

## Acquired Fallot's Physiology: The Nature that Kills, the Same Natures also Heals

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### Abstract

Natural history of some the congenital diseases are quite interesting. Thrombosis of patent ductus arteriosus (PDA) in intrauterine life is fatal to the foetus without ventricular septal defect or large arterial septal defect but thrombosis of PDA after birth is a blessing. Similarly, stenosis of aortopulmonary collaterals in their proximal part protect the pulmonary bed from over circulation in the cases of cyanotic congenital heart diseases. In the present case report, a boy with a large peri-membranous ventricular septal defect (PMVSD) with mild valvular pulmonary stenosis was followed from birth. The boy was pink at birth. However, by his 10<sup>th</sup> birthday child became cyanosed. The present echocardiography revealed almost spontaneously closed PMVSD with small residual right to left shunt because of the Gasulisation of the right ventricular outflow tract. The appropriate corollary in the context is the nature that kills (acquired right ventricular outflow tract obstruction), the same nature that heals (spontaneous closure of large perimembranous ventricular defect).

**Keywords:** *Large Ventricular Septal Defect; Gasulisation; Spontaneous Closure of Ventricular Septal Defect; Natural History*

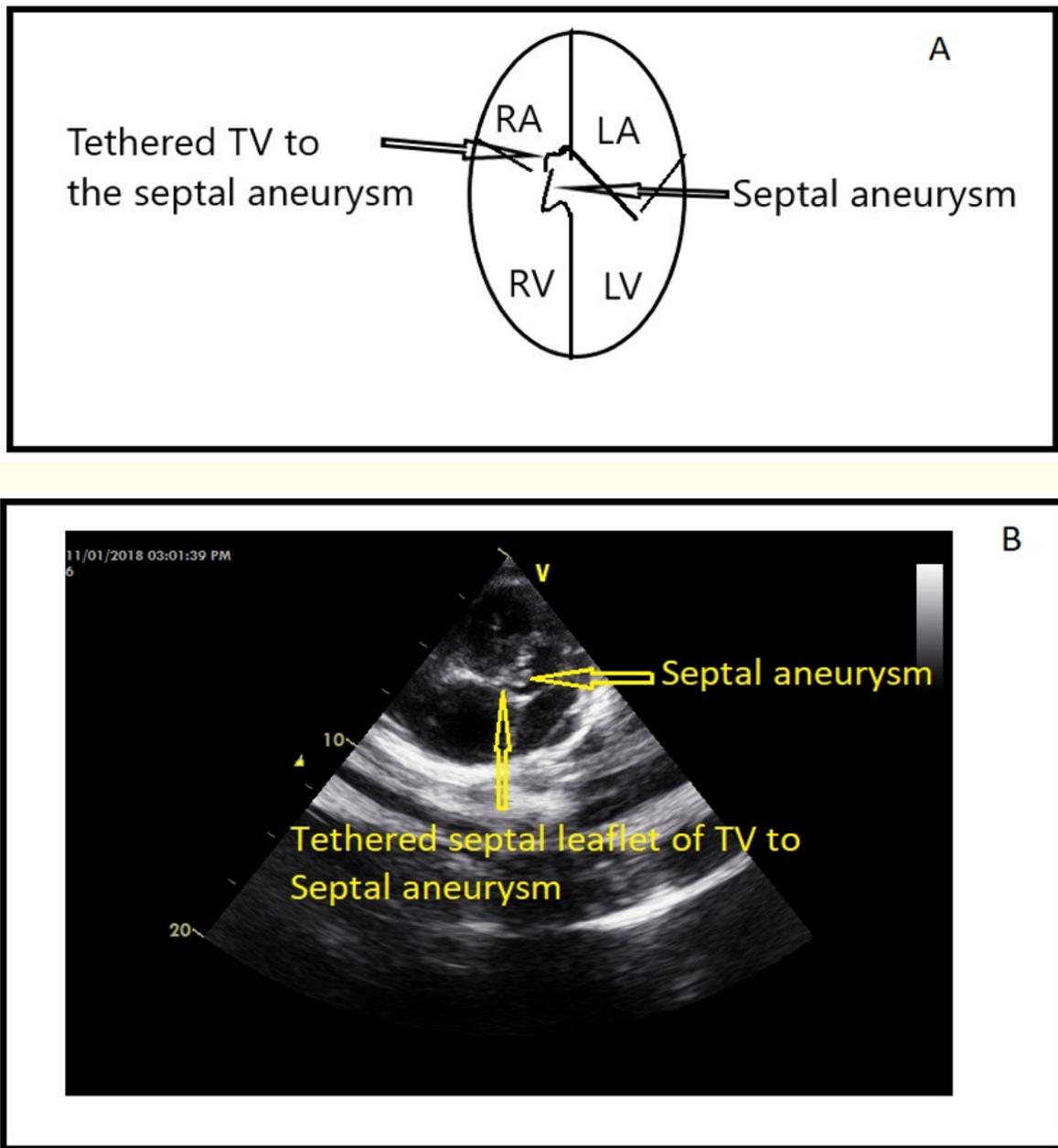
### Introduction

Tetralogy of Fallot is a congenital cardiac defect. It is due to the anterior and cephalad malalignment of the conal septum. However, a subset of the patient with a large ventricular septal defect with some amount of malalignment of their conal septum develop acquired infundibular obstruction during the follow up. It may be due to an unusual response to significant hemodynamic stress to the right ventricular outflow tract (RVOT). This phenomenon is called Gasulisation. This later situation may be confused with TOF if the case lacks continuous follow up of the natural history. This interesting case illustrates a rare case wherein a male child who had a large perimembranous VSD at birth, became cyanosed by 10<sup>th</sup> birthday due to spontaneous closure and Gasulisation of the ventricular septal defect in the course of time.

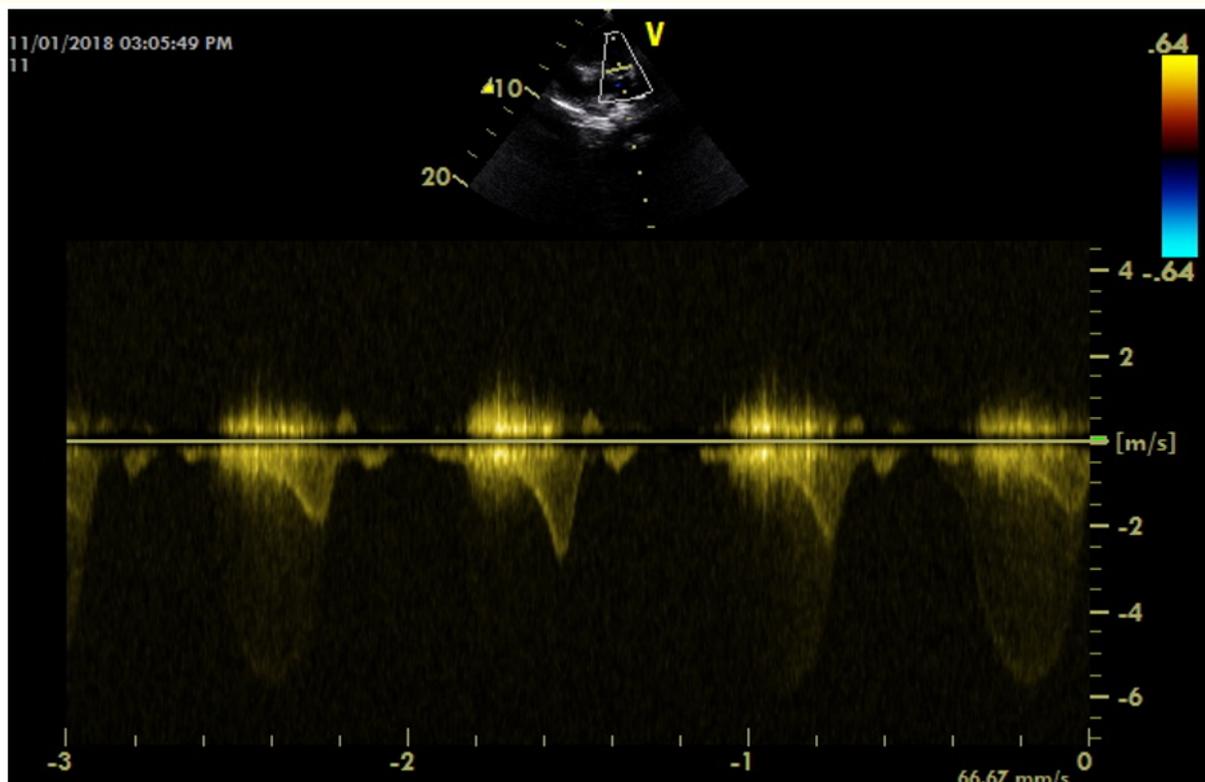
### Case Report

A 10-yrs old boy presented with breathlessness on exertion for last 2 years. He was a known case of a ventricular septal defect with mild pulmonary valve obstruction by birth and having the features of congestive heart failure in the early part of the childhood. The parents were not ready to accept early intracardiac repair. At present, he had central cyanosis in room air with SPO<sub>2</sub> of 82%. The pandigital clubbing was grade II. The "a" wave of jugular vein pulsation was prominent. The right arm sitting blood pressure was 97/68 mmHg. Pre-cordial examination showed grade V/VI ejection systolic murmur in the left parasternal border with selective propagation to towards left shoulder. Echocardiographic study was quite unusual. There was a large peri-membranous ventricular septal defect which was trying to be closed by mutual contribution from the crest of interventricular septum in the form of septal aneurysm and septal leaflet of tricuspid valve resulting in significant restricted ventricular septal defect (Figure 1). The tethering of septal leaflet of tricuspid valve to the aneu-

rysmal interventricular septum was causing mild to moderate tricuspid valve regurgitation. Right ventricular pressure by tricuspid valve regurgitation jet was 130 mmHg. There was no overriding of the aorta. The infundibular stenosis and the doming of pulmonary valve were obvious in the 2D echo. There was significant right ventricular outflow tract obstruction in the form of infundibular and valvular pulmonary stenosis with a gradient of 146 mm Hg (Figure 2). The shunt through the residual ventricular septal defect was right to right with a gradient of 40 mmHg between the right ventricle and left ventricle. The right ventricle (RV), right atrium (RA) and inferior vena cava were dilated.



**Figure 1:** (A)- The cartoon image shows the mechanism of closure of ventricular septal defect; (B)- 2D echo in parasternal short axis view is showing almost the complete closure of the large ventricular septal defect by mutual contribution from the septal aneurysm and tethered septal leaflet of the tricuspid valve (yellow coloured arrow marked).



**Figure 2:** There was significant right ventricular outflow tract gradient as measured using continuous wave Doppler in parasternal short axis view.

## Discussion

The natural history of certain congenital heart diseases is quite interesting and unique. Most of the congenital heart diseases when followed sincerely, deteriorate in course of natural history. However, there are unique instances of the cure of congenital cardiac defects with age. Therefore, the beautiful corollary is nature that kills, the same nature also heals. The spontaneous relief of pulmonary valvular obstruction by infective endocarditis, spontaneous closure of peri-membranous and muscular ventricular septal defect (VSD), spontaneous closure of patent ductus arteriosus (PDA) and small atrial septal defect, the survival of patient by a single coronary artery when there is congenital coronary atresia of one of the coronary arteries and significant collateral formation in cases of coarctation and pulmonary atresia etc. The natural history of congenital cardiac defect depends upon several factors like the age, gender, the size and site of the defect, the types of defect and the length of follow-up period [1]. The spontaneous closure of small VSD occurs up to the extent of 60 to 90% of cases, moderate size VSD can close spontaneously up to 10% of cases while the spontaneous closure of large VSD rarely occurs. The Gasulisation of VSD is seen in nearly 3 to 7% of cases of moderate to large size VSD [2]. However, simultaneous Gasulisation and spontaneous closure of large peri-membranous ventricular defect are very rare. The natural history of a large VSD which undergoes Gasulisation may follow the sequence of a pink baby by birth, features of congestive heart failure, a period relatively asymptomatic due to Gasulisation and finally cyanosis if the RVOT obstruction is very significant resulting in the reversal of ventricular level shunt [3,4]. In a very rare situation, these patients may become pink again because of complete closure of VSD. In my case, the large peri-membranous VSD has undergone significant spontaneous closure along with simultaneous Gasulisation which is quite rare. Echocardiography as a single imaging method is sufficient to document all these changes over time if the patient is followed sincerely as in this which tracks the morphological changes of the natural history of the ventricular septal defect. The management of this case is quite interesting. In the early childhood, when the

ventricular septal defect is large, intracardiac repair is a must. If the Gasulisation is very rapid as in my case, the follow-up results in Fallot's physiology. The later situation also requires intracardiac repair. However, if there is the simultaneous spontaneous closure of VSD is very rapid as in our case, it confuses the treating physician whether to wait and watch or go with immediate surgery.

### Conclusion

The spontaneous closure of large peri-membranous ventricular defect is unusual and so also simultaneous Gasulisation of the right ventricular outflow tract. However, getting either in one case history is quite rare resulting in acquired Fallot's physiology is an unusual experience as in this case report.

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### Conflict of Interest

None.

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