

CASE REPORT

Protein losing enteropathy after Fontan procedure

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SUMMARY

Fontan and Baudet described the procedure in 1971 and was very useful to improve the quality of life in many complex cyanotic heart diseases. It has gone through various modifications since then to improve the outcome. The mortality was reported as 2.1% and survival rate of at 5 years, 10 years, 15 years and 20 years were 91%, 80%, 73% and 69% respectively. Though it was a useful palliative procedure to improve the quality of life, it has complications which may affect the morbidity and mortality like Protein losing enteropathy, reduced exercise capacity, thromboembolism. The patients with protein losing enteropathy present with Pleural effusion, ascites, and edema and they need periodic replacement of Albumin and frequent reviews and close follow up in the management to reduce the mortality and to improve the quality of life. These complications we come across and they need our support in the management at district level hospitals.

CASE REPORT

On 11th October 2001, a 6-month-old boy was brought to Hospital Tawau with complaints of cyanosis during feeding and crying, which later became worsening with reduced effort tolerance. He was born at hospital via spontaneous vaginal delivery with the birth weight 3kg.

Echocardiogram was done in Jan 2002 and concluded as large ventricular septal defect, single atrium, two atrioventricular valves, patent ductus arteriosus. Thus he was started on anti-failure medications.

May 2005 he was referred to National Heart Institute and underwent a cardiac catheterization on 3rd February 2006. The procedure demonstrated bilateral superior vena cavae with bridging veins connecting both superior vena cavae, total anomalous pulmonary venous drainage with pulmonary veins drain into left superior vena cava, confluent pulmonary artery but slightly small left pulmonary artery, right pulmonary artery 12.2mm, descending aorta 10.9mm, McGoon ratio 1.7, right aortic arch and no collaterals.

Bilateral bidirectional Glenn shunt and correction of intracardiac total anomalous pulmonary venous drainage was done on 10th February 2006. Intraoperative findings are bilateral superior vena cava, small right atrium (almost single atrium), and hypoplastic tricuspid valve. He continued his regular follow-up in Paediatric Cardiology Clinic in hospital Tawau.

November 2008, he revisited National Heart Institute and both transoesophageal echocardiography and diagnostic cardiac catheterization were done. The findings are large veno-veno collateral (i.e., opened Azygous vein from right superior vena cava into inferior vena cava), collateral from under surface of the aorta, intact interatrial septum, mild tricuspid stenosis, mild tricuspid regurgitation, and mild aortic regurgitation. Owing to the findings, he underwent his second operation on 13th November 2008 – extra-cardiac Fontan operation with Goretex, ligation of azygous vein, division of main pulmonary artery and ligation of collateral artery to main pulmonary artery. After the surgery, he was started on warfarin and continued with anti-failure medication. His INR was maintained at 2-3. He was stable and symptom free till May 2012.

Since May 2012, patient complained of generalized body swelling with left pleural effusion and ascites, and occasionally passing out loose stools without blood stain. Investigations revealed hypoalbuminemia and hypocalcemia. Further investigations were carried out and he was diagnosed as post Fontan protein losing enteropathy in view of persistent hypoalbuminemia and required regular albumin transfusion.

Urine FEME showed normal, 24 hours urine protein estimation had no significant proteinuria. Liver and renal function test, alpha-fetoprotein level, ESR, C3, C4 and ANA were normal. Stool pH was alkaline. An ultrasound of abdomen demonstrated a normal liver without focal lesion, splenomegaly, both kidneys are normal, no obvious mass and there was free fluid intraabdominally. Thoracocentesis was done and pleural fluid was investigated. The results were suggestive of transudate.

Subsequently, he was referred to National Heart Institute in 2014. Multiple slice computed tomography of thorax was done on 23rd January 2014 and demonstrated post Fontan operation with large left pleural effusion, moderate pericardial effusion and collapse consolidation of the left lower lobe, patent left Glenn shunt, multiple venous collaterals around the left Glenn, however due to poor contrast, there was enhancement in the right Glenn and Fontan conduit. The presence of veno-veno collateral cannot be ruled out. Therefore cardiac catheterization was done on 27th January 2014 and confirmed that there was no obstruction in Fontan circuit, right pulmonary artery and left pulmonary artery were in good size, pulmonary artery branches pressure were high, bilateral Glenn shunt patent, follow through pulmonary artery gram did not show

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pulmonary venous obstruction, no flow seen through the main pulmonary artery, right ventricular good size and function, and aortogram showed one large major aortic pulmonary collateral artery from descending aorta to the right lungs. In view of redo surgery for fenestration of Fontan would be technically difficult with poor prognosis, he was advised to be managed medically.

His ascites and pulmonary effusion resolved with regular albumin infusion.

Currently, he is 14 years old and is managed medically. He requires a regular albumin transfusion and calcium supplement. Pulmonary artery pressure is controlled with Sildenafil. In addition, we continue his warfarin and anti-failure medication. Otherwise, he is stable.

DISCUSSION

Fonton and Baudet in 1971 described the surgical procedure to treat the patient for tricuspid atresia. The goal was to create a circulatory system in which systemic venous blood bypasses the right ventricle to enter the pulmonary circulation. Fonton and his colleagues initially used Glens shunt in which right pulmonary artery was connected to superior venacava (SVC) and SVC-atrial junction was ligated, a valve conduit connection between the right Atrium or right atrial appendages and the left pulmonary artery was then created with aortic homograft, which resulted in blood flow from SVC to right pulmonary artery and from inferior vena cava to left pulmonary artery. After the inception, it has undergone many variations that helped to improve the patient's outcome. Creation of Fonton is a palliative in nature and produced good results in improving the poor hemodynamics and substantial improvement in mortality and morbidity in several complex congenital heart abnormalities like Tricuspid atresia, double-inlet ventricle, Pulmonary atresia with intact ventricular septum, Hypoplastic left heart syndrome. MRI is the best to evaluate postoperatively, and cardiac transplant is the only definitive treatment for failure of Fonton.

Gaynor et al., reported mortality of 2.1%, Earing et al.,² reported actual survival rate for 203 operated patients at 5 year, 10 year, 15 year, 20 year as 91%, 80%, 73%, and 69% respectively. Yeh et al.³ suggested the average longevity of Fonton operation is 30-40 yrs.

The procedure is associated with many complications like; 1. Protein losing enteropathy, 2. Reduced exercise capacity, 3. Thromboembolism, 4. Reduced somatic growth, 5. Neurodevelopmental abnormalities, 6. Complications related to surgical techniques like atrial arrhythmias and bradythmeas.

Protein losing enteropathy was observed in Fonton operated patients over the period of ten years it carries a high mortality rate and its pathophysiology has still not been Various hypothesis including early elevation of post-operative central venous pressure, low pulmonary vascular compliance and elevated serum hepatocyte growth factor were reported.

Mertens L et al.⁵ in a multicentral European review of over 3000 patients has reported an incidence of 3.9%, three fourth of the patient had edema and effusion; however other studies have reported a higher incidence. In the European cohort of Fonton patients described above reported medical treatment was ineffective in 75% with a mortality of 46%. Surgical treatment was ineffective in 81% with a mortality of 62%. Since the outcomes were not very much encouraging the patients need frequent follow-up and care such as replacement of albumin to prevent edema and palliative care to improve the quality of life.

CONCLUSION

Protein losing enteropathy is not commonly seen in a routine pediatric practice however, we may come across them. A good follow-up and involvement in the care of the patient can reduce the morbidity and can postpone the mortality. They need emotional, medical socio economic and palliative support at the district level to reduce the work load of big hospitals and for the convenience of the patients.

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