

Patient/Family Lay Summary: FON Case Review Conference March 2022

Coordinated by: Mayo Clinic

Topic: Should this patient have a Fontan? Implication of AV valve interventions.

The Problem

Leakage (regurgitation) of the atrioventricular (AV) valve, located between the upper and lower chambers of the heart, is an important issue for many patients with single ventricles. Progressive leakage of the AV valve has been associated with poor outcome after a Fontan operation. Timing for repair or replacement of the valve relative to performing a Fontan operation has been controversial. In some patients, issues related to this valve may prohibit children from being a good candidate for a Fontan operation. In this group of patients, many of us are not sure what is the best long-term management strategy.

Clinical Course of the Cases Presented

We presented a case of a 6-year-old girl who was born with what is called an “unbalanced AV septal defect.” She had a common atrium, common AV valve, and common ventricle. She had 5 open heart surgeries. This started as a newborn when she required connection of the pulmonary artery to the aorta in order to improve blood flow to the body. During this surgery a shunt was placed to provide blood flow to the lungs. She had a Glenn operation at 5-months-old along with the 1st repair of the common AV valve. Subsequently, she had another repair of the AV valve when she was 18 month old. That repair also failed and at age 4 she had replacement of the valve with a mechanical valve. Unfortunately, that valve developed clot within 3 months and it needed to be replaced with a pig valve. She has had the pig valve for the last 2.5 years and done very well. She needs to take the blood thinner warfarin (Coumadin) on a daily basis. Her parents are fantastic about managing this difficult medicine in a young child. She is in the first grade and very smart. She keeps up with her friends and classmates. Her oxygen saturation is usually 80-85%. We have struggled with whether she should have a Fontan operation. The risks include that she may need another replacement of the valve afterwards. If she does not have a Fontan, she will eventually become more cyanotic (blue) and may require a shunt. Another option would be to consider heart transplant at that time or when the valve starts to fail.

Important Points, Lessons Learned, and Potential Solutions

- All AV valves are not the same in patients with single ventricles. Some patients have only a mitral valve, while others have only a tricuspid valve as in HLHS. The most difficult valves to manage are the ones, similar to our patient, where there is a common AV valve. The anatomy of the common valves is most difficult for the surgeons to repair. The surgical approach needs to be individualized for each patient based on specific valve anatomy.

- A recent large study from Australia/New Zealand showed that by age 25 years, 56% of AV valves in patients who had a Fontan had failed. The common AV valves did the worst.
- In those who had a bad AV valve, the Fontan circulation failed sooner. 20-year survival for these patients after Fontan was only 54%.
- If the AV valve needs repair, it is best to do this **prior** to Fontan operation. In a large report from Mayo Clinic, AV valve surgery *at the time of or after* the Fontan was associated with an increased risk of death or needing heart transplant.
- For patients with single ventricles who are not candidates for a Fontan operation it is controversial whether adding a shunt to improve oxygen saturation or going directly to heart transplant is the best option. This is an area that needs further study by a large group of centers such as those associated with the Fontan Outcomes Network.