National Pediatric Cardiology
Quality Improvement Collaborative

Research Explained

Hypoplastic Left Heart Stage I: No Norwood, No Hybrid

Dietmer Schranz, Anoosh Esmaeili, Roland Schrewer, Gunter Kerst, Hakan Akintuerk
Circulation 2020 Oct 6;142(14):1402-1404. doi: 10.1161/CIRCULATIONAHA.120.047668.

Clifford Cua MD (physician) and Steven Matthies (parent)

ABOUT THIS STUDY

Why is this study important?

- Currently, options for the first stage procedure for patients with HLHS include Norwood procedure with either a modified Blalock-Taussig shunt or right ventricle to pulmonary artery conduit, or a hybrid procedure with a PDA stent placement via catheter and surgically placed pulmonary artery bands.
- This brief communication reports on a totally non-surgical procedure via catheterization where the PDA is stented and blood flow to the lungs is decreased with vascular plugs/flow restrictors placed in both the right and left pulmonary arteries.
- This novel approach may allow patients to totally avoid open-heart surgery during the newborn period until their second intervention.

What is the goal of the study?

- The main goal of this study was to describe the outcome in patients with single ventricle anatomy who underwent a totally catheter based first stage procedure.

How was this study performed?

- Data from all patients with single ventricle anatomy who underwent the catheter based first stage procedure were reviewed.
- Baseline information, procedural data, hospital data, and follow up data were recorded.

What were the results of the studies?

- Total of 6 patients underwent this initial catheter based procedure.
    - 3 HLHS
    - 1 tricuspid atresia and transposed great arteries

    - 1 congenitally corrected transposition and hypoplastic systemic right ventricle
    - 1 interrupted aortic arch and hypoplastic aortic arch

- All patients are currently alive.
    - 4 patients have successfully undergone their second stage procedure at ~ 4 months of age.
    - The patient with an interrupted aortic arch underwent a two ventricle repair.
    - 1 patient with HLHS underwent cardiac transplantation at 83 days of age.

- Other procedures performed between the first stage procedure and the planned second stage procedure included:
    - Replacement of the vascular plugs/restrictors to a smaller plug/restrictor in 1 patient
    - because too much blood flow was going to the lungs.
    - Stenting of the atrial septum because the hole was getting too small in 2 patients.
    - Second stent placed in the PDA and aortic arch because of narrowing in 1 patient.
    - Placement of a left pulmonary artery stent because of a tear when the vascular plug/flow
    - Restrictor was removed during the 2nd procedure in 1 patient.

What are the limitations of the study?

- An extremely small group of patients were described, so data is limited in how generalizable this technique is to other patients with single ventricle physiology.
- No data that compared this procedure to more traditional procedures for patients with single ventricle physiology to compare results.
- Long-term data is not available.

What it all means

- This technique of PDA stent and vascular plugs/flow restrictors placement in the pulmonary arteries via catheterization is a novel way to totally avoid surgery for the first stage in patients with single ventricle physiology and obstructed systemic blood flow.
- Concerns about pulmonary blood flow, vascular plug/flow restrictor movement, and damage to the pulmonary artery need to be assessed.
    - Total Transcatheter Stage 1: A Word of Caution. Nageotte S, Shahanavaz S, Eghtesady P, Balzer D. Pediatr Cardiol. 2021 Aug;42(6):1410-1415.
- Larger studies with longer follow up are needed to determine the adverse effects, incidence of illness and death from this technique as well as how well it compares to the more traditional pathways for single ventricle palliation.