



## TOTAL CORRECTION IN ADULT TETRALOGY OF FALLOT WITH CHILD B LIVER CIRRHOSIS – A ROCKY PATH AHEAD

### Cardiology

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### ABSTRACT

At the present time few cases of pulmonary valve calcification have been cited in the literature and heavily calcified right ventricular outflow tract and the papillary muscles is not yet reported. Experience with a case of pulmonary valve calcification with extensive calcification of RVOT and papillary muscles in a patient with Tetralogy of Fallot (TOF) are presented herein. Moderate to severe congenital pulmonary stenosis has been present in all<sup>1</sup>. Associated defects, especially ventricular septal defect, atrial septal defect, and TOF are common. Longstanding elevation of right ventricular pressure appears essential for the deposition of calcium<sup>5</sup>. The role of aging and healed bacterial endocarditis in the aetiology is uncertain<sup>4,5</sup>.

### KEYWORDS

Tetralogy of Fallot, RVOT calcification, Liver cirrhosis

### INTRODUCTION

Tetralogy of Fallot's is a Cono-Truncal malformation with anterior and cephalad displacement of infundibular septum, later being responsible for all the morphological features of the Tetralogy of Fallot.

Natural history depends on the severity of the Right Ventricle and the Pulmonary artery outflow obstruction. 25% die in the first year of life, 20% by the age of 3 years, 70% by the age of 10 years and 95% by 40 years. 5% of the surviving cases die due to the congestive cardiac failure secondary to cardiomyopathy. Survival depends on the amount of the collateral circulation to the lungs.

Early repair is thus advised in all the children diagnosed to have TOF.

The morphological features of the pulmonary valve are as shown in the **chart 1** (GLH Experience). 72% cases have os infundibulum in transverse plane while 15% are in coronal plane. In early ages 6-9 months there is no infundibular fibrosis but later on there is acquired fibrosis leading to atresia. There is no evidence of any calcification of the pulmonary valve or the RVOT usually except in some post-mortem reports. Herein, we report a case with severe Pulmonary valve and RVOT calcification which is a rare feature of TOF and almost reported only in post-mortem patients.

Morphology	n	%
<b>Valve Configuration</b>		
Bicuspid	93	66
Three-cusp	21	15
Vestigial	14	10
Not recorded	13	9
TOTAL	141	100
<b>Valve Lesion</b>		
Tethering alone	89	63
Commissural fusion alone	20	14
Tethering + fusion	8	6
Vestigial valve	14	10
Atretic valve (acquired)	2	1
Not recorded	8	6
TOTAL	141	100

\*Based on 141 patients undergoing repair (GLH experience, 1968-1978) of tetralogy of Fallot with pulmonary stenosis (excluding patients with subarterial ventricular septal defect, absent pulmonary valve, or congenital pulmonary atresia).

### Case Report

A 28-year-old male had presented with cyanosis since childhood, exertional dyspnoea grade II and syncopal attacks on exertion. Was a chronic alcoholic and had developed grade II liver cirrhosis as on ultra-

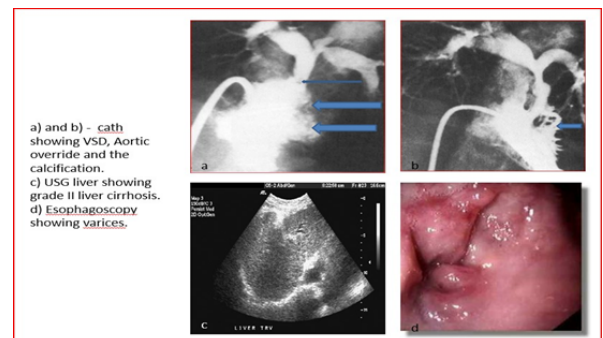
sonography, with esophageal varices grade II on upper Gastrointestinal endoscopy. The varices were sclerosed on several occasions for intermittent bleeding. Liver biopsy was advised for confirmation. Was investigated further by gastroenterologist from our hospital and diagnosed as alcoholic Child B liver cirrhosis with esophageal varices that bled intermittently. Since the past two years, the dyspnea was increased to Grade III, and developed pedal edema for which he was admitted.

On investigation his Hb was 20gm%, PCV was 55, LFT showed raised enzymes SGOT 135; SGPT 155; Bilirubin 3.2 mg% and RFT's were BUN 12 Creatinine 1; Na 143mmol/lit; K 3.9mmol/lit; Ca 1.2 mmol/lit. INR was 1.5 with control of 1.2.

ECG showed right axis deviation with sudden transition of R wave from V1 to V2.

2D-echo was suggestive of Tetralogy of Fallot with severe infundibular stenosis and calcific valvular pulmonary stenosis with PG of 70mm of Hg across RVOT. PV Annulus 15mm; MPA 15mm; RPA 10mm; LPA 10mm. SPO2 68% and mild RV dysfunction.

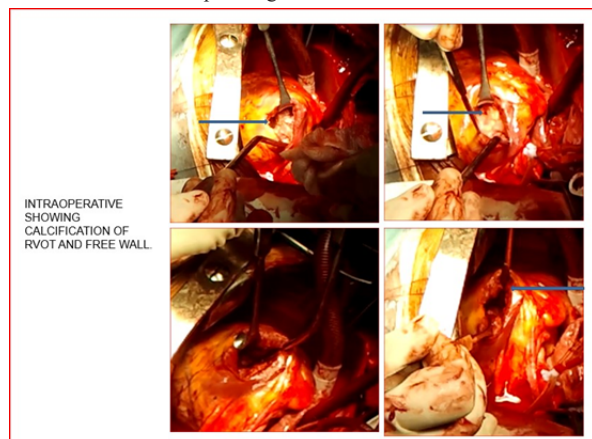
Cardiac catheterization (image 1) showed the same findings as echo; TOF with infundibular stenosis and pulmonary valve calcification with 3-4 minor MAPCA's.



With all the risks explained, was taken for elective total intra cardiac repair. Patients body surface area was 1.48 m<sup>2</sup>, height being 156cms and weight 51 kgs. The RV pressures were 120 mm of Hg with PG across PV being 100 mm of Hg. Patient was taken on full cardiopulmonary bypass with flow rate of 3.84 to 4.44 liters with aortic

cannula 20Fr and venous cannulae of 24 and 28 Fr respectively. Total CPB time was 2 hrs 55mins and cross clamp time of 2 hrs 15 mins with cold blood cardioplegia and ultrafiltration with moderate hypothermia.

Intraoperatively (image 2) found to have malaligned VSD of 30mm size, margins being free of calcification, with severe RVOT calcification extending to the papillary muscle and the free RVOT wall. Pulmonary valve anatomy could not be differentiated due to severe calcification. VSD was closed using dacron patch with interrupted mattress sutures of prolene 5-0 13mm needle, and trans annular pericardial patch augmentation of RVOT was done with hegar dilator of 18mm as the guide. Valved conduit was not used in view of severe calcification involving the free wall and RV cavity mostly appeared small, and suturing was difficult. The RV pressures were 50mm Hg even after transannular patching.



Patient was shifted to ICU with HR 120/min; BP 104/60 mm of Hg; CVP 7-8 cm of H<sub>2</sub>O and ABG showed acidosis with PH of 7.34, soda bicarbonate deficit of -7.5 and was corrected, PO<sub>2</sub> 253, PCO<sub>2</sub> 31.7 saturation of 100% on FiO<sub>2</sub> of 60% with inotropic support of dopamine 7 micro-gm/kg/min and adrenaline 0.1 micro-gm/kg/min. Patient was extubated on 1<sup>st</sup> post-operative day as his vitals were maintained and post extubation ABG showed no acidosis with PH of 7.41, PO<sub>2</sub> 251, PCO<sub>2</sub> 36.3 and saturation of 100%.

On the 2<sup>nd</sup> post-operative day, he developed acute pulmonary oedema and saturation of 70% and was ventilated noninvasively. Also had GI bleeding for which GI medicine opinion was taken and somatostatin infusion drip was started without altering the inotropic support and treated for pulmonary oedema. Was intubated on the same day for dropping saturation and aggravated pulmonary oedema. ABG after intubation showed severe metabolic acidosis with respiratory compensation PH 7.294, PO<sub>2</sub> of 71, PCO<sub>2</sub> 21.9, saturation 89%.

On post-operative day 3, his HR was 98/min, BP 70/48 mm of Hg, CVP 10-12 cm of H<sub>2</sub>O and frank frothy secretion from the endotracheal tube. Urine output dropped to 10-15 ml/hr as compared to earlier of 50 ml/hr and inotropic support was increased further dopamine 15 micro-gm/kg/min, adrenaline 0.2 micro-gm/kg/min with addition of isoprenaline 0.5 micro-gm/kg/min.

On post-operative day 4; Hb was 11 gm%, total count were 29000/cu mm, platelet count < 20000/cu mm, total protein 3.6 gm% and albumin 1.7 gm% ABG showed severe metabolic alkalosis with PH 7.550, PO<sub>2</sub> 60.5, PCO<sub>2</sub> 31.5, HCO<sub>3</sub> 25.7 saturation 80% on volume control mode. Urine out-put was nil with K<sup>+</sup> > 5.6mmol/lit for which K<sup>+</sup> lowering enema was started.

Portable 2D-echo showed patch in situ, persistent RVOT PG of 45mm of Hg, free PR, severe TR, severe RV dysfunction, no evidence of any vegetations, calcification in RVOT present. On day 5, early morning patient arrested, was resuscitated but failed to revive and was declared death due to cardio-respiratory failure with acute renal failure. Partial post-mortem was advised and found to have the features as mentioned, with patch in situ. There was no evidence of any healed bacterial vegetations.

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