



## Adult Nigerian with Untreated Pentalogy of Fallot: A Case Report

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**Received date:** 13 January 2020; **Accepted date:** 19 February 2020; **Published date:** 27 February 2020

**Citation:** Akinlade OM, Ogunmodede JA, Adeyemi OA, Yusuf WO, Awodun RO, Ayoola YA, Yusuf IA, Kolo PM. Adult Nigerian with Untreated Pentalogy of Fallot: A Case Report. *Asp Biomed Clin Case Rep.* 2020 Feb 27;3(1):62-66.

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### Abstract

Pentalogy of Fallot is a congenital cardiac defect comprising infundibular stenosis of the pulmonary artery, overriding aorta, ventricular septal defect, right ventricular hypertrophy and atrial septal defect. It is one of the most common cyanotic congenital heart defects with detrimental consequences if left untreated. We present the case of a 25-year-old female artisan and stroke survivor who was admitted with episodes of recurrent dizziness. She was diagnosed with the pentalogy of Fallot after echocardiography and awaiting cardiac repair surgery.

### Keywords

Infundibular Stenosis; Pentalogy of Fallot; Ventricular Septal Defects; Overriding Aorta

### Introduction

Pentalogy of Fallot is used to describe the occurrence of the tetralogy of Fallot (TOF) co-existing with atrial septal defects. TOF is described as one of the most common congenital heart disorders comprising: ventricular septal defect (VSD), right ventricular (RV) outflow tract obstruction (RVOTO), over-riding aorta and RV hypertrophy. The mortality rate in untreated patients is still very high. The description of TOF has been ascribed to the work of Stensen (first described it in 1672) and Fallot (first described the clinical and complete pathologic features of the defects). Helen Taussig and Alfred Blalock collaborated to work out the first type of palliation for these infants. Blalock in 1944 operated on an infant with tetralogy of Fallot and created the

first Blalock-Taussig shunt between the subclavian artery and the pulmonary artery. Several advances in the diagnosis and management have led to increasing survival outcomes for TOF patients [1].

If left untreated, TOF survivors are at risk of paradoxical emboli leading to stroke, pulmonary embolus, and subacute bacterial endocarditis. The mortality rate is said to be highest in the first year and then remains constant until the second decade. Without surgery, mortality rates gradually increase from 30% at age 2 years to 50% by age 6 years. No more than 20% of patients can be expected to reach the age of 10 years, and less than 5-10% of patients are alive by the end of their second decade. However, cases of survival of patients into their 80s have been

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reported. Due to advances in surgical techniques, a 40% reduction in deaths associated with tetralogy of Fallot was noted from 1979 to 2005.

We report a 25-year-old artisan with untreated TOF in north-central Nigeria.

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A 25-year-old artisan with a history of cyanosis and recurrent squatting since childhood presented with complaints of recurrent dizziness of 7 months duration. Dizziness was worse while sitting still or standing, had some episodes of syncopal attack which lasted about 2 minutes, No seizures. She had no history of palpitation, chest pain, dyspnea, orthopnea or paroxysmal nocturnal dyspnea. However, she had a history of progressive easy fatigability on mild exertion. No history of leg swelling. She had a prior history of stroke 11 months earlier with left hemiparesis from which she recovered completely over a period of two months.

General examination revealed an asthenic plethoric young lady, centrally cyanosed with grade 3 digital clubbing. Pulse was regular at a rate of 90 beats per minute. Her blood pressure was 134/80 mmHg. The precordium was hyperactive and the apex was located at 6<sup>th</sup> left intercostal space, lateral to midclavicular line with palpable systolic thrills over the left sternal

border and pulmonary area. A loud pulmonic component of the second heart sound and a pansystolic murmur over the left parasternal area and an ejection systolic murmur over the pulmonary area were heard. The lungs were clear. Abdomen showed no abnormality.

The chest radiograph showed a normal-sized boot-shaped heart without pulmonary oligemia. Electrocardiogram revealed sinus rhythm, normal PR interval, right QRS axis deviation, right atrial enlargement, right ventricular hypertrophy. The echocardiography revealed a non-restrictive ventricular septal defect with a large aorta overriding the muscular interventricular septum and atrial septal defect (Ostium Secundum) (**Fig-1**, **Fig-2** and **Fig-3**). She had eccentric LVH with impaired left ventricular systolic function (LVEF = 41%). The right ventricular EF was 62%, with right ventricular hypertrophy and grade 1 right ventricular diastolic dysfunction. There was no aortic or mitral regurgitation. Peak systolic pulmonary pressure was 52mmHg.

The packed cell volume was 72% and the blood film showed relative polycythemia. A final diagnosis of TOF with ASD (Pentalogy of Fallot) was made and the patient was placed on beta-blockers and clopidogrel. She has also had serial venesection to reduce her packed cell volume with a slight improvement in

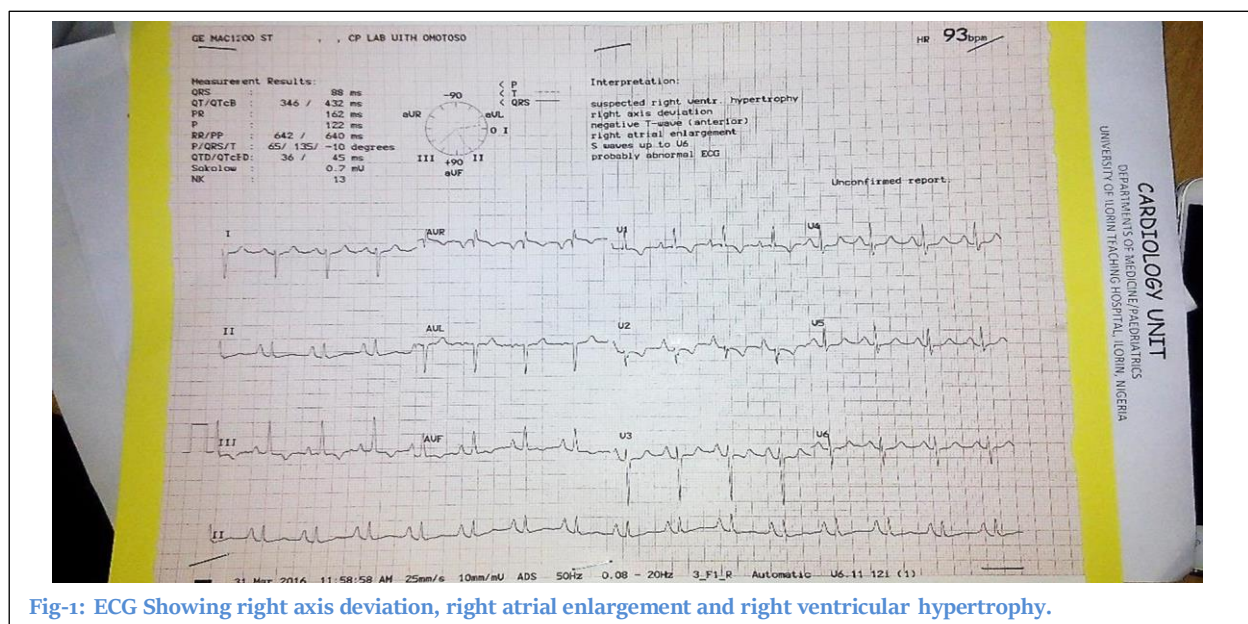


Fig-1: ECG Showing right axis deviation, right atrial enlargement and right ventricular hypertrophy.

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Fig-2: 2-D echo showing the ventricular septal defect and the huge overriding aorta (Parasternal long axis view)



Fig-3: Colour flow study showing flow across atrial septal defect (Subcostal view)

symptoms. She is awaiting total intra-cardiac repair.

### Discussion

Tetralogy of Fallot (TOF) is one of the cyanotic congenital heart disease associated with inadequate blood flow to the lungs for oxygenation (right-to-left shunt). Cyanosis is often first observed shortly after birth, thereby attracting early medical attention. Occasionally, a few children also have an atrial septal

defect (ASD), which makes up the pentad of Fallot as seen in this case report. The basic pathology of TOF is due to the underdevelopment of the RV infundibulum, which results in an anterior-leftward malalignment of the infundibular septum [2,3]. This malalignment determines the degree of right ventricular outflow tract obstruction (RVOTO).

Three main reasons have been described to account

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for the longevity in survivors with un-operated TOF. Firstly, a hypoplastic pulmonary artery with the slow development of sub-pulmonary obstruction [4-6]. A second common feature is that of left ventricular hypertrophy; presumably, this acts by delaying of shunting from the right to left ventricle [7,8]. Left ventricular hypertrophy (LVH) may be a late development in the natural history of TOF and any beneficial effect may not be seen until adult life. The index case had echocardiographic evidence of LVH (LVMI =128), this must have accounted for the reduced cyanotic spells she had. The last possible explanation has been extra-cardiac shunts including patent ductus arteriosus [9] reported in one of the survivors who died at age 77 or systemic to pulmonary artery shunting via aortopulmonary collaterals [7,10]. The co-existence of ASD in this patient might have equally contributed to her survival to date.

TOF surgeries are usually done around 6-12 months of life, which entail initial shunt procedures, thereafter a corrective surgery is done [2,11-13]. Though Mayo series reported 52 TOF patients, aged >40 years who had total correction with a 30-day mortality of 6% with an improved overall functional class [14]. Though our patient is yet to have surgery, current techniques for total surgical repair greatly improve the hemodynamic functions of the heart of TOF patients. Efforts are being made to ensure that our patients benefit from total intracardiac repair.

For unoperated adults, surgical repair is still recommended because the results are gratifying and the operative risk is comparable to that in pediatric series, provided that no serious coexisting morbidity is present [13-19].

## Conclusion

There is a need for increased suspicion of TOF even among adult patients with a recurrent history of dizziness and or stroke in the young. Surgical correction can improve outcomes among patients. Concerted effort and multidisciplinary collaboration is needed in patient management to identify those that will benefit from corrective surgery to prevent late complications and poor quality of life.

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