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# Transcatheter Closure of Large-Sized Coronary Artery Fistula

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#### ABSTRACT

We report our experience with the use of AMPLATZER® Vascular Plug for the closure of coronary artery fistulas. Three patients (age: 3, 12, 14 years) were diagnosed with coronary fistulas (pulmonary-to-systemic blood flow ratio: 1.5 to 3). Two of the fistulas originated each, from the proximal right and left coronary artery and had maximal diameter 9 and 10 mm respectively; their narrowest diameter (6 mm) was proximal to their entrance into the right atrium creating a form of a saccular aneurysm. The third fistula (maximal diameter: 16 mm) originated from the circumflex artery and entered the right atrium with unobstructed flow (narrowest diameter: 8 mm). Interventional closure was considered optimal and the decision was made to use devices sized twice the size of the narrowest diameter of the fistulas (12, 12 and 16 mm respectively). An arterio-venous loop was established through the fistula by snaring an exchange guide-wire. All plugs were implanted from the femoral vein with the use of a seven or eight French guide catheter, reaching the narrowest segment of the fistula and leading to complete closure of the two fistulas, immediately after the procedure. The fistula arising from the circumflex artery that received the largest plug continued to have residual flow up to 12 months after the procedure, when follow-up echocardiography revealed its complete occlusion. We present and consider the use of the AMPLATZER® Vascular Plug as a safe and effective method for the transcatheter closure of large-sized coronary fistulas. The plug potentially offers an alternative method to coil occlusion techniques as well as open heart surgery.

#### INTRODUCTION

Congenital coronary artery fistulas are rare malformations with an incidence rate of 0.002 percent and small fistulas up to 0.08 to 0.30% during coronary angiography [1]. Most of the patients are asymptomatic during childhood but in older patients, late symptoms and complications, like congestive heart failure, bacterial endocarditis, coronary ischemia or fistula rupture have been reported [2]. Surgical ligation of coronary artery fistulas, was reported for the first time in 1947 by Bjorck [3] and was the treatment of choice for many years. Transcatheter closure as a less invasive alternative to surgery was undertaken in the 1980's and early 1990's using different devices [4,5]. Today, transcatheter closure using coils is the most commonly used technique and is performed in many cardiac centers [6,7]. Even though the results of transcatheter closure of coronary artery fistulas are comparable with surgery, the procedure may be difficult and time consuming in

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Address for correspondence: Spyros Rammos, MD, Department of Pediatric Cardiology, Onassis Cardiac Surgery Center, Athens, Greece Email: srammos@gmail.com cases with large and unrestrictive fistulas where multiple coils are needed to occlude the communication. We report our experience with the use of the AMPLATZER® Vascular Plug, for the closure of large-sized coronary artery fistulas in three pediatric patients.

#### MATERIALS AND METHODS

## PATIENTS

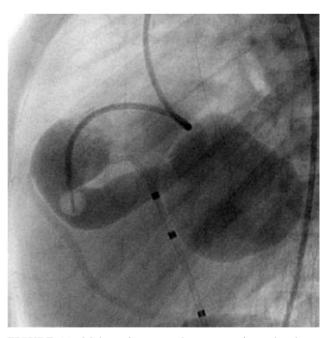
Three patients (age: 3, 12, 14 years) were diagnosed with coronary artery fistulas. Two of them were asymptomatic and the third presented with dyspnea on exertion, easy fatigue and required medication for congestive heart failure. Echocardiographic findings in all three patients demonstrated a dilated left ventricle and depending on the origin of the fistulas, a dilated proximal right, left or circumflex coronary artery. At catheterization, a pulmonary-to-systemic blood flow ratio of 1.5 to 3 confirmed the echocardiographic impression of shunt related left ventricular enlargement while the anatomical details of the coronary fistulas were enlightened by coronary angiography. Two fistulas arising from the proximal right and left coronary artery (maximal diameter: 9 and 10 mm) had their narrowest diameter (5.8 and 6 mm) proximal to the entrance into the right atrium via a saccular aneurysm like structure (Fig. 1a). The third fistula (maximal diameter: 16 mm), originated from the circumflex coronary artery and entered the right atrium with nearly unrestrictive flow (narrowest diameter: 8 mm) (Fig. 2a).

### THE DEVICE

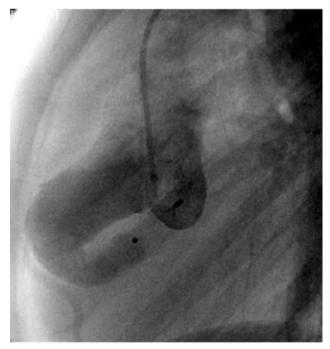
Originally designed for arterial or venous embolization in the peripheral vasculature, the AMPLATZER Vascular Plug (AMPLATZER® Vascular Plug) (AGA Medical Corp., Golden Valley, Minnesota) is a self-expanding, cylindrical device made from 144 Nitinol mesh wires secured on both ends with platinum marker bands. The AMPLATZER® Vascular Plug comes pre-loaded, attached to a 135 cm long stainless steel delivery cable. The plugs range in diameter from 4 to 16 mm (with 2 mm increments) and are 7 to 8 mm in length. For delivery, a 5 to 8 French standard coronary guide catheter is used (recommended internal diameter: 0.056 to 0.088 inch). Similar to other AMPLATZER<sup>®</sup> devices, the AMPLATZER® Vascular Plug can be repositioned or removed if the placement is not satisfactory. The recommended size for selection is that of an AMPLATZER® Vascular Plug that is 30 to 50% larger than the diameter of the target vessel.

## THE PROCEDURE

We performed all procedures under sedation. We evaluated all patients using standard catheterization techniques and performed selective coronary angiographies in order to study the anatomy of the fistulas and measure the exact

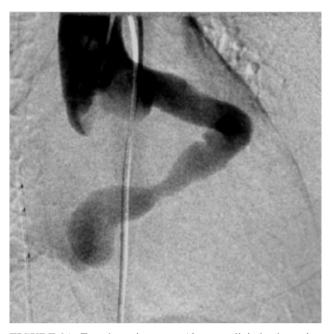


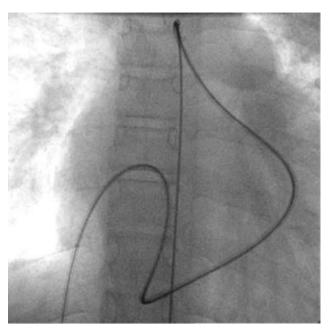
**FIGURE 1A.** Male patient, age: 3 years; angiography shows coronary artery fistula (diameter: 6 to 10 mm) originating from the proximal right coronary artery and draining into the right atrium via a saccular aneurysm.



**FIGURE 1B.** Angiography confirms complete occlusion immediately after placement of an AMPLATZER<sup>®</sup> Vascular Plug (diameter: 12 mm).

## TRANSCATHETER CLOSURE OF LARGE-SIZED CORONARY ARTERY FISTULA





**FIGURE 2A.** Female patient, age: 13 years; digital subtraction angiography with large coronary artery fistula (diameter: 8 to 16 mm) originating from the circumflex coronary artery and draining into the right atrium.

**FIGURE 2B.** Arterio-venous wire loop through the fistula with 8 French guide catheter which was advanced from the femoral vein to the ascending aorta.

diameter of the malformation. For interventional closure, we implanted vascular plugs 2 times the diameter of the narrowest segment of the fistula (12, 12 and 16 mm). We placed an arterio-venous wire loop through the fistula by snaring an exchange guide wire (Fig. 2b), which had been advanced from the arterial side into the right atrium via a standard 4 or 5 French multipurpose catheter. This allowed the exact placement of the plug via a 7 or 8 French guide catheter (internal diameter: 0.078 and 0.088 inch) (Cordis Corporation, Miami, USA), which was then advanced from the femoral vein while additional contrast injections in the aortic root or the coronary arteries confirmed the plug's correct position prior and after its release from the delivery cable. After the procedure (screening time: 20 to 38 min, procedure time: 100 to 120 min) aspirin (2 to 3 mg per kg per day) was administered for six months in order to prevent coronary thromboembolic events.

## RESULTS

We implanted all plugs at the narrowest segment of the fistula under fluoroscopic guidance; this was visually confirmed during the procedure as a slight compression of the middle part of the device. Immediately following placement of the device, complete closure of the two fistulas was confirmed by angiography (Fig. 1b) or transthoracic echocardiography. In the case of the circumflex coronary artery fistula a residual shunt was clearly demonstrated angiographically while the device was still connected with the delivery cable (Fig. 2c) and was consistent with turbulent flow through the plug's wire mesh on transoesophageal echocardiography. The decision to release the device was made, with the anticipation that the shunt will diminish and actually one hour later, angiography and color flow Doppler clearly showed diminished blood flow through the plug. Discrete ST-segment changes were identified in the electrocardiogram but angiography did not reveal compromise of major coronary side branches of the circumflex artery. The patient received heparin (400 IU per kg per day) and the electrocardiogram came back to normal 24 hours later. Follow-up examinations up to 9 months after device placement, revealed persistence of a small residual shunt but the patient presented with complete relief of cardiac symptoms with no need to continue anti-congestive medication. At 12 months follow-up examination, spontaneous complete occlusion of the residual shunt was demonstrated by echocardiography and color-flow Doppler. Left ventricular size normalized on transthoracic echocardiography in a follow-up period of 1 to 3 years.

#### DISCUSSION

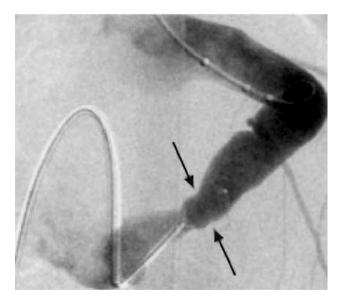
The new AMPLATZER® Vascular Plug has been used for transcatheter closure in a variety of malformations [8,11] There are only two reports to date regarding its use for closure of coronary artery fistulas, including no more than 3 patients worldwide [12,8]. Our experience with its successful use in pediatric patients with coronary artery fistulas demonstrates an alternative therapy to coil occlusion techniques or open heart surgery. Like other AMP-LATZER<sup>®</sup> devices the plug is easy to place, is retrievable and has the potential to become the method of choice for closure of moderate- to large-sized coronary artery fistulas. In comparison, coil occlusion sometimes requires complex catheter manipulation, as well as selection of various catheters and wires. In patients with complex anatomy or large fistulas, multiple coils may be required to occlude the malformation, which might lead to a prolonged procedure and radiation time. In such cases, the AMPLATZER® Vascular Plug offers the advantage of the use of a single device for effective closure.

A potential limitation of the plug technique, especially in pediatric patients, is the necessity to use relatively large guide catheters. Therefore, it might be essential to establish an arterio-venous wire loop through the coronary artery fistula. In our patients, this allowed the use of seven and eight French guide catheters for device insertion from the femoral vein. Nevertheless, it might be difficult or even impossible at times to establish an arterio-venous wire loop in patients with a very tortuous root of the fistula. Since it might be impossible to enter the coronary artery fistula from the venous side, the only alternative approach is the placement of the AMPLATZER® Vascular Plug via the arterial route. However, the arterial approach with the use of large guide catheters could be inadequate and therefore, coil occlusion technique with the insertion of coils through small 4 or 5 French catheters is the only option in cases that an arterio-venous wire loop cannot be established, as is the case in small sized pediatric patients.

The anatomy of the coronary artery fistula could be the limiting factor of either interventional technique, be it the plug or the coil technique. In cases with a very short and unrestricted route, for instance a fistula arising from the left coronary artery and draining into a left heart chamber, a device closure attempt bears the high risk of migration of the device into the systemic or even the coronary circulation. This risk could be a strong argument against any attempt of transcatheter closure and heart surgery is the safest and only option [13]. The decision to choose plugs of at least 2 times the narrowest diameter of the coronary artery fistula in our patients was made in order to prevent dislodgement. Our choice exceeded the manufacturer's recommendation to select an AMPLATZER<sup>®</sup> Vascular Plug 30 to 50% larger than the diameter of the target vessel. However, the fluoroscopic images showed only a slight compression of the middle part of the implanted devices, a fact that indicated that the chosen devices were not too big and also showed the correct and secure placement at the narrowest segment of the fistula (Fig. 2c).

We believe that an important recommendation for transcatheter occlusion technique is to assure that no vital branching vessels distal to the site of device placement are compromised. We performed test occlusion of the fistula and selective contrast injections using a wedge catheter prior to release of the device to secure uncompromised coronary flow. Test occlusion also offers the additional advantage to observe the electrocardiogram for any changes. In our patient, who developed transient changes in the ST-segment of the electrocardiogram, comparison of angiographic images before and after device placement confirmed that no major coronary side branch was compromised and the plug was finally released.

In conclusion, the AMPLATZER<sup>®</sup> Vascular Plug is easy to use even in pediatric patients with moderate to large sized coronary artery fistulas and offers an attractive alternative to open heart surgery or coil occlusion techniques.



**FIGURE 2C.** Residual shunt soon after placement of an AM-PLATZER<sup>®</sup> Vascular Plug (diameter: 16 mm) at the narrowest segment of the fistula. Arrows are showing the slight compression of the plug indicating the correct placement at the narrowest segment of the fistula. The plug is still connected to the delivery cable.

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