ORIGINAL ARTICLE

# Thoracoscopic Lobectomy for Congenital Pulmonary Airway Malformation with Prenatal Diagnosis

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## Abstract

**Introduction:** Congenital pulmonary airway malformations are increasingly diagnosed in the prenatal period raising concerns on the post-natal management of asymptomatic cases. Favoring early resection are the concepts of better compensatory lung growth as well as the prevention of the long-term risk of infection and malignancy. Our aim was to review the results of our conduct over the years that we promoted early thoracoscopic resection.

**Methods:** We have analyzed the series of patients with congenital pulmonary malformations submitted to thoracoscopic resection at our department, focusing on infants with a prenatal diagnosis of congenital pulmonary airway malformations.

**Results:** From March 2012 to March 2020, we performed 24 thoracoscopic pulmonary resections at our department for congenital pulmonary airway malformations, congenital lobar emphysema, pulmonary sequestration, bronchogenic cyst, and pleuropulmonary blastoma. Among these, 19 underwent thoracoscopic pulmonary lobectomy, including 14 children with a prenatal diagnosis of congenital pulmonary airway malformations that were operated on during infancy. In our series of thoracoscopic pulmonary lobectomy, we did not have any perioperative complications. Regarding the post-operative complications, there were two cases treated conservatively, but none in the subgroup of congenital pulmonary airway malformations with prenatal diagnosis. The median length of stay was three days. In the entire series, two patients reported recurrent wheezing, while in the prenatal diagnosed congenital pulmonary airway malformations subgroup, all of the patients had a completely uneventful follow-up.

**Discussion:** Early thoracoscopic resection of prenatal diagnosed congenital pulmonary airway malformations is safe and should be considered in cases with significant lung (morphological) malformation as the esthetic and clinical outcomes are excellent.

**Keywords**: Elective Surgical Procedures; Infant; Lung/abnormalities; Lobectomies; Portugal; Prenatal Diagnosis; Respiratory System Abnormalities; Thoracic Surgery, Video-Assisted/methods; Treatment Outcome

## Introduction

Thoracoscopy started to have a role in pediatric surgery in the early 1980s for the evaluation of thoracic lesions and small procedures like lung biopsies and limited pleural debridements.[1] As adequate instruments were developed, designed, and sized for infants and children, gradually more differentiated procedures had been performed. Thoracoscopic resection for congenital bronchopulmonary malformations is generally accepted, namely in congenital pulmonary airway malformations (CPAM), and in this sequence, thoracoscopic pulmonary lobectomy was first described in 2003 by Rothenberg.[2] The results of this strategy proved to be at least as safe as open surgery with the advantages of reduced postoperative pain, length of stay, and complications rate. Despite the controversy on the management of prenatally diagnosed congenital pulmonary airway malformations, elective surgery is performed in most cases on account of the risk of recurrent infection and malignant transformation.[3-5]then reviewed and included if they specifically addressed the proposed question. Results: 1040 articles were identified on initial search.

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After screening abstracts per eligibility criteria, 130 articles were used to answer the proposed questions. Based on the available literature, resection of an asymptomatic CPAM is controversial, and when performed is usually completed within the first six months of life. Lobectomy remains the standard resection method for CPAM, and can be performed thoracoscopically or via thoracotomy. There is no consensus regarding a monitoring protocol for observing asymptomatic lesions, although at least one chest computerized tomogram (CT It remains unclarified as to what is the ideal age for elective surgery in asymptomatic infants. During infancy, the safety and efficacy of resections have been questioned due to the smaller size of the structures, tissue fragility, and technical demands of unilateral lung ventilation.
Our aim was to review the results of our conduct over the years, in which we promote early thoracoscopic resection during infancy.

## Methods

### Population and data collection

We have analyzed the series of patients with congenital pulmonary malformations, referred to our department from various hospitals all over the country, submitted to thoracoscopic resection between March 2012 and March 2020, focusing on the infants with prenatal diagnosis (PND) who underwent thoracoscopic pulmonary lobectomy.
Demographic data and clinical details, including age, prenatal diagnosis, clinical presentation, surgical procedure, duration of the procedure, conversion to open repair, per and postoperative complications, postoperative time to extubation, thoracic drainage, and length of hospital stay, were gathered from the electronic files of the patients.

### Anesthetic considerations

All patients were operated on under combined anesthesia (general and thoracic epidural). Unilateral lung ventilation was achieved either by using a bronchial blocker positioned on the ipsilateral mainstem bronchus or, in smaller children, by a single lumen endotracheal tube advanced into the contralateral mainstem bronchus under fibroscopic guidance. When using the single lumen tube, namely when selecting the right lung, ultrasonographic auscultation was used to assure that the endotracheal tube was placed correctly, on the mainstem bronchus, not excluding the upper right bronchus. The identification of the sliding lung sign all along the hemithorax, together with its nonappearance on the contralateral hemithorax, was indicative of a successful selective lung ventilation.

### Operative technique

All thoracoscopic lobectomies were performed by a single experienced consultant assisted by a relatively fixed team. The patient was positioned in lateral decubitus with the affected side up. The surgeon stood at the patient front with the assistant at his side, and the monitor at the back of the patient. For thoracoscopic lobectomy, we introduced two 5 mm trocars (for a 30° optics, and Ultracision® or Ligasure® as available), a 3 mm trocar (additional working port), and sometimes a stab incision to aid in the mobilization of the pulmonary lobe. In Fig. 1, the trocar placement for a left upper lobectomy (LUL) is shown. Low pressure insufflation with carbon dioxide ($CO_2$) was set depending on the age of the patient and clinical tolerability. We used blunt and electrocautery hook dissection to isolate the arterial (Fig. 2A) and venous (Fig. 2B) segmental vessels and lobar bronchus (Fig. 2C). Vessels were controlled by intracorporeal ligation and sealing devices. Larger vessels were proximally ligated with clips. Bronchi were first ligated with nonabsorbable sutures and/or clips and then divided. For complete fissure division, we used the sealer for aerostasis. Specimens were exteriorized in piecemeal through a 5 mm port incision. A chest tube was placed under visualization.

### Data analysis

Data analysis was conducted using SPSS® software version 24.0 (SPSS, Chicago, IL, USA).

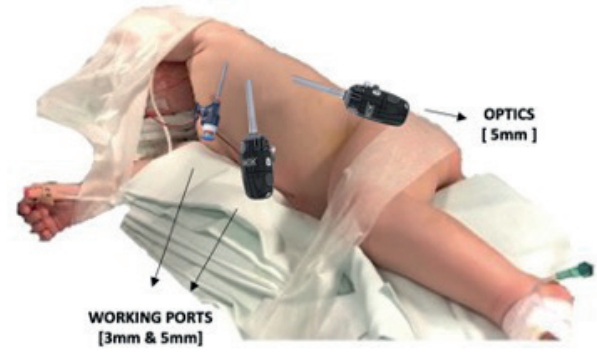



**Figure 1.** Patient positioning and trocar placement for thoracoscopic left upper lobectomy. The patient was positioned in lateral decubitus with the affected side up.

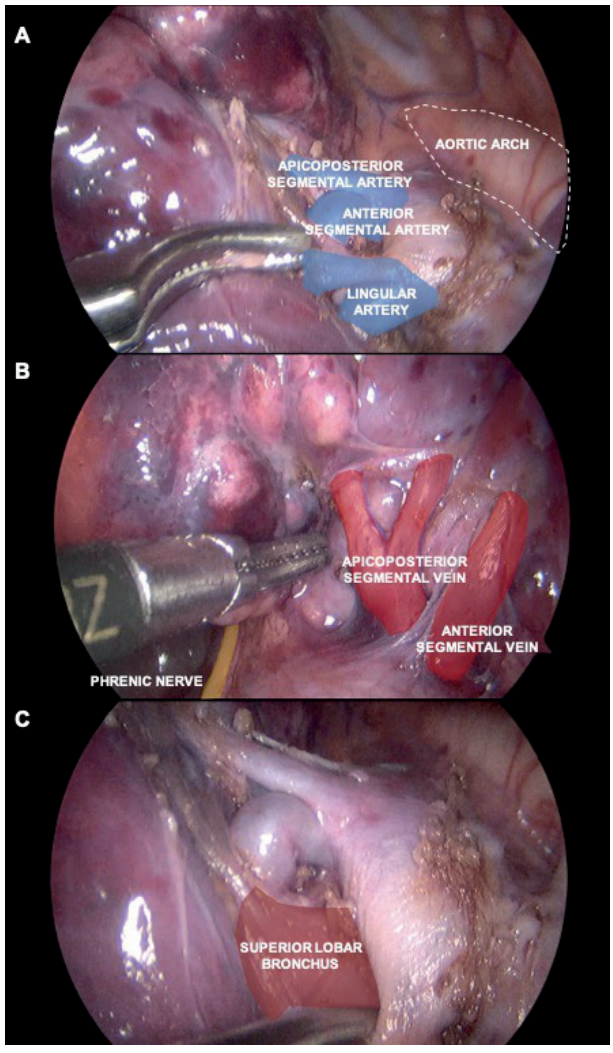



**Figure 2.** Surgical view after the dissection of the segmental pulmonary arteries (A), segmental pulmonary veins (B) and superior lobar bronchus (C).

## Results

We performed 24 thoracoscopic pulmonary resections at our department for CPAM (n = 16), congenital lobar emphysema (n = 2), pulmonary sequestration (n = 3), bronchogenic cyst (n = 2), and pleuropulmonary blastoma (n = 1). Nineteen undergone thoracoscopic lobectomy, being those with the diagnosis of CPAM, congenital lobar emphysema, and pleuropulmonary blastoma. Children with pulmonary sequestration were submitted to sequestrectomy, while bronchogenic cysts were excised. Seventeen (71%) infants had prenatal diagnosis of CPAM (n = 14) or bronchopulmonary sequestration (BPS) (n = 3). Eight (33%) children had symptomatic presentation (four presented with recurrent pneumonia and four had respiratory distress). Two (8%) had associated anomalies (*pectus excavatum* and interventricular communication) (Table 1).

**Table 1. Characteristics of children who underwent the resection of congenital bronchopulmonary malformation (n = 24)**

| | |
|---|---|
| **DEMOGRAPHIC CHARACTERISTICS** | |
| Age (years), median (IQR) | 1.3 (2.8) |
| **Pre-operative diagnosis, no. (%)** | |
| CPAM | 17 (70.8%) |
| Congenital lobar emphysema | 2 (8.3%) |
| Bronchopulmonary sequestration | 3 (12.5%) |
| Bronchogenic cyst | 2 (8.3%) |
| Prenatal diagnosis, no. (%) | 17 (70.8%) |
| Symptomatic, no. (%) | 8 (33.3%)* |
| Associated malformations, no. (%) | 2 (8.3%)† |
| **SURGICAL PROCEDURE** | |
| Lobectomy | |
| RUL, no. (%) | 1(4%) |
| RLL, no. (%) | 7(29%) |
| LUL, no. (%) | 7(29%) |
| LLL, no. (%) | 4(17%) |
| Sequestrectomy | |
| Intralobar (LLL), no. (%) | 2(8%) |
| Thoracoabdominal, no. (%) | 1(4%) |
| Bronchogenic cyst excision, no. (%) | 2(8%) |
| **HISTOLOGIC DIAGNOSIS** | |
| Type I CPAM, no. (%) | 8(33%) |
| Type II CPAM, no. (%) | 3(13%) |
| Type III CPAM, no. (%) | 2(8%) |
| CPAM (undetermined), no. (%) | 3(13%) |
| Pleuropulmonary blastoma, no. (%) | 1(4%) |
| Congenital lobar emphysema, no. (%) | 2(8%) |
| Bronchopulmonary sequestration, no. (%) | 3(13%) |
| Bronchogenic cyst, no. (%) | 2(8%) |

BC - bronchogenic cyst; BPS - bronchopulmonary sequestration; CLE - congenital lobar emphysema; CPAM - congenital pulmonary airway malformation; IQR – interquartile range; LLL - left lower lobe; LUL - left upper lobe; no - number; PPB - pleuropulmonary blastoma; RLL - right lower lobe; RUL - right upper lobe.
* Four patients had recurrent pneumonia and four had respiratory distress.
† *Pectus excavatum* and interventricular communication.

Single lung ventilation was attempted in all patients and effective in the great majority during the whole procedure. There were no conversions to open surgery and no perioperative complications.

Regarding thoracoscopic lobectomies (n = 19), the median duration of the procedure was 182 minutes; nonetheless, the duration of surgery tended to reduce along cumulative experience. All of the patients kept thoracic drainage after surgery for a mean of two (± 1) days. The median length of stay was three days with an interquartile range (IQR) of 3.0. Nearly all of the patients (95%) were extubated immediately after the procedure. Only one patient with the diagnosis of CPAM, with recurrent pneumonias before surgery, kept

endotracheal intubation for two days. Postoperative complications (n = 2, 10%) included two pneumothoraces (congenital lung emphysema, 15 days after surgery with spontaneous resolution; CPAM with previous recurrent pneumonia) and a pneumonia (CPAM with previous recurrent pneumonias).

Among these patients, there were 14 infants/children who underwent elective pulmonary lobectomy without having previous symptoms, after receiving the prenatal diagnosis of CPAM. As shown in Table 2, in this group, one can notice that the operative time was slightly reduced, all the patients were extubated at the end of surgery, there were no postoperative complications, and the length of stay was also diminished.

In a mean follow-up time of 51 months, there were two symptomatic patients (recurrent wheezing with SOS medication) who have had previous symptoms before surgery. For those with a prenatal diagnosis, the follow-up has been uneventful.

## Discussion

Thoracoscopic lobectomy for congenital bronchopulmonary malformations, although technically demanding, is now an accepted and well-described technique, namely for children and older infants. There are consensual advantages including less postoperative pain, shorter length of hospital stay, decreased long-term morbidity, and excellent cosmesis (Fig. 3).[6] The thoracoscopic approach that we perform, using two 5 mm and one 3 mm ports, differs from the single port approach that is mostly used in adult surgery. The single port procedure implies a small mini-thoracotomy scar that will eventually grow over the years. In addition, it does not allow for the accurate visualization of the whole thoracic cavity, hampering the control of a perioperative complication in case it occurs.[7] Therefore, pediatric surgeons tend to prefer and recommend the thoracoscopic approach using three working ports. Improvements in prenatal ultrasound lead to an apparent increase in the congenital bronchopulmonary malformation incidence. It is now estimated to be 1 in 2,500, while it was around one in 30,000 in previous reports.[5] Understanding the natural history of these lesions is essential to define an appropriate management. A small percentage of antenatally diagnosed bronchopulmonary malformations undergo intrauterine death (3%-4%), while up to 14% experience spontaneous pre- or post-natal regression.[8] Some lesions detected in the initial ultrasound screening will disappear in the following examinations, which do not imply that the lesion has resolved once its echogenicity might mimic normal lung tissue in advanced pregnancy. At our

**Table 2. Perioperative, post-operative, and follow-up assessment of the patients who underwent pulmonary lobectomy**

| | Global (n = 19) | Asymptomatic patients with CPAM and prenatal diagnosis (n = 14) |
|---|---|---|
| **PERIOPERATIVE ASSESSMENT/COMPLICATIONS** | | |
| Age (years), median (IQR) | 1.2 (1.9) | 1.3 (1.3) |
| Pulmonary exclusion, no. (%) | 17 (89%) | 13 (93%) |
| Operative time (minutes), median (IQR) | 182 (132) | 164 (99) |
| Conversion, no. (%) | - | - |
| Perioperative complications, no. (%) | - | - |
| Immediate extubating, no. (%) | 18 (95%) | 14 (100%) |
| **POST-OPERATIVE ASSESSMENT/COMPLICATIONS** | | |
| Thoracic drainage, no. (%) | 19 (100%) | 14 (100%) |
| Postoperative complications, no. (%) | 2 (10%)* | - |
| Length of stay (days), median (IQR) | 3.5 (3) | 3 (2) |
| **FOLLOW-UP ASSESSMENT** | | |
| Follow-up time, mean (SD), months | 50.8 (22.9) | 45.4 (23.5) |
| Symptomatic, no. (%) | 2 (10%)† | - |

CPAM - congenital pulmonary airway malformation; IQR – interquartile range; no = number; PND - prenatal diagnosis; SD - standard deviation.
* Pneumothorax and pneumonia.
† Recurrent wheezing.

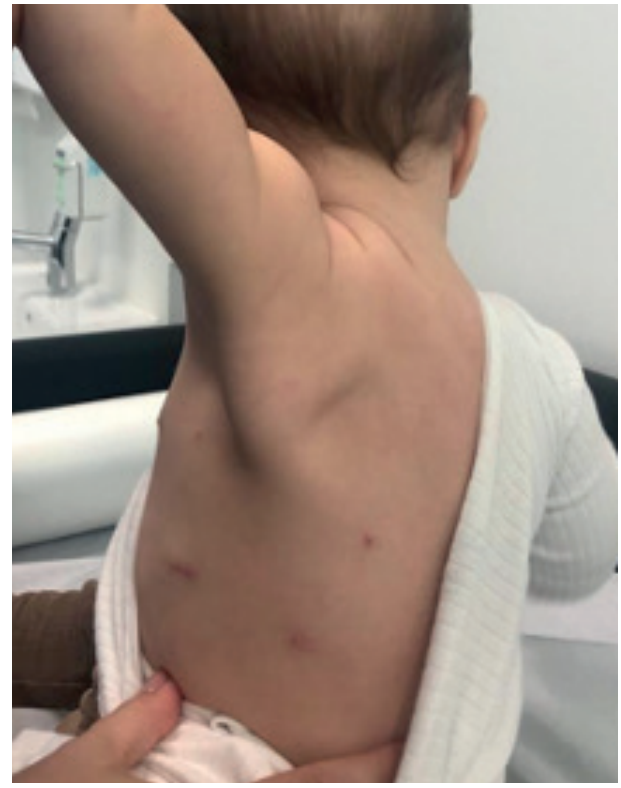


**Figure 3.** Postoperative esthetic result one month after a thoracoscopic left lower lobectomy.

department, whenever a CPAM was suspected during prenatal ultrasound screening (usually around 18-24 weeks), a management algorithm was applied (Fig. 4). Fetal magnetic resonance imaging (MRI) was considered if the ultrasound findings were equivocal or difficult to interpret, such as in late pregnancy or inaccessible fetal position. High-risk CPAM was defined by the presence of hydrops or a sonographic measurement of the CPAM volume ratio (CVR or index estimated dividing CPAM volume by the head circumference) greater than 1.6. In these cases, and before 32 weeks gestation, fetal intervention might be considered to alleviate the mass effect, prevent the progression of complications, and improve the outcome:

- In macrocystic lesions, decompression should be attempted by thoracoamniotic shunting or single needle thoracocentesis;
- In microcystic lesions, maternal betamethasone administration has been suggested to induce the regression of some lesions, namely on large microcystic CPAM, as we could document in two of our prenatally diagnosed cases.

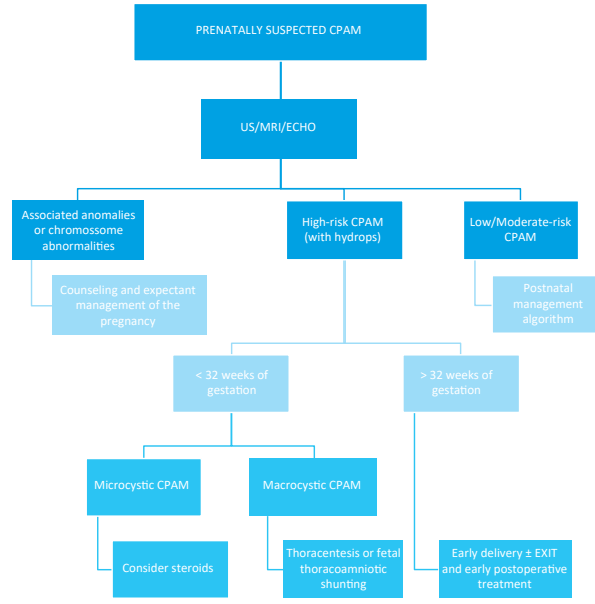
After 32 weeks of gestation, the *ex utero intrapartum* treatment (EXIT) might be an option namely in the presence of severe mediastinal shift and/or hydrops associated with persistently elevated CVR.[3,9,10] To date, no EXIT was performed at our department. After birth, all newborns with a history of prenatally detected CPAM, including those asymptomatic and whose lesion was not detected in the most recent prenatal ultrasound examinations should undergo an image exam (Fig. 5).[11] A small percentage of patients will present respiratory distress and require precocious/emergent surgery (10% at our series).[12] At this time, the development of symptoms is usually due to the fluid-filled interior of the lesion being replaced by air and compressing normal lung as well as other thorax structures, causing obstruction to gas exchange and leading to respiratory distress.[11] Around 25% of the previously asymptomatic will develop symptoms during the following years. At this stage, the symptoms usually relate to the fact that CPAM is greedy for infection, making patients susceptible to recurrent pneumonias and their accompanying symptoms as respiratory distress.[10] Therefore, the increasing number of cases with prenatal diagnosis (65% in our series) raise concerns on the post-natal management of asymptomatic cases. In favor of conservative treatment are[8]:

- The fact that the lesion is usually asymptomatic;
- Beliefs regarding spontaneous regression throughout the years;
- The inherent risks of the surgical procedure.

The arguments favoring early resection are[8]:

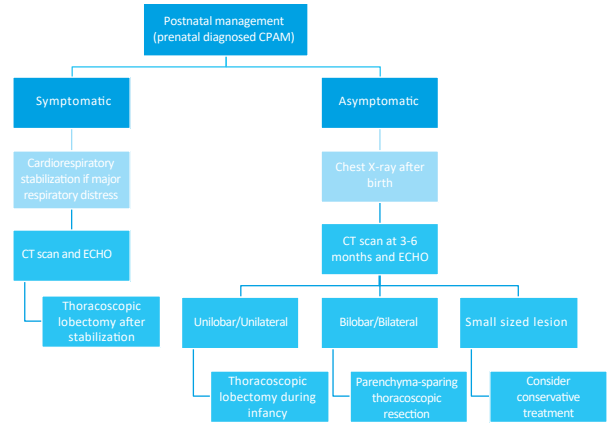
- The long-term risk of infection that hamper the surgical technique;
- The concept of a better compensatory lung growth than resection at a later age;
- Risk of malignancy.

Evidence suggests that elective surgery is performed in the majority of these cases (70%).[5] At our department, asymptomatic infants are proposed for surgical resection when the expected risk of infection is considerable as assessed by our team (pediatric pneumology and surgery). Hence, 60% of the bronchopulmonary malformations were operated with no previous clinical symptoms. We noticed that the surgical technique was more straightforward in these patients, as suggested by other proponents of elective surgery.[4,13,14]




CPAM - congenital pulmonary airway malformation; ECHO - fetal echocardiogram; EXIT - *ex utero intrapartum* treatment; MRI - fetal magnetic resonance imaging; US - ultrasound scanning.

**Figure 4.** Prenatal management algorithm.




CPAM - congenital pulmonary airway malformation; CT - computed tomography; ECHO - echocardiogram.

**Figure 5.** Postnatal management algorithm.

Actually, we hypothesized that it could have influenced the surgical outcomes, as all the postoperative complications in our series occurred in patients who were symptomatic before surgery, namely with repeated infections. Consistently, several studies, including a meta-analyses, found that there was a threefold higher risk of adverse effects following a surgery after infection compared with an elective surgery in asymptomatic neonates and infants.[4,12] After an infection, the tissue turns friable and fibrotic, which hampers the dissection and identification of structures, and it increases the risk of bleeding and accidental lesion of nearby structures. The lack of perioperative complications in our series was in line with those described in the literature for thoracoscopic surgery (1%-5%),[6,12] or open resection (up to 10%).[15,16] Moreover, when it comes to postoperative complications, our results (10%) fit other thoracoscopic series (3%-10%)[6,17] and are, therefore, better than other open resection series (10%-25%).[16-18] Thoracoscopic lobectomies were performed in a median time of 182 minutes (164 minutes in the group with prenatal diagnosis), which is not different from other studies of thoracoscopic lobectomies (115 to 279 minutes).[6,15,17] It was also comparable to other series of open surgery (169 to 205 minutes).[15,17] It should be highlighted that the operation time decreased consistently along the cumulative experience of our team, reaching the mean of 120 minutes in the last three cases. Median length of stay was three days, which is in line with the literature (3-5 days).[15,19] Among those with pre-operative diagnosis of CPAM, histopathologic analysis revealed one pleuropulmonary blastoma (PPB). Pleuropulmonary blastoma (types I-III) is a rare lung tumor in childhood that presents under 6 years of age. Commonly, it is considered if there is a prenatal diagnosis of CPAM, as pleuropulmonary blastoma and congenital pulmonary airway malformations cannot be easily distinguished by clinical and radiologic exams prior to resection. It appears that pleuropulmonary blastoma is a distinct histological entity and not a benign CPAM that undergoes malignant transformation.[20] Some clinical and image features suggest the diagnosis of pleuropulmonary blastoma such as symptomatic presentation, bilateral involvement, complex morphology, or mediastinal shift.[20] Nevertheless, there is at least one report of a case that was asymptomatic at the time of surgery. This remains an argument favoring early resection rather than the conservative observation of prenatal diagnosed CPAM.[21]

Nowadays, more and more surgeons are performing early bronchopulmonary malformations excision. We found that, under 6 months of age, tissue fragility turned out to be an advantage and dissection was easier than in older infants, which other authors have previously stated. Some series reported higher complication and conversion rates in patients less than 5 months of age, but that might be explained by a lack of systematized technique or appropriate instruments. In addition, the surgeon experience clearly plays a role in the outcome.[22] Some authors reported shorter operative times and length of stay as well as a lower complication rate in patients under 10 kg.[22] Concerns regarding the prolonged time on capnothorax, hypercapnia, and cerebral hypoxemia, especially in smaller infants, were not an issue in the reported series. In our study, among those patients under 6 months of age (n = 6), we did not find intraoperative complications nor conversion to open surgery. No capnothorax related intercurrences were identified. Although controversy remains on the ideal time of surgery, there is little evidence to suggest that delayed resection improves outcome.[4,6] The risk of recurrent infection and associated technical difficulties, risk of malignancy, and possibility of compensatory lung growth, all support an early intervention.[4] In Fig. 5, there is the postnatal management algorithm that we follow at our department. When asymptomatic after birth, a chest radiography was performed, and at 4-6 months of age, a computed tomography scan to confirm the presence of the lesion and characterize its location and potential extra-pulmonary blood supply. Emergent or urgent surgery was indicated in case of respiratory symptoms or alarming findings on chest radiography, such as mediastinal shift or multilobar lesion. Asymptomatic infants were assessed by our team (pediatric pneumology and surgery) to make an individual case-by-case decision, in which parents were involved, considering the risk of infection in case of conservative management. Asymptomatic small sized CPAM as well as extra lobar small sized pulmonary sequestrations could be treated conservatively (parents are involved in this decision). Those proposed for thoracoscopic lobectomy were operated during infancy (tendentially around 6 months of age). In the event of a bilobar or bilateral lesion, parenchyma sparing thoracoscopic resection should be considered.[9]

In conclusion, early thoracoscopic resection of prenatal diagnosed CPAM is safe and should be considered in cases with significant lung (anatomic) malformation as the esthetic and clinical outcomes are excellent.

**WHAT THIS STUDY ADDS**

- A clinical reflective practice exercise from the pioneering group in Portugal performing thoracoscopic pulmonary lobectomy in children.
- Highlights the importance of a systematic multidisciplinary approach of patients with a prenatal diagnosis of bronchopulmonary malformations.
- Early thoracoscopic resection of prenatal diagnosed congenital pulmonary airway malformations is safe in patients with significant lung (anatomic) malformation with excellent esthetic and clinical outcomes.

## Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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There were no external funding sources for the realization of this paper.

## Protection of human and animal subjects

The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

## Provenance and peer review

Not commissioned; externally peer reviewed

## Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

## Acknowledgements

The senior author of this manuscript would like to share a posthumous tribute to Prof. Tiago Henriques-Coelho for his huge contribution to science in the uncovering of prenatal mechanisms underlying the pathophysiology of congenital pulmonary airway malformation development as well as all of his enthusiastic and unconditional collaboration for the early clinical steps to develop and promote the program to implement the technique of thoracoscopic pulmonary lobectomy in children in Portugal.

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**Lobectomia Toracoscópica por Malformação Congénita das Vias Aéreas Pulmonares de Diagnóstico Prénatal**

**Resumo:**

**Introdução:** As malformações congénitas da via aérea pulmonar são frequentemente alvo de diagnóstico pré-natal originando controvérsia quanto à abordagem clínica. Favorecem a resseção pós-natal precoce o conceito de crescimento pulmonar compensatório, o risco de infeção pulmonar recorrente e malignidade. O objetivo foi avaliar os resultados do protoloco seguido no nosso hospital, em que promovemos a resseção toracoscópica precoce do lobo pulmonar afetado.

**Métodos:** Analisamos a série de doentes com malformações pulmonares congénitas, focando-nos naqueles com diagnóstico pré-natal submetidos a lobectomia toracoscópica.

**Resultados:** Desde março de 2012 a março de 2020, realizámos 24 resseções pulmonares toracoscópicas por malformações pulmonares congénitas da via aérea, enfisema lobar congénito, sequestro pulmonar, cisto broncogénico e blastoma pleuropulmonar. Destas, 19 crianças foram submetidas a lobectomia pulmonar toracoscópica incluindo 14 com diagnóstico pré-natal que foram operadas enquanto lactentes. Na nossa série de lobectomias toracoscópicas não se verificaram complicações intra-operatórias. Relativamente a complicações pós-operatórias, houve dois casos tratados conservadoramente, mas nenhum no subgrupo das malformações pulmonares congénitas da via aérea com diagnóstico pré-natal. A mediana do tempo de internamento foi de três dias. Na série global, dois pacientes reportam sibilância recorrente durante o seguimento pós-operatório, enquanto que este período decorreu sem intercorrências naqueles com diagnóstico pré-natal de malformações pulmonares congénitas da via aérea.

**Discussão:** A resseção toracoscópica de malformações pulmonares congénitas da via aérea em lactentes é segura e deve ser ponderada em casos de malformação pulmonar congénita considerável uma vez que os resultados funcional e estético são excelentes.

**Palavras-Chave:** Anomalias do Sistema Respiratório; Cirurgia Torácica Vídeoassistida/métodos; Diagnóstico Pré-Natal; Lactente; Pneumonectomia/métodos; Portugal; Procedimentos Cirúrgicos Eletivos; Pulmão/anomalias; Resultado do Tratamento