases in group IV. Forty-four malformations located in the right side, 14 in the left. Two cases had bilateral malformations. The right to left ratio was 3.1:1. The blood supply of abnormal lung tissue was reported in 39 cases, with from pulmonary artery in 17 and systematic artery in 22 respectively.

Gestation and family history

Polyhydramnios were reported in 10 cases (3 cases in Group I, 3 cases in Group II, 3 cases in Group III and 1 case in Group IV). Fetal MRI detected the tubular structure directing to the gastroesophageal junction during the second-trimester in 5 cases. These 5 cases belonged to Group III. A mass was detected in the fetal chest cavity in 2 cases. The gestation of the other cases was reported as uncomplicated or not mentioned.

There were four cases from three twins. Two cases were monochorionic, monoamniotic twins. One of them was group IA with VACTERL association, but the other one was group II without associated malformation. The other two cases were from two twins, with one of the twins affected but the siblings were normal.

Symptoms

Twenty patients presented with respiratory distress after birth. All patients with group I had drooling, feeding intolerance and failure to pass nasogastric tube because of esophageal atresia. Recurrent respiratory infection, presenting with cough and/or fever, were the main symptoms in older children and adults (10/22, 45.5%). Other symptoms included cough/choking following food intake (n=5), hemoptysis (n=2), nocturnal cough (n=1), epigastric pain (n=1).

Associated malformations

CBPFM group I was associated esophageal atresia and tracheoesophageal fistula (EA/TEF). So, EA/TEF was not counted as associated malformation in our review. Cardiovascular anomalies were the most common associated malformation (n= 11, 18.0%), followed by VACTERL association (n=6, 9.8%), skeletal malformation (n=6, 9.8%), anorectal malformation (n=2, 3.3%), diaphragmatic hernia (n=2, 3.3%) and other less frequently malformations.

Diagnosis

For group I, chest radiographs were obtained in 12 cases. The hazy hemithorax and mediastinal shift with dextrocardia were revealed in 9, hypoaerated or normal lung in 3. Only one case of CBPFM was suggested by CT scan initially. One case without chest X-ray was demonstrated during the surgery for EA/TEF. Fourteen cases with group I CBPFM were not diagnosed correctly and received operation for EA/TEF initially (ligation of tracheoesophageal fistula, primarily esophageal anastomosis or gastrostomy). Definitive diagnoses were confirmed by further evaluations that were prompted by persistent atelectasis of one lung, refractory respiratory distress or routine postoperative UGI.

Opacification of ipsilateral lung and mediastinal shift were also the uniform presentations in chest radiograph of group II (18/18). Plain X-ray in group III and group IV were reported in 11 cases. The presentations were various, including mediastinal or pulmonary mass (n=5), consolidation of part of the lung (n=6).

The diagnoses of CBPFM were confirmed by UGI in 38 case (62.3%), CT in 7 cases (11.5%), bronchoscopy in 1 case (1.6%) and intraoperative finding in 8 cases (13.1%). There were five cases diagnosed prenatally by ultrasonography and fetal MRI.

Treatment

Fifty-five cases underwent surgeries. Unilateral pneumonectomies were performed in six cases with group I, 11 cases with group II and one case with group IV. Three cases with Group IA and four cases with group II received the reimplantation of esophageal bronchus. Lobectomies or resection of aberrant lung tissue and bronchus were done in 24 cases with group IB, group III or group IV. The procedures of the other 6 cases were unknown.

Outcome

Eight patients died in this series with 4 in group I, 2 in group IB, 1 in group II and group III respectively. The mortality was 12.9%. In survivors, 27 cases were reported as doing-well or uneventful. Eight cases had respiratory morbidities such as air way stenosis, tracheomalacia, repeat respiratory infection or difficult in weaning from ventilator. Postpneumonectomy syndrome occurred in two cases. The outcomes of 15 cases were unknown.

Discussion

The term of congenital bronchopulmonary foregut malformation was proposed by Gerle et al. in 1968 to describe either an intralobar or an extralobar sequestration communicating with the gastrointestinal tract⁴¹. Srikanth et al. combined 30-year (1959 to 1989) experience of 6 cases of CBPFM with 51 reported patients and proposed the classification².

The embryogenesis of CBPFMs is not clear though it was treated as a sub-type of bronchopulmonary foregut malformation including foregut vascular abnormalities, lung parenchymal abnormalities and airway anomalies⁴². This malformation was proposed to arise when a mass of lung bud connecting to the esophagus through a mesodermal defect. The aberrant lung tissue was separated with the rest of the lung and trachea during the rapid elongating of the esophagus, which accounted for the missing portion for the corresponding bronchi tree²⁷. CBPFM was also occurred in adriamycin-induced rat model of

esophageal atresia. The observations were consistent with the hypothesis that CBPFM and EA were variations of spectrum of abnormalities and may have a similar etiology ⁴³.

Our series were in agree with that reported by Srikanth et al. in the female and right-side preponderance. The distribution of numbers among groups was similar. But the mortality in Srikanth's series was 24.6% compared with 13.1% after 1992. The increasing of survival rate may attribute to the improvement of critical care of the newborn. Nevertheless, delay of diagnosis was still common for the rarity of the entity. Especially for group I, the existence of CBPFM was always obscured by the presentation of EA/TEA. Though the abnormal pulmonary tissue could be aerated through the communication of the esophagus before the inflammatory commenced, collapse of the ipsilateral lung was the typical presentation. Hence the persistent or worsening unilateral lung collapse with or without EA/TEF should raise the suspicion and warranted further examinations to exclude CBPFM. UGI was the first choice to delineate the bronchial tree connected to esophagus, but the value was limited in group I for the atresia of the proximal esophagus. In this circumstance, bronchoscopy should be considered. Computed tomography (CT) is helpful in evaluating the pulmonary damage and vascularization.

Prenatal diagnosis was possible in some high-volume institution by experienced radiologist ²⁰. In fetal MRI, a tubular T2 hyperintense structure (bronchocele) directed from the lung lesion to the gastroesophageal junction can be seen. The congenital lung lesions are now more commonly recognized on screening anatomic ultrasound, and CBPFM should be as a differential diagnosis.

No predisposing factors have been identified in the etiology of CBPFM, although the observed concordance of this lesion in a pair of twins arguing in favor of an underlying genetic predisposition. Screening for associated malformations was important for the high possibility. Echocardiography was mandatory preoperatively because the cardiovascular malformation occurred in 18.0% of the patients. VACTERL association was not uncommon and a comprehensive examination should be performed. Accompanying airway stenosis was the most difficult problem which always resulted in long-term morbidity. The management needed extensive experiences.

Surgery was the only method to cure this malformation. Resection of the anomalous lung or lobe through an open or thoracoscopic approach was straightforward. Pneumonectomy was performed in most patients with group IA or group II. Unilateral lung resection in neonates and infants was well tolerated. But the prevalence of long-term consequences, such as chest wall deformation, scoliosis and postpneumonectomy syndrome were unclear. One patient developed postpneumonectomy syndrome in our review. The tissue expander was inserted, and the symptom subsided with growth. The other choice was reconstruction of the esophagus bronchus. This was the theoretically preferential procedure to preserve the affected lung and prevent the complications of pneumonectomy. The first successful reconstruction effort was reported by Michael et al. in 1997 ¹⁶. Five reconstruction procedures were included in our study with one underwent pulmonary artery reimportation simultaneously. The results were dismal due to the airway stenosis or accompanying malformations. Therefore, attempts to

reconstruction of the bronchus should be confined to stable patients with non-infectious lung and significant associated malformations excluded.

Because most of the included studies were single case reports. The Selection bias cannot be ignored for the possible tendency of more successful interventions being preferred for publication. But the distribution of sex, classification and malformation side was similar with previous reported series. Our study samples may be representative.

Conclusions

CBFPM is rare and the outcome depends on the associated malformations and early recognition. Persistent opaque hemithorax with ipsilateral mediastinal shift on chest radiograph should raise the suspicion of CBFPM in infants. UGI is the preferred method to delineate the abnormal bronchus communicating with the esophagus. CT and bronchoscopy are also valuable as a complementary method or screening for accompanying malformations. Echocardiography is mandatory for the high frequency of associated congenital heart disease. Resection of abnormal pulmonary tissue, lobe and even unilateral lung is the reasonable treatment in most cases. Decision of parenchymal sparing surgery, such as reimplantation of bronchus, should be prudent for the high possibility of long-term complications.

Declarations

List of abbreviations

CBPFM, Congenital bronchopulmonary foregut malformation

UGI, upper gastrointestinal series

EA/TEF, esophageal atresia and tracheoesophageal fistula

Ethics approval and consent to participate

This article does not contain any studies with human participants or animals performed by any of the authors.

Consent for publication

Informed consent was waived by our IRB due to the fact that the data collected for this review were retrospective and de-identified.

Availability of data and materials

All data generated or analyzed during this study are included in this published article and its supplementary information files.

Competing interests

The authors have no conflict of interest to disclose.

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Authors' contributions

GY collected data and wrote the draft. YL contributed to the elaboration of the ideas developed in the manuscript and made critical amendments. CX and LNC contributed to the data collection and interpretation. MY provided the statistical analysis. All authors read and approved the final manuscript.

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Not applicable

References

1. Lallemand D, Quignodon JF, Courtel JV. The anomalous origin of bronchus from the esophagus: report of three cases. *Pediatr Radiol.* 1996;26(3):179-82.
2. Srikanth MS, Ford EG, Stanley P, Mahour GH. Communicating bronchopulmonary foregut malformations: classification and embryogenesis. *J Pediatr Surg.* 1992 Jun;27(6):732-6.
3. Moher D, Liberati A, Tetzlaff J, Altman DG; PRISMA Group. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA Statement. *Open Med.* 2009;3(3):e123-30.
4. Takamizawa S, Yoshizawa K, Machida M, *et al.* Successful tracheobronchial reconstruction of communicating bronchopulmonary foregut malformation and long segment congenital tracheal stenosis: a case report. *J Pediatr Surg.* 2012 Sep;47(9):E41-6.
5. Yutaka Y, Omasa M, Shikuma K, Taki T. Bronchopulmonary foregut malformation in an adult. *Gen Thorac Cardiovasc Surg.* 2007 Nov;55(11):476-8.
6. Verma A, Mohan S, Kathuria M, Baijal SS. Esophageal bronchus: case report and review of the literature. *Acta Radiol.* 2008 Mar;49(2):138-41.
7. Usui N, Kamata S, Ishikawa S, *et al.* Bronchial reconstruction for bronchopulmonary foregut malformation: a case report. *J Pediatr Surg.* 1995 Oct;30(10):1495-7.
8. Tsuchiya T, Mori K, Ichikawa T, *et al.* Bronchopulmonary foregut malformation diagnosed by three-dimensional CT. *Pediatr Radiol.* 2003 Dec;33(12):887-9.

9. Tsugawa J, Tsugawa C, Satoh S, *et al.* Communicating bronchopulmonary foregut malformation: particular emphasis on concomitant congenital tracheobronchial stenosis. *Pediatr Surg Int.* 2005 Nov;21(11):932-5.
10. Ren H, Duan L, Zhao B, Wu X, Zhang H, Liu C. Diagnosis and treatment of communicating bronchopulmonary foregut malformation: Report of two cases and review of the literature. *Medicine (Baltimore).* 2017 Mar;96(11):e6307.
11. Sumner TE, Auringer ST, Cox TD. A complex communicating bronchopulmonary foregut malformation: diagnostic imaging and pathogenesis. *Pediatr Radiol.* 1997 Oct;27(10):799-801.
12. Sugandhi N, Sharma P, Agarwala S, Kabra SK, Gupta AK, Gupta DK. Esophageal lung: presentation, management, and review of literature. *J Pediatr Surg.* 2011 Aug;46(8):1634-7.
13. Segulier-Lipszyc E, Dager S, Malbezin S, Aigrain Y, de Lagausie P. Reimplantation of oesophageal bronchus following a type III oesophageal atresia repair. *Pediatr Surg Int.* 2005 Aug;21(8):649-51.
14. Singal AK, Kumar VR, Rao M, Matthai J. Bilateral communicating bronchopulmonary foregut malformations in a child. *Ann Thorac Surg.* 2006 Jul;82(1):330-2.
15. Saydam TC, Mychaliska GB, Harrison MR. Esophageal lung with multiple congenital anomalies: conundrums in diagnosis and management. *J Pediatr Surg.* 1999 Apr;34(4):615-8.
16. Michel JL, Revillon Y, Salakos C, *et al.* Successful bronchotracheal reconstruction in esophageal bronchus: two case reports. *J Pediatr Surg.* 1997 May;32(5):739-42.
17. Patil M, Sutagatti J, Bhavikatti M, Nayak PV. A rare case of esophageal lung in a neonate. *Indian J Radiol Imaging.* 2016 Apr-Jun;26(2):206-9.
18. Partridge EA, Victoria T, Coleman BG, *et al.* Prenatal diagnosis of esophageal bronchus—first report of a rare foregut malformation in utero. *J Pediatr Surg.* 2015 Feb;50(2):306-10.
19. Matsusaka K, Kinoshita Y, Udagawa H, Fukayama M, Ohashi K. Squamous cell carcinoma arising in a communicating bronchopulmonary-foregut malformation. *Hum Pathol.* 2010 Nov;41(11):1650-4.
20. Park T, Jung K, Kim HY, Jung SE, Park KW. Thoracoscopic management of a communicating bronchopulmonary foregut malformation in a 23-month-old child. *J Pediatr Surg.* 2012 Mar;47(3):e21-3.
21. Nakaoka T, Uemura S, Yano T, *et al.* Successful reconstruction of communicating bronchopulmonary foregut malformation associated with laryngotracheoesophageal cleft. *J Pediatr Surg.* 2009 May;44(5):e29-32.
22. Murray ME, Given-Wilson RM, Christopher JA, Jeffrey IJ. Bilateral communicating bronchopulmonary foregut malformations in an infant with multiple congenital anomalies. *Pediatr Radiol.* 1994;24(2):128-30.
23. Matsumoto Y, Ohi R, Hayashi Y, Chiba T. Right pneumonectomy syndrome: report of two cases. *Surg Today.* 1995;25(3):278-80.
24. Lucaya J, Garel L, Martín C. Clinical quiz. Extralobar sequestration, esophageal bronchus (bronchopulmonary foregut malformation). *Pediatr Radiol.* 2003 Sep;33(9):665-6.

25. Linnane BM, Canny G. Congenital broncho-esophageal fistula: A case report. *Respir Med.* 2006 Oct;100(10):1855-7.
26. He QM, Xiao SJ, Zhu XC, *e al.* Communicating bronchopulmonary foregut malformation type IA: radiologic anatomy and clinical dilemmas. *Surg Radiol Anat.* 2015 Dec;37(10):1251-6.
27. Lee P, Westra S, Baba T, McCauley R. Right pulmonary aplasia, aberrant left pulmonary artery, and bronchopulmonary sequestration with an esophageal bronchus. *Pediatr Radiol.* 2006 May;36(5):449-52.
28. Kim CW, Kim DH. Single-incision video-assisted thoracic surgery lobectomy in the treatment of adult communicating bronchopulmonary foregut malformation with large aberrant artery. *J Thorac Dis.* 2016 Jan;8(1):E148-51.
29. Katz R, Pitt R, Kim D, Wingrove B. Thoracoscopic pneumonectomy for communicating bronchopulmonary foregut malformation in a 4-month-old child. *J Pediatr Surg.* 2010 Feb;45(2):427-9.
30. Katayama Y, Kusagawa H, Komada T, Shomura S, Tenpaku H. Bronchopulmonary foregut malformation. *Gen Thorac Cardiovasc Surg.* 2011 Nov;59(11):767-70.
31. Jamieson DH, Fisher RM. Communicating bronchopulmonary foregut malformation associated with esophageal atresia and tracheo-esophageal fistula. *Pediatr Radiol.* 1993;23(7):557-8.
32. Eom DW, Kang GH, Kim JW, Ryu DS. Unusual bronchopulmonary foregut malformation associated with pericardial defect: bronchogenic cyst communicating with tubular esophageal duplication. *J Korean Med Sci.* 2007 Jun;22(3):564-7.
33. Colleran GC, Ryan CE, Lee EY, Sweeney B, Rea D, Brenner C. Computed tomography and upper gastrointestinal series findings of esophageal bronchi in infants. *Pediatr Radiol.* 2017 Feb;47(2):154-160.
34. Chung JH, Lim GY, Kim SY. Esophageal lung diagnosed following the primary repair of esophageal atresia with tracheo-esophageal fistula in a neonate. *Surg Radiol Anat.* 2014 May;36(4):397-400.
35. Alsaadi A, Alsufiani HA, Allugmani MD, Gora AH. Esophageal lung with rare associated vascular and anorectal malformations. *Radiol Case Rep.* 2018 Feb 28;13(2):444-448.
36. Bokka S, Jaiswal AA, Behera BK, Mohanty MK, Khare MK, Garg AK. Esophageal lung: A rare type of communicating bronchopulmonary foregut malformation, case report with review of literature. *J Indian Assoc Pediatr Surg.* 2015 Apr-Jun;20(2):92-4.
37. Becker J, Hernandez A, Dipietro M, Coran AG. Identical twins concordant for pulmonary sequestration communicating with the esophagus and discordant for the VACTERL association. *Pediatr Surg Int.* 2005 Jul;21(7):541-6.
38. Boersma D, Koot BG, van der Griendt EJ, van Rijn RR, van der Steeg AF. Congenital bronchopulmonary foregut malformation initially diagnosed as esophageal atresia type C: challenging diagnosis and treatment. *J Pediatr Surg.* 2012 Oct;47(10):e59-62.
39. Borsellino A, Alberti D, Vavassori D, Pericotti S, Cheli M, Locatelli G. Communicating bronchopulmonary foregut malformation involving a mixed sequestration/cystic adenomatoid

- malformation: a case report. *J Pediatr Surg.* 2002 Nov;37(11):E38.
40. Rahman GF, Bhardwaj N, Suster B, Arliss JJ, Connery CP. Communicating bronchopulmonary pancreatic foregut malformation. *Ann Thorac Surg.* 1999 Dec;68(6):2338-9.
41. Gerle RD, Jaretzki A 3rd, Ashley CA, Berne AS. Congenital bronchopulmonary-foregut malformation. Pulmonary sequestration communicating with the gastrointestinal tract. *N Engl J Med.* 1968 Jun 27;278(26):1413-9.
42. Choo JY, Hwang J, Lee JH, Lee KY. Bronchopulmonary foregut malformation presenting as extralobar pulmonary sequestration associated with a bronchogenic cyst: an unusual clinical and radiological feature in an adolescent patient. *J Thorac Dis.* 2017 Jul;9(7):E632-E635.
43. Qi BQ, Beasley SW. Communicating bronchopulmonary foregut malformations in the adriamycin-induced rat model of oesophageal atresia. *Aust N Z J Surg.* 1999 Jan;69(1):56-9.

Tables

Table 1 Clinical characteristics of included patients in the systematic review

Clinical Features	Group					Total
	IA	IB	II	III	IV	
Sex						
Male	7	2	8	9		26
Female	6	2	10	9	3	30
Not stated				5		5
Age						
Newborn	13	3	10	12		38
Infant (<1 yr)			5	2	1	8
Child (<18 yr)		1	3	3	1	8
Adult				6	1	7
Not Stated			1			1
Side						
Right	9	4	14	14	3	44
Left	3		4	7		14
Bilateral				2		2
Not stated	1					1
Arterial supply						
Systemic	2	1	2	17		22
Pulmonary	3		11	2	1	17
Not stated	8	3	5	4	2	22
Diagnostic method						
UGI	10	2	14	10	2	38
CT	2	1	4			7
Bronchoscopy	1					1
Intraoperative		1		7		8
Other				6	1	7
Outcome						
Survive	7	2	12	14	3	38
Death	4	2	1	1	0	8
Not stated	2		5	8		15

UGI, upper gastrointestinal series.

Figures

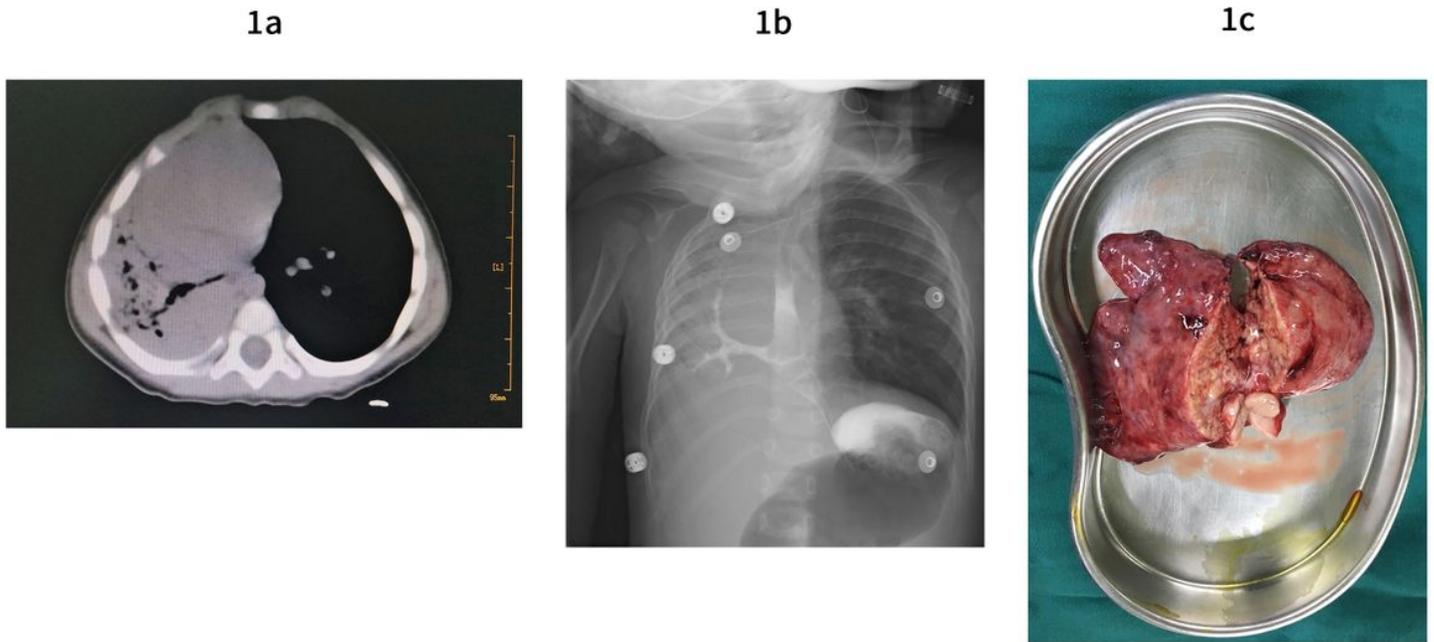


Figure 1

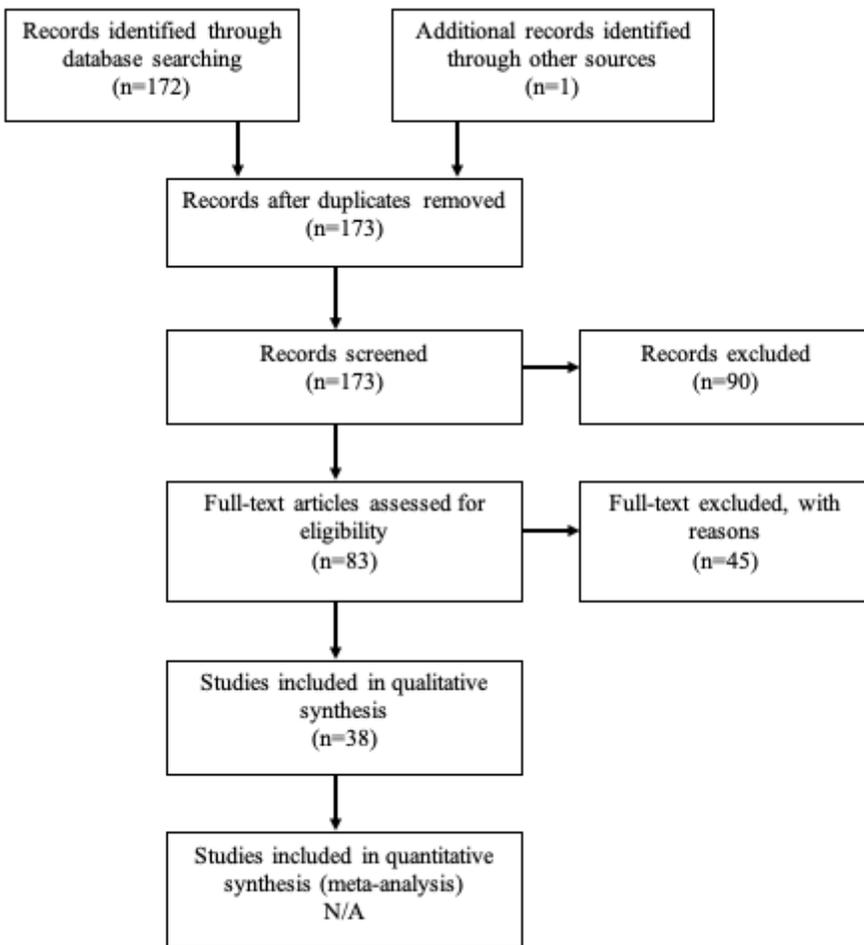


Figure 2

Preferred reporting items for systematic reviews and meta-analyses flowchart.

Supplementary Files

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