

# Heart failure complicating tetralogy of Fallot

\*O. O. Ogunkunle, S. I. Omokhodion, R. E. Oladokun and A. A. Odutola

Department Of Paediatrics,  
University College Hospital, Ibadan, Nigeria.

## Summary

**Background:** Heart failure is considered to be an unusual complication of uncorrected tetralogy of Fallot.

**Patients and method:** Three adolescents with tetralogy of Fallot, presenting in congestive cardiac failure are presented. Two died. Myocardial infarction was found in the only patient that underwent autopsy, and is thought to be an important aetiological factor in the development of the heart failure in the patients.

**Conclusion:** The cases presented illustrate the fact that heart failure complicating tetralogy of Fallot may not be as uncommon as was previously thought to be the case, especially in older children. There is a need for early diagnosis and expeditious institution of appropriate management before such complications occur.

**Keywords:** Tetralogy of Fallot, Heart failure, Myocardial infarction

## Résumé

L'arrêt cardiaque est considéré d'être une complication peu commune de la tetrélogie non corrigée et Fallot. Trois adolescents atteints de la tetrélogie de Fallot, avec l'arrêt cardiaque congestif sont l'objet de cet étude. Deux étaient mort. L'infarctus du myocarde était trouvé chez le seule patient qui a subi l'autopsie, et il est envisagé d'être un facteur étiologique très considérable dans le développement d'arrêt cardiaque chez des patients.

Les cas ici présentes montrent le fait que la complication de la tetrélogie de Fallot d'arrêt cardiaque pourrait ne pas être peu commun comme on l'avait envisagé auparavant, surtout chez des enfants plus âgés. C'est nécessaire d'avoir un diagnostic précoce et la prise en charge rapide et prendre les mesures indiquées avant que telle complication eut lieu.

## Introduction

Tetralogy of Fallot (TOF), a cyanotic congenital heart disease comprising of right ventricular outflow tract obstruction (RVOTO), right ventricular hypertrophy (RVH), ventricular septal defect (VSD) and overriding of the aorta, constitutes about 10% of congenital heart diseases seen in Nigeria.<sup>1</sup> Congestive heart failure is not a usual feature of patients with this condition, but may occur in infants with acyanotic or "pink" TOF before the development of severe RVOTO.<sup>2</sup>

In countries where advanced medical facilities are available, uncorrected TOF is increasingly less common in older children and adults, because most patients undergo total correction during infancy or early childhood. This report illustrates three cases of TOF presenting with heart failure in adolescence. The unusual association seen in 3 patients within the space of 18 months prompted the report.

## Case 1

O. O. a 13-year-old boy was admitted into the University College Hospital (UCH) Ibadan in April 2001 with a 3-month history of facial swelling, easy fatigability and breathlessness. He was first seen at the age of 11 years in another teaching hospital, where a diagnosis of TOF had been made. Drug history before UCH presentation included digoxin and frusemide.

Physical examination revealed a small-for-age boy (weight - 26kg = 60% of expected) with central cyanosis, digital clubbing, facial and bilateral pedal oedema. He was dyspnoeic with a respiratory rate of 44/min but the chest was clinically clear. The jugular venous pressure (JVP) was raised to the angle of the jaw. The pulses were of normal volume with a rate of 112/min; BP was 105/65 mmHg. The first heart sound was normal, the second heart sound was single and there was a grade 2/6 systolic murmur loudest at the left upper sternal edge. There was a tender hepatomegaly of 6cm; the spleen was not palpably enlarged and there was no demonstrable ascites.



Fig. 1 Chest X-ray of Case 1, showing an enlarged, globular cardiac silhouette

Chest X-ray (figure 1) revealed an enlarged, globular cardiac silhouette with reduced pulmonary vascular markings. Electrocardiogram demonstrated right atrial enlargement, biventricular hypertrophy, right axis deviation and non-specific S-T segment changes. 2-Dimensional (2-D) echocardiography confirmed Fallot's anatomy with reduced left ventricular systolic function and a dyskinetic wall. The full blood count showed packed cell volume (PCV) - 64% (normal = 37 - 50%) white cell count (WBC) - 6,000/mm<sup>3</sup> (normal = 3,000-9,000/mm<sup>3</sup>) Platelets - 154,000/mm<sup>3</sup> (normal = 100,000-300,000/mm<sup>3</sup>). Urinalysis was normal.

Based on the above findings, a diagnosis of TOF in heart failure, with possible myocardial infarction (MI) was

\* Correspondence

made, and he was placed on antifailure regime. Subsequent cardiac catheterisation confirmed Fallot's anatomy, but also revealed multiple aorto-pulmonary collaterals in the lungs, which together with the poor left ventricular function contraindicated the desirable corrective surgery. The patient is currently being followed up on conservative management in the Children's Outpatient Department, but attending irregularly.

### Case 2

T. A. a 15-year-old student, was admitted into the UCH, Ibadan in June 2001 with a 3-week history of cough, breathlessness, fever and generalized body swelling. He had been seen first at the age of 4 years, at which time a diagnosis of TOF had been confirmed by 2-D echocardiography. He however defaulted from UCH thereafter, but was allegedly being followed up in a private medical centre. Drug history during the present illness before presentation at UCH included digoxin, frusemide, Ampiclox, and aspirin.

The essential physical findings were a small-for-age boy (wt-33kg - 60% of expected for age) with central cyanosis, digital clubbing, periorbital and bilateral pedal oedema up to the knees and a temperature of 38°C. Oral hygiene was poor. The respiratory rate was 40/min with dyspnoea and orthopnoea, but the chest was clinically clear. The pulses were of small volume but regular, with a rate of 108/min. The blood pressure was  $100/70$  mmHg. The JVP was raised to the angle of the jaw. The first heart sound was normal, the second heart sound was single and there was a grade  $3/6$  systolic murmur loudest at the left upper sternal edge. The liver was palpable 9cm below the costal margin and tender; the spleen

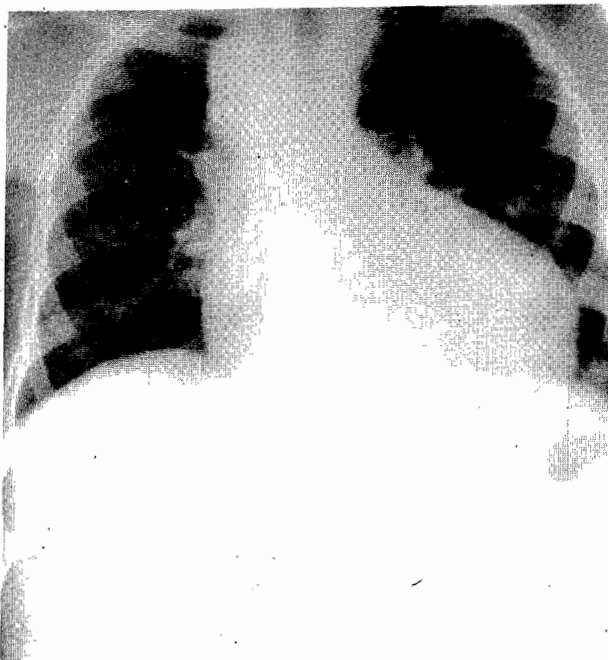


Fig. 2 Chest X-ray of Case 2, showing a boot shaped heart, right aided aortic arch, and reduced pulmonary vasculature

was palpable 3cm below the left costal margin and there was demonstrable ascites.

The chest radiograph (Figure 2) showed the characteristic boot shaped cardiac silhouette with a right-sided aortic arch and decreased pulmonary vascular markings. The electrocardiogram showed sinus rhythm, right axis deviation, RVH and non-specific T wave abnormality. Echocardiography, apart from the Fallot's anatomy did not reveal any vegetation. There was however, poor left ventricular systolic function with marked dyskinesia and paradoxical septal motion.

The full blood count showed a PCV of 69%, WBC 4,600/mm<sup>3</sup> with normal differentials and platelets 133,000/mm<sup>3</sup>. Serial blood culture yielded coagulase negative *staphylococcus*, sensitive to Cloxacillin, Gentamicin, Augmentin and Amoxicillin. Blood film for malaria parasites was negative. Serum electrolytes and urea (E/U) and creatinine (Cr) were normal. Urinalysis revealed blood 3+, protein 2+. Urine microscopy and culture showed 18 - 20 RBC/high power field, but yielded no growth, Haemoglobin genotype was A.

A diagnosis of TOF with heart failure secondary to infective endocarditis (IE) was made, following which he was commenced on digoxin, hydrochlorothiazide, potassium supplements, crystalline penicillin, cloxacillin and gentamicin. On the 7<sup>th</sup> day on admission while awaiting erythropheresis (on account of a rise in PCV to 82%), he suffered an episode of generalised tonic-clonic seizures and subsequently lapsed into coma. An impression of cerebrovascular accident was made. A brain C. T. scan was suggestive of cerebral oedema with no focal lesions. Repeated erythropheresis lowered the PCV to 68%. His general condition however continued to deteriorate until he succumbed to the illness after 20 days on admission.

Post mortem examination confirmed the diagnosis of TOF, with left ventricular MI and apical mural thrombus. The brain did not show any evidence of cerebral oedema, haemorrhage or infarction.

### Case 3

LA, a 16-year-old boy, was admitted to UCH in November 2002 with a history of intermittent generalised body swelling associated with cough of 3 months' duration, and chest pain of 2 weeks' duration. The body swelling and chest pain had worsened in the week preceding presentation and was associated with reduced urine output and worsening breathlessness. A diagnosis of TOF had been made in another tertiary institution at the age of 6 years, but was only confirmed by 2-D echocardiography at UCH at the age of 12 years (his first presentation here) following which he had attended follow-up very infrequently.

Physical examination revealed an ill-looking boy weighing 55kg. There was facial puffiness, pedal oedema up to the knees, moderate cyanosis and digital clubbing. Axillary temperature was 35.1°C. He was coughing and wheezing, with a respiratory rate of 22/min. There was a few crepitations in the left mid- and basal zones of the lung fields posteriorly.

Cardiovascular examination revealed a heart rate of 96/min with fair volume pulses, a normal first heart sound, a single second heart sound, with a grade  $3/6$  ejection systolic murmur loudest at the left lower sternal edge. Blood pressure was  $90/60$  mm Hg. The liver was palpable 6cm below the right costal margin and tender; the spleen was not palpable and



Fig. 3 Chest X-ray of Case 3 showing an enlarged almost globular cardiac silhouette and pulmonary oedema.

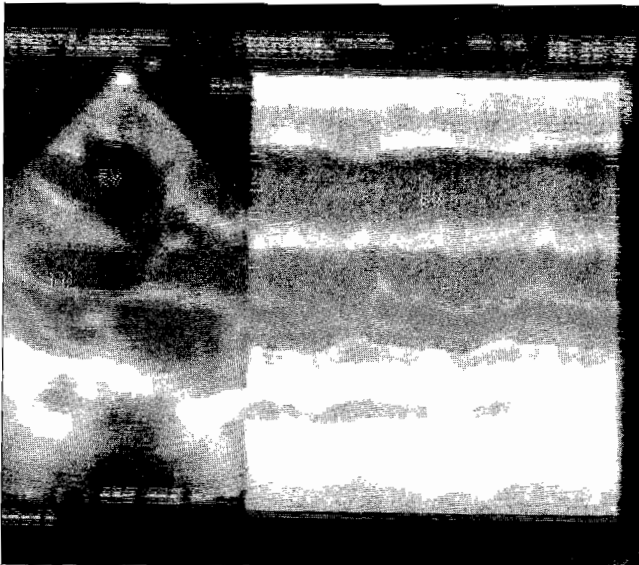


Fig. 4 2-D and M-Mode echocardiograph of Case 3, showing a dilated, poorly contracting left ventricle

there was demonstrable ascites.

Chest radiograph (Figure 3) showed cardiomegaly with an almost globular configuration of the heart, as well as pulmonary oedema. ECG showed sinus tachycardia, right axis deviation and RVH. T waves were inverted in Lead aVF.

2 - D echocardiography revealed Fallot's anatomy with a dilated, poorly contracting LV (Figure 4), a thick walled, poorly contracting RV and a small pericardial effusion. The PCV was 61%. Urinalysis showed proteinuria 3+ but no blood; serum E/U and Cr were normal.

He was placed on diuretics and made initial improvement, but died suddenly, 2 days after admission. Post mortem examination was not possible for technical reasons.

### Discussion

The three cases illustrated present similar clinical features. A correct diagnosis of TOF had been made in each case a number of years before their presentation in heart failure. Subsequent follow-up was however unsatisfactory. Severe cardiac decompensation, with extremely poor myocardial function co-existing with chronic hypoxia and polycythaemia, were common to all three.

The cause of the heart failure may be multifactorial. Severe polycythaemia predisposes to thrombo-embolic events, including coronary occlusion and may lead to myocardial ischaemia or infarction, which might precipitate heart failure.

Watson et al<sup>3</sup> reported the case of a boy who was first diagnosed as having TOF when he presented at the age of 3 years with hypoxia and heart failure. The heart failure was attributed to cardiomyopathy associated with severe hypoxia, and improved following an aorto-pulmonary shunt. Choudhary et al<sup>4</sup> reported a similar experience with a 4-year-old boy, postulating severe hypoxia to be responsible for severe myocardial dysfunction. The theory sounds attractive in the cases illustrated. However, given the fact that many of our patients with TOF do present with severe cyanosis and polycythaemia, relatively late, one would have expected heart failure to be encountered much more frequently than is the case. It is therefore reasonable to assume that in the 3 cases presented, there may have been some precipitating event, such as MI, that tipped them into heart failure. Notably, the ECGs of all 3 patients showed some abnormality in the ST segments or T waves, and could have alerted one to the possibility of underlying myocardial pathology. MI was found at autopsy in Case 2 but could not be confirmed in the other 2.

IE may cause heart failure in patients with TOF.<sup>5</sup> It is also a known cause of MI. The diagnosis of IE requires a high index of suspicion, but Case 2 was a likely candidate, with 2 predisposing factors (congenital heart disease and poor oral hygiene) being present. The hallmark of diagnosis ante-mortem is a positive blood culture.<sup>6</sup> Isolation of coagulase-negative *staphylococcus* (normally a contaminant and not commonly a cause of IE) could not be ignored, since the patient was symptomatic. Failure to demonstrate vegetations on 2-D echocardiography was not enough evidence against the diagnosis,<sup>5,6</sup> however, vegetations were not found at autopsy.

The 3 cases posed very challenging management problems. Digoxin combined with diuretics is the usual choice of treatment of heart failure in children with cardiac defects in our environment. Experience with the newer options, e.g. ACE inhibitors is not widespread. Concerns have been raised

about the safety of digoxin in the setting of severe hypoxia, extremely poor myocardial function and MI.<sup>7</sup> With hindsight, treatment with ACE inhibitors may have been more appropriate.<sup>8</sup> Diuretic therapy, needed to relieve pulmonary and systemic congestion or oedema, would only worsen polycythaemia, (as was the experience in Case 2) and increase the risk of thrombo-embolic events. The importance of monitoring the PCV and hydration status in such a patient cannot be overemphasised. Without gainsaying, any form of surgical intervention (apart from cardiac transplant\_ undertaken at this stage, would most certainly have had a poor outcome, as the already compromised ventricles would not have been able to sustain adequate cardiac output following surgery.

Definitive treatment of TOF consists of relief of the RVOTO and patch closure of the VSD, following which, patients are able to lead unrestricted lives. Where this is not possible, a palliative systemic-to-pulmonary artery shunt procedure is carried out with temporary relief of the symptoms.

Clinical outcome of children with cyanotic congenital heart disease after cardiac surgery is known to be inferior to that of those with acyanotic lesions, and is related to the degree of cyanosis before surgery.<sup>9</sup> Some patients with TOF do survive into adulthood without surgical intervention. Although correction in adulthood is reported to show excellent results with normal life expectancy for patients,<sup>10</sup> early repair is surely the preferred option,<sup>11,12</sup> since mortality rate is high in childhood, and quality of life in unoperated patients is severely compromised. The 3 cases here presented illustrate the point poignantly. Had surgery been performed promptly at the time of their first presentation at a tertiary hospital, the outcome is likely to have been different.

It is common for cases of congenital heart disease in developing countries to present late, by which time complications would have occurred.<sup>13</sup> Ignorance concerning the disease and lack of competent personnel and diagnostic facilities may contribute to delay in the diagnosis and institution of appropriate management.

Fortunately, there are now treatment outlets for total corrective cardiac surgery abroad in collaboration with Non-Governmental Organisation (NGO)s in a number of countries, including Nigeria.<sup>14</sup> With greater awareness of this fact, it is hoped that patients suspected to have cardiac defects will be referred early to tertiary centres, where they can be appropriately evaluated and managed so that corrective surgery can be performed at the earliest possible opportunity before complications occur.

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