Successful occlusion of a coronary arteriovenous fistula using an Amplatzer[™] duct occluder

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Abstract In an 11-year old boy with a large coronary arteriovenous fistula between the left coronary artery and the right atrium, we achieved successful closure using a 10–8 Amplatzer Duct Occluder introduced from the right internal jugular vein. Angiography 6 weeks later showed complete occlusion of the fistula, with normal opacification of the left coronary arterial system. This technique may be used as an alternative to coils and surgery in selected patients.

Keywords: Interventional cardiology; therapeutic interventions; arteriovenous fistula

CONGENITAL CORONARY ARTERIOVENOUS fistula is a rare congenital malformation. When seen, such fistulas arise from the left coronary artery in about one-third of patients.¹ The majority of the fistulas drain into the chambers of the right heart or its venous tributaries, and only rarely to the pulmonary trunk or arteries. About one-quarter drain to the right atrium.¹ Crosssectional and transoesophageal echocardiography² have improved their non-invasive diagnosis, but cardiac catheterization, aortography and selective coronary angiography are still necessary for definitive diagnosis and for the planning of the mode of treatment.^{2,3} Surgery has been the standard treatment over the years, and is safe and effective.⁴ With advances in interventional catheterization techniques, nonetheless, embolization has been used for treatment of selected cases, taking account of the site, size and morphology of the fistulous connections. Detachable balloons, conventional stainless steel coils, and controlled release coils have all been used.4-7

In this report, we describe the successful occlusion of a large fistula arising from the left coronary artery and draining into the right atrium using an Amplatzer Duct Occluder, and the angiographic appearance as seen at follow-up 6 weeks later.

Case report

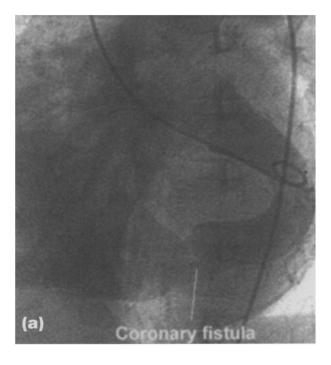
An 11-year old boy was referred for the assessment of a cardiac murmur. He had suffered repeated chest infections since infancy, and had recently developed shortness of breath on exertion. The clinical signs suggested a large left-to-right shunt, with a continuous murmur heard along the left sternal border being associated with cardiomegaly. The cardiothoracic ratio on the chest radiograph was 0.65, with right atrial prominence. The electrocardiogram demonstrated sinus rhythm with right atrial and biventricular hypertrophy. Crosssectional echocardiography showed a dilated right atrium, with turbulent flow near the dilated coronary sinus. Continuous-wave Doppler at the entry point in the right atrium showed a peak velocity of 4.2m/s. The dilated and tortuous channel could be traced to its origin from the ascending aorta, with the proximal part of the fistula being as large as the ascending aorta.

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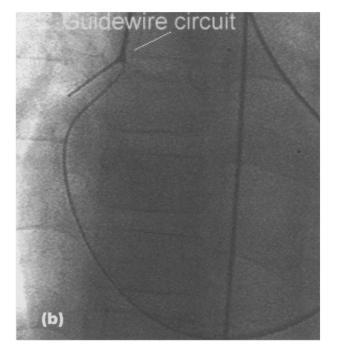
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The patient underwent cardiac catheterization under general anesthesia, when the right femoral artery and the right internal jugular vein were cannulated. Oximetry confirmed a left-to-right shunt, with a ratio of pulmonary to systemic flow of 2.6:1. Ascending aortography, and selective coronary angiography, confirmed the presence of a large fistula draining from the circumflex coronary artery to the right atrium (Fig. 1a). The right coronary artery was normal. Small marginal branches were seen arising from the fistula. The



narrowest point at its entry into the right atrium measured 6mm, with the diameter proximal to the point of entry being 14 mm.

A 0.035' extra-stiff exchange guidewire was used to cross from the fistula into the right atrium via the femoral arterial catheter. The venous catheter was used to snare the guidewire in the right atrium using a gooseneck snare (Microvena, Vadnais, MN) (Fig 1.b). An 8Fr delivery sheath (AGA Medical Corp.) was introduced from the internal jugular vein over the guidewire and passed into the fistula.



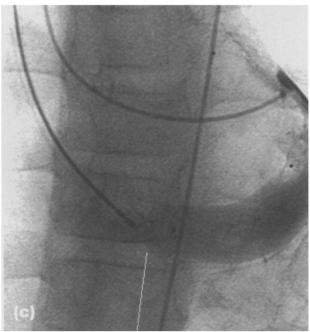


Figure 1.

Selective left coronary arteriography (a) demonstrates a large fistula extending from the circumflex coronary artery to the right atrium. An arteriovenous guidewire circuit was established (b) by snaring a guidewire in the right atrium and withdrawing it through the right internal jugular vein. Angiography was performed (c) in the left coronary artery prior to the release of the Amplatzer Duct Occluder, the device being released at the narrowest point in the fistula.

The guidewire circuit was then broken. A 10-8 Amplatzer Duct Occluder (AGA Medical Corp., Golden Valley, MN) was loaded on a delivery cable and passed through the sheath. Using angiography from a catheter placed in the left coronary artery for control, the distal disk of the occluder was opened in the fistula and the occluder was withdrawn slowly until the retention flange was deployed close to the point of entry to the right atrium. The sheath was withdrawn, and the position checked angiographically whilst the occluder was still attached to the delivery system (Fig 1.c). The initial placement was deemed unsatisfactory, the body of the device being within the wide portion of the fistula, rather than in its narrow point of entry to the right atrium. The device, therefore, was partially withdrawn into the sheath, and the assembly pulled back until the distal disc was adherent firmly against the narrow point. The proximal part of the occluder was then redeployed, and showed a 'waist' at the point of entry. Angiography prior to release then showed substantial residual shunting through the mesh of the device (Fig 1.c). The occluder was released successfully. Angiograms in the left coronary artery repeated 20 minutes later showed improved perfusion of the branches of the circumflex artery with complete occlusion of the

fistula (Fig 2.a). Serial electrocardiograms and cardiac enzymes showed no evidence of ischaemia over the next three days.

At follow-up six weeks later, there was marked symptomatic improvement, with a reduction in the heart size as seen radiologically, the cardiothoracic ratio being reduced to 0.55. Repeated cardiac catheterization and selective coronary angiography showed improved perfusion of the left coronary system with obliteration of shunting. The fistula was now seen as a blind-ending sack (Fig. 2b). The arterial branches arising proximally from the fistula had increased in size because of improved perfusion. The left ventricular angiogram was normal.

Discussion

Coronary arteriovenous fistulas constitute 0.2–0.4% of all congenital cardiac defects.¹ The fistulas originate from the right coronary artery or its branches in over half the cases, from the left coronary artery in one-third, and from both coronary arteries in about one-twentieth.¹ Fistulas have a predilection for drainage into the chambers of the right heart and its venous tributaries.¹ Except for small fistulas, which may regress sponta-

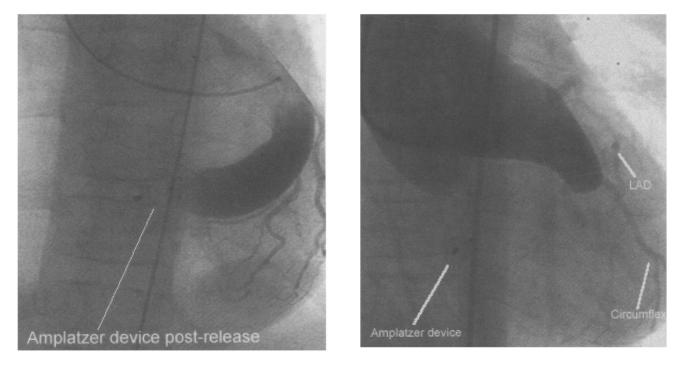


Figure 2.

Selective angiography immediately subsequent to the procedure showed complete occlusion of the fistula by the occluder, with improved filling of the other branches of the left coronary artery. Follow-up angiography six weeks after the initial procedure confirmed complete occlusion of the fistula (b), which now appeared as a blind ending sack, with normal opacification of the branches of the left coronary. LAD=left anterior descending coronary artery. Circumflex=left circumflex coronary artery

neously, most fistulas should be closed to prevent infective endocarditis, congestive heart failure, or myocardial ischemia in later life. Until the mid 1980s, surgery was the only form of treatment, and was associated with a low morbidity and mortality.⁴ With advances in interventional techniques, and the availability of various types of occlusion devices. an increasing number of these fistulas can now be closed in the cardiac catheterisation laboratory, thus avoiding surgery.4-7 Ischaemia may occur due to a coronary 'steal' phenomenon, and has been reported after both spontaneous thrombosis and after surgical closure.8 Transient changes in the T waves have been reported after coil occlusion, but more significant ischaemia has not been reported after closure using devices.⁶

Most of the reports in the literature document closure of fistulas using controlled-release, or other, coils or detachable balloons. Detachable balloons require a large introducing catheter, with the additional small risk of premature deflation and embolisation.^{5,6} Conventional coils, and controlled-release coils, can be used to close the majority of fistulas.⁷ The main technical difficulty is encountered in large fistulas with high flow. In these situations, coils may embolise to the lungs.^{6,7} A balloon catheter, placed at either end of the fistula, may be used to reduce or stop flow during placement.

In our patient, the fistula was large and had a high flow. It was considered that whilst coils could have been used, there would have been a high risk of embolisation. For this reason, we chose to insert an Amplatzer Duct Occluder into this fistula. This occluder is a self-expanding mushroom shaped device, made from nitinol mesh, and designed to close the persistently patent arterial duct.9 It has previously been used in a single case to close a coronary arterial fistula.¹⁰ It offers the advantages that the delivery and release of the occluder are controlled, test occlusion can be performed prior its release, and it can be retrieved if to misplacement occurs. It can be delivered through a small sheath, thus permitting closure of large fistulas in small children.

Availability of this device adds to the armamentarium for closure of coronary arterial fistulas. We recommend that all patients with such fistulas, irrespective of their size, should be assessed for possible interventional closure before being referred for surgery.

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