Journal Pre-proofs

Review

A clinical guideline for structured assessment of ct-imaging in congenital lung abnormalities

Sergei M. Hermelijn, Bernadette B.L.J. Elders, Pierluigi Ciet, René M.H. Wijnen, Harm A.W.M. Tiddens, J. Marco Schnater

PII: S1526-0542(20)30019-1
DOI: https://doi.org/10.1016/j.prrv.2019.12.004
Reference: YPRRV 1362

To appear in: *Paediatric Respiratory Reviews*

Received Date: 16 August 2019
Revised Date: 11 December 2019
Accepted Date: 13 December 2019

Please cite this article as: S.M. Hermelijn, B.B.L. Elders, P. Ciet, R.M.H. Wijnen, H.A.W. Tiddens, J. Marco Schnater, A clinical guideline for structured assessment of ct-imaging in congenital lung abnormalities, *Paediatric Respiratory Reviews* (2020), doi: https://doi.org/10.1016/j.prrv.2019.12.004

A CLINICAL GUIDELINE FOR STRUCTURED ASSESSMENT OF CT-IMAGING IN CONGENITAL LUNG ABNORMALITIES

Sergei M. Hermelijn[1] MD, Bernadette B.L.J. Elders[2,3] MD, Pierluigi Ciet[2,3] MD, PhD René M.H. Wijnen[1] MD, PhD, Harm A.W.M. Tiddens[2,3] MD, PhD , J. Marco Schnater[1] MD, PhD

[1] Department of Paediatric Surgery, Erasmus University Medical Centre, Sophia Children's Hospital, Rotterdam, The Netherlands
[2]Department of Paediatric Pulmonology, Erasmus University Medical Centre, Sophia Children's Hospital, Rotterdam, The Netherlands
[3]Department of Radiology, Erasmus University Medical Centre, Rotterdam, The Netherlands

Corresponding Author:

J.M. Schnater, MD, PhD
Erasmus MC – Sophia Children's Hospital
Dr. Molewaterplein 40. 3015 GD Rotterdam

Mailing address: Postbus 2060. 3000 CB Rotterdam
Email: j.schnater@erasmusmc.nl
Telephone number: (+31)107036214

Acknowledgements: We would like to thank Wichor Bramer, biomedical information specialist, for his expertise with the literature search. We thank Ko Hagoort for critically reviewing the manuscript and providing editorial advice.

Conflicts of Interest and Source of Funding: We declare no conflicts of interest and this research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Abbreviations:

CLA	Congenital Lung Abnormalities
CPAM	Congenital Pulmonary Airway Malformation
BPS	Bronchopulmonary Sequestration
CLE	Congenital Lobar Emphysema
BC	Bronchogenic Cyst
CT	Computed Tomography
ILS	Intralobar Sequestration
ELS	Extralobar Sequestration
PPB	Pleuropulmonary Blastoma

Educational aims

The reader will be able:

- To comprehend the broad spectrum of CT-imaging findings as well as variable nomenclature in congenital lung abnormalities
- To understand why the overlap between abnormalities on pathology make distinction on CT-imaging difficult and unreliable
- To understand why radiological appearances should not be categorized using a pathology based classification system
- To structurally assess and report chest CT findings of congenital lung abnormalities utilizing uniform nomenclature

Future research directions

- Prospective imaging studies utilizing uniform nomenclature may make comparison between studies more reliable and expand our understanding of congenital lung abnormalities
- Validate the use of the structured report for clarity of radiological reports

Keywords:

- Ceongenital lung disease, Computerised Tomography scan, lung malformation, pediatric pulmonology, lung surgery

Abstract

Objectives: To develop a clinical guideline for structured assessment and uniform reporting of congenital lung abnormalities (CLA) on Computed Tomography (CT)-scans.

Materials and Methods: A systematic literature search was conducted for articles describing CT-scan abnormalities of congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar emphysema (CLE) and bronchogenic cyst (BC). A structured report using objective features of CLA was developed after consensus between a paediatric pulmonologist, radiologist and surgeon.

Results: Of 1581 articles identified, 158 remained after title-abstract screening by two independent reviewers. After assessing full-texts, we included 28 retrospective cohort-studies.

Air-containing cysts and soft tissue masses are described in both CPAM and BPS while anomalous arterial blood supply is only found in BPS. Perilesional low-attenuation areas, atelectasis and mediastinal shift may be found in all aforementioned abnormalities and can also be seen in CLE as a cause of a hyperinflated lobe. We have developed a structured report, subdivided into five sections: Location & Extent, Airway, Lesion, Vascularization and Surrounding tissue.

Conclusions: CT-imaging findings in CLA are broad and nomenclature is variable. Overlap is seen between and within abnormalities, possibly due to definitions often being based on pathological findings, which is an unsuitable approach for CT imaging. We propose a structured assessment of CLA using objective radiological features and uniform nomenclature to improve reporting.

Introduction

Structured prenatal screening and technical advances in prenatal ultrasound imaging have led to an increase in the detection of congenital lung abnormalities (CLA) [1]. Postnatally, computed tomography (CT) is the gold standard for diagnosis in pre-operative patients due to its high spatial resolution and short acquisition times. Yet, the diagnosis can be challenging due to overlapping features and inconsistent use of nomenclature [2].

The most common CLA in order of prevalence are congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar emphysema (CLE) and bronchogenic cyst (BC).

CPAM is a cystic malformation with aberrant connection to the tracheobronchial tree caused by overgrowth of terminal bronchioles. Its appearance on postnatal imaging is dependent on the subtype (type 0 - 4) of CPAM, and varies from a mass consisting of multiple small cysts to large air-filled cysts (figures 1-2) [4]. BPS is an non-~~a~~functional lung mass without a connection to the bronchial tree, and vascularized by systemic arteries (figures 3-4). A distinction is made between intra-lobar sequestration (ILS), which is located within the pleura of the lung, and extra-lobar sequestration (ELS) which is contained within its own pleura. Both types receive systemic arterial blood supply and often differ in venous drainage as ILS commonly drain to the pulmonary vein, while ELS often drain to the azygos system. A hybrid BPS is reported when a sequestration is seen in conjunction with CPAM [5]. CLE is a hyperinflation of lung tissue that usually occupies a whole lobe and is caused by abnormal bronchial wall development or external compression on an airway (figure 5) [6]. Some prefer the term hyperinflation instead of emphysema as overdistention of intact alveoli is found on pathology opposed to destructed alveoli in emphysema [7]. We encourage the use of the newer term Congenital Lobar Overinflation (CLO) as this more accurately describes this disease~~,~~ but will use CLE throughout this paper to prevent confusion in the literature review. BC is a fluid-filled cyst, typically located in the mediastinum near the carina, but also occurring within the lung parenchyma [8]. Contents may have a mucoid substance or appear milky due to calcium depositions within the cyst [8,9].

Most CLA are asymptomatic and thus are often missed if not detected prenatally. In some cases they may be an incidental finding, such as BC, which may cause dysphagia, persistent cough or retrosternal chest pain [9]. ELS and CPAM type 2 may be diagnosed in association with other congenital anomalies such as congenital diaphragmatic hernia, vertebral anomalies and congenital heart disease [10,11]. However, most BPS remain asymptomatic, especially ELS [7,12,13]. In contrast, CLE usually causes respiratory distress within the first 6 months and earlier presentation is associated with worse distress [14]. More recently, bronchial atresia is increasingly being reported in conjunction with CPAM and CLE as a diagnosis depicting an abrupt interruption of the airway with a distal mucus plug. However, this is more likely a feature which may be found in combination with the underlying CLA rather than a diagnosis on its own [15,16].

While surgical resection is the generally accepted treatment in symptomatic patients, the management in asymptomatic patients is still under debate [17]. Standardized guidelines for describing radiological CLA-related features on chest CT are lacking, although a clear pathological classification exists [11]. We aimed to summarize the radiological appearance of parenchymal abnormalities in CLA on chest CT and devised a standardized, objective manner of assessing and reporting them.

Materials and Methods

Systematic literature search

To assess current descriptions in the literature we conducted a systematic literature search in accordance with PRISMA guidelines [18]. A search of all published reports on postnatal CT-imaging of CLA was carried out in the PubMed, EmBase, Medline, Web of Science, Cochrane and Google Scholar database in August 2017 and updated in July 2019. The search strategy is summarized in figure 6 and search terms are supplied in the online supporting information.

All articles published in English until August 2018 and describing a paediatric cohort with a median/mean age ≤ 18 years were included. Articles and case-series exceeding 5 cases, describing parenchymal CT-abnormalities of CPAM, BPS, CLE and BC, were studied in detail. Articles with a cohort consisting of multiple CLA and < 5 cases for each individual diagnosis were included as well. Articles that only described prenatal imaging and articles reporting findings of other than the aforementioned CLA were excluded. Articles reporting only confirmation of diagnosis by means of CT-imaging without describing specific parenchymal abnormalities in detail were excluded as well.

Data extraction & construction of guide

From each article we extracted all CT-imaging descriptions for the CLA concerned~~,~~ and based our approach on distinctive features which may be useful in diagnosis and clinical decision-making. The paediatric surgeon and pulmonologist screened for clinically relevant features with implications for further management and the paediatric radiologist added essential features for diagnosis. Using the information extracted from these articles, a structured way of reporting was developed by consensus (Figure 7). Reporting of these features was translated into objective radiological terms with help of the Fleischner Society lexicon [19].

Results

The updated literature search resulted in 1581 citations of which 158 remained after title/abstract screening and 28 after assessing full texts (figure 6). Articles were independently assessed by two reviewers (SH, BE) and discordant selections were discussed to reach a consensus. All 28 articles reported a retrospective study; no randomized controlled trials were found.

Article characteristics and CT-imaging features can be found in Ttables 1a-d; bar charts visualizing the weighted prevalence of each feature are displayed in Ffigure 8. The prevalence of features for each abnormality alongside corresponding bar charts are shown in descending order in the online supplements.

CPAM was described in 16 articles and identified as air-containing cysts in all 16 [7,20-34]. Thirteen of these articles distinguish between a single cyst and multiple cysts [7,20-31]; four also describe fluid-filled cysts [7,24,27,30] and four describe air-fluid levels [21,24,30,33]. Ten articles describe a soft tissue mass which is dependent on the type of CPAM [7,20-22,26,29-32,34]. Perilesional low-attenuating lung parenchyma (four articles) [23-25,34], emphysema (four articles) [21,26,27,33], atelectasis (six articles) [21,23,24,26,32,33] and hyperinflation (five articles) [22,23,30-32] are described, as well as mediastinal shift (six articles)[22,23,28,30,32,33].

BPS was described in ten articles and identified as a soft-tissue mass with an anomalous arterial blood supply in all ten [5,7,12,13,20,23,27,34-36]. Aerated cysts are described in six articles [12,13,20,23,35,36] and three articles distinguish between a single cyst and multiple cysts [13,23,35]. Two articles describe air-fluid levels [12,35] and one article describes fluid filled cysts [13]. Perilesional emphysema is described in two articles [5,12] and perilesional low-attenuating lung parenchyma in four [7,13,23,34]. Two articles describe atelectasis [23,35] and one described mediastinal shift [23]. Low-attenuation [5] and air within the mass [12] are each described in one article.

CLE was described in ten articles and identified as hyperinflation of the affected segment in nine of the ten articles [6,7,14,20,28,34,37-39]. One article describes only emphysematous changes in the lung parenchyma [40], whereas five describe this in addition to hyperinflation [6,7,14,34,38]. These abnormalities are variably

reported in combination with mediastinal shift (eight articles) [6,7,14,28,37-40], herniation of lung through the mediastinum (seven articles) [6,14,28,37-40], atelectasis of neighbouring lobes (six articles) [6,14,37-40] and decreased lung vessel attenuation (five articles) [7,20,28,37,40]. A bronchial stenosis is described in six articles [6,14,34,37,38,40]. One described a normal bronchial tree [39] and three did not report these findings [7,20,28].

Mediastinal and parenchymal BC was described in four articles [7,8,20,27] and identified as a solitary, round or ovoid cystic structure in three articles [7,8,20]. Two describe a soft tissue mass [8,27], two a well-defined, smooth border [7,20] and one article describes atelectasis of neighbouring structures [8].

Anomalous artery in BPS, contralateral herniation and vessel attenuation in CLE were the only exclusive features.

We assessed all the features described in our literature search, categorized them based on anatomical distribution as is customary in lung CT imaging [41] and selected features relevant for radiological diagnosis and/or surgical planning. These features were divided into five clinically important main categories: the location and extent, airway connection, characterization of the lesion, vascularization and aspect of surrounding lung tissue.

Discussion

Our results show a broad range of CT- imaging findings in CLA and inconsistent use of nomenclature. Imaging features clearly overlap between and within various abnormalities. This is partly due to definitions being based on pathological findings, which might not be the best approach for CT-imaging [2,34,42,43]. A glossary of terms for thoracic imaging is provided by the Fleischner Society, but it is not uniformly used and not specifically developed for CLA [19].

Overlap in CT-imaging features

In CPAM, various cystic abnormalities corresponding to the respective subtypes are seen. Depending on the composition of the cohort, either single aerated cysts are described [32-34] or multiple cysts [7,20-31]. In CPAM type 3, a soft tissue mass can be seen, also one of the most common features in BPS. The systemic arterial blood supply to the lung mass in BPS distinguishes this entity from a CPAM although this is not always easily discernible. Perilesional features such as low-attenuation, mediastinal shift, and atelectasis may be seen in all CLA due to mass effect and are dependent on the size and location of the lesion[30,44]. In both BPS and CPAM the terms hypodense area and emphysema are variably used to describe an area of low attenuation although emphysema suggests structural lung damage. Similarly, in CLE the terms hyperinflation or overinflation are sometimes preferred instead of emphysema because pathological examination shows overdistended but intact alveoli in contrast to the destructed alveoli seen in emphysema [7]. Previous studies assessing correlations between CT findings and pathology show a good correlation and accurate diagnosis of the type of CLA on CT. Yet, distinguishing CPAM subtypes and hybrid lesions proved to be more difficult [8,23-25,29,34]. Previous studies suggested different classification systems for the various CLA based on either clinical relevance, pathological appearance or pathogenesis [2,43,45-47]. Each classification system has its merits and limitations and, so far, no consensus has been reached on the optimal classification of these CLA.

In our opinion, a more objective description depending on the most relevant findings is needed. This will ensure that future studies use the same parameters, which facilitates comparison of study findings. In

order to achieve this, we have constructed a structured radiologic report which can be used as a guide for structured assessment and uniform reporting of CT-imaging findings in CLA. We have subdivided this into five sections (Location & Extent, Airway, Lesion, Vascularization and Surrounding tissue) and explain the relevance of the features.

Location & Extent (figure 7, section A)

The location and extent of the CLA is essential for the clinical management and potential surgical plan [34]. For instance, it is known that upper lobe involvement is associated with more severe symptoms [14,37,38]. Similar to reporting CT-imaging of cystic fibrosis, we suggest reporting the affected lobes as well as the extent in each lobe expressed as the affected volume in thirds (up to ⅓ lobe, up to ⅔ lobe, complete lobe; see figure 7, section A) [48]. This lobe-oriented approach is clinically relevant as a lobectomy is preferred over a wedge resection, due to more frequent postoperative complications such as persistent air leakage, residual disease in up to 15% of cases [49] and risk of malignancy in the residual tissue [50,51].

We suggest reporting the affected lobes as well as the extent in each lobe expressed as the affected volume in thirds

Airway (figure 7, section B)

Different CLA can be distinguished by looking at the connection to the bronchial tree. In BPS and BC no airway connection with the bronchial tree is found whereas in CPAM and CLE an aberrant connection may be identified [52]. An aberrant connection can either be a bronchomalacia or bronchial atresia, both of which can be diagnosed with bronchoscopy, inspiratory and expiratory CT-scan, cine-CT, dynamic MRI and on pathology [53,54]. On CT-imaging, bronchomalacia can be diagnosed in cooperative children by computing the change in cross-sectional area between inspiration and expiration [7,53]. In non-cooperative children, CT is performed in cine-mode or by changing lung volume under anaesthesia [55]. Atresia is an abrupt complete interruption of the bronchus which can be identified on CT-imaging as a mucus plug distal from the atresia and a local hypodense region distally. The mucus plug is formed due to

accumulated mucus produced in the patent distal bronchus (mucocele) [15,16,56]. The hypodense region is a result of hyperinflation of the excluded lung parenchyma by collateral ventilation through the pores of Kohn [57]. Especially in smaller children, atresia may be difficult to diagnose due to the small diameter of distally located airways in relation to the resolution of the CT-scanner [53].

We suggest noting the airway connection as normal, bronchomalacia or bronchial atresia.

Lesion (figure 7, section C)

CLA lesions are variable in appearance and can be characterized as cystic, solid or hybrid lesions. Cystic lesions can be seen in CPAM, BPS and BC and can be distinguished by number, size and content of the cyst(s). Hybrid lesions exhibit features of both CPAM and BPS.

In CPAM, the average cyst size and size of the largest cyst are used to determine the subtype. A cut off at 2 cm for largest cyst size is suggested for distinguishing CPAM types 1 and 4 from type 2, while a soft tissue mass is seen in CPAM types 0 and 3 [7,20,23-25,27,30,31,33,34]. Classification on the basis of cyst size and aspect showed a good correlation with final histological diagnosis [23-25,30,34], although cyst size may be dependent on age and growth of the child and the cut-off values are arbitrarily chosen. Due to the association between CPAM type 4 and pleuropulmonary blastoma (PPB), classification is desirable to determine treatment and follow-up. PPB is a rare, aggressive, paediatric lung tumour which appears as a distally located cystic or solid mass [58,59]. The risk of PPB type 1 is significantly increased in CPAM type 4; still it is often thought to be the same entity [60,61]. Distinction on imaging is difficult, although cystic PPB lesions seem to have a bigger largest cyst, are septated and more often contain solid components [22,23].

In BPS, the appearance of ILS ranges from aerated cystic lesions to homogeneous or heterogeneous solid lesions. This variation may be the result of collateral airflow to the homogeneous solid lesion through the 'pores of Kohn'. Depending on the degree of this airflow, the appearance might become more heterogeneous with hypodense areas or air-filled cysts [13,62-66]. In contrast, ELS rarely contains air as it is

separated from the normal lung parenchyma by its own pleura. Thus, the presence or absence of air may be used to distinguish ELS from ILS [5,7,12,13].

In BC, the common mediastinal localization makes it difficult to characterize the cyst and to distinguish it from malignant neoplasms and lymphadenopathy [7]. However, BC may occur in the lung parenchyma as well and present with a smooth well-defined cyst wall, opposed to a thick irregular wall. Contrast agents usually enhance surrounding tissue rather than the BC. Furthermore, opposed to homogeneous BC, a heterogeneous mass with central contrast-enhancement is more suggestive for a mediastinal neoplasm [7,8].

BC are commonly fluid filled and as such can objectively be distinguished from a soft tissue mass if the region of interest measurement is lower than the cut-off value of 20 Hounsfield units [23] or by comparing content to fluid in the gall bladder or spinal canal [7,8,20,27]. An internal air-fluid level and enhanced thick cyst wall are signs of infection [4].

For cystic lesions, we suggest noting the structure as well as the average cyst size and size of the largest cyst. Content should be noted as air-filled, fluid-filled or air-fluid level.

For solid lesions, we suggest noting the size expressed as volume in thirds of the affected lobe as well as the homogeneity of the lesion.

Vascularization (figure 7, section D)

Abnormal vascularization is an important distinctive feature of BPS and is often the only difference between BPS and CPAM type 3 on CT imaging. Particularly for planning surgery or interventional radiology, the number, size and origin of abnormal vessels is important. Spontaneous regression of ELS is more likely with arterial blood supply by multiple small arteries opposed to a large feeding artery [67]. Venous drainage to the pulmonary vein is common in ILS, while ELS often drain to the azygos system. This may help distinguishing between different types of BPS [35]. Hypervascularity is often seen in BPS and is caused by a high systemic blood pressure in the low-resistance lung circulation [5,13,36].

We suggest reporting both the arterial and venous vascularization as well as the origin and number of aberrant vessels.

Surrounding tissue (figure 7, section E)

Surrounding lung tissue may be affected by the lesion and show abnormalities as well. Mediastinal shift and atelectasis may be seen in large lesions and increase the risk of becoming symptomatic [30]. Growth and increasing solid appearance are other concerning features [44,68]. In PPB , mediastinal shift, hyperdensity and lack of hypodensity [23] is seen more often as well as pleural effusion and right side predominance [69]. Hyperinflation is seen in both CPAM and PPB, although studies contradict each other on its predictive value [22,23].

Perilesional low-attenuation areas indicate poorly perfused or ventilated regions [70]. It is hypothesized these regions may be CLE alongside CPAM, which would support the hypothesis that all CLA share a common pathogenesis [15,16,34,45].

Worsening of symptoms may occur after chest tube insertion when CPAM or CLE is misdiagnosed as a pneumothorax. Although commonly misdiagnosed as such, a pneumothorax is rarely associated with CLE [22,24,37,38,40,60]. Vascular markings and low-attenuation surrounding the cyst suggest a CPAM [28], while cranial or caudal lung compression is seen in CLE opposed to lung collapse towards the hilum.

We suggest noting abnormalities of surrounding parenchyma such as mass effect, mediastinal shift or herniation. Atelectasis, low-attenuation areas or air-trapping and emphysema should be noted as volume in thirds of the affected lobe [48].

Conclusion

Due to the wide variety of CT-imaging features in CLA, assessment and reporting can be difficult. Distinction may be hard and depends on the quality and type of scan. The inconsistent nomenclature

when describing these abnormalities leads to confusing, non-uniform and incomplete reports [34]. Radiological appearances should not be categorized using a pathological classification as the overlap between abnormalities in both imaging features and pathology make distinction on CT-imaging difficult and unreliable [2]. When a structured approach is used, uniform and complete reports with well-defined terms are made, which is beneficial in clinical decision-making [71,72]. In our structured report we have captured key features of structural changes required for diagnosing the different CLA and clinical decision-making. Structured assessment and reporting of objective radiologic features utilizing uniform nomenclature is time-efficient and could improve the diagnostic accuracy. Using this method and avoiding subjective interpretation could lead to better quality of reports in CT- imaging of CLA. We aim to test the proposed structured report in a large multi-centre longitudinal study.

References

1. Stocker LJ, Wellesley DG, Stanton MP, Parasuraman R, Howe DT. The increasing incidence of foetal echogenic congenital lung malformations: an observational study. Prenat Diagn 2015;35(2):148-153.

2. Bush A. Congenital lung disease: a plea for clear thinking and clear nomenclature. Pediatr Pulmonol 2001;32(4):328-337.
3. Baez JC, Ciet P, Mulkern R, Seethamraju RT, Lee EY. Pediatric Chest MR Imaging: Lung and Airways. Magn Reson Imaging Clin N Am 2015;23(2):337-349.
4. Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. Radiology 2008;247(3):632-648.
5. Hang JD, Guo QY, Chen CX, Chen LY. Imaging approach to the diagnosis of pulmonary sequestration. Acta Radiol 1996;37(6):883-888.
6. Karnak I, Şenocak ME, Ciftci AO, Büyükpamukçu N. Congenital lobar emphysema: Diagnostic and therapeutic considerations. J Pediatr Surg 1999;34(9):1347-1351.
7. El-Medany S, El-Noueam K, Sakr A, Galal A. Role of multi slice computed tomography in the evaluation of congenital anomalies of tracheobronchial tree and lungs. Egypt J Radiol Nucl Med 2011;42(3):315-325.
8. McAdams HP, Kirejczyk WM, Rosado-de-Christenson ML, Matsumoto S. Bronchogenic cyst: imaging features with clinical and histopathologic correlation. Radiology 2000;217(2):441-446.
9. Sarper A, Ayten A, Golbasi I, Demircan A, Isin E. Bronchogenic cyst. Tex Heart Inst J 2003;30(2):105-108.
10. Durell J, Lakhoo K. Congenital cystic lesions of the lung. Early Hum Dev 2014;90(12):935-939.
11. Stocker JT. Congenital pulmonary airway malformation - a new name for and an expanded classification of congenital cystic adenomatoid malformation of the lung. Histopathology 2002(41):424-431.
12. Kang M, Kh, elwal N, Ojili V, Rao KLN, Rana SS. Multidetector CT angiography in pulmonary sequestration. J Comput Assisted Tomogr 2006;30(6):926-932.
13. Ikezoe J, Murayama S, Godwin JD, Done SL, Verschakelen JA. Bronchopulmonary sequestration: CT assessment. Radiology 1990;176(2):375-379.
14. Choudhury SR, Chadha R, Mishra A, Kumar V, Singh V, Dubey NK. Lung resections in children for congenital and acquired lesions. Pediatr Surg Int 2007;23(9):851-859.
15. Kunisaki SM, Fauza DO, Nemes LP, Barnewolt CE, Estroff JA, Kozakewich HP, Jennings RW. Bronchial atresia: the hidden pathology within a spectrum of prenatally diagnosed lung masses. J Pediatr Surg 2006;41(1):61-65; discussion 61-65.
16. Riedlinger WF, Vargas SO, Jennings RW, Estroff JA, Barnewolt CE, Lillehei CW, Wilson JM, Colin AA, Reid LM, Kozakewich HP. Bronchial atresia is common to extralobar sequestration, intralobar sequestration, congenital cystic adenomatoid malformation, and lobar emphysema. Pediatr Dev Pathol 2006;9(5):361-373.
17. Morini F, Zani A, Conforti A, van Heurn E, Eaton S, Puri P, Rintala R, Lukac M, Kuebler JF, Friedmacher F, Wijnen R, Tovar JA, Pierro A, Bagolan P. Current Management of Congenital Pulmonary Airway Malformations: A "European Pediatric Surgeons' Association" Survey. Eur J Pediatr Surg 2018;28(1):1-5.
18. Moher D, Liberati A, Tetzlaff J, Altman DG, Group P. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. BMJ 2009;339:b2535.
19. Hansell DM, Bankier AA, MacMahon H, McLoud TC, Muller NL, Remy J. Fleischner Society: glossary of terms for thoracic imaging. Radiology 2008;246(3):697-722.
20. Bondioni MP, Gatta D, Lougaris V, Palai N, Signorelli M, Michelini S, Di Gaetano G, Tessitore P, Mascaro L, Tironi A, Boroni G, Maroldi R, Alberti D. Congenital cystic lung disease: prenatal ultrasound and postnatal multidetector computer tomography evaluation. Correlation with surgical and pathological data. Radiol Med 2014;119(11):842-851.
21. Chow PC, Lee SL, Tang MHY, Chan KL, Lee CP, Lam BCC, Tsoi NS. Management and outcome of antenatally diagnosed congenital cystic adenomatoid malformation of the lung. Hong Kong Med J 2007;13(1):31-39.
22. Feinberg A, Hall NJ, Williams GM, Schultz KAP, Miniati D, Hill DA, Dehner LP, Messinger YH, Langer JC. Can congenital pulmonary airway malformation be distinguished from Type i pleuropulmonary blastoma based on clinical and radiological features? J Pediatr Surg 2016;51(1):33-37.

23. Griffin N, Devaraj A, Goldstraw P, Bush A, Nicholson AG, Padley S. CT and histopathological correlation of congenital cystic pulmonary lesions: a common pathogenesis? Clin Radiol 2008;63(9):995-1005.
24. Kim WS, Lee KS, Kim IO, Suh YL, Im JG, Yeon KM, Chi JG, Han BK, Han MC. Congenital cystic adenomatoid malformation of the lung: CT-pathologic correlation. AJR Am J Roentgenol 1997;168(1):47-53.
25. Lanza C, Bolli V, Galeazzi V, Fabrizzi B, Fabrizzi G. Cystic adenomatoid malformation in children: CT histopathological correlation. Radiol Med 2007;112(4):612-619.
26. Lee HJ, Song MJ, Cho JY, Lee YH. Echogenic fetal lung masses: Comparison of prenatal sonographic and postnatal CT findings. J Clin Ultrasound 2003;31(8):419-424.
27. Mata JM, Caceres J, Lucaya J, Garcia-Conesa JA. CT of congenital malformations of the lung. Radiographics 1990;10(4):651-674.
28. Prabhu S, Choudhury S, Solanki R, Shetty G, Agarwala S. Inadvertent chest tube insertion in congenital cystic adenomatoid malformation and congenital lobar emphysema-highlighting an important problem. Indian J Radiol Imaging 2013;23(1):8-14.
29. Shimohira M, Hara M, Kitase M, Takeuchi M, Shibamoto Y, Kurono K, Shimizu S. Congenital pulmonary airway malformation: CT-pathologic correlation. J Thorac Imaging 2007;22(2):149-153.
30. Tawil MI, Pilling DW. Congenital cystic adenomatoid malformation: Is there a difference between the antenatally and postnatally diagnosed cases? Pediatr Radiol 2005;35(1):79-84.
31. Winters WD, Effmann EL, Nghiem HV, Nyberg DA. Disappearing fetal lung masses: Importance of postnatal imaging studies. Pediatr radiol 1997;27(6):535-539.
32. Lima JS, Camargos PAM, Aguiar RALP, Campos AS, Aguiar MJB. Pre and perinatal aspects of congenital cystic adenomatoid malformation of the lung. J Matern -Fetal Neonatal Med 2014;27(3):228-232.
33. Zhang ZJ, Huang MX. Children with congenital cystic adenomatoid malformation of the lung CT diagnosis. Int J Clin Exp Med 2015;8(3):4415-4419.
34. Kyncl M, Koci M, Ptackova L, Hornofova L, Ondrej F, Snajdauf J, Pychova M. Congenital bronchopulmonary malformation: CT histopathological correlation. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub 2016;160(4):533-537.
35. Ou J, Lei X, Fu Z, Huang Y, Liu E, Luo Z, Peng D. Pulmonary sequestration in children: A clinical analysis of 48 cases. Int J Clin Exp Med 2014;7(5):1355-1365.
36. Ren JZ, Zhang K, Huang GH, Zhang MF, Zhou PL, Han XW, Duan XH, Li Z. Assessment of 64-row computed tomographic angiography for diagnosis and pretreatment planning in pulmonary sequestration. Radiol Med 2014;119(1):27-32.
37. Thakral CL, Maji DC, Sajwani MJ. Congenital lobar emphysema: experience with 21 cases. Pediatr Surg Int;17(2):88-91.
38. Özçelik U, Göçmen A, Kiper N, Doğru D, Dilber E, Yalçin EG. Congenital lobar emphysema: Evaluation and long-term follow-up of thirty cases at a single center. Pediatr Pulmonol 2003;35(5):384-391.
39. Cataneo DC, Rodrigues OR, Hasimoto EN, Schmidt Jr AF, Cataneo AJM. Congenital lobar emphysema: 30-year case series in two university hospitals. J Bras Pneumol 2013;39(4):418-426.
40. Song ZW, Ge W, Xu CY, Fang BD, Yang CY, Zhang HL, Zhao YP. The diagnostic value of multislice computed tomography in children with congenital lobar emphysema. Expert Rev Med Devices 2011;8(4):435-439.
41. Raju S, Ghosh S, Mehta AC. Chest CT Signs in Pulmonary Disease: A Pictorial Review. Chest 2017;151(6):1356-1374.
42. Epelman M, Kreiger PA, Servaes S, Victoria T, Hellinger JC. Current imaging of prenatally diagnosed congenital lung lesions. Semin Ultrasound CT MR 2010;31(2):141-157.
43. Newman B. Congenital bronchopulmonary foregut malformations: concepts and controversies. Pediatr Radiol 2006;36(8):773-791.
44. van Leeuwen K, Teitelbaum DH, Hirschl RB, Austin E, Adelman SH, Polley TZ, Marshall KW, Coran AG, Nugent C. Prenatal diagnosis of congenital cystic adenomatoid malformation and its

postnatal presentation, surgical indications, and natural history. J Pediatr Surg 1999;34(5):794-798; discussion 798-799.
45. Langston C. New concepts in the pathology of congenital lung malformations. Semin Pediatr Surg 2003;12(1):17-37.
46. Seear M, Townsend J, Hoepker A, Jamieson D, McFadden D, Daigneault P, Glomb W. A review of congenital lung malformations with a simplified classification system for clinical and research use. Pediatr Surg Int 2017;33(6):657-664.
47. Lee ML, Tsao LY, Chaou WT, Yang AD, Yeh KT, Wang JK, Wu MH, Lue HC, Chiu IS, Chang CI. Revisit on congenital bronchopulmonary vascular malformations: a haphazard branching theory of malinosculations and its clinical classification and implication. Pediatr Pulmonol 2002;33(1):1-11.
48. Brody AS, Klein JS, Molina PL, Quan J, Bean JA, Wilmott RW. High-resolution computed tomography in young patients with cystic fibrosis: distribution of abnormalities and correlation with pulmonary function tests. J Pediatr 2004;145(1):32-38.
49. Stanton M, Njere I, Ade-Ajayi N, Patel S, Davenport M. Systematic review and meta-analysis of the postnatal management of congenital cystic lung lesions. J Pediatr Surg 2009;44(5):1027-1033.
50. Baird R, Puligandla PS, Laberge JM. Congenital lung malformations: informing best practice. Semin Pediatr Surg 2014;23(5):270-277.
51. Muller CO, Berrebi D, Kheniche A, Bonnard A. Is radical lobectomy required in congenital cystic adenomatoid malformation? J Pediatr Surg 2012;47(4):642-645.
52. Correia-Pinto J, Gonzaga S, Huang Y, Rottier R. Congenital lung lesions--underlying molecular mechanisms. Semin Pediatr Surg 2010;19(3):171-179.
53. Ciet P, Wielopolski P, Manniesing R, Lever S, de Bruijne M, Morana G, Muzzio PC, Lequin MH, Tiddens HA. Spirometer-controlled cine magnetic resonance imaging used to diagnose tracheobronchomalacia in paediatric patients. Eur Respir J 2014;43(1):115-124.
54. Ngerncham M, Lee EY, Zurakowski D, Tracy DA, Jennings R. Tracheobronchomalacia in pediatric patients with esophageal atresia: comparison of diagnostic laryngoscopy/bronchoscopy and dynamic airway multidetector computed tomography. J Pediatr Surg 2015;50(3):402-407.
55. Lee EY, Zucker EJ, Restrepo R, Daltro P, Boiselle PM. Advanced large airway CT imaging in children: evolution from axial to 4-D assessment. Pediatr Radiol 2013;43(3):285-297.
56. Gipson MG, Cummings KW, Hurth KM. Bronchial atresia. Radiographics 2009;29(5):1531-1535.
57. Desir A, Ghaye B. Congenital abnormalities of intrathoracic airways. Radiol Clin North Am 2009;47(2):203-225.
58. Papaioannou G, Sebire NJ, McHugh K. Imaging of the unusual pediatric 'blastomas'. Cancer Imaging 2009;9:1-11.
59. Feinberg A, Hall NJ, Williams GM, Schultz KA, Miniati D, Hill DA, Dehner LP, Messinger YH, Langer JC. Can congenital pulmonary airway malformation be distinguished from Type I pleuropulmonary blastoma based on clinical and radiological features? J Pediatr Surg 2016;51(1):33-37.
60. MacSweeney F, Papagiannopoulos K, Goldstraw P, Sheppard MN, Corrin B, Nicholson AG. An assessment of the expanded classification of congenital cystic adenomatoid malformations and their relationship to malignant transformation. Am J Surg Pathol 2003;27(8):1139-1146.
61. Hill DA, Dehner LP. A cautionary note about congenital cystic adenomatoid malformation (CCAM) type 4. Am J Surg Pathol 2004;28(4):554-555; author reply 555.
62. Mirza B, Raza AB, Ijaz I, Ijaz L, Naz F, Sheikh A. Intralobar pulmonary sequestration masquerading as congenital lobar emphysema. J Indian Assoc Pediatr Surg 2011;16(1):15-17.
63. Kim M, Woo JJ, An JK, Jung YY, Choi YS. A Pulmonary Sequestered Segment with an Aberrant Pulmonary Arterial Supply: A Case of Unique Anomaly. Korean Journal of Radiology 2016;17(2):302-305.
64. Ferretti GR, Jouvan FB, Coulomb M. MDCT Demonstration of Intralobar Pulmonary Sequestration of the Right Upper Lobe in an Adult. American Journal of Roentgenology 2005;185(6):1663-1664.
65. Stern EJ, Webb WR, Warnock ML, Salmon CJ. Bronchopulmonary sequestration: dynamic, ultrafast, high-resolution CT evidence of air trapping. AJR Am J Roentgenol 1991;157(5):947-949.
66. Cooke CR. Bronchopulmonary sequestration. Respir Care 2006;51(6):661-664.

67. Yoon HM, Kim EAR, Chung SH, Kim SO, Jung AY, Cho YA, Yoon CH, Lee JS. Extralobar pulmonary sequestration in neonates: The natural course and predictive factors associated with spontaneous regression. Eur Radiol 2017;27(6):2489-2496.
68. Cook J, Chitty LS, De Coppi PD, Ashworth M, Wallis C. The natural history of prenatally diagnosed congenital cystic lung lesions: Long-term follow-up of 119 cases. Arch Dis Child 2017.
69. Naffaa LN, Donnelly LF. Imaging findings in pleuropulmonary blastoma. Pediatr Radiol 2005;35(4):387-391.
70. Tiddens HA, Rosenow T. What did we learn from two decades of chest computed tomography in cystic fibrosis? Pediatr Radiol 2014;44(12):1490-1495.
71. Larson DB, Towbin AJ, Pryor RM, Donnelly LF. Improving consistency in radiology reporting through the use of department-wide standardized structured reporting. Radiology 2013;267(1):240-250.
72. Ganeshan D, Duong PT, Probyn L, Lenchik L, McArthur TA, Retrouvey M, Ghobadi EH, Desouches SL, Pastel D, Francis IR. Structured Reporting in Radiology. Acad Radiol 2018;25(1):66-73.