Pulmonary artery and Lung findings in Absent Pulmonary Valve Syndrome from the Prenatal to Postnatal Period

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Introduction

- Absent pulmonary valve syndrome (APVS) is defined by absent or rudimentary pulmonary valve leaflets and aneurysmal dilation of the main pulmonary artery (MPA) and/ or branch pulmonary arteries (RPA, LPA).
- While some neonates present with severe respiratory distress, many have no symptoms and optimal timing of surgery is unclear.

Objective

- Hypothesis: PA dilation worsens with time in unoperated patients, and that this information along with prenatal MRI can inform prenatal counseling and optimal timing for surgical repair of APVS.

Methods

- 8 prenatally diagnosed APVS patients were evaluated at our institution with echocardiography +/- fetal MRI (n= 6) between 2010 and 2020.

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Methods

- All patients had surgical intervention, ranging from 20 to 488 days of life (median= 62).
- Using the prenatal and postnatal echocardiograms prior to surgery, maximum diameters of the MPA, RPA and LPA were measured and normalized to z scores.
- Fetal MRI findings of segmental lung T2 hyperintensity consistent with fluid trapping were noted.

Results

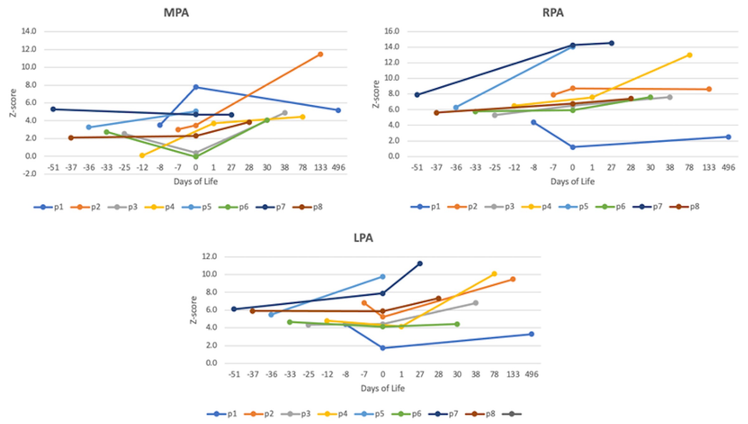

Figure 1. Main pulmonary artery, right and left pulmonary artery maximal diameters measured via echocardiography over time and normalized to z-score values

Results

	Rate of change from fetal period to surgery- Average change in Z-score/day (SD)	**Postnatal rate of change-** Average change in Z-score/day (SD)	**Rate of change with neonatal surgical repair-** Average change in z-score/day (SD)	**Rate of change with infant surgical repair-** Average change in z-score/day (SD)
MPA	0.030 (0.024)	0.054 (0.057)	0.023 (0.024)	0.037 (0.025)
RPA	0.059 (0.070)	0.027 (0.027)	0.089 (0.088)	0.028 (0.034)
LPA	0.040 (0.041)	0.051 (0.042)	0.051 (0.054)	0.029 (0.026)

Figure 2. Pulmonary arteries Z-score measurement rate of change over time; SD= standard deviation

Results

- All patients exhibited a postnatal increase in size and z score of the MPA, LPA, and/or RPA prior to surgery
- The rate of change in z score per day from the fetal period to surgery was 0.03, 0.06 and 0.04 for MPA, RPA, LPA
- Patients with neonatal repair (n=4) and infant repair (n=4) had similar rates of change.

Results

- Neither prenatal nor postnatal MPA, LPA, and RPA size correlated with timing of intervention
- All 6 patients with MRI had normal fetal lung volumes; 3 had a regional T2 hyperintensity subsequently noted to be abnormal on postnatal radiographs.
- 2 of these patients developed bronchomalacia requiring positive pressure ventilation and one required bronchopexy after initial surgery.

Conclusions

- In APVS patients whose surgical repair was delayed beyond the neonatal period, MPA, RPA, and LPA dilation worsens with time
- Given this finding and the potential impact of PA dilation on airway compression and development, it may be prudent to consider fetal lung MRI and early elective surgical repair for minimally symptomatic APVS

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