A rare case of Tetralogy of Fallot (TOF) with Potts shunt - 4th decade of survival

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Abstract:
Potts shunt is one of the palliative shunt done before the corrective surgery era for TOF. It is done by side to side anastomosis between LPA and descending thoracic aorta. Due to long term risk of increased pulmonary vascular resistance with potts shunt, it is not done as a primary modality of surgery nowadays but in olden days it has saved many cyanotic heart disease patients like TOF and tricuspid atresia. The first Blalock-Thomas-Taussig shunt surgery was performed on 15-month old Eileen Saxon on November 29, 1944 with dramatic results. The Potts shunt and the Waterston-Cooley shunt are other shunt procedures which were developed for the same purpose. For unoperated patients with TOF of all degrees of severity, 11 percent are alive at age 20 years, 6 percent at age 30 years, and 3 percent at age 40 year. In 1946 Potts demonstrated the efficiency of anastomosing the descending aorta to the left pulmonary artery in relieving cyanotic congenital heart disease, and showed it to be especially valuable in young babies in whom a Blalock shunt is not technically feasible. With potts shunt life is definitely prolonged with improved symptomatology. Here we are going to present one such case of TOF with potts shunt surgery done at the age of 4 years (1983) and surviving up to 32 years of age with minimum discomfort.

Keyword: Pott's shunt, Tetralogy of Fallot, survivor

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Introduction:
Pott's shunt is one of the palliative shunt done before the corrective surgery era for TOF. It is done by side to side anastomosis between LPA and descending thoracic aorta. Due to long term risk of increased pulmonary vascular resistance with pott's shunt, it is not done as a primary modality of surgery nowadays. But in olden days it has saved many cyanotic heart disease patients like TOF and tricuspid atresia. The first Blalock-Thomas-Taussig shunt surgery was performed on 15-month old Eileen Saxon on November 29, 1944 with dramatic results. The Potts shunt and the Waterston-Cooley shunt are other shunt procedures which were developed for the same purpose. For unoperated patients with TOF of all degrees of severity, 11 percent are alive at age 20 years, 6 percent at age 30 years, and 3 percent at age 40 year. In 1946 Potts demonstrated the efficiency of anastomosing the descending aorta to the left pulmonary artery in relieving cyanotic congenital heart disease, and showed it to be especially valuable in young babies in whom a Blalock shunt is not technically feasible. With potts shunt life is definitely prolonged with improved symptomatology. Here we are going to present one such case of TOF with potts shunt surgery done at the age of 4 years (1983) and surviving up to 32 years of age with minimum discomfort.

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Case report:
Mr. Vedaraj 32 year male presented with NYHA class II breathlessness to dept of cardiology, Madras medical college. He gave history of cyanotic spells atleast 2-3 episodes per day before he was taken up for surgery. At the age of 4 years (1983) he was done pott’s shunt surgery at the department of cardio thoracic surgery, Pediatrics, Government general hospital. Post operative period was uneven full and patient was discharged with T.digoxin 0.25 mg ½ od. Then on 1994 patient was readmitted in Stanley medical college cardiology dept for NYHA class II symptoms. Catheterization study was done on 24-06-1994 which revealed normally functioning pott’s shunt. Echo done at December 1994 also showednormally functioning pott’s shunt. Patient was apparently normal till November 2012 except for frequent history of lower respiratory tract infection. He was now admitted for NYHA class II symptoms. Patient had ESM in pulmonary area and continuous murmur due to pott’s shunt. Patient also has mild cyanosis and grade 2 clubbing. He was taken CXR PA view and lateral view, electrocardiogram, echocardiogram and cath study was done. CXR PA (Fig 1) view revealed scoliosis with concavity towards right side, no cardiomegaly, left aortic arch with unfolding of aorta. Lateral X RAY revealed right ventricular enlargement. Electrocardiogram (Fig 2) revealed sinus rhythm with rate of 75 / min , right axis deviation, and right ventricular enlargement. Echocardiogram revealed large perimembranous VSD with bidirectional shunt, 50% over riding of aorta, and normal flow across pott’s shunt (Fig 3). Cath study showed normal flow across potts shunt. Proximal right pulmonary artery 7.2 mm and proximal left pulmonary artery 7.6 mm. Descending aorta at diaphragm 19.1 mm. Mcgoon’s ratio 0.75 left pulmonary artery distal to pott’s shunt is 16.5 mm (Fig 4). Patient was discharged after LRI treatment.

Left pulmonary artery systolic pressure 120 mm Hg Discussion:
Survival of patients till 32 years with palliative pott’s shunt for TOF is very rare. Most of the patients without corrective surgery die due to CCF and infective endocarditis. But our patient has survived up to 32 years and still living with improved symptoms. Since Mcgoon’s ratio is 0.75 in this patient, he was not referred for total corrective surgery.
Conclusion:
The pott's shunt which is technically easy to perform is rarely done nowadays. But 30 years before when corrective surgery was not that famous, this procedure has remarkably improved the quality of life and prolonged the life till date.

References:

2 Evaluation of patients with tetralogy of Fallot and Potts' anastomosis. American Journal of Cardiology, 25, 259.


