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Anastasia Karaseva
45-1 Lenina Prospekt, Apt. 7
Oktyabriy City, Russia 452600

RE: 2nd Opinion: Roman Karasev—DOB: January 18, 2017

Dear Mrs. Karaseva:

We received the clinical information as well as CT scan and echocardiogram studies done on Roman who is now five years of age and was born with the diagnosis of double outlet right ventricle with a large ventricular septal defect and obstruction to the pulmonary outflow tract. From the clinical notes, Roman underwent a palliative Blalock shunt at ten months of age followed by a bidirectional Glenn procedure and subsequently at four years of age, he underwent Fontan operation with subsequent catheter closure of the main pulmonary artery with a device. At four years of age, he also underwent fenestration of the Fontan connection presumably because of intolerance to the Fontan physiology. Since that time, his oxygen saturations had been approximately 19%, his initial operation for the Blalock shunt was complicated by a middle cerebral stroke from which he still has residual paresis of the right sided extremities.

We have reviewed the imaging studies and I have asked Dr. David Schidlow from our Cardiology Service to review the studies with me. We concur with the diagnosis of double outlet right ventricle, and tetralogy of Fallot type. There is a large ventricular septal defect with attachments of the tricuspid valve to the septum; however, we did not see straddling of this valve. Both ventricles are of good size, although the left ventricle does appear to have some diminished systolic function of unknown and unclear cause. He otherwise has good size pulmonary arteries and appears to be potentially a good candidate for biventricular conversion. However, in order to fully assess this option he would require first a cardiac MRI study with contrast to assess the ventricular function, particularly of the left side and also to determine whether there is any scarring in the left side of the heart that would suggest that it has been previously injured and therefore maybe a concern to use as a systemic ventricle. We also noted that there is significant tricuspid valve regurgitation and the mechanism of the tricuspid valve regurgitation would need to be determined with the MRI study. If this study suggests that he would still be a candidate for biventricular repair, then cardiac catheterization prior to a surgical intervention would also be required. The surgical procedure would then involve tunneling the left ventricle to the native aorta and in this process, closing the ventricular septal defect. The right ventricular outflow tract towards the pulmonary would need to be re-opened as there is a device occluding it. The pulmonary valve still does exist however, it is relatively small and one of the assessments that would have to be made would be whether the valve is salvageable or whether a new pulmonary valve would be required. The MRI and cardiac catheterization studies would help determine this prior to surgery.

As far as the timing of such an intervention, given that Roman has already reached the age of five, there is no advantage to waiting and we would recommend proceeding with the initial imaging studies within the next six months.

I hope this information is helpful and if you like for Roman to be evaluated at our center, please let us know and we can initiate that process.

Sincerely,

Pedro J. del Nido, MD

cc: David Schidlow, MD
Department of Cardiology
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