

Potts Anastomosis: Transthoracic Echocardiographic Features – Tetralogy of Fallot, Doubly Committed Sub-arterial Ventricular Septal Defect, Anomalous Left Anterior Descending Coronary Artery Crossing Right Ventricular Outflow Tract

S Williams-Phillips

ABSTRACT

Potts anastomosis is a central systemic-pulmonary surgical shunt between the descending aorta and the left pulmonary artery, developed and subsequently disbanded in the 1950s to provide pulmonary blood flow in patients with tetralogy of Fallot. Blalock-Taussig shunt is a peripheral systemic pulmonary communication which was varied to make the modified Blalock-Taussig shunt, which is now the standard of surgical care for temporary or permanent blood flow to the right or left pulmonary artery from the subclavian artery. The central shunts were disbanded in the 1950s as early development of pulmonary hypertension and its sequelae were the major prohibitive complications. This is a case report of a patient with tetralogy of Fallot with a rare combination of doubly committed sub-arterial ventricular septal defect, anomalous left anterior descending coronary artery crossing the right ventricular outflow tract and patent ductus arteriosus, who developed pulmonary hypertension within four years of Potts anastomosis and then required cardiac and lung transplantation. The transthoracic echocardiographic images are the focussed feature in this paper, confirming clearly defined structural anatomy in complex structural congenital heart disease.

Keywords: Blalock-Taussig shunt, palpitations, patent ductus arteriosus, pulmonary hypertension, tetralogy of Fallot, valved conduit, Waterston anastomosis

Anastomosis de Potts: características ecocardiográficas transtorácicas – tetralogía de Fallot, defecto septal con compromiso doble subarterial ventricular, y arteria coronaria descendente anterior izquierda anómala a través del tracto de salida ventricular derecho

S Williams-Phillips

RESUMEN

La anastomosis de Potts – desarrollada y disuelta posteriormente en los años 50 – es una derivación quirúrgica sistémico-pulmonar central entre la aorta descendente y la arteria pulmonar izquierda, cuyo fin es proporcionar flujo de sangre pulmonar en pacientes con la tetralogía de Fallot. La derivación de Blalock-Taussig es una comunicación pulmonar sistémica periférica. Este procedimiento sufrió cambios que condujeron a la derivación modificada de Blalock-Taussig, que es ahora el procedimiento quirúrgico estándar para tratar el flujo de

From: Tai Wing, Andrews Memorial Hospital, Kingston, Jamaica, West Indies.

Correspondence: Dr S Williams-Phillips, Tai Wing, Andrews Memorial Hospital, 27 Hope Road, Kingston 10, Jamaica, West Indies. Email: sandrap@cwjamaica.com

sangre temporal o permanente hacia la arteria pulmonar derecha o izquierda desde la arteria subclavia. Las derivaciones centrales fueron disueltas en los años 50, cuando el desarrollo temprano de la hipertensión pulmonar y sus secuelas eran las complicaciones prohibitivas principales. Éste es un reporte de caso de un paciente con la tetralogía de Fallot con una rara combinación de defecto septal con compromiso doble subarterial ventricular, arteria coronaria descendente anterior izquierda anómala a través del tracto de salida ventricular derecho, y conducto arterioso persistente. El paciente desarrollo hipertensión pulmonar a los cuatro años de una anastomosis de Potts, y requirió entonces trasplante cardíaco y pulmonar. Las imágenes ecocardiografías transtorácicas constituyen el aspecto central de este trabajo, que confirma la anatomía estructural claramente definida de la enfermedad cardíaca congénita, estructuralmente compleja.

Palabras clave: Blalock-Taussig shunt, palpitaciones, conducto arterioso persistente, hipertensión pulmonar, tetralogía de Fallot, conducto con válvula, anastomosis de Waterston

West Indian Med J 2018; 67 (3): 275

INTRODUCTION

The central systemic pulmonary artery surgical shunt between the descending aorta and the left pulmonary artery developed by Potts *et al* was used in the 1940s and 1950s for temporary connection for reduced pulmonary blood flow in patients with tetralogy of Fallot (1). On long-term follow-up, Potts and other central shunts (such as Waterston anastomosis) showed statistically increased incidence of the development of pulmonary vascular hypertension and disease and were technically difficult to close when total correction was needed, leading to an increase in morbidity and mortality compared to the current surgical choice with the modified Blalock-Taussig anastomosis (1–3). These were developed when it was noted that the persistent patency of the arterial duct prevented the sequelae of cyanosis from reduced pulmonary blood flow in the tetralogy of Fallot. Two-dimensional echocardiography was not invented until the latter half of the 20th century. Hence, the classical echocardiographic features of Potts anastomosis are not available, which are the focussed features of this article (1–3). Echocardiography, non-invasively, shows the direct visualization of a surgically created shunt and Doppler evaluation of the shunt patency, with blood flow velocities (1, 2, 4). This index case had a rare combination of tetralogy of Fallot with a non-restrictive doubly committed sub-arterial ventricular septal defect (VSD) and an anomalous left anterior descending artery from the right coronary artery crossing the severely stenosed valvar right ventricular outflow tract, which makes it necessary to use a right ventricle to the pulmonary artery valved conduit or other means, to increase the

pulmonary artery blood flow in corrective or palliative surgery (1–3).

CASE REPORT

The index case was a 16-year-old Afro-Caribbean male with a two-year history of palpitations with increasing frequency, exacerbated by exertion, easy fatigability and increasing central cyanosis. His height was 165 cm, and his weight was 57 kg, with a body mass index of 21.5 kg/m² and a body surface area of 1.6 m². He had plethoric mucous membranes, central cyanosis, and mild clubbing of the fingers and toes. His steady-state oxygen saturation was 67% with blood pressure of 113/42 mmHg, showing a wide pulse pressure of 73. He was four years post-Potts cardiac surgery where he preoperatively had central cyanosis with saturations of 63%. After the cardiac surgery, he was noted to be pink and was well initially for one year. He functioned at NYHA1 immediately post-surgery but became NYHA 11-111 with palpitations exacerbated by exertion three years post-surgery. There was no history of syncope or deafness. He had developmental delay with seizures controlled with medication. His growth parameters had been maintained between the 25th and 50th percentiles during childhood.

There was no history of caffeine ingestion, energy drinks, high-dose steroids, stimulants or illicit drugs. There was no family history of deafness, palpitations or use of pacemakers.

Cardiovascular examination revealed vertical central and left lateral anterior chest sternotomy scars, palpability of all peripheral pulses, collapsing pulse, precordial

bulge, parasternal heave, displaced apex beat in the sixth left intercostal space in the anterior axillary line, normal first heart sound, single second heart sound, and a long systolic murmur 3/6 at the upper-left sternal border. There was an ejection systolic murmur 2/6 at the apex. There were no continuous or diastolic murmurs anteriorly or posteriorly, no additional heart sounds, and no signs of heart failure. His haemoglobin was 16 g/dL, and his packed cell volume was 51 L/L.

His electrocardiogram showed sinus rhythm with paroxysmal supraventricular tachyarrhythmia complexes, extreme right axis deviation, right atrial enlargement and biventricular hypertrophy. There were inverted T waves in the V1–V5, RSR¹ in the V1, V2, L11, L111 and AVF, and prolonged QRS duration. There was no significant ST segment anomaly, with normal P axis and T axis. He had no pre-excitation syndromes (*eg* Wolff-Parkinson-White, Long-Ganong-Levine, Mahaim, ion channelopathy), no Epsilon wave suggestive of arrhythmogenic ventricular dysplasia, and no saddle back deformity which ruled out Brugada syndrome and takotsubo cardiomyopathy.

A 24-hour Holter assessment recorded eight patient-events with supraventricular tachyarrhythmia and frequent isolated premature ventricular contractions. The R-R interval was 1.48 seconds.

His chest X-ray showed an increase in the cardiothoracic ratio and normal lung vascularity. His transthoracic echocardiography showed situs solitus (Fig. 1), atrio-ventricular concordance (Figs. 1 and 2) and ventricular arterial concordance (Fig. 2).

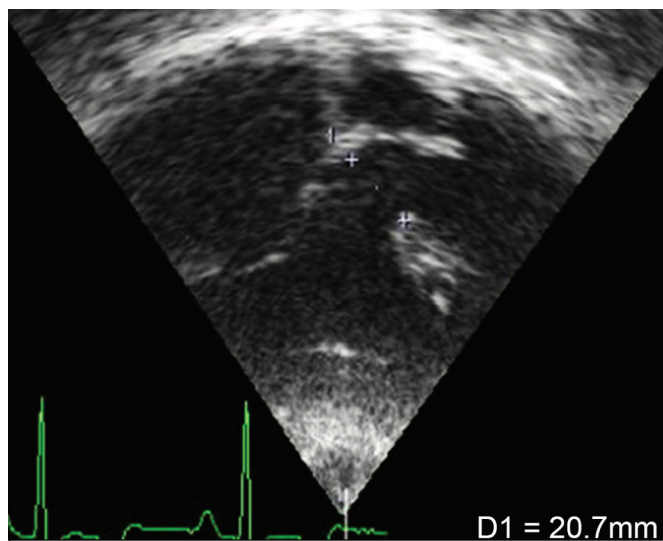


Fig. 1: Non-restrictive ventricular septal defect.

There was a large non-restrictive doubly committed sub-arterial VSD with aortic over-ride less than 50% (Figs. 1 and 2). The aortic valve was in continuity with the pulmonary valve and formed the rim of the VSD (Fig. 3). There was severe pulmonary valve and supra-valvar stenosis with minimal flow noted on his colour Doppler across the pulmonary valve. There was normal origin of the left (Fig. 4) and right coronary artery and anomalous left anterior descending artery from the right coronary artery crossing the right ventricular outflow tract (Figs. 4 and 5).

In the suprasternal view, the ligated patent ductus arteriosus (PDA) was seen on colour Doppler with no

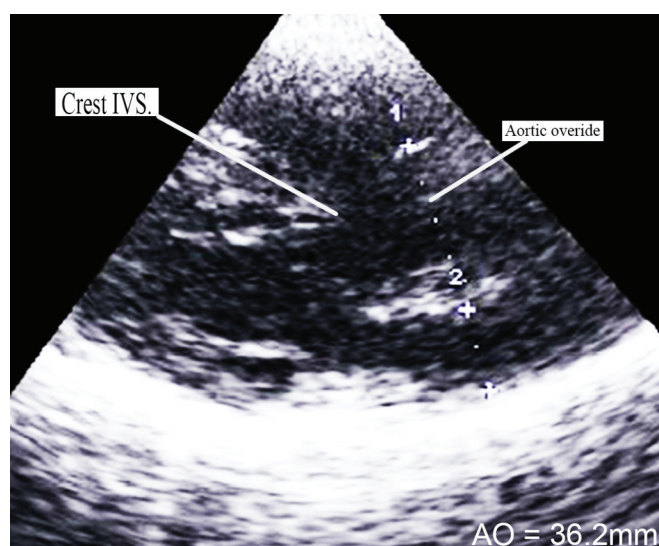


Fig. 2: Aortic over-ride of crest of interventricular septum. Large non-restrictive ventricular septal defect.

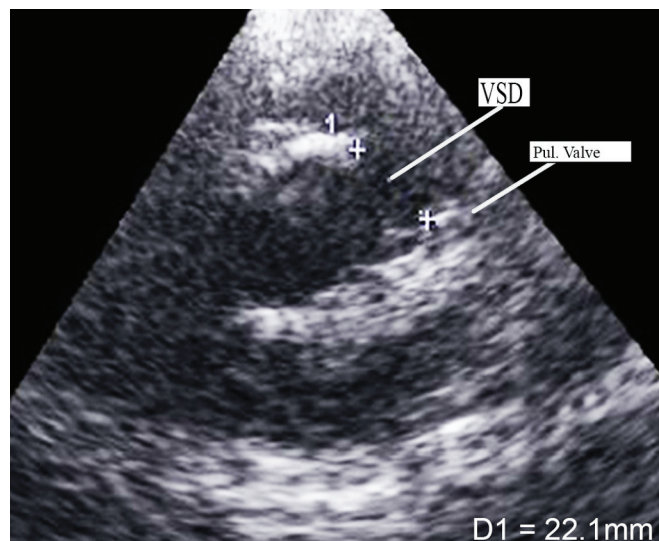


Fig. 3: Doubly committed subarterial ventricular septal defect adjacent to the pulmonary valve.

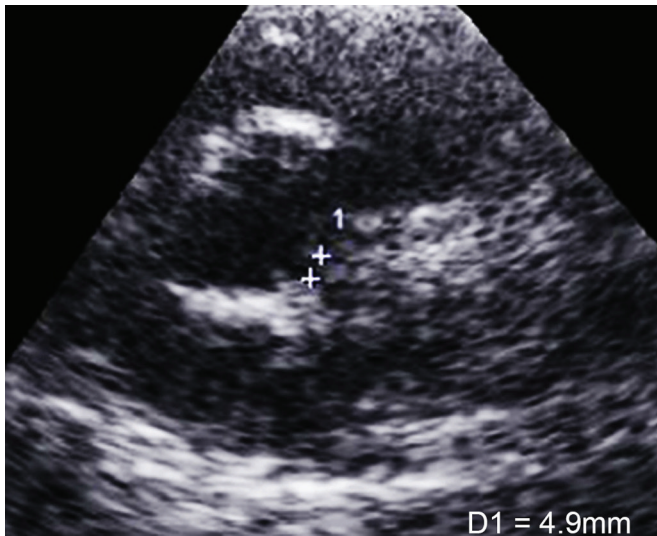


Fig. 4: Origin of the left coronary artery.

flow beyond a millimetre from the descending aorta (Figs. 6, 7).

Just below the ligated PDA was a flow on colour Doppler between the descending aorta and the left pulmonary artery using angled non-traditional views (Figs. 6, 7). Systemic-pulmonary collaterals and PDA would have been ligated when he had bypass surgery. Colour and Pulse wave Doppler to his left and right pulmonary arteries showed continuous Doppler flow from the descending aorta confirming Potts anastomosis (Fig. 8). In the suprasternal view, there was a left-sided aortic arch, and there was no flow from the right or left subclavian artery to the right or left pulmonary artery.

The patient's transthoracic echocardiogram showed hypertrophied interventricular septal dimension in diastole 21.7 mm left ventricular posterior wall diastolic dimension of 38.3 mm, with normal fractional shortening of 37% and ejection fraction of 71%. There was mild mitral and mild tricuspid regurgitation. Cardiac magnetic resonance imaging was not available in the index country.

Cardiac and pulmonary transplantations were the definitive corrective recommendation. Medications were recommended for the control of dysrhythmia and pulmonary hypertension in the patient. There continued to be exercise restrictions with no competitive sports or sustained exertion.

DISCUSSION

This index case had a rare combination of tetralogy of Fallot with a non-restrictive doubly committed subarterial defect and an anomalous left anterior descending

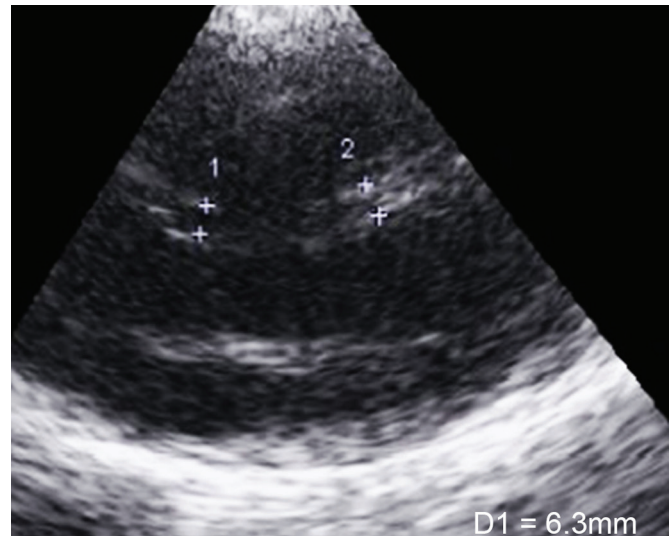


Fig. 5: Left coronary artery (marked 2) crossing the right ventricular outflow tract. Origin of coronary artery (marked 1).

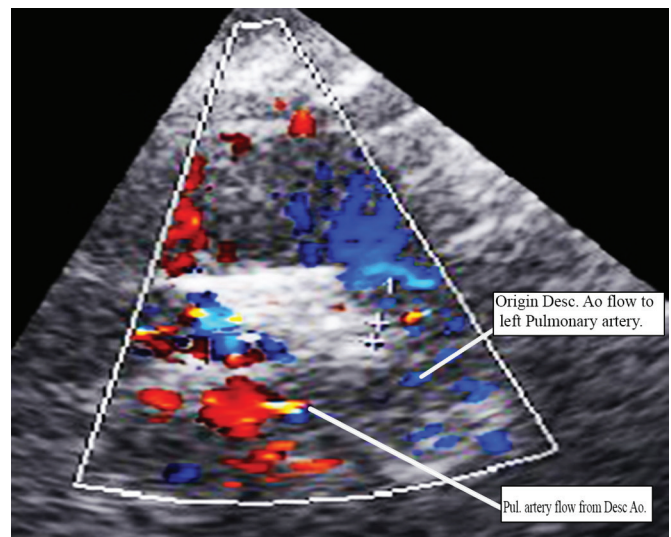


Fig. 6: Ligated origin of patent ductus arteriosus with no flow (marked 1). Flow from the descending aorta (Desc Ao) with flow to the left pulmonary artery (Pul artery).

artery crossing the right ventricular outflow tract, whose echocardiographic features of the Potts anastomosis are being documented for the first time in an Afro-Caribbean, in the English medical literature (1–5). The desaturation, collapsing pulse with wide pulse pressure, redevelopment of central cyanosis post-surgery, increase in haemoglobin levels, and only a long systolic murmur and not a continuous murmur clinically indicated the development of pulmonary vascular disease. The patient's echocardiography confirmed that the central shunt was still patent with continuous unobstructed flow of the Potts anastomosis on colour and pulsed wave Doppler. On the preceding bypass surgery, the systemic pulmonary collaterals and

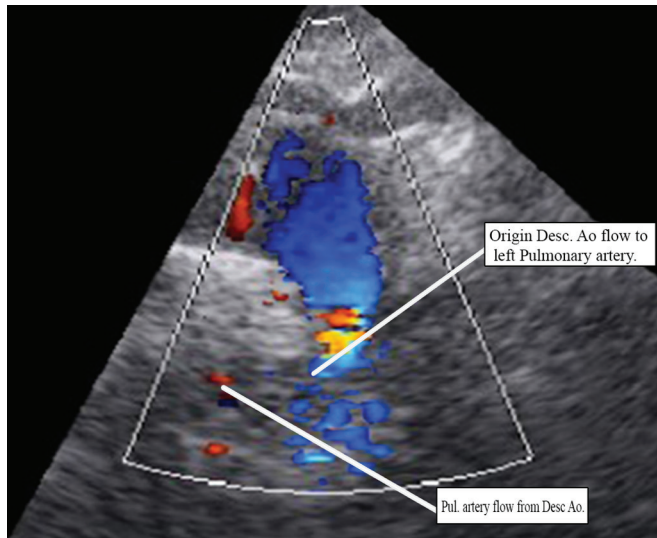


Fig. 7: Suprasternal view showing the flow from the descending aorta (Desc Ao) to the left pulmonary artery (Pul artery).

PDA would have been ligated to prevent pulmonary haemorrhage, and the nominal flow across the pulmonary valve would not have been the cause of the continuous flow in the pulmonary artery documented on colour and pulse wave Doppler. The continuous flow was reduced in diastole, hence the explanation clinically for a long systolic but not a continuous murmur. There were no signs of right or left Blalock-Taussig shunts. The ethics and medical management of this index case are not the feature of this article but require deep consideration as this is not the preferred corrective methodology of choice based on data worldwide and the confirmed morbidity and mortality associated with central shunts (1, 5). This index case emphasizes the need for knowledge of the history of the development of medical and surgical management, which would caution future approach of handling patients

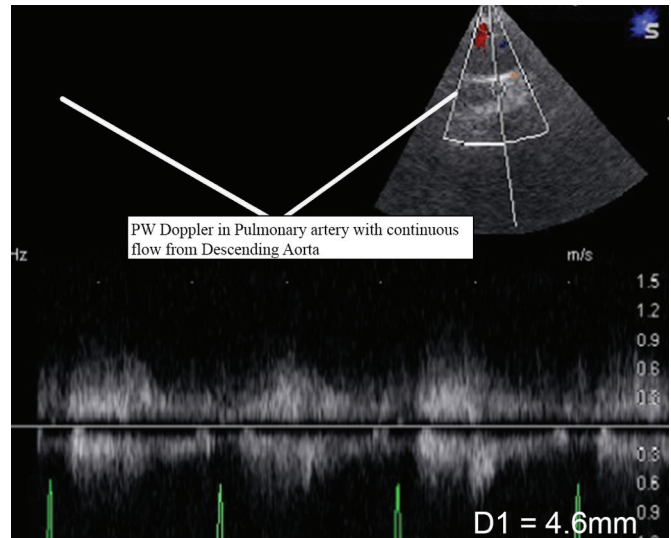


Fig. 8: Suprasternal view showing continuous flow from the descending aorta (Desc Ao) to the pulmonary artery (Pul artery).

with complex and rare structural anatomy (1–5). This index case has made available documented echocardiographic findings of Potts shunt for the first time in an Afro-Caribbean.

REFERENCES

1. Kirklin JW, Barrett-Boyes BG. Truncus arteriosus. Cardiac surgery, vol 2. New York: Churchill Livingstone Inc; 1993: 1131, 1399–402.
2. Jordan SC, Scott O. Heart disease in paediatrics. Oxford: Butterworth-Heinemann Ltd; 1989: 58.
3. Anderson RH, Macartney FJ, Shinebourne EA, Tynan M. Truncus arteriosus. Paediatric cardiology, vol 2. New York: Churchill Livingstone Inc; 1987: 788.
4. Seward JB, Tajik AJ, Edwards WD, Hagler DJ. Two-dimensional echocardiographic atlas. Vol 1: congenital heart disease. New York: Springer-Verlag; 1987: 541–5.
5. Redington A, Shore D, Oldershaw P. Congenital heart disease in adults. London: WB Saunders Company Ltd; 1994: 213–8, 234–6.