A Rare Complication of Hepatopulmonary Syndrome after a Fontan Cardiac Surgery

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Introduction

- Fontan procedure was first performed in 1971 and has been used to treat congenital heart defects involving a single ventricle
- Performed by Frances Fontan and Eugene Baudet- an anastomosis is created directly to the pulmonary artery from the right atrium, bypassing the ventricle, allowing for systemic venous blood flow into the lungs (Figure 1)
- This rare procedure is performed for less than 0.02% of live births per year
- Known pulmonary risks associated with this procedure include the development of pulmonary AVM, plastic bronchitis; associated liver damage (FALD) specifically hepatic fibrosis and hepatocellular carcinoma
- Currently, there are only 6 known Fontan cases that suffered from hepatopulmonary syndrome (HPS) as it's main complication (Choi et al 2009). We present the seventh such case.

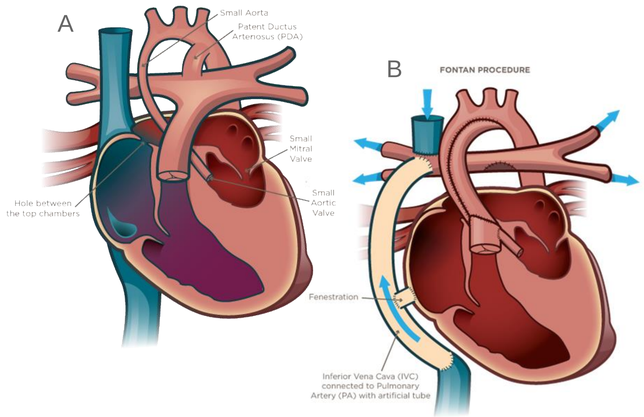

Figure 1:
Figurative illustration of how a patient presents at the time of birth (A). Where there may be congenital defects of the aorta, resulting in a hypoplastic left ventricle, and possible septal defects. The Fontan procedure (B), is shown with the presence of an artificial circuit, that connects the IVC with the pulmonary artery. From SSM Health Cardinal Glennon Children's Hospital

Case Report

- 26-year-old female with a history of coarctation of the aorta, hypoplastic left heart status post Fontan procedure at Day 2 of life.
- Admitted with acute hypoxic respiratory failure and decompensated liver cirrhosis, following recent cruise travel with binge drinking episode.
- Physical exam revealed clubbing of her digits, LE edema, abdominal pain, ascites, central and peripheral cyanosis (Figure 2)

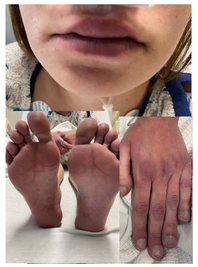

Figure 2:
Physical exam revealing clubbing of the digits, along with central cyanosis.

- Labs revealed elevated ALP, bilirubin and a secondary polycythemia
- CT Chest (Figure 3) and echocardiogram revealed endocardial cushion defect and dilated right ventricle, with hypoplastic left ventricle.
- Peritoneal fluid was positive for SBP, and pleural fluid was positive for spontaneous bacterial pleuritis
- Patient was treated with antibiotics, thoracentesis and evaluated for possible heart transplant.

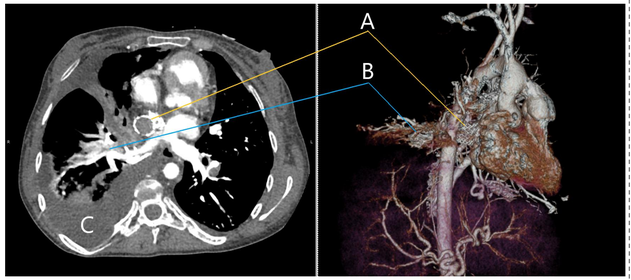

Figure 3:
CT Chest revealed the Fontan conduit (A), dilated sub-pleural pulmonary vessels concerning for AVMs (B), right pleural effusion (C), portosystemic venous shunt, esophageal and inferior epigastric varices.

Discussion

- There is a 85% survival rate that is estimated post Fontan procedure
- Extra-cardiac manifestations post Fontan circulation include protein enteropathy, pulmonary vascular disease, restrictive lung disease, secondary erythrocytosis, renal abnormalities, sexual dysfunction, and pregnancy complications (Biissett et al, 2022)
- Those patients with an increased risk of Fontan circulation failure, especially those with tricuspid atresia, double inlet left ventricle, and hypoplastic left heart; would benefit from surgery such as Fontan conversion and heart transplantation (Hassan et al 2021).
- Limited data on the post transplant survival outcomes as a result of small sample sizes, although meta-analysis completed by Hassan et al. showed a 74.5% survival rate 1 year post cardiac transplant in the pediatric population vs a 69.2% 5 year survival post transplantation.

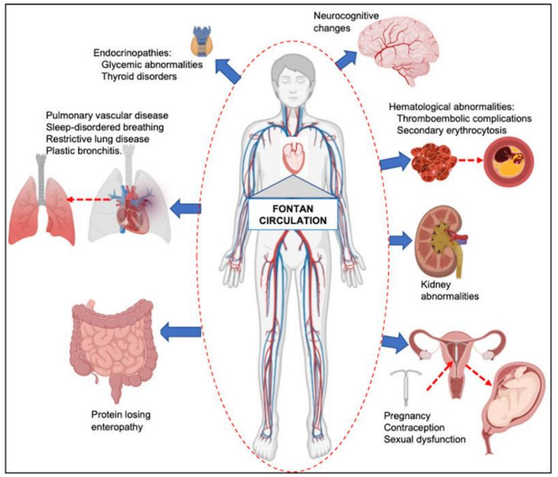

Figure 4:
Illustrates the extra cardiac manifestations post Fontan Circulation
Adapted from International Journal of Cardiology Congenital Heart Disease

Conclusion

- There is limited data in the development of HPS status post-Fontan surgery. Rodriguez et al (2018) suggests an increased risk of mortality in those with HPS, and a correlation between mortality and disease severity of HP
- Given there is an overlap of HPS, PPHTN (portopulmonary hypertension), and FALD; this case serves to highlight the complexity of managing similar patients

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