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Natalia Maksimova
Petra Smorodina Bldg. 6, Apt 115
St. Petersburg, Russia 195248

2ND OPINION
NAME: ANDREI FEDOROV
MRN: 71054225
DOB: 06/07/2019

Dear Mrs. Maksimova,

We received the clinical information as well as echocardiogram and cardiac catheterization studies done on Andrei, who is now five years of age and was born with hypoplastic left heart syndrome with mitral stenosis and aortic stenosis. From the clinical notes, he initially underwent a hybrid procedure with pulmonary artery banding and then subsequently had a comprehensive Stage II procedure with bilateral bidirectional Glenn's procedures as well as a connection of the main pulmonary artery and the aorta. He developed re-coarctation and has required multiple balloon dilations and, most recently, stenting of the re-coartation because of recurrent aortic arch obstruction. He has been followed because of moderate tricuspid regurgitation, although he does appear to have reserved right ventricular function. However, his oxygen saturation has been decreasing now in the high 60s to low 70s, and he is exercise-limited.

I have reviewed the imaging studies with Dr. Sunil Ghelani from our cardiology service, and we both concur with the diagnosis of hypoplastic left heart syndrome with a moderate to severe hypoplastic left ventricle with very little inflow. There is a very large connection between the main pulmonary artery and the ascending aorta, and there is still significant narrowing of the left pulmonary artery. This has been stented at a previous cardiac catheterization, and nevertheless, there appears to be very little blood flow going into the left lung at this time. There is moderate tricuspid regurgitation with preserved ventricular function, and the aortic arch appears to be improved in caliber after the stenting procedure.

Based on these findings, our assessment is that due to the very small size of the left ventricle in a 5-year-old and very little antegrade flow into that ventricle that he would not be a candidate for a reverse 1½ ventricle procedure. As to whether he would be a candidate for the Fontan, he would need to have improved blood flow into his left pulmonary artery as currently there is very little flow going into that left lung, and for a Fontan procedure to be successful, both lungs would need to be well perfused. In order to accomplish this, the only options that we have seen to be successful have been to first reduce the size of the neoaorta, which is quite large and compressing the left pulmonary artery, then add a shunt that goes into the left pulmonary artery to improve left lung profusion, and recruit the left lung for future surgery. At the same time, the tricuspid valve would need to be repaired in order to improve its function and to prevent further worsening of the tricuspid regurgitation because of the volume load from the shunt. This would be an interim procedure, and if the left lung does successfully recruit, then reassess as to whether a Fontan should be the next step. At this initial procedure, the left ventricle could also be evaluated to see if there is any possibility of recruiting it to use it as a subpulmonary ventricle. For this stage type of approach, we have usually performed the first stage relatively soon, and it typically would be one to two years before the second stage that could be considered.

I hope this information is helpful. If you have any questions, please do not hesitate to contact me.

Sincerely,

Pedro J. del Nido, MD

cc: Sunil Ghelani, MD | Department of Cardiology | Boston Children's Hospital | BADER 2
Andrei Borisov, MD | St. Petersburg, Russia

PDN:lw