

MANAGEMENT OF CORONARY ARTERIOVENOUS FISTULA BY DEVICE

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SUMMARY

Coronary arteriovenous fistulas can be managed surgically or lately by transcatheter occlusion using various devices. We report a child where it was closed percutaneously and review the literature. An 11 -year old boy had clinical and echocardiographic features of a fistulous communication between the aorta and the right atrium. Selective angiography showed a large fistula between the left coronary artery and the right atrium. Access to the fistula was obtained via the right femoral artery and the right internal jugular vein. An extra-stiff 0.035" exchange guidewire was passed from the femoral artery through the fistula into the right atrium and snared from the jugular venous approach. A 10-8 Amplatzer Duct Occluder was advanced from the internal jugular vein through an 8 Fr Mullins sheath placed over the guidewire antegradely into the fistula. The fistula was occluded successfully with no evidence of ischemia on ECG or on serial measurement of cardiac enzymes. Repeat angiography six weeks later showed the occluder in position with complete occlusion of the fistula. The right coronary artery was the dominant vessel while the left anterior descending coronary artery and the obtuse marginal artery arising from the fistula had increased in size. The cardiac size regressed with no left ventricular akinesia or dysfunction on echocardiography. Occlusion of coronary arteriovenous fistula with an Amplatzer Duct Occluder is a safe and effective alternative to surgery in selected patients.

KEYWORDS

Coronary artery, arteriovenous fistula, Amplatzer occluder.

INTRODUCTION

A congenital coronary arteriovenous (AV) fistula, first described by Krause in 1865(1), is a rare congenital malformation (2). The left coronary artery is the involved vessel in about 35% of the patients (2). The majority of the fistulas drain into the right heart chambers or its connecting veins (rarely to the pulmonary artery), and about 25% of these drain to the right atrium (3). Cross-sectional and transesophageal echocardiography (4) have improved the non-invasive diagnosis of the coronary arteriovenous fistulas but cardiac catheterization, aortography and selective coronary angiography are still necessary for the definitive diagnosis and the planning of the mode of treatment (5,6). Surgery has

been the standard treatment over the years and is safe and efficient (7,8). With advances in interventional catheterization techniques, embolization of these fistulas has been used for treatment of selected cases (taking account of their site, size & morphology). (9,10,11).

We describe the successful occlusion of a large left coronary artery fistula draining into the right atrium using an Amplatzer Duct Occluder device and the follow up appearances on repeat angiography six weeks later.

PATIENT DETAILS

An 11 -year old boy was referred for the assessment of a cardiac murmur. The clinical signs suggested a large left to right shunt with a continuous murmur along left sternal border associated with cardiomegaly. He had a history of repeated chest infections since infancy and had developed shortness of breath on exertion recently. A chest x-ray showed cardiomegaly (cardiothoracic ratio of 0.65) with right atrial prominence. The electrocardiogram

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demonstrated sinus rhythm with right atrial and biventricular hypertrophy. Cross-sectional echocardiography showed a dilated right atrium with turbulent flow in the right atrium near the coronary sinus. The coronary sinus was dilated. Continuous-wave Doppler at the entry point in the right atrium showed a peak velocity of 4.2m/s. The dilated channel was traced posteriorly and had a tortuous course with the origin from the ascending aorta. The size of this abnormal fistula at its origin from the aorta was as large as the ascending aorta. The head and neck vessels and the aortic arch were normal with no aortic regurgitation.

Because of a suspicion of a coronary arteriovenous fistula the patient underwent cardiac catheterization under general anesthesia. The right femoral artery and the right internal jugular vein were cannulated. Oximetry confirmed a left to right shunt with a Qp/Qs of 2.6: 1. An ascending aortogram and a selective coronary angiogram confirmed a large left circumflex coronary artery fistula draining to the right atrium (Fig 1). The right coronary artery was normal. Small marginal branches were seen to be arising from the fistula. The narrowest point at the entry into the right atrium measured 6mm. The diameter of the fistula in its distal portion, proximal to the narrower entry point to the right atrium, was approximately 14 mm. A 0.035" extrastiff exchange guidewire was used via the femoral arterial catheter to cross from the fistula into

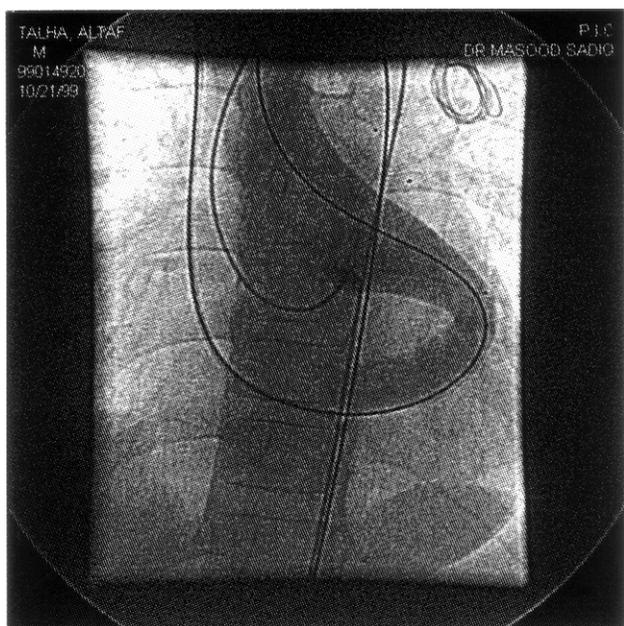


FIGURE - 1: Selective Arteriovenous Fistula Angiogram showing a large fistula between aorta and right atrium.

the right atrium. The right jugular venous catheter was used to snare the guidewire in the right atrium using a gooseneck snare (Microvena, Vadnais, MN) (Fig 2). An 8Fr Mullins transseptal sheath (AGA Medical Corp.) was introduced from the right jugular

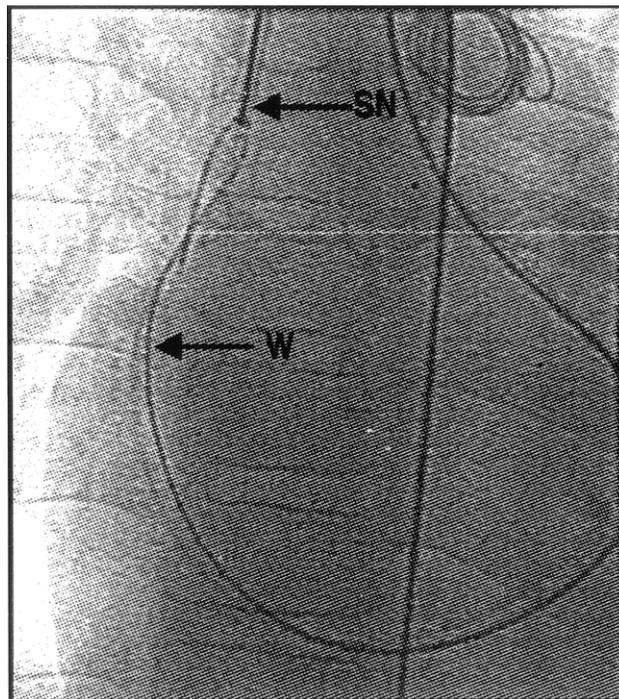


FIGURE - 2: Amplatzer Extra Stiff wire (0.035), being snared from the Right Atrium by juggler approach (W - Exchange wire, SN - Microvena Gooseneck Snare).

vein over the guidewire and was passed into the fistula antegradely. A 10-8 Amplatzer Duct Occluder (AGA Medical Corp., Golden Valley, MN) was loaded into the delivery system and passed through the sheath. Using check angiography from the catheter placed in the left coronary artery, 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 and pulled firmly against the entry of the fistula. The sheath was withdrawn and the device position was checked by an angiogram whilst the occluder was still attached to the delivery system (Fig 3). The initial device placement was unsatisfactory as the proximal part of the device was within the wide portion of the fistula. For this reason the device was partially withdrawn into the sheath and the assembly was pulled back towards the right atrium until the distal occluder disc was firmly up against the narrow point at the distal end of the fistula. The proximal part of the occluder was then redeployed and showed a 'waist' at the

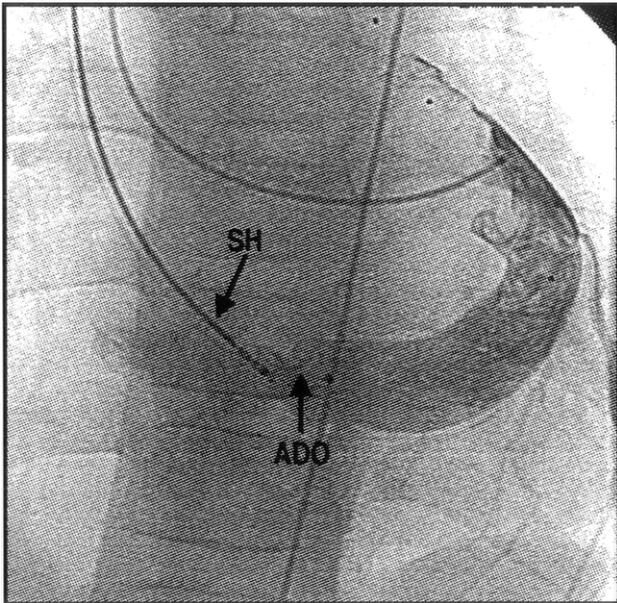


FIGURE - 3: Coronary Arteriovenous Fistula Angiogram showing Amplatzer Duct Occluder still attached to the delivery system introduced through a sheath passed from Right Jugular Vein. Almost complete occlusion achieved (ADO - Amplatzer Duct Occluder, SH - Mullen Sheath)

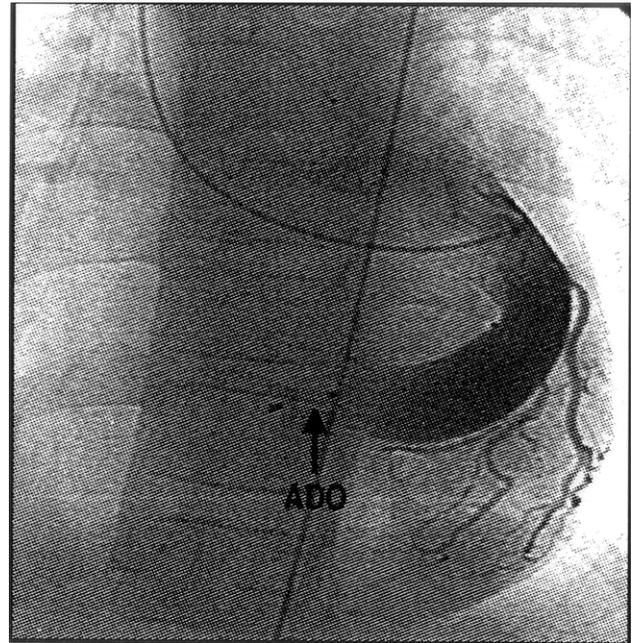


FIGURE - 4: Repeat Angiogram after duct occluder has been released showing complete, occlusion. Branch coronary arteries are better visualized (ADO - Amplatzer Duct Occluder).

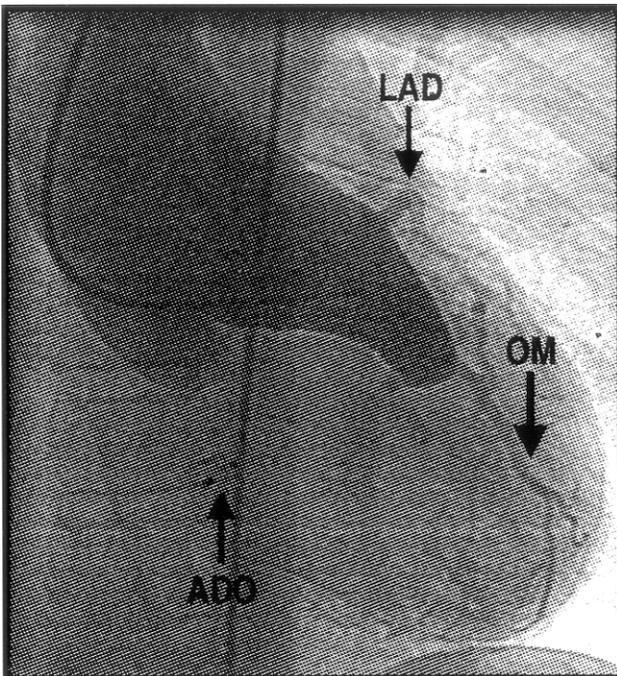


FIGURE - 5: Aortogram showing complete obliteration of the distal part of the fistula. Left anterior descending and obtuse marginal are much better visualized. The duct occluder is seen in position (LAD - Left Anterior Descending, OM - Obtuse marginal, ADO - Amplatzer Duct Occluder).

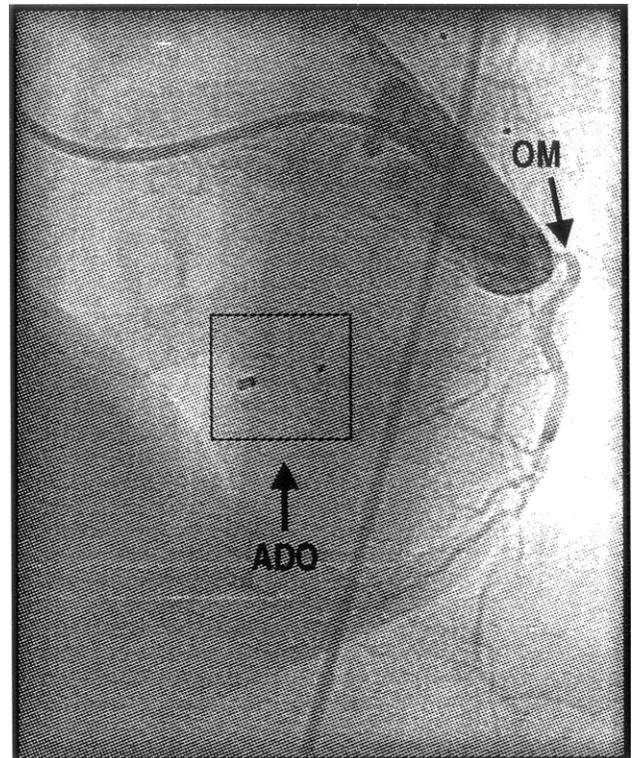


FIGURE - 6 : Selective coronary fistula angiogram showing the remaining stump. Obtuse marginal branch is well seen. Amplatzer duct occluder is shown in position (OM - Obtuse marginal, ADO - Amplatzer Duct Occluder).

'entry point'. Initial angiography, prior to release, showed substantial residual shunting through the mesh of the device. It was anticipated that this would resolve as thrombus developed on the Polyester membranes within the device. The occluder was released successfully. A repeat selective left coronary angiogram 20 minutes later showed no residual flow (Fig 4). Serial electrocardiograms and cardiac enzymes, checked for 72 hours after the procedure, showed no ischaemic changes or a rise in cardiac enzymes.

At follow-up six weeks later, there was marked symptomatic improvement with a reduction in the heart size radiologically (CT ratio 0.55). Cardiac catheterization and selective coronary angiogram was repeated. The right coronary artery filled better with complete disappearance of the shunt - the 'fistula' now appearing as a blind ending sac (Fig 5). The branches arising proximally from the fistula (the left anterior descending and obtuse marginal coronary arteries) had increased in size because of improved perfusion (Fig 6). The left ventricular angiogram showed no akinetic segment.

DISCUSSION

Coronary arteriovenous fistula, a direct communication between a coronary artery and the lumen of any one of the four cardiac chambers or the coronary sinus or its tributaries or the great arteries or veins adjacent to the heart, constitutes 0.2-0.4% of all congenital heart defects (2,3,5). The right coronary artery or its branches are the site of origin of the fistula in over 50% of the cases, whilst the left coronary artery is the site of origin in 35% and both coronary arteries in 5% (2,5). These fistulas have a predilection for drainage into the right heart chambers or its connecting vessels (3). Except for the very small coronary artery to pulmonary artery fistulas, some of which may close spontaneously, 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 (7,8). With advances in interventional techniques and the availability of various types of occlusion devices, an increasing number of these fistulas can be closed in the cardiac catheterisation laboratory thus avoiding a sternotomy and cardiopulmonary bypass (8,9,10,11,12). Ischemia has occurred after spontaneous thrombosis and after surgical closure of

such fistulas (13,14) but has not been reported after device closure, except transient T wave changes reported in one of the patients after coil occlusion (15).

Most of the reports in the literature consist of closing of the coronary artery fistulas using controlled-release coils or detachable balloons. These have some technical advantages in some types of fistulas and disadvantages in others. Detachable balloons require a large introducing catheter and have the additional small risk of premature deflation and embolisation (15, 16). Conventional coils and controlled-release coils can be used in the majority of coronary artery fistulas. The main technical difficulty that may be encountered with these is in those fistulas, which have a high flow. In these coils may not stay in place and may embolise to the lungs and necessitate removal by snare catheters (15,16,17). A flow-directed catheter can be used to stop the flow whilst the coils are implanted to avoid these complications (18).

In our patient, the fistula was large and had a high flow. Whilst coils could have been used to occlude the fistula, there would have been a higher risk of coil embolisation and larger diameter coils without a controlled-release mechanism may have been required. It was felt that the Amplatzer Duct Occluder was more suitable for closure of this fistula. The Amplatzer Duct occluder is a self-expandable, mushroom shaped device made from Nitinol wire mesh (0.004") designed to close large patent arterial ducts and is available in several sizes. (19). It has been reported to have been used once before in the literature to close a coronary artery fistula (20). It offers certain advantages. The delivery and the release of the occluder are controlled and test occlusion can be performed prior to its release. It can be retrieved if misplacement occurs. A small delivery system is available and large fistula can be closed with the larger device in small children. Availability of this device adds to the armamentarium used to close coronary artery fistulas and with a large number of different types of devices, we recommend that all patients with coronary artery fistulas, irrespective of size, should be assessed for catheter closure before being considered for surgery.

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