CASE REPORTS

Post Ross Procedure Aortic Right Sinus of Valsalva Fistula to Right Ventricle

S Williams-Phillips

ABSTRACT

Post Ross procedure complications have been limited predominantly to neoaortic valvular dilatation (10–30%) and insufficiency, right ventricular prosthetic deterioration or right ventricular pulmonary artery conduit obstruction. Arrhythmia has been documented to occur in a third of these patients. This is the first time that neoaortic right sinus of valsalva dissection and rupture to the right ventricle with a fistulous communication has occurred and been described, as far as the author is aware.

Keywords: Dissection, Marfan's syndrome, neoaorta, Ross procedure, rupture

Seno de Valsalva Aórtico Derecho con Fístula a Ventrículo Derecho tras el Procedimiento Ross

S Williams-Phillips

RESUMEN

Las complicaciones tras el procedimiento Ross han estado limitadas predominantemente a la dilatación (10–30%) e insuficiencia de la válvula neoaórtica, el deterioro prostético ventricular derecho, o la obstrucción del conducto de la arteria pulmonar derecha. Se ha documentado la presencia de arritmia en un tercio de estos pacientes. Hasta donde el autor conoce, esta es la primera vez que se produce y se describe la disección y ruptura de un seno de Valsalva aórtico derecho a ventrículo derecho con comunicación fistulosa.

Palabras claves: Disección, síndrome de Marfan, neoaorta, Procedimiento Ross, ruptura

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INTRODUCTION

The Ross procedure, named after its inventor Donald Ross, is a surgical technique used in a diseased aortic valve with the pulmonary valve forming the neoaorta; it has been used since 1967. In underdeveloped countries, there is still a high level of rheumatic heart disease (RHD) in the paediatric population and the ability of the neoaorta to grow seems to be the panacea in this age group for patients with irreparable aortic valve disease (1– 4). Females in particular will be able to have the choice to have a family as no anticoagulation is required with the attendant bleeding and teratogenic effects of warfarin, as is needed for mechanical valves, and the neoaortic valves are believed to be more resistant to infective endocarditis. In poor third world countries where RHD is still prevalent, and considering the high cost of the total procedure and the need for replacement of homografts every 4–6 years, the use of the Ross procedure is advantageous and more cost effective.

Dilatation of the neoaorta has been shown to occur particularly in the first two years post Ross procedure and studies from India suggest that patients with RHD tend to develop more dysfunction than other groups of patients with aortic valve disease. These two concerns lead to the need for re-operation. In this specific case, the aortic to right ventricular fistulous communication was diagnosed four years after implantation accompanied by moderate to severe aortic

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

regurgitation and has never been reported before in the English literature. There was an accompanying dissection with an intimal elevation occurring within the ascending aorta. The pulmonary homograft is still intact and functioning well (5– 8).

Aortic dissection with elevation of the intima in the sinus of valsalva is of concern post Ross procedure but has never been complicated with a rupture into the right ventricle. This general concern for dilatation has led to the publication by Ross *et al* (2), Kollar *et al* (9) and Luciani *et al* (10) of the modified Ross procedure which would help to negate the development of this specific complication. As far as the author is aware, this is the first ever case now documented of aortic dissection and rupture of right sinus of valsalva into the right ventricle occurring post Ross procedure.

CASE REPORT

In 2005, a 9-year old girl was noted by a doctor at another medical institution to have a murmur on routine examination. She was never admitted. There was no history of joint pain, signs of arthritis, erythema marginatum, erythema nodosum, chest pain or choreiform movements. She was noted on echocardiogram to have severe aortic valvular disease. She was presumed to be a case of RHD and was started on monthly prophylactic penicillin. Within one year of her initial diagnosis, the aortic regurgitation deteriorated where she needed aortic valve replacement and she had the Ross procedure done. In a report three years later, there was mild residual aortic regurgitation with good biventricular function. She came to the Adolescent Cardiac Clinic at the University Hospital of the West Indies (UHWI) in 2010 for an evaluation after referral from her previous institution. She gave a history of sharp sticking, intermittent left mid-sternal chest pain with pinpoint accuracy at the third and fourth left intercostal spaces in the four years following the Ross surgical procedure. This pain initially occurred daily and gradually became less frequent until it subsided spontaneously. She had not experienced any pain for two months prior to her visit to the UHWI. She functioned at NYHA 1 level but when the pain occurred she functioned at NYHA 1V; she had to stop what she was doing because of the severity until it subsided spontaneously within 1–2 minutes. Pain only occurred on exertion and there was no pain in the remainder of the praecordium, chin, neck, arm or epigastrium. There was no accompanying or otherwise palpitations, nausea, vomiting, dizziness, syncope, fainting or seizures.

Medications prescribed post Ross procedure was 28 day Penadur 1.2 mega units. There was no occurrence of any symptoms or signs of rheumatic fever. She exhibited good exercise tolerance by participating in physical education at school and cross-country races without any undue symptoms except what was mentioned above. She was never prohibited from participation in competitive or weight bearing sporting activities.

On examination, she was a tall 14-year old who has parents of similar stature. Weight was 58.5 kg, arm span was greater than height (> 1.05), blood pressure was 127/83 mmHg, pulse rate was 70/minute and respiratory rate was 20/minute. She was in no cardiorespiratory distress. Oxygen saturation was 98%. A high arched palate, arachnodactyly and a midline scar from a previous sternotomy were noted. The apex beat was in the fifth left intercostal space at the midclavicular line and was thrusting in character. The heart sounds were normal. An ejection systolic murmur 3/6 at mid left sternal border and a short diastolic murmur 1-2/6 at mid left sternal border were heard. There were no continuous murmurs heard.

She satisfied the Ghent criteria for diagnosis of Marfan's syndrome. Investigations were as follows: Hb 12.2 g/dL, HCT 36.2%, ESR 13, WBC 6.13 10 x 9/L, ASTO titre: < 80 (negative), C-reactive protein: < 0.5 mg/dL (negative), ECG: normal, chest X-ray: normal. Transthoracic echocardiogram: aortic regurgitation 3 m/s with reversal of flow in descending aorta. Mild aortic stenosis 2 m/s. Aortic root at level of sinuses 4.18 cm. Aortic valve at hinge point 1.79 cm. Left atrial diameter 2.18 cm; AO: LA ratio 2:1 (N I: I.I).

There were three intimal tears 0.3–0.4 cm in length, at intervals from each other, starting at hinge point with dissection into the ascending aorta and one rupture in the adventitia from the right coronary sinus into the right ventricle. These findings were visualized in parasternal long and short axis views of the aorta. The origin of the right coronary artery was 0.279 cm and the left coronary artery, 0.623 cm.

Transoesophageal echocardiogram confirmed findings above and showed signs suggestive of dissection of the posterior aspect of the ascending aorta. A mild supravalvar aortic stenosis of 2 m/s and a "waist" at the site of suture of the neoaortic sleeve to the ascending aorta was noted.

A 64-slice computerized axial tomography (CAT) scan confirmed diagnosis of right sinus of valsalva with fistulous communication to the right ventricle with five intimal tears within the aorta anteriorly and posteriorly at different levels on the ascending aorta.

Medications started after echocardiogram were a β-blocker, atenolol 50 mg once daily, an angiotensin receptor blocker, losartan 50 mg once daily. On review, her blood pressure was 80/52 mmHg and pulse rate 68/minute with no symptoms or signs of postural hypotension.

DISCUSSION

There was no clinical evidence of occurrence of rheumatic fever or carditis post Ross procedure and the ASTO titre was not significant. There was also no record postoperatively of the aorto-right ventricular (Ao – Rv) fistula and dissection of the ascending aorta which puts to question the possibility of a silent reactivation of the rheumatic fever; however, this

possibility was rejected because the ASTO titre was not significant and showed by its level that there was never a previous β-haemolytic streptococcal infection. Dehiscence along the suture line postoperatively, especially if there was an infection, is a possibility, even though the operative data were not available and there was no history of sepsis postoperatively.

Neoaortic dilatation with rupture and dissection is the most likely possibility which has been shown to occur in the first two years post Ross procedure. There has been proven growth and dilatation of the neoaorta, but this may have been excessive, as it has been documented that there is intimal hyperplasia and medial degeneration caused by elastin loss and breakage in post Ross patients. This would explain the five intimal tears noted. Defect in the fibrillin-1 in the neoaortic valve made it more susceptible to dissection and rupture.

The possibility of this child not having had rheumatic fever with carditis now seems clear. In the third world population, there is still a high incidence of rheumatic fever and it is expected that the ASTO titre would have shown by its level a previous infection with β-haemolytic strepto-coccus, which was not significant, and hence did not satisfy the modified Jones criteria for rheumatic fever.

This child is now believed to have Marfan's syndrome; phenotypic traits do not always manifest themselves in the young, as she was 9-years old when she was diagnosed with a presumed missed rheumatic fever. Given the tall stature of first degree relative in a child, without symptoms and signs of rheumatic fever and with sole involvement of the aortic valve, the diagnosis of Marfan's syndrome should have been considered. With the history of Marfan's syndrome, this rupture was possibly due to the inherent weakness of the pulmonary valve (Fig. 1) which also is not designed to withstand systemic pressures because it has a different stress-strain curve than the aortic root. Continuous wave Doppler flow (Fig. 2) confirmed flow across the defect in systole and aortic regurgitation in diastole. Colour Doppler (Fig.1) showed left to right shunt during systole, followed by start of aortic regurgitation in diastole (Fig. 4) with increased flow filling the aortic root with concomitant aortic regurgitation (Fig. 3).

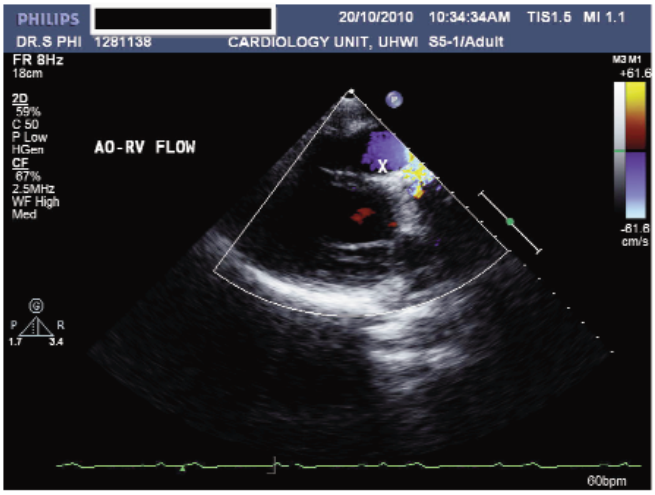

Fig. 1: Colour Doppler showing flow from right sinus of valsalva sistula to right ventricle in systole.

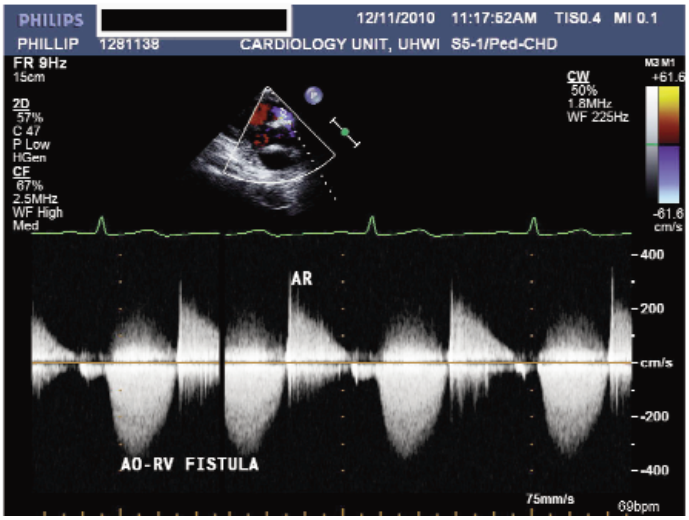

Fig. 2: Continuous Wave Doppler at site of fistula showing flow across the aorto-right ventricular (Ao-Rv) fistula below the baseline and concomitant aortic regurgitation above the baseline.

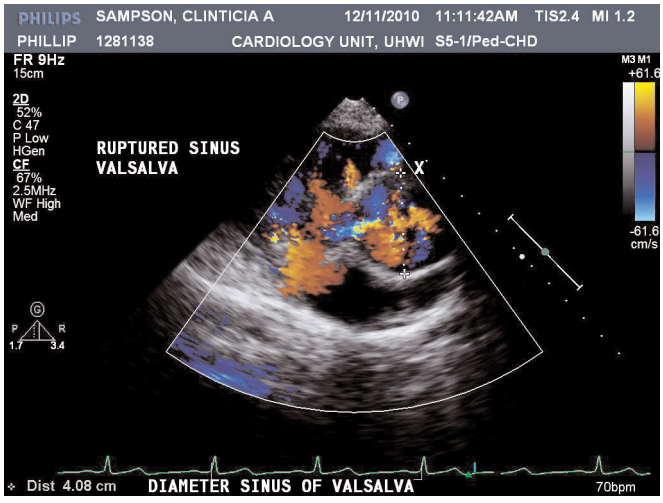

Fig. 3: Colour Doppler in diastole showing concomitant flow across ruptured right sinus of valsalva fistula and aortic regurgitation.

In view of the possible dilatation or the likelihood of this child having Marfan's syndrome and the worrisome occurrence of pulmonary dilatation, there is an obvious case for prophylactic use of β-blockers and losartan in all patients

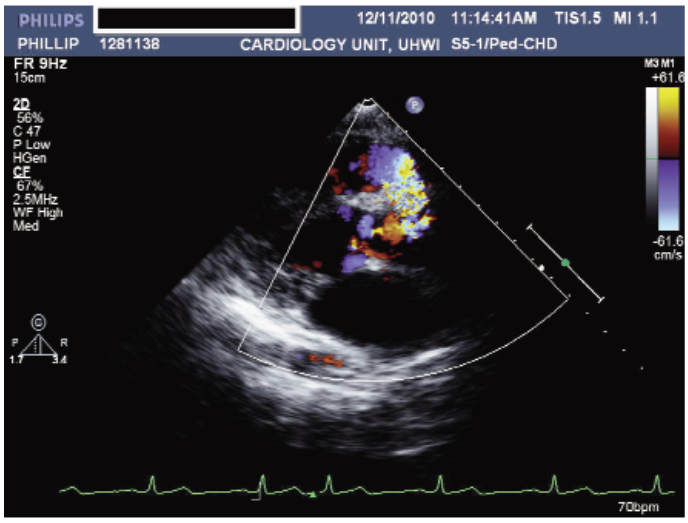

Fig 4: Colour Doppler of flow across right sinus of valsalva fistula into right ventricle at onset of diastole when aortic regurgitation commences.

following the Ross procedure. In addition, keeping the systolic pressure in children over 12 years of age at or below 110 mmHg and those of younger children at levels dependent on the norm for their age is important to prevent this complication. This has been proven in Marfan's syndrome.

A modified Ross procedure, where the autograft/neoaorta is encircled within a Dacron graft prior to suturing to prevent pulmonary dilatation as set out by Ross *et al* in 2010, may be able to resolve or reduce the development of neoaortic dilatation, dissection or rupture of the neoaorta (2, 9, 10).

The chest pain in this case could be due to the dissection slowly occurring over a period of time, as well as impaired coronary flow because of distortion of the anatomy of the right coronary sinus of valsalva, from which the artery arises. She would have benefitted from β-blockers prophylactically. The possibility existed that these intimal tears and rupture could have been avoided or allayed. This case is one from which we can learn and improve on long term postoperative management of patients who have the Ross procedure where prophylactic β-blockers and, if contraindicated, some other antihypertensive medication should be used, as tolerated.

We also need to be reminded to substantiate that there is RHD or other clear reasons for a diseased aortic valve. The Ross procedure is contraindicated by some authors in Marfan's syndrome because of the inherent problems with aortic dissection and rupture. In the paediatric age group in third world countries, the cost-effectiveness of this procedure is without question (1). The alternative surgery would be the use of homografts which need replacement every four to six years, with the attendant increased morbidity and mortality of reoperations. Hence, this procedure should still be considered in the very young with Marfan's syndrome, where growth of the neoaorta is a valuable asset until they are old enough to accommodate a mechanical valve or after childbearing in the female with careful follow-up of the neoaorta. The dissection and rupture can be detected with new modalities now available, such as a computed tomography (CT) scan every three to six months because of the pinpoint accuracy it provides. Early intervention would be possible to avoid rupture.

In view of the well documented pulmonary dilatation that occurs post Ross procedure (11–21), all patients, irrespective of the underlying cause of the diseased aortic valve, should be prohibited from all competitive and weight bearing sporting activities as the long term concern about this problem has not yet been ascertained, and this would be elucidated by following these patients over the next two to three decades.

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