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February 26, 2014

Firth, Aiden
DOB: 6/2/05
Date: 2/26/14

Patricia McQuilken, MD
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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 Worcester on 2/26/2014. As you know, Aiden is an 8½ 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).

Aiden was diagnosed with intestinal malrotation S/P surgical repair (Ladd's procedure) with an appendectomy at Children's Hospital Boston (10/13/06) without any postoperative intestinal obstruction. 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 (as reviewed by Dr. Keuker) did not demonstrate any abnormal Howell-Jolly bodies.

Of note, he developed mild sinus node dysfunction associated with an intermittent junctional escape rhythm. His most recent Holter exam done a year ago rare ectopy (< 1% of total heartbeats), sinus pause with junctional escape rhythm at lower HRs (without any correlating symptoms) and resumption of normal sinus node activity (with sinus tachycardia) at faster HRs.

He now presents with at least two recent episodes of dizziness (while at rest) with a pulse rate of 47-58 bpm - that lasted for hours - without any vomiting, diarrhea, onfoing febrile illness, significant pallor, prolonged lethargy or progression to frank

syncope. Mom also thinks that he has become more tired of late - especially when he gets home from school - which appears to be more noticeable compared to this time last year. He denies any chest palpitations or sensed tachycardia.

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 riding his bike, swimming, soccer, karate, ice-skating without any particular difficulties. 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. 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.

On physical examination, Aiden was a well-appearing well-developed well-nourished boy in no respiratory distress. His weight was 24.1 kg (18 %ile). His height was 127 cm (23 %ile). His BMI was 14.9 (16 %ile). His heart rate was 59 BPM (stable from last year). His blood pressure was 101/53 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 good air exchange. His abdomen was non-distended without any tenderness, palpable mass or enlargement of the spleen or liver. The back and chest revealed no tenderness.

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 2/6 low-pitched systolic murmur heard best at the right upper sternal border. His pulses were full and regular in the radial and posterior tibial arteries.

An ECG performed 2/26/2014 revealed junctional rhythm with a rate of 51 with a normal QTc interval of .37 sec. QRS axis was normal (+62). R wave progression was normal across the left precordium.

An echocardiogram was perfomed 02/26/2014 - primarily to assess cardiac function. 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. 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 right SVC flow obstruction seen. There was no significant RA chamber dilation. The left atrium was normal. There was non-disturbed flow from at least 2 of 4 pulmonary veins into the left atrium. There was no significant residual ASD seen in this f/u exam. The right ventricle appeared normal in size. There was no RV infundibular narrowing seen. There was mild RV apical muscle bundle prominence seen. The left ventricle appeared top normal in size. There was no left ventricular wall hypertrophy or subaortic obstruction. There was mild endomyocardial noncompaction seen towards the LV apex. The interventricular septum was intact. The tricuspid valve was normal. There was trivial tricuspid valve regurgitation. The mitral valve was normal. There was no mitral regurgitation or LV inflow obstruction. There was no mitral valve prolapse. 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. There was no residual PDA shunt. 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 37%. Qualitative right ventricular wall contraction appeared normal.
LA = 28.6 mm (13.9 - 33.2, z = +1.0)
LVs = 27.5 mm (19.7 - 29.0, z = +1.3)
LVd = 43.8 mm (32.4 - 43.3, z = +2.1)
PWTd = 7.1 mm (5.0 - 8.4, z = +0.4)
IVSd = 5.7 mm (5.1 - 9.0, z = -1.3)
FS = 37% (28% - 42%, z = +0.5)

A 24 hr Holter recording was initiated at the end of this visit (02/26/2014). The heart rate varied between 37 and 142 BPM, mean 59 BPM. There was normal circadian variability. There was normal sinus arrhythmia. Cardiac rhythm varied between sinus bradycardia, ectopic atrial rhythm and sinus pauses with a junctional escape rhythm - all at relatively lower HRs. Conversely, the prevailing rhythm was sinus in nature at faster HRs (i.e. sinus rhythm / sinus tachycardia at HR > 85 bpm). There were occasional isolated ventricular, junctional and atrial premature beats (1.5 % of total heartbeats) with variable aberrancy. There were no atrial/ventricular couplets or runs of narrow or wide complex tachycardia seen. There were no symptoms reported. This is a stable exam (with similar HR range) compared to his last Holter study (March 2013).

In summary, Aiden has a stable f/u cardiac exam with adequate systemic perfusion.

Firth, Aiden

His EKG and Holter study do not indicate progressive or increased sinus node dysfunction), high-grade atrial/ventricular ectopy or significant AV block. His echocardiogram supports stable mild LV apical noncompaction with normal RV/LV wall contraction and the absence of any significant MR/AI or pulmonary hypertension.

Mom is aware that patients with heterotaxy syndrome with polysplenia can develop progressive sinus/AV node dysfunction. I don't think we have reached the clinical threshold for pacemaker placement - but his occasional dizziness needs closer attention.

As such, I am recommending a (baseline) exercise EKG cycle test with metabolics through the exercise lab at Children's Hospital Boston.

In the absence of any significant concerns with this exam, I would like to see Aiden back in cardiology clinic in 6-9 months, or sooner if he should have any signs or symptoms attributable to his cardiovascular system that you or his family find concerning.

Due to a change in the guidelines by the American Heart Association in 2007, endocarditis prophylaxis is no longer recommended for most children and adults with congenital heart disease. Consequently, it is not necessary for Aiden to take SBE prophylaxis at times of increased risk, such as dental visits. If you or his dental provider have any questions about this change in recommendations, please do not hesitate to give me a call.

It has been my great pleasure to participate in the care of this very pleasant boy and his family. 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

cc: Nancy Hagberg, FNP