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February 18, 2016

Firth, Aiden
DOB: 6/2/05
Date: 2/18/16

Patricia McQuilken, MD
UMass Medical Center Group Practice
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Dear Trish,

Thank you for requesting a consultation for Aiden Firth. I had the pleasure of seeing him at Child Heart Associates in Fitchburg on 2/18/2016. As you know, Aiden is a 10½ year old boy with heterotaxy / polysplenia syndrome (S,D,S) associated with a secundum atrial septal defect, partial anomalous pulmonary venous return (with ipsilateral pulmonary veins), bilateral SVCs (absent bridging vein) and interrupted inferior vena cava (with hemiazygous communication to a left-sided SVC that drains into a dilated coronary sinus). He underwent surgical closure of his ASD with re-alignment of his atrial septum (allowing right-sided pulmonary veins to drain into the left atrium) along with resection of redundant mitral annular tissue last 02/21/06 (Dr. Pedro del Nido). His postoperative clinic evaluations of late have supported (asymptomatic) mild sinus node dysfunction with junctional escape rhythm - without any residual ASD shunt, significant mitral regurgitation or pulmonary vein stenosis.

He was last seen in clinic a year ago. Since then, mom has not observed of any recent exercise intolerance, exertional dyspnea, frequent dizziness, near-syncope or prolonged malaise.

Aiden was also diagnosed with intestinal malrotation as an infant S/P surgical repair (Ladd's procedure) with an appendectomy at Children's Hospital Boston (10/13/06). He had prior evidence of mild right-sided hydronephrosis with bilateral nephrocalcinosis, without any VCUG-evidence of reflux. An abdominal US at birth demonstrated splenic tissue. A peripheral blood smear did not demonstrate any abnormal Howell-Jolly bodies.

He was also recently diagnosed with right kidney ureteral obstruction (from abnormal lower pole crossing vessels) S/P right-sided pyeloplasty procedure (July 2015, CHB) accompanied by surgical repair of a small right-sided diaphragmatic hernia (that was incidentally found intraoperatively).

Aiden has a history of asthma and chronic enlargement of his tonsills and adenoids with obstructive sleep apnea S/P tonsilloadenoidectomy (Dec 2011). He enjoys soccer, basketball and karate - without any particular difficulties. He is particularly good with math and science in school. He has had good growth and development.

Other than as mentioned above, no significant issues were related with regard to the ophthalmologic, ENT, musculoskeletal, integumentary, neurological, immunological or hematological systems.

At present, he remains on prn Albuterol, daily multi-vitamins and prn Miralax. He is allergic to MORPHINE SULFATE.

There was a family history of coronary artery disease (great-grandmother and great-grandfather), asthma (mother) and hypertension(father). The rest of the family history was unremarkable specifically for heart disease in childhood, Wolff-Parkinson-White syndrome, cardiomyopathy, congenital deafness and early sudden death of unclear etiology.

He was accompanied to clinic by his mother and older sister, Ciara.

On physical examination, Aiden was a well-appearing well-developed well-nourished boy in no respiratory distress. His weight was 29.5 kg (19 %ile). His height was 138 cm (28 %ile). His BMI was 15.4 (11 %ile). His heart rate was 52 BPM. His blood pressure was 99/72 in the right arm. His pulse oximeter saturation was 100%. His skin was warm, dry and well-perfused. The conjunctivae were clear. His head, ears, nose and mouth were grossly normal. His neck appeared unremarkable without jugular venous congestion or neck mass. His lungs were clear with equal breath sounds over both lung fields. His abdomen was non-distended and non-tender. There was no audible abdominal bruit or hepatosplenomegaly.

His cardiovascular exam revealed a quiet precordium without lifts or thrills. The apical impulse was non-displaced. The first and second heart sounds were normal in intensity. There were no gallops, clicks or rubs. There was a grade 1-2/6 low-to medium- pitched systolic ejection murmur heard best at the left mid-sternal border - that became softer from the supine to the upright position. His pulses were full and regular in the radial and posterior tibial arteries.

An ECG performed 2/18/2016 revealed junctional rhythm with a rate of 47 and a normal QTc interval of .41 sec. The QRS axis was 91°. There was no evidence of RVH or LVH.

An echocardiogram was perfomed 02/18/2016. This patient has heterotaxy with polysplenia syndrome, bilateral SVCs, interrupted IVC with hemiazygous connection to the left SVC, secundum ASD, PAPVR with ipsilateral pulmonary veins S/P ASD

patch repair with septal patch diversion of right pulmonary vein flow return to the left atrium and mitral annuloplasty repair.

There was levocardia with concordant atrioventricular and ventriculoarterial relationships. This patient has bilateral SVCs with an absent bridging vein. The hepatic portion of the inferior vena cava was absent - with hepatic veins seen entering the right atrium. There was a left hemiaygous connection of the IVC to the left SVC that drained into a moderately dilated coronary sinus. The right SVC communicated directly with the right atrium. There was no significant RA chamber dilation. The left atrium was normal in size. There was limited interrogation of pulmonary venous flow return into the left atrium. There was no residual ASD seen (from parasternal windows, very limited subcostal views). The right ventricle appeared normal in size. There was no RV infundibular narrowing seen. The left ventricle appeared top normal in size. There was no concentric or asymmetric left ventricular wall hypertrophy or subaortic obstruction. There were coarse endocardial trabeculations seen towards the LV apex. The interventricular septum was intact. Septal wall contour was normal. The tricuspid valve was normal. There was trivial tricuspid valve regurgitation. The mitral valve was normally attached to (2) LV papillary muscles. There was no significant mitral regurgitation or mitral stenosis. There was no mitral valve prolapse or supravalve mitral membrane. The aortic valve was trileaflet in nature. There was no aortic valve stenosis or aortic valve regurgitation. The aortic aortic arch was unobstructed. There was no pulmonary valve stenosis. There was no discrete proximal PA branch stenosis. The origin of the left main coronary artery was normal. The origin of the right main coronary artery was not well-imaged. There was no evidence of any pericardial effusion, valve vegetations, intracardiac mass or thrombus. The left ventricular function was normal with a fractional shortening of 38% and LVEF 66%. Qualitative right ventricular wall contraction appeared normal.
AoAn = 18 mm (13.1 - 18.9, z = +1.4)
AoSin = 23 mm (16.4 - 25.2, z = +1.0)
AoAs = 19.9 mm (14.1 - 22.3, z = +0.8)
LA = 33 mm (14.7 - 35.0, z = +1.6)
LVs = 28.1 mm (20.9 - 30.8, z = +0.9)
LVd = 45.3 mm (34.3 - 45.9, z = +1.7)
PWTd = 6.5 mm (5.3 - 9.0, z = -0.7)
IVSd = 7.6 mm (5.4 - 9.6, z = +0.1)
LPA = 13.5 mm (7.5 - 14.0, z = +1.7)
RPA = 12 mm (8.2 - 14.6, z = +0.4)
FS = 38% (27% - 43%, z = +0.8)

A f/u 24 hr Holter recording was initiated after today's visit (2/19/2016). The heart rate varied between 42 and 154 BPM, mean 61 BPM (z = -2.2 range 63.3 - 107.9 for age.) There was normal circadian variability. Cardiac rhythm alternated between an ectopic atrial rhythm and junctional (escape) rhythm (especially at the low- to mid-range of his HRs). There were occasional isolated ventricular premature beats and

less frequent atrial premature beats with aberrancy (total ectopy approx. 2% of his total heartbeats. There was evidence of sinus tachycardia at his maximum HR (140-150's) - noted during an eating contest in church. There was no high-grade (type 2 second degree or third degree) AV block seen. There were no symptoms reported. This study is stable compared to his previous exam (done Feb 2015).

In summary, Aiden has stable mild (asymptomatic) sinus node dysfunction - with a preserved ability to increase his HR with exertion and (expected) top normal LV chamber size and normal LV systolic function - that warrants continued serial monitoring for now (i.e. no strong clinical criteria for pacemaker placement at this point in time). His f/u echocardiogram also supports coarse LV apical trabeculations (not quite classic for LV noncompaction) - without any significant mitral stenosis, mitral regurgitation or pulmonary hypertension. Mom was reassured accordingly.

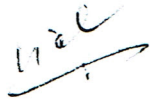
I would like to see Aiden back in cardiology clinic in one year, or sooner if he should have any signs or symptoms attributable to his cardiovascular system that you or his family find concerning.

In the meantime, Aiden does not require SBE prophylaxis at times of increased risk, nor need he be restricted in his activity.

It has been my great pleasure to participate in the care of this very pleasant young man. Please do not hesitate to contact me if I can be of additional assistance.

Thanks!

Sincerely,



M. Victoria T. Tantengco, MD

cc: Family

cc: Gerald Marx, MD