Treatment of branch pulmonary stenosis secondary to pathologic vascular calcification with serial balloon angioplasty

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Introduction

- Generalized arterial calcification of infancy (GACI) is characterized by ectopic mineralization
- Biallelic mutations in ENNP1 or ABCC6 result in low levels of inorganic pyrophosphate (PPi), an inhibitor of mineralization
- Wide spectrum of clinical manifestations due to arterial, soft tissue, and periarticular calcifications

Case Presentation

- Patient with GACI like disorder born at 34 weeks via c-section for maternal pre-eclampsia
- Low serum PPi levels with negative trio whole exome sequencing (negative for ENNP1 and ABCC6 mutation)
- CT scan on DOL 7 with no calcifications or stenosis in the aorta, great vessels, or aortic visceral branches. Calcifications noted in branch pulmonary arteries
- Echocardiogram prior to discharge on DOL 19 with near systemic RV pressure estimate and mild flow acceleration across branch pulmonary arteries



Figure 1: CT scan completed on DOL 7. On the left, axial view of branch pulmonary arteries with IV contrast demonstrating mild narrowing of the distal right pulmonary artery. On the right, axial view without IV contrast demonstrating subtle calcifications in the branch pulmonary arteries, right greater than left.

Case Presentation



Figure 2: CT scan at 3 months of age. On the left, axial view with IV contrast demonstrating high-grade bilateral branch pulmonary artery stenosis. On the right, axial view without IV contrast demonstrating improvement in calcifications in the branch pulmonary arteries.





Figure 3: Cardiac catheterization at 3 months of age. On the left, RAO/caudal projection demonstrating stenosis of the lobar branches of RPA with additional segmental branch stenosis and long segment narrowing of the RLPA. On the right, repeat angiography after balloon angioplasty of RUPA, RMPA, and RLPA with improvement in vessel caliber.



Figure 4: On the left, LAO projection demonstrating stenosis of the lobar branches of the LPA. On the right, after balloon angioplasty of LLPA with improvement in vessel caliber.







Figure 5: Cardiac catheterization at 7 months of age. On the left, RAO/caudal projection following balloon angioplasty of proximal anterior segmental RLPA with improvement in vessel caliber. On the right, LAO projection following balloon angioplasty of proximal superior segmental LLPA and proximal LUPA with improvement in vessel caliber.

- may require intervention

Patients with pulmonary artery tree stenosis related to ectopic calcification may be candidates for catheter-based interventions Unclear if treatment success would differ in patients with

- typical GACI
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Case Presentation

Discussion

Progressive calcification associated with RV hypertension

Previous report of a 13 month old with GACI undergoing surgical intervention with bilateral pulmonary arterioplasty Patients with multi-vessel lobar and segmental arterial stenosis may not be as amenable to surgical intervention Catheter based intervention may be a reasonable alternative

Conclusion

References

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