



Boston Children's Hospital Heart Center

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HARVARD MEDICAL SCHOOL TEACHING HOSPITAL

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May 19, 2021

Natalia Erofeeva
8/38, Abramtsevskaia Ul.
Moscow 127576
Russia

RE: 2nd Opinion: Victoria Erefeeva—DOB: 9/12/2017

Dear Mrs. Erefeeva:

We received the clinical information as well as echocardiogram, cardiac catheterization and CT scan performed on Victoria who is now three years of age and was born with complex congenital heart disease with an unusual variant of complete AV canal defect, double outlet left atrium, likely inverted ventricles with the left ventricle being inferior and rightward. From the clinic notes, she underwent coarctation repair and pulmonary artery banding in September 2017, and then bidirectional Glenn and Damus Stansel operation in August 2018. In December of 2018, she underwent left ventricular valve plasty and also in 2019 had stenting of the left pulmonary artery. In January 2021, she underwent repeat left atrial ventricular valve plasty. At present she has moderate to severe regurgitation still from the left-sided superior portion of the common AV valve with the right-sided portion of the valve functioning well. There are two ventricular chambers with the left ventricle being inferior and rightward and is larger than the anatomic right ventricle which is leftward and superior. The right ventricle appears to be mildly to moderately hypoplastic. Ventricular function is well preserved. CT scan also shows severe narrowing of the left pulmonary veins with very little blood flow going in to left lung despite the stenting.

We have reviewed all the imaging studies and I have asked Dr. Kevin Friedman from our Cardiology Service to review the studies with me. We both concur with the above diagnosis. Victoria has complex anatomy. The major complicating factor at present is the fact that the left pulmonary veins near totally occluded and this fact will preclude the Fontan procedure as a surgical option. We do note however that she does have two ventricles with the right one being smaller, however the exact size is not well determined from the echocardiogram studies that we received. Our assessment is that she may be a candidate for potentially a biventricular repair or at least a one and a half ventricle repair, the small right ventricle receiving only inferior vena cava blood flow. This would require partitioning of the common AV valve, as well as an arterial switch procedure and re-routing of the venous return. Such a procedure is quite complex and is highly dependent on the specific details of the anatomy. In order for us to assess whether Victoria would be a potential candidate for such a procedure, we would need to perform a cardiac MRI, which would give us the required information to assess both the size of the two ventricles as well as the potential pathway between the left ventricle and the aorta. Thus we would recommend obtaining a cardiac MRI with specific emphasis on the size and position of the ventricular chambers which would allow us to determine whether a complete correction could be achieved in Victoria.

Sincerely,

Pedro J. del Nido, MD

cc: Kevin Friedman, MD
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