

**Patient/Family Lay Summary:** FON Case Review Conference November 2021

**Coordinated by:** Children's Hospital of Philadelphia

**Topic:** The Growing Problem of Aortopathy in the post-Norwood Reconstructed Aorta

### **The Problem**

There is currently a very small, but growing, number of patients with single ventricle and a Norwood-type aortic reconstruction who are experiencing increased dilation of the aorta. A significantly increased size aorta that is unusually dilated, can compress important adjacent structures. With dilation also comes wall weakness which may increase the risk for vessel tearing in its length (dissection), or rupture, which can be life-threatening.

### **Clinical Course of the Cases Presented**

Case 1 is a young adult who exhibited progressive enlargement of his reconstructed aorta with compression of the left pulmonary artery. A catheterization was undertaken to stent the left pulmonary artery, and this caused severe compression of the left sided airway. The stent was removed and likely due to the extensive manipulation, there was infection (endocarditis). With progressive enlargement of the aorta there was increasing leakage of the neo-aortic valve. The aorta reached nearly 7 cm in size (normal aorta is less than 3 cm and most post-Norwood reconstructed aortas are 5 cm or less). Surgery was performed with replacement of the aorta and placement of a mechanical artificial valve. The patient did very well and has resumed all life activities with improvement in oxygen levels. Case 2 is a 15-year-old with a dilated reconstructed aorta (6 cm) who suffered a severe tear (dissection) of the aorta, likely related to an event of breaking his forearm while skateboarding, and a sudden, significant elevation of blood pressure. He was asymptomatic of the dissection which was identified during one of his routine MRI screens for a dilated aorta. The patient underwent surgery with an endovascular aortic stent graft and is awaiting further surgery for aortic root replacement.

### **Important Points, Lessons Learned, and Potential Solutions**

- The reconstructed aorta is made up of many components, including: natural aortic tissue, natural main pulmonary artery, and a patch of pulmonary homograft tissue. At the time of Norwood operation, the surgeon carefully sews all of these components together creating a "new" reconstructed aorta.
- Increasing aortic enlargement causes increased wall stress which can progressively weaken the walls of the reconstructed aorta. Weakened walls of a dilated aorta increase the risk of a tear or rupture of the vessel walls.
- The reconstructed aorta after Norwood surgery in infancy starts out larger than any normal aorta, but in some select patients the aorta continues to progressively enlarge over time. Significant enlargement appears to occur in some in the adolescent years. Aortic enlargement can compress the left pulmonary artery causing unequal blood flow

and diverting blood away from the left lung which can lead to cyanosis (blueness) and increased work of breathing.

- Serial MRI evaluation with focused attention on aortic size is of value in those with Norwood procedure as patients enter the teenage years and should be part of general surveillance.
- Who is at greatest risk is unknown. Factors leading to risk of significant aortic dilation after aortic reconstruction need to be explored (i.e. native aortic anatomy? rate of growth in childhood? genetic basis to aortic dilation).
- In our institution, consideration is given to surgical intervention once the aorta demonstrates: 1) progressive enlargement and reaches more than 6 cm in diameter, 2) compresses the left pulmonary artery, and 3) when there is moderate or greater amount of aortic valve leakage.
- Risk factors for development of this aortic disease (aortopathy) and optimal criteria for intervention are in need of multicenter exploration and further research.