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February 19, 2015

Firth, Aiden
DOB: 6/2/05
Date: 2/19/15

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 2/19/2015. As you know, Aiden is a 9½ 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 have supported mild (asymptomatic) sinus node dysfunction without any residual ASD shunt, significant mitral regurgitation or right-sided pulmonary venous flow obstruction.

Aiden was also 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.

He was last seen in clinic almost six months ago with a soft systolic ejection flow murmur and remaining in junctional rhythm (while at rest). An EKG treadmill test in March 2014 revealed a mildy reduced chronotropic response to exercise (peak exercise HR 155, 75% of predicted normal) with evidence of sinus rhythm / sinus tachycardia towards peak exercise.

Since then, he has had almost cyclical bouts of (RUQ) abdominal pain, vomiting and

constipation (every 4-6 weeks) - without any fever, recent weight loss or progressive deterioration in his overall exercise tolerance. He denies any frequent or associated dizziness, dimming of his vision, near-syncope, chest palpitations or sudden-onset 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 ice skating, gym class and plays soccer 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, 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.

On physical examination, Aiden was a well-appearing well-developed well-nourished boy in no respiratory distress. His weight was 26.8 kg (20 %ile). His height was 132 cm (23 %ile). His BMI was 15.3 (17 %ile). His heart rate was 53 BPM. His blood pressure was 101/56 in the right arm. His pulse oximeter saturation was 99%. 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 with normoactive bowel sounds - without any tenderness, palpable mass or enlargement of the spleen or liver.

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 left mid-sternal border through the base - 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 2/19/2015 revealed sinus pause with a junctional (escape) rhythm with a rate of 53 and a QTc interval of .42. QRS axis was normal (+89). There was no manifest pre-excitation, significant ST segment elevation / depression, pathologic q waves, atrial enlargement, RVH, LVH or RBBB with ST segment elevation over the right precordial leads.

An echocardiogram was perfomed 02/19/2015. 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 heptic 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 right SVC flow obstruction seen. There was no significant RA chamber dilation. The left atrium was normal. There was non-disturbed venous flow return from all 4 pulmonary veins into the left atrium. There was no residual ASD seen. The right ventricle appeared normal in size. There was no RV infundibular narrowing seen. There were RV apical coarse trabeculations (normal finding). The left ventricle appeared top normal in size. There was no concentric or asymmetric left ventricular wall hypertrophy or subaortic obstruction. There were trabeculations seen towards the LV apex (not quite consistent with classic LV noncompaction). The interventricular septum was intact. The tricuspid valve was normal. There was trivial tricuspid valve regurgitation. The mitral valve was normal. There was no significant 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 left-sided 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. The origin of the left main coronary artery was normal. LCA = 3.8 mm (2.0 - 3.8, z = +2.1). 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 36%. Qualitative right ventricular wall contraction appeared normal.

AoAn = 17.8 mm (12.6 - 18.2, z = +1.7)
AoSin = 26 mm (15.9 - 24.3, z = +2.8)
AoAs = 21.2 mm (13.7 - 21.6, z = +1.8)
LA = 25 mm (14.3 - 34.1, z = +0.1)
LVs = 28 mm (20.3 - 29.9, z = +1.2)
LVd = 44 mm (33.4 - 44.6, z = +1.7)
PWTd = 7 mm (5.2 - 8.7, z = +0.1)
IVSd = 7 mm (5.2 - 9.3, z = -0.3)
LCA = 3.8 mm (2.0 - 3.8, z = +2.1)
PV = 21.8 mm (13.9 - 22.4, z = +1.7)
LPA = 12.7 mm (7.2 - 13.5, z = +1.5)
RPA = 12.6 mm (7.9 - 14.1, z = +1.0)
FS = 36% (28% - 43%, z = +0.3)

Firth, Aiden

A 24 hr Holter recording was initiated at the end of this visit (2/19/2015). The heart rate varied between 43 and 92 BPM, mean 53 BPM (z = -3.1 range 64.8 - 108.9 for age.) There appears to be an attenuated or reduced circadian variability - with awake HRs mostly in the 60-70's range, asleep HR in the 40-50's range. Cardiac rhythm appeared to be predominantly junctional in nature (sinus node dysfunction with junctional escape rhythm, accelerated junctional rhythm,) - with junctional rhythm that alternated with an ectopic atrial rhythm at lower HRs. There was appropriate recovery of sinus node function at faster HRs (i.e., sinus rhythm noted at his maximum recorded HR in the low 90s). There were occasional ectopic atrial premature beats (< 1 % of total heartbeats) and rare ventricular premature beats (<< 1% of total heartbeats) - without any ventricular couplets/triplets or runs of atrial- or ventricular- tachycardia seen. There was no third degree AV block seen. There were no symptoms reported.

In summary, Aiden has a stable f/u cardiac exam with asymptomatic mild sinus node dysfunction (i.e., mildly reduced resting HRs now in the 50's, junctional in nature). His f/u echocardiogram supports normal biventricular wall contraction, no residual ASD shunt, non-obstructed right-sided pulmonary veins, top-normal LV chamber size with normal LV wall contraction and coarse LV apical trabeculations (not quite fulfilling criteria for LV noncompaction or spongiform cardiomyopathy). His Holter exam demonstrated appropriate recovery of sinus node function at faster HRs - without any AV dissociation or high-grade AV block seen.

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 boy. 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