

CASE REPORTS

Postoperative Tetralogy of Fallot with Concomitant Atrial Fibrillation and Ventricular Tachycardia

S Williams-Phillips

ABSTRACT

Postoperative tetralogy of Fallot patients, since the first corrective surgical correction in the 1960s, have borne a burden of postoperation arrhythmias and sudden death. This is confirmed to be secondary to supraventricular arrhythmias and predominantly ventricular arrhythmias. It is rare to have both types of arrhythmias in the same patient, which the index case developed. Attempts to stratify patients in groups to predict which patients are susceptible to sudden death and arrhythmias have not been unanimous, despite multiple retrospective and prospective analysis of this group of patients.

Keywords: Arrhythmias, atrial fibrillation, infundibular resection, sudden death, tetralogy of fallot, ventricular tachycardia

Tetralogía de Fallot Postoperatoria con Fibrilación Atrial Concomitante y Taquicardia Ventricular

S Williams-Phillips

RESUMEN

Los pacientes de tetralogía de Fallot postoperatoria, desde la primera corrección quirúrgica en los años sesenta, ha soportado una pesada carga de arritmias postoperatorias y muerte súbita. Esto se confirma como secundario a las arritmias supraventriculares y las arritmias predominantemente ventriculares. Es raro encontrar ambos tipos de arritmias en el mismo paciente, como ocurrió en el caso del paciente índice. Los esfuerzos por estratificar a los pacientes en grupos para predecir qué pacientes son susceptibles a la muerte súbita y las arritmias no han sido unánimes, a pesar de los múltiples análisis prospectivos y retrospectivos de este grupo de pacientes.

Palabras claves: Arritmias, fibrilación atrial, resección infundibular, muerte súbita, tetralogía de Fallot, taquicardia ventricular

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INTRODUCTION

“La Maladie bleu” was documented by Louis Fallot in 1888, but was first described by Nicholas Steno in 1673. Fallot described four morphologic features, all from post mortem cases: overriding aorta, large ventricular septal defect, pulmonary

stenosis and right ventricular hypertrophy. Lev and Eckner, in 1964, documented that not all cases had those four features. The most consistent features were ventricular septal defect and pulmonary stenosis (1–3).

Becker *et al* in 1995, noted that all hearts had one solitary feature which was confirmed with the advent of trans-thoracic echocardiography. This is anterior and cephalad deviation of the outlet septum, which can be of varying degrees, producing a spectrum of patients who present with a ventricular septal defect, also called the “Pink Tetralogy of Fallot”, to the severely cyanosed newborn, called originally, “La Maladie bleu”. Tetralogy of Fallot (TOF) is one of the few congenital

From: Adolescent Cardiac Clinic, Cardiology Unit, Department of Medicine, University Hospital of the West Indies, Kingston 7, Jamaica, West Indies.

Correspondence: Dr S Williams-Phillips, Adolescent Cardiac Clinic, Cardiology Unit, Department of Medicine, University Hospital of the West Indies, Kingston 7, Jamaica, West Indies. E-mail: sandrap@cwjamaica.com

cardiac anomalies that are progressive. Anterior deviation of the outlet septum in the pink TOFs becomes progressively more severe and usually leads to cyanosis by nine months to two years of age. The patient in this report is an example of this feature (1–3).

Until the advent of palliative shunts by Blalock and Taussig in 1945 (1–3) and the first successful total correction in 1954 by Lillehei and Varco at the University of Minnesota, 25%, 40% and 70% died at first, third and 10th year of life respectively (3).

Corrective surgery was thought to be the panacea of this medical problem for 10 years until in the 1970s, a decade after the first surgeries, postoperative tetralogy of fallot patients were the most common group of children with sudden death (1–4).

Attempts have been made to stratify the group of patients at risk for sudden death, using age at operation, type of surgical technique, whether there was a two-staged procedure with a palliative shunt prior to total correction or one stage correction. Use of technology has helped stratify the patients: assessment of electrocardiograms (ECG), Holter monitoring, treadmill stress test to assess induced arrhythmias but which can also cause suppression, and with the advent of electrophysiological studies and echocardiogram assessment of cardiac dimensions and Doppler flow and ventricular function became possible. There has been no unanimity on all risk factors that have been identified by varied and many researchers, for sudden death (4–6).

The only effective way that has been used to reduce sudden death is use of anti-arrhythmic agents, if sudden death is not the presenting feature of an arrhythmia (5–16). The patient to be discussed does not fulfil all the criteria that have been used to stratify the patients used by many researchers, and has a rare combination of arrhythmias occurring concomitantly.

CASE REPORT

A 27-year old male who was diagnosed at three months of age with a ventricular septal defect (VSD) and pulmonary stenosis was presented to the cardiac clinic, University Hospital of the West Indies (UHWI), with congestive cardiac failure requiring anti-failure therapy. Electrocardiograms showed biventricular hypertrophy, chest X-ray confirmed clinical cardiomegaly and plethoric lung fields. Anti-failure therapy was discontinued by 10 months of age when he started having hypercyanotic spell requiring admission on three occasions and prophylactic propranolol. He developed polycythemia with maximum haemoglobin of 18.1 g/dL. Electrocardiograms at this time showed right axis deviation (RAD), right atrial enlargement and right ventricular hypertrophy (RVH). Cardiac catheterization confirmed ventricular septal defect, infundibular, valvular and supavalvular pulmonary stenosis and a right-sided aortic arch. One-stage, total corrective surgery was done overseas, at two years and eight months of age. He had patch of the VSD and infundibular resection through the tricuspid

valve *via* a right atriotomy, with no transannular patch, because of an anomalous origin of left anterior descending artery crossing the right ventricular outflow tract (RVOT). Mild residual RVOT obstruction remained postoperatively with no significant pulmonary regurgitation and mild pulmonary stenosis.

It is important to note that the only complication of his mother's pregnancy was a urinary tract infection. There was no gestational diabetes, phenylketonuria or intake of retinoic acid. There were no phenotypic traits of chromosomal abnormalities or Digeorge syndrome. There was no family history of congenital heart disease. No fetal echocardiography was done. His birthweight was 3.15 Kg with an uncomplicated spontaneous vaginal delivery with an Apgar score of 10 at one and five minutes. There was no history of drug ingestion except for medications prescribed, no ingestion of highly caffeinated beverages, high dosed steroid therapy or use of narcotic substances.

He defaulted, but remained well, functioning at NYHA 1. He went to a tertiary institution of learning and is employed. In December 2010, at age 27, (25 years post-surgery) he developed generalized weakness, diaphoresis, shortness of breath on exertion, paroxysmal nocturnal dyspnoea, palpitations and near fainting episodes 2–3 times daily with a duration of approximately five minutes. He was now functioning at NYHA 111. His ECG was noted by his general practitioner to have signs of atrial fibrillation, RAD and RVH; QRS duration on ECG was 126 ms. Chest X-ray showed cardiomegaly with plethoric lung fields. He was admitted at a peripheral hospital where he was given anti-failure and anti-arrhythmic therapy which included digoxin, lasix furosemide, aldactone, verapamil, and the anticoagulant, warfarin. There was unsatisfactory improvement of palpitations and signs of failure. His reduced exercise capacity remained the same post discharge.

When seen in the cardiac clinic in January 2011, ECG showed no signs of atrial fibrillation. Echocardiography done showed signs of congestive cardiomyopathy secondary to arrhythmia with reduced ejection fraction of 33%, pulmonary regurgitation of 2.5 m/s, mild pulmonary valve stenosis, the anomalous coronary artery was seen crossing the RVOT and the VSD patch was intact with no residual leak. Right ventricular systolic pressure was normal. His anti-failure regime was optimized by increasing diuretics, and atenolol was started.

He had no more palpitations except for one week in February 2011, whilst having gastroenteritis. In March 2011, his CCF and palpitations were controlled. He was functioning at NYHA 1. Repeat echocardiogram showed normal ventricular function and dimensions. There was complete reversal of the arrhythmogenic cardiomyopathy.

Treadmill stress test using the bruce protocol induced atrial fibrillation in stage 1 (Fig. 1) and ventricular tachycardia in stage 4 (Fig. 2), with no palpitations noted by patient.

In June 2011, verapamil and diuretics were tapered over a three-month period with no recurrence of symptoms. He is currently maintained symptom free, on digoxin, atenolol

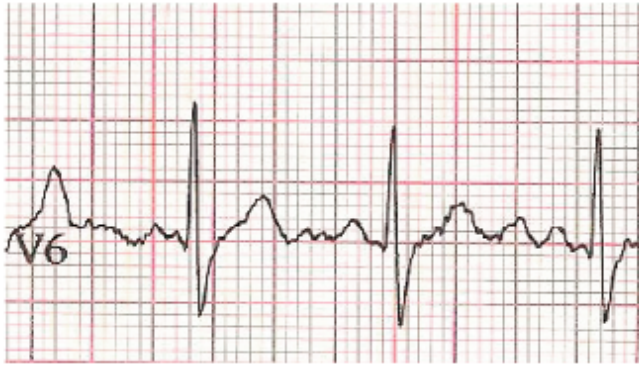


Fig. 1: Atrial fibrillation in stage 1 of treadmill stress test with (R) bundle branch block.



Fig. 2: Ventricular tachycardia in stage 4 of treadmill stress test with (R) bundle branch block.

(*ie* Class V and 11 anti-arrhythmic agents, respectively) and warfarin. His exercise is restricted with no competitive sports or sustained exertion. Monitoring of his international normalized ratio (INR) continues as he is still maintained on warfarin. He was advised to maintain endocarditis prophylaxis and good dental hygiene.

DISCUSSION

Separate multi-institutional studies (5–7), especially one done by Gatzoulis *et al* (6) show that the most common arrhythmias are atrial fibrillation and ventricular tachycardia postoperation for TOF. There was no history of drug ingestion except for medications prescribed, no ingestion of highly caffeinated beverages, high dosed steroid therapy or use of narcotic substances, all of which can cause atrial fibrillation (17).

Gatzoulis *et al* found only four patients with combined supraventricular and ventricular arrhythmias from 793 patients. This combination of atrial fibrillation and ventricular tachycardia, which was not accompanied by palpitations, hence not

evident to the patient during the stress test when ventricular function was back to normal, indicated that these arrhythmias could have been occurring long before the patient became symptomatic. Some postoperative TOF patients present for the first time with sudden death. In order to minimize this happening, there are some researchers who advocate screening of the patients at intervals with assessment of the areas they identify in their study which has statistical significance in helping to identify such patients, before they become symptomatic (5–12). Burns *et al* (12) used elevated right ventricular systolic pressure and Karamlou *et al* (11) found prolonged QRS duration > 180 msec statistically significant in determining the risk of occurrence of arrhythmia. There, however, is no unanimity on these specific tests that need to be done and sudden death continues to occur in the various groups despite routine ECGs, Holter monitoring, transthoracic echocardiograms and treadmill stress tests (6–18).

The factors considered to be important to stratify risk for arrhythmia are age at repair, infundibula resection, one stage corrective surgery, no transannular patch, normal right ventricular systolic pressure, mild pulmonary regurgitation, left ventricular function and QRS duration post-surgery; his QRS duration was 123 ms (*ie* > 180 ms is significant). The prevalence of sudden death depends on the type of arrhythmia, supraventricular or ventricular, this being more prevalent with ventricular arrhythmias. Atrial fibrillation was more prevalent with patients who had re-interventions and also with moderate tricuspid regurgitation. Moderate to severe pulmonary regurgitation preceded ventricular arrhythmia and sudden death and was more likely in patients who had a transannular patch and older age at repair, in excess of one year of age (5–16).

Gatzoulis *et al* indicate that optimal time for surgery, which reduces risk of arrhythmia is less than a year of age. Some authors indicate that the ideal time is between three months to one year of age, which most centres adhere to. The index patient was two years of age. Infundibula resection is associated with an increased incidence of ventricular arrhythmia. An atriotomy puts patients at an increased risk of atrial fibrillation but is more commonly seen after 40 years of age. The vast majority of TOF patients would have both these procedures done, based on the anatomy and approach of the cardiothoracic surgeon. The index case had no transannular patch, no significant pulmonary or tricuspid regurgitation, normal left ventricular function, all of which are in his favour, against arrhythmias. Two-stage operations and re-interventions were more common in patients who had both types of arrhythmias. He had one operation with total correction (5–18).

This case report highlights the dilemma cardiologists face in identifying patients with postoperative TOF who are likely to develop arrhythmias, and if the arrhythmia is uncontrollable, to think of separate arrhythmias occurring concurrently, requiring anti-arrhythmic agents from different classes.

Arrhythmias, if detected and managed early, can prevent or reverse secondary cardiomyopathy as in this case. New modalities of therapy of atrial fibrillation in uncontrollable

cases of arrhythmia are transcatheter electrophysiological studies or minimally invasive epicardial ablation which can be curative but unfortunately is not available at the reporting institution (19, 20).

This case report highlights the importance of lifelong monitoring of not only the postoperative TOF patients, but all post-surgery congenital cardiac patients.

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