SHORT CASES IN CARDIOLOGY

Interventricular septal expansion of a sinus of Valsalva aneurysm: a rare cause of complete heart block

M I Walters, D Ettles, L Guvendik, G C Kaye

We present the case of a 64 year old man, previously of good health, who presented in November 1995 with a short history of presyncope and one witnessed episode of syncope. On admission he was clinically in complete heart block with a ventricular rate of 40 beats/min but had a well maintained blood pressure at 150/80 mm Hg. Auscultation revealed a soft systolic murmur in the aortic area. The remainder of the examination was unremarkable. Resting ECG confirmed complete AV block with a slow regular broad complex ventricular escape rhythm. A Paragon III (Pacesetter, Coventry, UK) dual chamber pacemaker was subsequently implanted with complete resolution of cardiac symptoms.

He remained well for three months when he presented with sudden onset dyspnoea in January 1996. Further examination at this time revealed signs of pulmonary congestion and a cardiac murmur throughout systole and diastole that had changed in character from the previous evaluation. There were no peripheral signs of infective endocarditis. Initial management with intravenous diuretics resulted in rapid clinical improvement and allowed the following sequence of investigations:

- ECG confirmed normal AV sequential pacing.
- Transthoracic echocardiography revealed a left ventricle of normal dimension and function. A subaortic membrane of uncertain aetiology was seen and appeared to be attached to the interventricular septum. Aortic regurgitation was confirmed.
- Transoesophageal echocardiography revealed an abnormal aortic valve with a large apparently cystic mass under the aortic root, attached inferiorly to the interventricular septum (fig 1). This structure was felt to be a large aneurysm of the right coronary sinus extending into the interventricular septum.
- Cardiac catheterisation demonstrated a normal coronary arterial tree but confirmed the presence of a large saccular aneurysm of the right coronary sinus, shown best in the lateral position. Severe aortic regurgitation was seen.

The patient was referred for cardiac surgery and at operation a large aneurysm of the right coronary sinus invading the interventricular septum was confirmed. The walls of the sac were intact. The aortic valve itself was tricuspid and normal except that the right coronary cusp was noticeably redundant and was sagging into the left ventricular cavity (fig 1). The aortic valve and aneurysm were excised and the valve replaced with a Carbomedics mechanical prosthesis (East Crawley, West Sussex, UK) under full cardiopulmonary bypass. The patient made an uncomplicated recovery and remains well.

Discussion

Sinus of Valsalva aneurysm is an uncommon condition usually caused by a congenital defect of continuity between the aortic media and the aortic fibrous ring. Secondary causes are rare but include bacterial endocarditis, syphilis, tuberculosis, and Behçet’s disease.1,2 Surgical treatment is required in nearly all cases with the long term results being favourable; one large series showed a 95% 20 year survival.3

The classic presentation of a ruptured sinus of Valsalva aneurysm is described as chest pain of sudden onset associated with the clinical findings of a continuous “to and fro” murmur best...
heard at the base of the heart. As the aneurysm often ruptures into the right heart structures (right atrium or ventricle), right sided cardiac failure often follows quickly. The pattern of presentation does however vary widely, and unruptured aneurysms are often “silent” or present only with vague non-specific symptoms. However, cerebrovascular embolisation and angina have been reported. In our patient the unruptured aneurysm had extended into the interventricular septum and, presumably through compression, had compromised normal AV nodal/His bundle function and resulted in complete heart block. Case reports of the latter association are very rare, and to our knowledge only one case of an antemortem diagnosis has been reported previously. Therefore, sinus of Valsalva aneurysm deserves to be considered as a rare cause of complete heart block.


SHORT CASES IN CARDIOLOGY

Ventricular fibrillation triggered by a ruptured sinus of Valsalva aneurysm

J Hoshino, F Naganuma, R Nagai

A 40 year old Japanese man fainted suddenly while fishing and was brought to hospital. He fainted again in the outpatient department. Electrocardiography showed ventricular fibrillation (fig 1A), and sinus rhythm was recovered by defibrillation with 200 J. He had no history of syncope or life threatening arrhythmias. A grade V/VI continuous murmur was accompanied by a diastolic thrill along the left sternal border. Electrocardiography showed negative T waves in leads II, III, aVF, and V4–6. Transoesophageal echocardiography with colour Doppler imaging showed an aneurysm of the right coronary sinus of Valsalva, which displaced towards the right ventricle, and had systolic–diastolic turbulent flow from the aorta into the right ventricle (fig 2).

Catheterisation of the right heart revealed moderate increases of the pulmonary arterial and right ventricular systolic pressures. Blood oxygen saturation was increased by 12%, from 79.9% at the right atrium to 91.8% at the right ventricle. The left to right shunting rate was 42% and the pulmonary to systemic flow ratio was 1.7. Aortography revealed a jet of contrast medium flowing out of the right coronary sinus of Valsalva into the right ventricle (fig 2).

The patient had surgery on the seventh hospital day. Intraoperatively, a strong thrill caused by the ruptured sinus of Valsalva aneurysm was palpated directly under the pulmonary arterial valve. The aneurysm was Konno I type with a diameter of 6–7 mm. A ventricular septal defect type I, 6–7 mm in diameter, was also found at the site. Postoperative electrophysiological examination showed that ventricular fibrillation was induced by continuous stimulation of the outflow route of the right ventricle (fig 1B). The postoperative course was favourable. Amiodarone 200 mg daily was initiated to prevent ventricular fibrillation.

Rupture of an aneurysm of the sinus of Valsalva is rare, accounting for only 0.26% of all
surgical cases of congenital cardiac disease. Sinus of Valsalva aneurysm may result from a weak sinus of Valsalva wall. Recent studies have emphasised the wide variety of clinical manifestations of this lesion depending on its location and course. Because complications such as ventricular septal defect are relatively frequent, the aetiology is assumed to be congenital weakness of the sinus of Valsalva, except in cases resulting from inflammation, infection, or necrosis. The aetiology of the our case was assumed to be congenital because the patient was only 40 years old and had a ventricular septal defect; there were no inflammatory findings.

There have been two reported cases of ventricular fibrillation caused by rare mechanical complications in sinus of Valsalva aneurysms. In one report, the mechanism of ventricular fibrillation was myocardial ischaemia caused by compression of the left circumflex artery induced by a large aneurysm of the left sinus of Valsalva. In the other report, a ruptured aneurysm of the sinus of Valsalva had dissected into the ventricular septum and was complicated by ventricular fibrillation. No such complications were found in our patient. The origin of the aneurysm, the chamber into which it ruptured, and the association with a ventricular septal defect were typical for aneurysm of the sinus of Valsalva. However, the left to right shunting rate (42%) was enough to cause a rapid change in circulatory dynamics. Although ventricular fibrillation was easily induced by extrastimulation, our patient had no history of syncope or life threatening arrhythmias. These findings suggest that, in the absence of a rare mechanical complication, a ruptured aneurysm of the sinus of Valsalva may precipitate ventricular fibrillation because of an abrupt change in circulatory dynamics, if the patient has a tendency towards ventricular fibrillation. No other similar cases have been reported.

We thank the following physicians for their help in the management of this patient: Tatsuo Kaneko, Akihiko Nogami, Hiroshi Kamiyama, Shigeru Oshima, and Tadashi Suzuki.


Figure 2 Transoesophageal echocardiography with colour flow Doppler showing systolic-diastolic turbulent flow from the aorta into the right ventricle.
Acquired coarctation of the aorta in William’s syndrome

R Dhillon, T D Reddy, A Redington

The arterial lesions of William’s syndrome are characteristic: supravalvar aortic stenosis, coarctation of the aorta, peripheral pulmonary artery stenosis, and stenosis of the renal and coronary arteries. The development of slowly progressive coarctation has been described in only one patient, in whom angiography at 3 years of age showed a diffusely hypoplastic ascending aorta but no coarctation. Seven years later, angiography showed long segment coarctation distal to the left subclavian artery. We describe a patient in whom coarctation of the aorta developed over a much shorter time period.

Case report

William’s syndrome was diagnosed in this boy at the age of 4 years on the basis of “elfin facies” comprising epicanthic folds, a shallow nasal bridge, anteverted nares, long philtrum, and prominent lips. Hypercalcaemia had been noted during infancy. At routine follow up aged 5 years his femoral pulses were weak, with normal upper limb pulses. Echocardiography showed supravalvar aortic stenosis but no discrete coarctation; however, in view of the compelling clinical signs, cardiac catheterisation was performed in April 1993. There was no demonstrable gradient between the ascending and descending aorta. Aortic angiography showed no coarctation, although the descending aorta was diffusely hypoplastic (fig 1). The narrowest luminal point distal to the left subclavian artery measured 6.1 mm compared to the widest diameter of 8.3 mm.

In December 1996, clinical review again revealed weak femoral pulses with upper limb hypertension (right arm blood pressure 160/104 mm Hg). Echocardiography showed mild supravalvar aortic stenosis with a peak Doppler velocity of 2.0 m/s, moderate left ventricular hypertrophy, and suggested severe coarctation of the aorta distal to the left subclavian artery. Cardiac catheterisation was repeated in February 1997 when the patient was 9 years old. On this occasion, his systolic pressure gradient on withdrawal from the ascending to descending aorta was 44 mm Hg. Aortic angiography showed a long segment severe coarctation distal to the left subclavian artery (fig 1). Measurements performed at corresponding sites to the previous aortic angiography showed progressive coarctation, with a narrowest diameter of 2.6 mm, although there had been continued growth of the descending aorta distally measuring 10.9 mm. Five months later, he underwent end to end repair of coarctation with an interposition graft. Postoperative recovery was uncomplicated.

Discussion

The speed of development of aortic coarctation in this child is remarkable (less than four years). This does not represent a failure of growth. There was progressive narrowing of the lumen of the aorta, with the narrowest diameter falling from 6.1 to 2.6 mm, although
the widest lumen diameter distal to the site of coarctation increased over the same time interval, from 8.3 to 10.9 mm. Furthermore, the histopathological examination of the resected segment of aorta revealed an external diameter of 7.0 mm compared with an internal diameter of 2.6 mm. This severe luminal stenosis was largely the result of intimal proliferation, without obvious major medial hypertrophy, which is often described as an additional component of arterial wall thickening in this condition.5

Although the peripheral pulmonary artery stenoses of William’s syndrome tend to improve, other vascular lesions become worse.1 Acquired coarctation of the aorta, however, is rare.2 3 5 This case illustrates that the finding of weak femoral pulses in this situation may not be a reliable guide to the progression of coarctation, because of distal aortic hypoplasia. A high level of suspicion and regular echocardiographic follow up are necessary for the diagnosis of this unusual problem.


SHORT CASES IN CARDIOLOGY

Transcatheter recanalisation and stenting of a closed ductus arteriosus in duct dependent lung perfusion

C Kampmann, C-F Wippermann, F-X Schmid

In patients with the congenital cardiac malformation of tetralogy of Fallot, occasionally one pulmonary artery, usually the left, seems angiographically to be absent.1 This pulmonary artery is usually present, but discontinuous with the pulmonary trunk, having originally been supplied by a patent arterial duct. With closure of the duct, the receiving flow to that pulmonary artery is by small collateral vessels, which leads to reduced growth of the involved pulmonary vessels and impedes definite surgical repair.

We report a case of a 2 day old, 1890 g, premature, cyanotic boy (oxygen saturation 82%) with tetralogy of Fallot, right sided aortic arch, and discontinuity between the pulmonary trunk and the left pulmonary artery with a distance of 6 mm, and a complete left sided pulmonary perfusion deficiency. Six hours after starting prostaglandin E, (PGE,) infusion (100 ng/kg/min) there was still no flow to the left pulmonary artery or any increase in saturation.

Antegrade angiography from the right femoral vein confirmed the diagnosis. A closed patent arterial duct, originating from the left subclavian artery, could be seen selectively after passing the right ventricle into the overriding aorta (fig 1). To recanalise the closed patent arterial duct, an additional 50 ng/kg/min PGE, was given over 10 minutes into the origin of the patent arterial duct, while it was attempted to pass a 0.014 inch floppy guidewire through the patent arterial duct. After successfully passing the wire, PGE, infusion was stopped and dilatation was performed up to 3 mm. The length of ductal obliteration was 11 mm. To maintain ductal patency, a 9 mm long coronary NIR stent was mounted on a 3.5 mm balloon catheter and implanted at the pulmonary end of the duct (fig 2). Postinterventional angiography showed a well developed left sided pulmonary arterial system, but without a main left pulmonary artery (fig 3).

Figure 1 Selective posterior-anterior angiogram of the origin of the closed arterial duct from the left subclavian artery.
Immediately after stenting, the child developed left sided lung oedema with pleural effusion that necessitated drainage. After stenting, dilatation of the right ventricular outflow tract was performed up to 7 mm. All interventions were without complication. Total fluoroscopy time was 18.6 minutes. Before stenting, antithrombin III was substituted to a blood level of 100% and a bolus of 100 U/kg heparin was given intravenously followed by continuous infusion of 400 U/kg/day for the next three days. The child was extubated on the next day and discharged 14 days later in very good condition (oxygen saturation 91%); he was treated with 2 mg/kg/day propranolol and dipyridamole.

Redilatation of the stent up to 4.2 mm was necessary after 235 days. Selective angiograms of the right and left pulmonary arteries showed adequate development with diameters of 8 and 7.6 mm, respectively, and mean right and left pressures of 10 and 13 mm Hg. At the age of 1 year the child had successful transatrial–transpulmonary repair of the tetralogy of Fallot with connection of the left pulmonary artery to the pulmonary trunk.

Discussion

In symptomatic newborns with tetralogy of Fallot, percutaneous balloon dilatation of the right ventricular outflow tract and the application of propranolol is used palliatively to provide ideal conditions for repair, although the dysplastic pulmonary valve may be damaged. Stenting of the patent arterial duct to maintain patency was first reported by Gibbs et al in patients with pulmonary atresia but required redilatation. In our case with duct dependent lung perfusion, adequate left pulmonary artery growth could be established by stenting the patent arterial duct. Interventional recanalisation combined with stenting of a closed patent arterial duct in man has not been reported previously. Early interventional recanalisation of closed patent arterial ducts is feasible even in premature neonates, and the procedure supports age appropriate development of pulmonary arteries. This should contribute to a reduction in perioperative mortality.

Interventricular septal expansion of a sinus of Valsalva aneurysm: a rare cause of complete heart block

M I Walters, D Ettles, L Guvendik and G C Kaye

*Heart* 1998 80: 202-203
doi: 10.1136/hrt.80.2.202

Updated information and services can be found at:
http://heart.bmj.com/content/80/2/202

These include:

**References**

This article cites 5 articles, 1 of which you can access for free at:
http://heart.bmj.com/content/80/2/202#BIBL

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Topic Collections**

Articles on similar topics can be found in the following collections

- Drugs: cardiovascular system (8839)
- Clinical diagnostic tests (4778)
- Echocardiography (2126)
- Hypertension (3004)
- Interventional cardiology (2932)
- Aortic valve disease (414)
- Bradyarrhythmias and heart block (242)

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/