

The Aortic Perfusion Score: A Novel Scoring System to Predict Death or Transplant in Children with Pulmonary Atresia with Intact Ventricular Septum

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Purpose

Pulmonary atresia with intact ventricular septum (PAIVS) is a unique congenital malformation of the heart in which patients may undergo single-, two-, or 1 ½- ventricle repair. Size of the tricuspid valve annulus, morphology of the right ventricle, and presence of ventriculo-coronary connections (VCCs) can all impact the selection of appropriate palliative strategy. We developed the aortic perfusion score (APS), a novel scoring system based on antegrade coronary perfusion with the aim of being able to identify patients at risk for death or transplant.

Methods

A retrospective study of patients at our institution was conducted. Patients were included if an initial catheterization was done prior to any intervention. Each patient was assigned an APS. In this system, each of the named coronaries (left anterior descending, left circumflex, right main, and posterior descending) had a maximum of 100 points they could be assigned. If the entire length of the coronary was being supplied via the aortic root then the coronary would receive 100 points, if only a percentage of it were receiving supply from the aortic root then it would receive that many points. If there was no flow in the coronary due to VCCs then the coronary artery's score was halved. These were then summated for a maximum score of 400. ROC analysis was done to determine a cutoff point predictive of a composite endpoint of death or transplant. A univariate logistic regression was conducted with the endpoint as the dependent variable and the APS as the independent variable.

Results

A total of 64 patients were included in the analysis with 10 reaching the endpoint of death or transplant. At most recent follow-up 31% of patients had undergone single ventricle, 13% 1 ½ ventricle repair, 30 two-ventricle, 17% shunt, and 9% transplant palliation. An APS of 227.5 predicted the endpoint with a sensitivity of 90% and a specificity of 83%. For each 1-point increase in the APS, the odds of death or transplant decreased by 1.7%.

Conclusion

The APS is a scoring method which can be used to predict a composite endpoint of death or transplant in patients with PAIVS and may be helpful in selecting patients that should be listed for transplant. Additional multicenter studies may help further refine the scoring system.