An Unusual Cause of Progressive Cyanosis Post Fontan Operation: Congenital Extra-Hepatic Porto-Systemic Shunt

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Objective: *To report an unusual case of progressive cyanosis post Fontan operation due to porto-systemic venous shunt and the result of its treatment.*

Material and Method: *A patient with diagnosis of progressive cyanosis post Fontan operation from porto-systemic venous shunt at QSNICH.*

Results: *This is a case of twelve years old girl, who had diagnosis of situs solitus, levocardia, atrio-ventricular concordant, ventriculo-arterial concordant, hypoplastic right ventricle with large ventricular septal defect. She had pulmonary artery banding at 4 months of age followed by a non-fenestrated extra-cardiac conduit Fontan performed at 7 years and 7 months of age. During the first year of post operation, her systemic oxygen saturation (SpO_2) was 93-94% after which it decreased to 87%, 84%, 75% at 1.5, 2.5 and 3 years after surgery, respectively. Clinically she also had progressive dyspnea on exertion. Diffuse pulmonary arterio-venous malformation was demonstrated by contrast echocardiogram during cardiac catheterization. Cardiac magnetic resonance angiography showed abnormal extra-hepatic portal vein to inferior vena cava shunt. After balloon test occlusion in the cath lab, which showed no change in the portal venous pressure, complete occlusion of this porto-systemic venous shunt was performed by using Amplatzer Vascular Plug II. Her systemic oxygen saturation increased to 83% with functional class I at one-year post occlusion.*

Conclusion: *The present report an unusual case of progressive cyanosis post Fontan operation due to pulmonary arterio-venous malformation, which was secondary to congenital extra-hepatic porto-systemic shunt. The venous blood from the intestinal and splenic veins was partially bypassing the liver into inferior vena cava. The patient's clinical condition and SpO_2 improved after transcatheter occlusion of the shunt with the device.*

Keywords: *Fontan operation, Cyanosis, Congenital extra-hepatic porto-systemic shunt, Pulmonary arterio-venous malformation*

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Fontan type operation is a palliative treatment for univentricular heart disease. Progressive cyanosis is an unusual post Fontan operation[1,2]. Most reports have shown it to be due to systemic venous collateral drainage into the heart causing right to left shunt from the higher systemic venous pressure to the lower pulmonary venous pressure. Reports of persistent cyanosis post Fontan are mostly from fenestrated or venous collateral channels to the heart[1,3,4], which commonly occur in the supracardiac than cardiac and infracardiac type[5,6]. These venous collaterals increase in size post op with time[5] and cause progressive cyanosis. Pulmonary ateriovenous malformation (pulmonary AVM) is another cause of cyanosis but mostly developed after bidirectional cavopulmonary shunt[7,8], post Kawashima or Fontan operation which exclude hepatic flow[9,10]. There have been reports of porto-systemic venous shunt as a cause of pulmonary AVM or hepatopulmonary syndrome[11-15]. Acute severe cyanosis after Kawashima operation resulting from congenital porto-systemic shunt and cyanosis, which improved after surgical division of this porto-systemic shunt had been reported[16]. Recently there have been reports of endovascular treatment for closure of this shunt instead of surgery[11,17].

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Material and Method

This is a case report of a twelve years old girl, who had diagnosis of situs solitus, levocardia, atrio-

ventricular concordance, ventriculo-arterial concordance, hypoplastic right ventricle with large ventricular septal defect since birth. She had pulmonary artery banding done at the age of 4 months due to her congestive heart failure. Her first cardiac catheterization performed at the age of 6 years confirmed the above detailed diagnosis and the angiogram showed migration of the banding site from MPA to PA bifurcation causing mild RPA stenosis. Her RPA pressure was 9/5 (mean 7) mmHg, LPA pressure was 26/6 (mean 15) mmHg and the aortic oxygen saturation was 81%. The calculated Fontan index on her right lung was 2.3 and left lung was 5. She underwent total cavo-pulmonary connection (TCPC) with non-fenestrated extra cardiac conduit at the age of 7 years and 7 months. After the TCPC operation, she had been asymptomatic with her systemic oxygen saturation of 93-94% during the first year post operative which then gradually decreased to 87%, 84% at 1.5 and 2.5 years post surgery, respectively. Clinically she also had mild progressive dyspnea on exertion. The second cardiac catheterization performed showed that her systemic oxygen saturation was 92% and the angiogram of SVC, innominate vein, IVC, pulmonary arteries could not identify any cause of cyanosis.

By 3 years post TCPC, her systemic oxygen saturation had progressively dropped to 75%. Her cardiac magnetic resonance angiography (MRA) showed no abnormal systemic venous connection to the heart, normal hepatic veins and supra hepatic IVC connected to Fontan conduit and no major collateral artery. However, there was a huge collateral vein in the abdomen from the extra hepatic portal vein running tortuously and connecting to inferior vena cava at supra renal vein (porto-systemic venous shunt). The third cardiac catheterization demonstrated multiple small pulmonary AVM using micro air bubble saline injection selectively in pulmonary arteries along with low oxygen saturation at right upper lobe pulmonary vein detected via retrograde catheterization from left ventricle to left atrium across the mitral valve.

After extensive reviewed literature search and deliberation, her progressive cyanosis was attributed to pulmonary AVM, which was thought to be secondary to porto-systemic shunt. Her liver enzymes were normal and gastroscopic examination showed no signs of portal hypertension. The fourth catheterization was performed retrogradely from IVC into the collateral vein, and revealed venous blood shunting from the portal system to IVC. The patency of portal system was confirmed by selective injection of contrast media at the portal vein. The pressure in portal venous system was 9 mmHg and the IVC pressure was 9 mmHg. After the balloon occlusion test with pressure monitoring at distal end of the catheter did not show any change in the portal venous pressure, the shunt was completely occluded with 14 mm Amplatzer vascular plug II device. There was no complication during and after the procedure. Her systemic oxygen saturation did not immediately change. However, at follow-up, her symptoms and systemic oxygen saturation gradually improved. At 1-year post occlusion, her systemic oxygen saturation increased to 83% and she was in a clinically functional class I status.

Discussion

In complex congenital heart disease (complex CHD) and CHD, which have only one functioning ventricle, the aim of treatment is to proceed with Fontan type operation. Venous blood from superior vena cava and inferior vena cava will be diverted to pulmonary arteries; only venous blood from the coronary system will drain to the heart. Therefore, after Fontan operation, there may be mild or no clinical cyanosis detected. However due to high systemic venous pressure, early and late complication after Fontan operation may occur. Fenestrated Fontan or leaving some systemic venous blood drainage into the heart can cause a small right to left shunt resulting in mild systemic oxygen desaturation. Acquired right to left shunt from the systemic venous blood to the heart may occur and cause progressive cyanosis after Fontan[1,3,4]. Supracardiac type of the venous collateral to the heart was reported to occur more commonly than cardiac and infracardiac types[5,6].

Pulmonary arteriovenous malformation (pulmonary AVM) is another cause of cyanosis. Deoxygenated blood from the pulmonary arteries will shunt to pulmonary veins before oxygenation in the alveoli. Pulmonary AVM is a common late complication of bidirectional cavopulmonary shunt or hemi-Fontan, where venous blood from only SVC be oxygenated in both lungs[7,8]. Lack of hepatic blood flow to either or both lungs can result in pulmonary AVM in that lung(s)[10,18]. After Fontan conversion, venous blood from IVC drain to both pulmonary arteries and can resolve the problem of pulmonary AVM[7]. Pulmonary AVM can also develop secondary to portal hypertension and abnormal liver function. There are numerous reports of pulmonary AVM in patients with left isomerism or interrupted IVC with azygous continuity to SVC[19] post Kawashima operation[16,20].

In these patients, both SVC and IVC, not hepatic blood will drain to pulmonary arteries after bidirectional cavopulmonary shunt. Pulmonary AVM post Kawashima will improve after additional hepatic blood flows to both pulmonary arteries[7].

Congenital extra-hepatic porto-systemic malformation or Abernethy malformation is a veno venous shunt from portal vein to inferior vena cava[21-24], It was first reported by Abernethy in 1793. Congenital porto-sytemic malformation is a rare congenital anomaly, and is divided into extra and intra hepatic porto-systemic shunt[25]. Congenital extra-hepatic porto-systemic shunt is classified into type I (absence of intrahepatic portal veins) and type II (patent intrahepatic portal veins)[26]. Park et al described 4 types of congenital intra-hepatic porto-systemic shunt[27] and some author adds patent ductus venosus as the fifth type. The patients with porto-systemic venovenous shunt may present with signs of hepatic shunting: encephalopathy, pulmonary hypertension, hepatic pulmonary syndrome and/or hypoglycemia. Liver function tests are usually preserved and there are no clinical signs of portal hypertension (e.g. splenomegaly, ascites, varices) in patients with porto-systemic shunt. Clinical symptoms are less dependent on the anatomical classification[25]. The pathophysiology of those symptoms are explained by venous blood shunting from the liver causing less detoxification leading to hyperammonemia and encephalopathy, vasoactive substance from splancnic area leading to pulmonary hypertension, lack of hepatic factor leading to pulmonary AVM or hepatic pulmonary syndrome. Currently, there are many reports of congenital porto-systemic shunt in heterotaxy polysplenic syndrome in which there is interruption of IVC and azygous continuity to SVC[16,19,20,28].

In the present case, the patient had situs solitus, and all hepatic veins joined the IVC and so the hepatic venous blood flowed to both the lungs and since the patient had no bidirectional cavopulmonary shunt done before the Fontan operation, she was not at risk for developing pulmonary AVM. Shortly after the Fontan operation, her systemic oxygen saturation of 93-94% meaning that there was no significant right to left shunt. Progressive cyanosis after Fontan led to intensive work up for the location of right to left shunt and abnormal vein of porto-systemic veno venous shunt was eventually located. Cardiac MRA is very useful for screening of abnormal vessel, which may not be diagnosed by usual cardiac catheterization unless there is clinical suspicious. Veno-venous blood

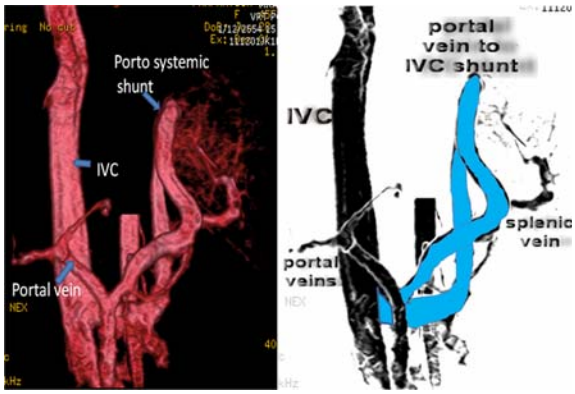

Fig 1. Picture and diagram from MRA showed Extrahepatic portosystemic shunt connecting from portal vein with tortous course upward and downward then connected to inferior vena cava.

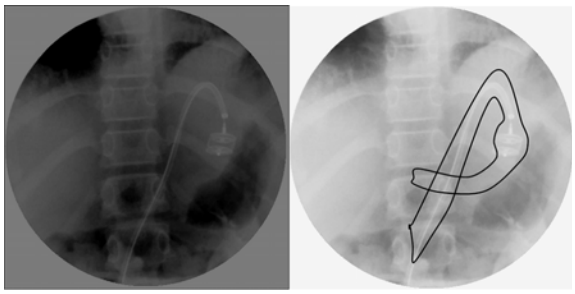

Fig 2. Cardiac catheterization in antero posterior projection showed the 14 mm AVP II occluded the portosystemic shunt retrogradely from inferior vena cava.

shunting from portal system to IVC per se does not produce right to left shunt. Air bubble saline contrast echo can easily diagnose pulmonary AVM and deoxygenated blood from pulmonary veins can confirm the diagnosis of right to left shunt from pulmonary AVM. Porto-systemic shunt can be found secondary to portal hypertension. Congenital porto-systemic shunt can be differentiated from secondary porto-systemic shunt by the absence of ascites, splenomegaly esophageal and gastric varices or other venous collaterals.

Diagnosis of congenital extra-heaptic porto-systemic shunt by ultrasound may not be difficult for experienced radiologist but it would be a big problem for pediatric cardiologist who are not familiar with abdominal organs and vessels. The use of non-invasive methods of computed tomography angiogram (CTA) and MRA can make the diagnosis of porto-systemic venous shunt easier.

Conclusion

The authors report an unusual cause of progressive cyanosis post Fontan operation from pulmonary AVM secondary to congenital extra-hepatic porto-systemic shunt. The venous blood from the intestinal and splenic veins was partially bypassing the liver into inferior vena cava. Clinical signs and symptoms improved after transcatheter occlusion of the shunt with Amplatzer vascular plug II device.

Potential conflicts of interest

None.

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สาเหตุที่พบไม่บ่อยของอาการเขียวที่ค่อย ๆ เพิ่มมากขึ้นหลังการผ่าตัด Fontan: Congenital Extra-hepatic Porto-systemic Shunt

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วัตถุประสงค์: เพื่อรายงานเด็กที่มีอาการเขียวที่ค่อย ๆ เพิ่มมากขึ้นหลังการผ่าตัด Fontan จากหลอดเลือดดำลัดวงจรจาก extra-hepatic porto-systemic ที่เป็นมาตั้งแต่กำเนิดและผลการรักษา

วัสดุและวิธีการ: ผู้ป่วยเด็กที่ได้รับการวินิจฉัยว่ามีอาการเขียวค่อย ๆ เพิ่มมากขึ้นหลังการผ่าตัด Fontan โดยมีสาเหตุของการเขียวจากหลอดเลือดดำลัดวงจร extra-hepatic porto-systemic shunt ที่สถาบันสุขภาพเด็กแห่งชาติมหาราชินี

ผลการศึกษา: ผู้ป่วยเด็กหญิงไทยอายุ 12 ปี ได้รับการวินิจฉัยว่าเป็นโรคหัวใจพิการแต่กำเนิดชนิด situs solitus, levocardia, AV concordant, VA concordant, hypoplastic RV with large VSD เด็กได้รับการรักษาด้วยการทำ pulmonary artery banding ตั้งแต่อายุ 4 เดือนและได้รับการผ่าตัด non-fenestrated extra-cardiac conduit Fontan เมื่ออายุ 7 ปี 7 เดือน หลังการผ่าตัดในช่วงปีแรกพบว่ามีค่าความอิ่มตัวของออกซิเจนที่วัดทางผิวหนังได้ 93-94% หลังจากนั้นค่าออกซิเจนลดลงเหลือ 87%, 84%, 75% ที่ 1.5, 2.5 และ 3 ปี หลังการผ่าตัดตามลำดับโดยที่เด็กมีอาการเหนื่อยง่ายมากขึ้นร่วมด้วย จากการตรวจคลื่นเสียงสะท้อนหัวใจโดยการฉีดน้ำเกลือที่เขย่ากับฟองอากาศเล็ก ๆ เข้าทางหลอดเลือดดำ และจากการสวนหัวใจช่วยให้การวินิจฉัยภาวะ pulmonary ateriovenous malformation ผลการตรวจด้วยคลื่นไฟฟ้าแม่เหล็กพบว่ามีหลอดเลือดดำลัดวงจรผิดปกติต่อจาก extra-hepatic portal vein ไปยัง inferior vena cava เด็กได้รับการสวนหัวใจ ทำการทดสอบอุดหลอดเลือดดำที่ผิดปกตินี้ด้วยสายสวนหัวใจที่มีลูกโป่งซึ่งพบว่าไม่มีการเปลี่ยนแปลงความดันของ portal vein จึงทำการอุดหลอดเลือดดำลัดวงจรที่ผิดปกตินี้ด้วย AVP II จากการติดตามที่ 1 ปีหลังการรักษาพบว่าค่าความอิ่มตัวของออกซิเจนทางผิวหนังเพิ่มขึ้นเป็น 83% และอยู่ใน functional class I

สรุป: ได้รายงานกรณีผู้ป่วยเด็กหลังการผ่าตัด Fontan แล้วมีอาการเขียวค่อย ๆ เพิ่มมากขึ้นเรื่อย ๆ จากการตรวจค้นหาความผิดปกติเพิ่มเติมพบว่ามีสาเหตุจาก pulmonary AVM ซึ่งเป็นผลตามหลังมาจากหลอดเลือดดำลัดวงจร extra-hepatic porto-systemic shunt ซึ่งเป็นความพิการแต่กำเนิด ผลของการลัดวงจรทำให้เลือดดำจากบริเวณลำไส้ และม้ามถูกลัดวงจรไปยังหลอดเลือดดำ inferior vena cava ไม่ผ่านไปยังตับอาการเด็กดีขึ้นและค่าความอิ่มตัวของออกซิเจนเพิ่มขึ้นหลังการอุดหลอดเลือดดำลัดวงจรนี้ด้วยอุปกรณ์อุดผ่านสายสวนหัวใจ