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Prenatal Growth Characteristics and Pre/Postnatal Management of Bronchopulmonary Sequestrations

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Abstract

Purpose—The prenatal natural history of intralobar and extralobar bronchopulmonary sequestrations (BPSs), including lesion growth patterns and need for prenatal intervention, have not been fully characterized. We review our series of BPSs to determine their natural history and outcomes in the context of the need for prenatal intervention.

Methods—A retrospective review of the pre/postnatal course of 103 fetuses with an intralobar (n=44) or extralobar BPS (n=59) managed at a single institution between 2008 and 2015 was performed. Outcomes included prenatal lesion growth trajectory, presence of hydrops, need for prenatal intervention, survival, and postnatal surgical management.

Results—Most extralobar (71%) and intralobar BPSs (94%) decreased in size or became isoechoic from initial to final evaluation. Peak lesion size occurred at 26–28 weeks gestation. Eight fetuses developed hydrothorax, four of which (all extralobar BPSs) also developed hydrops. All four hydropic fetuses received maternal betamethasone, and three hydropic fetuses underwent thoracentesis and/or thoracoamniotic shunt placement with subsequent hydrops resolution. All fetuses survived. Forty-one intralobar (93%) and 35 extralobar BPSs (59%) were resected after birth.

Conclusions—BPSs tend to decrease in size after 26–28 weeks gestation and rarely require fetal intervention. Lesions resulting in hydrothorax ± hydrops can be effectively managed with maternal steroids and/or drainage of the hydrothorax.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Keywords

bronchopulmonary sequestration; hydrothorax; hydrops; thoracentesis; thoracoamniotic shunt; laser coagulation

1.0 Introduction

Bronchopulmonary sequestrations (BPSs) are the second most common prenatally-diagnosed congenital lung lesions [1]. They are composed of non-functional lung tissue disconnected from the tracheobronchial tree with arterial blood supply from an anomalous systemic feeding vessel [2–4]. They can be classified as intralobar or extralobar based on pulmonary versus systemic venous drainage and their location within or outside of the pleura of an anatomical lung lobe [5]. While historically it has been difficult to distinguish intralobar from extralobar BPSs prior to postnatal resection, advances in prenatal color Doppler ultrasound now allow pulmonary versus systemic venous drainage to be differentiated and thus intralobar versus extralobar BPSs to be diagnosed before birth [6].

The prenatal natural history of congenital cystic adenomatoid malformations (CCAMs) including their prenatal growth profiles and response to prenatal interventions has been well documented [7–10], but that of BPSs is less well-defined. In a limited number of cases, BPSs have been shown to result in hydrops fetalis, a condition characterized by pleural and pericardial effusions, skin and scalp edema, and ascites that is usually lethal in the absence of fetal intervention [8, 11]. Fetuses with hydrops secondary to BPSs have been managed with maternal steroids, fetal thoracentesis, thoracoamniotic (TA) shunts, and – in rare cases – open fetal surgery [4, 8, 12–13]. Ablation of the systemic arterial supply by ultrasound-guided laser or sclerotherapy has also been proposed to manage BPSs associated with large size, hydrothorax, or hydrops [12, 14–17]. In the current study, we review the prenatal growth characteristics, prenatal therapeutic interventions, and postnatal course of our large series of BPSs to evaluate the natural history of these lesions and the need for prenatal interventions.

2.0 Methods

This is a retrospective review of all cases of fetuses referred to the Center for Fetal Diagnosis and Treatment between August 2008 and December 2015 with a congenital lesion confirmed on postnatal surgical/pathological analysis and/or postnatal imaging to be a BPS. This study was approved by The Children's Hospital of Philadelphia Institutional Review Board (IRB#16-012681). All patients underwent a detailed evaluation and nondirective multidisciplinary counseling at the time of fetal evaluation. Initial prenatal imaging consisted of a detailed fetal ultrasound in all patients and an ultrafast fetal MRI in 84% of patients. Serial sonographic imaging was performed at intervals dictated by the fetal status. On postnatal imaging (computed tomography angiography (CTA), ultrasound, and/or MRI), extralobar lesions were defined by the presence of systemic arterial supply and systemic venous drainage, versus intralobar lesions which were defined by the presence of systemic arterial supply and pulmonary venous drainage. These lesions were also classified at surgery as extralobar or intralobar based on the presence or absence of a separate investing pleura.

Operative and pathology reports were reviewed to identify the pleural investment and to assess the vascularity. Cases with coexisting conditions or insufficient follow-up were excluded.

Prenatal records and imaging were reviewed to determine the gestational age (GA) at presentation, lesion side and location with respect to the diaphragm, the presence of mediastinal shift, cardiac compression, hydrothorax, hydrops, and polyhydramnios as well as the need for prenatal interventions including maternal betamethasone (BMZ) (12 mg intramuscularly, 2 doses, 24 hours apart), fetal thoracentesis/TA shunt, and lesion resection via open fetal surgery or ex utero intrapartum treatment (EXIT) procedure. Hydrops was defined as 2 or more of the following: ascites, skin/scalp edema, pleural effusion or pericardial effusion. Resolution of hydrops was defined as a reduction in hydrops criteria from 2 to 1. Resolution of hydrothorax following thoracentesis/TA shunt was defined as at least one subsequent ultrasound detecting no more than trace residual pleural effusion without recurrence prior to delivery.

Serial prenatal ultrasounds were reviewed for lesion size defined by the CCAM volume ratio (CVR) (length x width x height x 0.52/head circumference) [7]. Two strategies were employed to model the typical prenatal growth of BPSs. First, individual CVR measurements were aggregated into GA intervals of two weeks, and a mean size was calculated for each interval. Second, quadratic regression was performed, and the apex of the curve was used to estimate the GA corresponding with peak CVR. Data points from ultrasounds reporting isoechoic lesions were excluded.

Peri- and postnatal records were reviewed to note the GA at birth, location and mode of delivery, respiratory symptoms requiring positive pressure support, postnatal management via surgical resection, and postoperative length of stay (LOS). Postnatal CTAs were reviewed for lesion volume defined as the volume of a prolate ellipse (cranial-caudal x anterior-posterior x transverse x 0.52), and surgical pathology of each lesion was reviewed for evidence of systemic arterial blood supply and possible CCAM features.

Binary outcomes were compared using Pearson's Chi-square or Fisher's exact test. Normally-distributed continuous outcomes were first analyzed for equal variance between groups, and Student's t test was then performed assuming equal or unequal variance when appropriate. Continuous outcomes that were not normally distributed were compared using the Mann-Whitney test. Statistical analysis and graphical modeling were performed using Stata/1C, version 14.2 (StataCorp, College Station, TX). All statistical tests were two-sided with an alpha level set at 0.05 for statistical significance.

3.0 Results

3.1 Study Population

One hundred and twenty-one fetuses with BPSs were evaluated at our center during the study period. Eighteen patients were excluded from the study due to coexisting congenital diaphragmatic hernia (n=8); concurrent, separate CCAM (n=2); multiple gestation pregnancy (n=5); both intralobar and extralobar BPSs (n=2); or insufficient follow-up (n=1).

Among the remaining 103 cases, 59 fetuses had an isolated extralobar BPS, and 44 fetuses had an isolated intralobar BPS. Lesion characteristics by prenatal imaging are reported in Table 1. Intralobar BPSs were most commonly located in the left lower lobe (77%).

3.2 Prenatal Growth

The natural history of BPS growth was determined by assessing CVR measurements in fetuses not undergoing any prenatal intervention. Serial measurements were available for 80 of 98 patients that were not treated with maternal BMZ and/or thoracentesis/TA shunting (49 extralobar and 31 intralobar). In 64 cases (80%), CVR decreased or the lesion became isoechoic from initial to final evaluation. The GA associated with the highest mean CVR for all BPSs was 26 ± 1 weeks with subsequent decrease in mean CVR thereafter (Figure 1A). Quadratic regression of CVR by GA generated the growth curve $\text{CVR} = -0.0043(\text{GA})^2 + 0.25(\text{GA}) - 2.91$ with the apex at 28.4 weeks gestation.

Extralobar BPSs were less likely than intralobar BPSs to decrease in CVR or become isoechoic from initial to final evaluation (71% vs. 94% of lesions, respectively, decreased in CVR/became isoechoic, $P = 0.021$). The GA associated with highest mean CVR was 26 ± 1 weeks for both extralobar and intralobar lesions (Figure 1B and 1C). There was no difference in the apices of the growth curves between extralobar and intralobar BPSs (28.9 weeks vs. 27.6 weeks, respectively).

3.3 BPSs Associated with Hydrothorax ± Hydrops

The incidence of mass effect, including hydrothorax with and without hydrops, is reported in Table 1. Among the 7 extralobar BPSs associated with hydrothorax, 6 were intrathoracic and 1 was transdiaphragmatic. All 4 extralobar BPSs associated with hydrops were intrathoracic, and their prenatal management and outcomes are described in Table 2. Among the 3 fetuses with hydrothorax in the absence of hydrops, 2 resolved without prenatal intervention. The third fetus was treated with maternal BMZ, and the hydrothorax remained stable until scheduled delivery at 37 weeks gestation. One fetus with an intralobar BPS developed a small hydrothorax that resolved prior to delivery without intervention.

The peak CVRs of extralobar BPSs associated with hydrops were significantly higher than those not associated with hydrops (mean: 0.98 vs. 0.42, $P = 0.015$). The peak CVR associated with hydrothorax ± hydrops was similarly elevated (mean: 0.87 vs. 0.40, $P = 0.008$). By simple linear regression, $\text{CVR} > 0.75$ increased the odds of hydrothorax ± hydrops by a ratio of 28.7 (95% confidence interval (CI): 3.1 – 268, $P = 0.003$). The negative predictive value of $\text{CVR} < 0.75$ for hydrothorax ± hydrops was 98%. While BPSs associated with hydrothorax ± hydrops tended to have higher CVRs early in gestation, this difference resolved later in gestation following fetal intervention in multiple cases (Figure 2). Notably, extralobar BPSs associated with hydrothorax ± hydrops did not have an increased prevalence of CCAM features on pathology (33% vs. 35%, $P = 1.0$).

3.4 Peri- and Postnatal Course

Peri- and postnatal outcomes are reported in Table 3. Among the 8 extralobar BPSs associated with respiratory symptoms at birth, 7 were intrathoracic and 1 was

transdiaphragmatic. By univariate analysis, the need for positive pressure support was associated with a higher peak CVR (mean: 0.80 vs. 0.40, $P = 0.021$), hydrothorax $\pm$ hydrops during gestation (57% vs. 7.7%, $P = 0.004$), and lower GA at birth (35.0 weeks vs. 38.8 weeks, $P = 0.026$). By multivariate regression controlling for GA, $\text{CVR} > 0.75$ increased the odds of respiratory symptoms at birth by a ratio of 20.6 (95% CI: 2.0 – 212, $P = 0.011$). The negative predictive value of the $\text{CVR} < 0.75$ for respiratory symptoms at birth was 96%.

Seven of the 8 newborns with respiratory distress at birth required surgical resection. The median age at surgery was 32 days (range: 0 – 100 days) with a median postoperative LOS of 23 days (range: 1 – 81 days). One patient with hydrops did not require resection (Case 1, Table 2), and one patient without hydrothorax or hydrops required resection immediately after birth for a large lesion (peak CVR: 1.4) associated with significant mediastinal shift and cardiac compression.

Twenty-seven of 35 (77%) intrathoracic, 3 of 18 (17%) intraabdominal, and 5 of 6 (83%) transdiaphragmatic extralobar BPSs were resected. The volume of the lesion, as calculated on postnatal CTA, was available for 70% of the extralobar BPS cases ($n=41$). Infants undergoing surgical resection had significantly larger lesions (12cm^3 (standard deviation (SD): 15cm^3) vs. 3.3cm^3 (SD: 3.1cm^3), $P = 0.007$). There was no difference in the mean age at which imaging was performed between those that did and did not undergo surgery (45 days vs. 59 days, $P = 0.48$). A thoracoscopic or laparoscopic approach was utilized to resect 13 of 27 (48%) intrathoracic, 3 of 3 (100%) intraabdominal, and 3 of 5 (60%) transdiaphragmatic lesions.

Three premature infants (mean GA: 33.8 weeks; range: 32.0 – 35.5 weeks) with an intralobar BPS had respiratory symptoms at birth. Peak CVR was not significantly higher among these patients versus patients without respiratory symptoms (0.53 vs. 0.49, $P = 0.88$), and they did not require early resection. Two patients with relatively large intralobar BPSs (peak CVRs: 1.6 and 1.2), however, underwent surgery within the first 3 days of life. Although these patients did not have any respiratory symptoms or signs of vascular steal, surgery was performed soon after birth secondary to the large lesion size, large systemic feeding vessel, and increased risk for the development of high output cardiac failure. While intralobar BPSs were less likely than extralobar BPSs to be resected thoracoscopically (Table 3), this difference resolved when intraabdominal and transdiaphragmatic extralobar BPSs were excluded from analysis (29% vs. 48%, $P = 0.11$).

4.0 Discussion

BPSs are congenital lung lesions with the potential to cause fetal hydrothorax and hydrops. The purpose of this study was two-fold: first, to characterize the typical prenatal growth pattern of intra and extralobar BPSs and the incidence of fetal hydrothorax/hydrops associated with these lesions, and, second, to evaluate the outcomes of our pre- and postnatal management of these patients. The majority of both intra and extralobar BPSs decreased in CVR or became isoechoic sonographically as gestation progressed with the mean CVR peaking at 26 ± 1 weeks. These findings are similar to the reported growth trajectory of CCAMs, for which mean CVR peaks at 25 weeks gestation [7]. In our series, BPSs were

rarely associated with hydrothorax and hydrops. Extralobar BPSs were more likely to cause hydrothorax than intralobar BPSs, and all cases of hydrops were associated with an extralobar BPS. The rate of hydrops among fetuses with extralobar BPSs in our series (6.8%) was similar to a smaller series reported previously by Adzick et al. where 4 of 39 (10%) fetuses developed hydrops [8]. Interestingly, the peak CVR in patients with extralobar BPSs associated with hydrops was less than the CVR previously associated with hydrops in patients with CCAMs (0.98 vs. 3.1) [7]. This likely reflects a difference in the pathophysiology: in contrast to CCAMs in which hydrops is secondary to the mass effect of the lesion itself, hydrops in the setting of an extralobar BPS is due to lymph-like fluid secreted into the pleural cavity with consequent tension hydrothorax. The tension hydrothorax causes the “mass effect” with consequent contralateral mediastinal shift, compression of the heart and vena cavae, and resultant right heart failure [18]. The tension hydrothorax may also compress the ipsilateral lung including the lesion, which may contribute to the smaller lesion size associated with hydrops in extralobar BPSs. Together, these data support serial monitoring by ultrasound as a reasonable strategy for the management of prenatally-diagnosed BPSs with a heightened awareness of the potential for hydrothorax/hydrops to develop in relatively small lesions.

In the current study, all 4 hydropic fetuses survived. Three had hydrops resolution following thoracentesis/TA shunting. In the fourth case, a TA shunt was planned, but before it could be performed the mother suffered a placental abruption secondary to abdominal trauma. Overall, the favorable outcomes of our management of hydropic fetuses with thoracentesis/TA shunts are similar to those previously reported [8]. While effective in resolving hydrops, these interventions are not without risks. Both patients treated with TA shunts had preterm premature rupture of membranes and were born at a mean GA of 34.3 weeks. This is consistent with the mean GA at delivery of fetuses receiving TA shunts (36 weeks) that we previously reported [9]. Additionally, 1 TA shunt migrated out of the chest wall with subsequent hydrothorax recurrence. This outcome is not uncommon, as the reported rate of shunt failure ranges from 8.3% to 45% [9, 19–21].

Respiratory symptoms after birth were infrequent in our series, occurring most often in association with larger lesions or hydrothorax $\pm$ hydrops. While we resect almost all intralobar BPSs secondary to an increased risk of infection, we do not routinely resect extralobar BPSs if the infant is asymptomatic and the lesion is small without evidence of cysts on postnatal CTA. In these cases, routine follow-up without further imaging is sufficient. In the current study, 41% of extralobar BPSs, including one extralobar BPS associated with hydrops, were not resected. When surgery was indicated, postoperative LOS was generally short, and survival was 100%. These favorable postnatal outcomes are important to consider when weighing the potential risks and benefits of prenatal interventions for BPSs, including ultrasound-guided ablation of the arterial blood supply [12, 14–17].

Study limitations include the fact that the frequency of serial ultrasound measurements was variable and dependent on lesion size. As a result, the growth curves generated through quadratic regression may be shifted slightly to the right, explaining the difference in the GA associated with peak CVR obtained with the two modeling strategies. Additionally, the small

number of fetuses with hydrothorax and hydrops limits our ability to make strong conclusions about the relative efficacy of our prenatal interventions compared to other series for this rare patient population [12, 15].

Despite these limitations, this study has yielded three important outcomes. First, our modeling of BPS prenatal growth provides evidence to support decreasing lesion size as measured by CVR after 26–28 weeks gestation, a trend previously suggested by clinical experience but never before shown empirically. Second, this study has demonstrated that, for extralobar BPSs, a CVR cutoff of 0.75 is a useful predictor of pre- and postnatal complications. Finally, this study has demonstrated that the vast majority of BPSs do not require prenatal intervention while providing a robust characterization of the pre- and postnatal course of intralobar and extralobar BPSs which will be valuable for counseling families and against which future studies of prenatal interventions for BPSs can be compared.

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Figure 1.
Prenatal Growth Trajectory. (A) All BPSs, (B) extralobar BPSs, and (C) intralobar BPSs.

Figure 2.
Prenatal Growth Trajectory of BPSs Associated with Hydrothorax $\pm$ Hydrops

Table 1

Prenatal Characteristics and Interventions for Extralobar and Intralobar BPSs

Prenatal Characteristics and Interventions	Extralobar BPSs n=59	Intralobar BPSs n=44	P Value
Lesion Characterization by Prenatal Imaging			
GA at Presentation (weeks)	24.6 (19.9 – 36.9)	22.7 (18.9 – 31.1)	0.004
CVR at Presentation	0.35 (0.03 – 1.9)	0.46 (0.03 – 1.6)	0.12
Left-sided	46 (78)	35 (80)	0.85
Intrathoracic	35 (59)	44 (100)	<0.001
Intraabdominal	18 (31)	0 (0)	
Transdiaphragmatic	6 (10)	0 (0)	
Lesion Mass Effect			
Mediastinal Shift	31 (53)	16 (36)	0.10
Cardiac Compression	8 (14)	4 (9.1)	0.55
Hydrothorax without Hydrops	3 (5.1)	1 (2.3)	0.63
Hydrothorax with Hydrops	4 (6.8)	0 (0)	0.13
Polyhydramnios	5 (8.5)	1 (2.3)	0.24
Prenatal Interventions for Hydrothorax ± Hydrops			
Maternal BMZ	5 (8.5)	0 (0)	0.070
Thoracentesis/TA Shunt	3 (5.1)	0 (0)	0.26
Open Fetal Surgery	0 (0)	0 (0)	1.0
EXIT Procedure	0 (0)	0 (0)	1.0

Data are represented as mean (range) or n (%).

GA = gestational age, CVR = CCAM volume ratio, BMZ = betamethasone, TA = thoracoamniotic, EXIT = ex utero intrapartum treatment

Table 2

Prenatal Interventions and Outcomes of Extralobar BPSs associated with Hydrops

Intervention/Outcome	Case 1	Case 2	Case 3	Case 4
GA at Presentation (weeks)	23.0	27.0	34.4	29.9
CVR at Presentation	0.58	0.80	0.57	0.79
Hydrops Criteria				
Ascites	yes	no	yes	yes
Skin/Scalp Edema	yes	yes	yes	yes
Pleural Effusion	yes	yes	yes	yes
Pericardial Effusion	no	no	no	no
Maternal BMZ	yes	yes	yes	yes [‡]
Thoracentesis	yes	no	yes	no
TA Shunt	yes	yes	no	no [†]
Hydrops Outcome	resolved	resolved	resolved	did not resolve
Hydrothorax Outcome	resolved	recurred [§]	recurred	did not resolve
Survival	yes	yes	yes	yes
GA at Birth (weeks)	35.0	33.6	36.6	30.7
Required Positive Pressure Support	no	yes	yes	yes
Postnatal Chest Tube	no	yes	yes	yes
Postnatal Surgery – Age (days)	no – n/a [¥]	yes – 32	yes – 47	yes – 100
Postoperative LOS (days)	n/a	21	44	81

[‡]Patient received maternal steroids prior to presentation to our center.

[†]Patient was scheduled for TA shunt placement; upon arrival for the procedure, the mother was found to have a placental abruption secondary to recent abdominal trauma, and an emergency cesarean delivery was performed.

[§]Hydrothorax recurred after TA shunt migration.

[¥]Infant was followed closely after birth and had no respiratory symptoms. Repeat imaging at 7 months of age demonstrated complete resolution of the lesion.

GA = gestational age, CVR = CCAM volume ratio, BMZ = betamethasone, TA = thoracoamniotic, LOS = length of stay

Table 3

Peri- and Postnatal Outcomes of Extralobar and Intralobar BPSs

Peri- and Postnatal Outcomes	Extralobar BPSs n=59	Intralobar BPSs n=44	P Value
Survival to Birth and Discharge	59 (100)	44 (100)	1.0
GA at Birth	38.3 (28.0 – 41.0)	38.2 (30.6 – 41.0)	0.89
Delivered in Our Institution	12 (20)	5 (11)	0.23
Cesarean Delivery	22 (58) [§]	7 (35) [‡]	0.097
For Fetal Indication	2 (9.1)	1 (13)	1.0
Neonatal Resuscitation			
Required Positive Pressure Support	8 (14)	3 (6.8)	0.35
CPAP	3 (38)	1 (33)	1.0
Intubation	5 (62)	2 (67)	1.0
Postnatal Surgery	35 (59)	41 (93)	<0.001
Open	16 (46)	28 (68)	0.036
Laparoscopic/Thoracoscopic	19 (54)	12 (29)	
Converted to Open	0 (0)	1 (2.4)	
Age at Surgery (days) [¥]	49 (0 – 331)	61 (2 – 1877)	0.091
Postoperative LOS (days) [¥]	2 (1 – 81)	2 (1 – 16)	0.43
Pathology Reported	35 (59)	41 (93)	
CCAM Features	12 (34)	7 (17)	0.084

Data are represented as mean (range) or n (%) unless otherwise specified.

[§]Delivery method data was available for only 38 of 59 extralobar BPS patients.[‡]Delivery method data was available for only 20 of 44 intralobar BPS patients.[¥]Data are represented as median (range).

GA = gestational age, CPAP = continuous positive airway pressure, LOS = length of stay