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PRENATAL GROWTH CHARACTERISTICS AND PRE/POSTNATAL MANAGEMENT OF PRENATALLY-DIAGNOSED EXTRALOBAR BRONCHOPULMONARY SEQUESTRATIONS

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Aim of Study: Laser coagulation of the systemic vascular supply has been proposed to manage extralobar bronchopulmonary sequestrations (BPS) associated with hydrops. We review the prenatal longitudinal growth characteristics, prenatal therapeutic interventions, and postnatal management of our series of extralobar BPSs to evaluate whether recent outcomes warrant exploration of this novel intervention.

Methods: An IRB-approved retrospective review of 58 fetuses diagnosed between August 2008 and June 2015 with an isolated extralobar BPS based on postnatal CT scan alone (n=22) or in combination with surgical pathology (n=36). Serial ultrasounds were reviewed for lesion size defined by CVR (lesion volume x 0.52/head circumference). Quadratic regression was performed to model typical extralobar BPS growth trajectory.

Results: The CVR of 60% of lesions decreased from initial to final evaluation. CVR tends to increase prior to 29 weeks gestation and decrease thereafter. Seven fetuses developed hydrothorax; four of these also had hydrops (Table 1). All 4 hydropic fetuses received maternal betamethasone. Three fetuses, all hydropic, underwent thoracocentesis and/or thoracoamniotic shunt placement. Hydrops resolved in 3 of 4 cases, and thoracocentesis/thoracoamniotic shunting resolved the hydrothorax in 2 of 3 cases. Both fetuses that failed to respond to treatment presented late in gestation (mean: 32.1 weeks) and were born shortly thereafter. No fetus required open fetal surgery or an EXIT to resect the lesion. All fetuses survived to birth and hospital discharge. Eight infants had respiratory symptoms attributed to prematurity and/or the BPS. Thirty-six patients underwent postnatal BPS resection (mean age: 66 days; range: 0-331). The majority of BPSs were resected thoracoscopically/laparoscopically (55.6%) with a mean post-operative stay of 7.9 days (range: 1-81).

Conclusion: Extralobar BPSs tend to decrease in size after 29 weeks gestation and rarely require fetal intervention. Lesions resulting in hydrothorax and/or hydrops can be effectively managed without the need for laser coagulation.

Table 1: Pre- and Postnatal Outcomes of Extralobar BPS

PRE- AND POSTNATAL OUTCOMES	MEAN (RANGE) OR FREQUENCY (%)
Lesion Characterization by Prenatal Imaging	
Gestational Age at Initial Evaluation (weeks)	24.64 (19.86 - 36.86)
Left-sided	44 (83.0)
Intrathoracic	39 (67.2)
Intraabdominal	14 (24.1)
Transdiaphragmatic	5 (8.6)
Lesion Mass Effect	
Mediastinal Shift	28 (48.3)
Cardiac Compression	8 (13.8)
Hydrothorax	7 (12.1)
Early Hydrops (1 hydrops criterion)	3 (5.2)
Hydrops (2 or more hydrops criteria)	4 (6.9)
Polyhydramnios	4 (6.9)
Fetal Interventions	
Steroids	5 (8.6)
Thoracocentesis	2 (3.4)
Thoracoamniotic (TA) Shunt	2 (3.4)
Open Fetal Surgery	0 (0)
Survival to Birth	**58 (100)**
Gestational Age at Birth	38.29 (28 - 41)
Delivered at Our Institution	12 (20.7)
C-Section	22 (37.9)
For Fetal Indication	*2 (9.1)*
EXIT Procedure	0 (0)
Neonatal Resuscitation	
Positive Pressure Support Required	8 (14.0)
CPAP	*3 (37.5)*
Intubation	*5 (62.5)*
Transferred to NICU after Birth (all indications)	19 (32.8)
Duration of NICU stay (days)	*22.6 (0 - 91)*
Postnatal Surgery	**36 (62.1)**
Open	16 (44.4)
Laparoscopic/Thoracoscopic	20 (55.6)
Age at Surgery (days)	66.1 (0 - 331)
Post-operative Duration of Stay (days)	7.9 (1 - 81)
Pathology Reported	**36 (62.1)**
CCAM Features	12 (33.3)
Microcystic or Maldevelopment	11 (30.5)