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Longitudinal Acute Vasoreactivity Testing in Pediatric Pulmonary Hypertension

Patrick Quinn B.A; Patrick Evers M.D; Paul Critser M.D; Ben Frank M.D; Mohammed Alnoor

Student Researcher; Pediatric Cardiologist; Medical Fellow

Keywords

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Abstract

Introduction

Pediatric Pulmonary hypertension is a rare chronic disease of the lungs and often results in numerous challenges including breathing abnormalities. Upon diagnosis, standard procedure for Pediatric PH involves Acute Vasoreactivity Testing (AVT), a strong predictor of prognosis and treatment options. The goal of this longitudinal study was to identify AVT results as a potential prognostic factor in the treatment of pediatric PH patients.

Methods

A retrospective chart review was performed for pediatric patients diagnosed with PH between 2008-2021. Inclusion criteria consisted of >2 catheterizations with corresponding AVT testing. Study cohorts included patients who were AVT negative upon initial catheterization but AVT positive (AVT-/+) in additional catheterizations and those that were AVT negative for all catheterization procedures (AVT-/-). AVT positive results were defined by the standard Sitbon criteria. Analyzed outcomes included event-free survival as a composite of death, atrial septostomy, lung transplantation, and unplanned PHrelated hospitalization. A relationship between study cohort and event-free survival was analyzed to control for cofounders.

Results

Of the 36 patients who met inclusion criteria, 10 tested as AVT negative on their first catheterization and AVT positive on additional catheterizations (AVT -/+) while 18 tested AVT negative for all catheterizations (AVT-/-). From a statistical standpoint, AVT -/+ patients exhibited significantly better event-free survival than AVT -/- patients (p-value = 0.004). In addition, controlling for initial pulmonary artery pressure and calcium-channel

blocker use led to the finding that subsequent AVT positive results amongst patients originally AVT negative indicated a positive prognostic factor: hazard ratio 0.11 (95% CI: 0.01 – 0.91).

Conclusion

AVT testing is a positive prognostic method of predicting future treatment for pediatric pulmonary hypertension patients. The study indicates that no matter initial AVT catheterization results, subsequent AVT positive tests can predict improved expectation for event-free survival. Continual AVT testing for pediatric PH patients is strongly recommended.