Transcatheter embolization of congenital coronary arterial fistulas in adults

Antônio M. Kambara¹, Carlos A. C. Pedra², Cesar A. Esteves², Manoel N. Cano², Sérgio L. N. Braga², Amanda G. M. R. Souza³, J. Eduardo M. R. Souza³, Valmir F. Fontes⁵

¹Director of Radiology Department, ²Assistant of Invasive Cardiology Department, ³Director of Invasive Cardiology Department, ⁴Director of “Instituto Dante Pazzanese de Cardiologia”, ⁵Director of Diagnostic and Therapeutic Methods Division. Departments of Interventional Radiology and Cardiology of “Instituto Dante Pazzanese de Cardiologia”, São Paulo, Brazil

Abstract In this report, we describe our experience with transcatheter occlusion of congenital coronary arterial fistulas in adults. From November 1992 to November 1996, 5 symptomatic patients, aged from 47 to 70 years, underwent transcatheter occlusion of fistulas using a retrograde arterial approach. All had chest pain or dyspnea on exertion. Detachable balloons were used in 4 patients, and Gianturco coils in 1. Detachable balloons were implanted through a Debrun system, while the coils were implanted through a 5 French right coronary Judkins catheter. Both were passed through an 8 French guiding catheter (Amplatz II). One patient had a single fistula. The fistulas originated from the right coronary artery in 3 patients, and from the circumflex artery in 2. They drained into the pulmonary trunk in 3 patients, into the right atrium in 1, and into a bronchial artery in the other. All fistulas were occluded completely in the catheterization laboratory, and the procedures were uncomplicated. At follow up, 3 patients underwent coronary angiography, and there was no evidence of recanalization. Transcatheter embolization in adults of single congenital coronary fistulas with detachable balloons and coils is safe and effective and can be regarded as an acceptable alternative to surgery.

Keywords: Coronary artery fistulas; transcatheter embolization; treatment

Coronary arterial fistula is an anomalous connection between a coronary artery and one of the cardiac chambers or vessels around the heart.¹,² Such fistulas are uncommon anomalies, found in approximately 1 of every 50,000 patients with congenital heart disease, and in 1 of every 500 patients who undergo coronary arteriography.²⁻⁵ They generally occur in isolation, and are the most common hemodynamically significant congenital anomaly of the coronary arteries.⁶ Children with congenital coronary arterial fistulas are usually asymptomatic, but the incidence of symptoms and complications increases with the age of the patient.¹,²,³,⁵⁻⁷ Because they rarely close spontaneously,⁸ most fistulas should be closed so as to prevent late complications.²⁻⁸⁻¹³ To date, surgical closure has been the standard treatment, with low rates of mortality and morbidity.²⁻⁷⁻¹¹⁻¹³ With the recent development of interventional procedures in congenital heart disease, and the increasing use of embolization techniques, several investigators¹²⁻²² have described catheter-based therapy as a potential alternative in the management of this disease. In this study, we report our overall experience with transcatheter embolization of congenital coronary fistulas in adults.

Correspondence to: Carlos A. C. Pedra, Instituto “Dante Pazzanese” de Cardiologia, Av. Dr. Dante Pazzanese 506, CEP 04012–180 São Paulo – SP, Brazil. Tel.: 5511(510-8541); Fax: 5511(571-5621); E-mail: pedraic@manic.com.br

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Methods

Patients (Table 1)

From November 1992 to November 1996, five patients underwent percutaneous transcatheter embolization of congenital coronary arterial fistulas. Our first patient has already been previously reported. Ages ranged between 47 and 70 years. There were 4 males and one female. All were symptomatic, suffering from angina and/or dyspnea, and had a continuous murmur best heard at the middle left sternal border. In one patient (case 2), color-Doppler echocardiography detected anomalous continuous flow in the pulmonary trunk, suggesting the diagnosis. Reversible myocardial ischemia was found in 2 patients undergoing exercise thallium-201 scan.

Coronary angiography (Table 2)

Routine right and left cardiac catheterization, as well as the interventional procedure, were performed by percutaneous entry to the femoral vein and artery, with local anesthesia, after having obtained informed consent. No heparin was given before the procedure. Selective coronary angiograms were performed, and angled views were employed to detail the anatomy of the anomalous vessel and coronary arteries. Each patient had a single fistula. The sites of origin and drainage of the fistulas are shown in Table 2. Fistulas originated from the circumflex artery in 2 cases, and from the right coronary artery in the other three. Three of them drained into the pulmonary trunk, one into the right atrium, and the one into a bronchial artery. In all cases, except in #5, coronary vessels were markedly dilated and tortuous (compare Figs 1 and 2).

Embolization procedures (Table 2)

A detachable balloon was inserted using a coaxial Debrun system in 4 patients. The Debrun delivery system is composed of a latex balloon (Ingenor) attached to the tip of a 2 French teflon catheter by an elastic mechanism. This catheter is itself passed through another 3.5 French polyethylene catheter. From a retrograde approach, a guiding catheter for angioplasty (Amplatz II – 8 French) was positioned at the origin of the coronary artery supplying the fistula. The coaxial delivery system was passed through this guiding catheter to the most distal segment of the feeding vessel. The attached balloon was inflated with isos-

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<th>Case</th>
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M: male; F: female; DE: dyspnea on exertion; Ang: angina; Pulp: palpitation; CE: cardiac enlargement; CM: continuous murmur; Rev isch: reversible ischemia; +: provided the diagnosis; #: not available

<table>
<thead>
<tr>
<th>Case</th>
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<th>Technique</th>
<th>Complications</th>
<th>Occlusion</th>
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LCxA: left circumflex artery; PT: pulmonary trunk; CS: coronary sinus; RCA: right coronary artery; LAD: left anterior descending artery; RA: right atrium; Angio: coronary angiography; occl: occluded; m: months; yrs: years; asympt: asymptomatic; no control angiography; +: death in a car accident.
Figure 1.
A: The right coronary artery in right anterior oblique projection before occlusion; the bizarre course of the fistula is conspicuous. The arrow shows the distal narrowing of the fistula. B: Right coronary angiography in the same view after the procedure: total occlusion of the fistula. The arrow shows the detachable balloon inflated.

Figure 2.
A: The right coronary artery in left anterior oblique projection before occlusion. A small coronary fistula arising from the initial third of the right coronary artery, draining into a bronchial artery in the right lung, is demonstrated. B: The arrow displays the single 35-5-5 Gianturco coil that was delivered in the fistula. C: Coronary angiography in the same view showing complete closure of the fistula. The arrow displays the position of the coil.
mocic contrast medium and, being flow directed, the whole system could be progressed smoothly to the chosen location. The position and the occlusive effect of the balloon were evaluated by repeat small injections of contrast material through the guiding catheter. If the position was not adequate, or if there was electrocardiographic signs of myocardial ischaemia, the balloon was immediately deflated and repositioned. The release mechanism was activated by pulling the 2 French teflon balloon catheter against the 3.5 French polyethylene catheter. The balloon was released only when it was known to be positioned distal to any normal myocardial branches. In one patient (case 5), the fistula was closed using one Gianturco coil (35–5–5, Cook Inc) delivered via a 5 French right coronary Judkins catheter passed through the guiding catheter (Fig. 2).

Immediately after embolization, repeat coronary angiograms were performed in order to assess the position of the devices and the presence of residual flow (Figs 1 & 2).

Follow up

Patients were observed in the intensive care unit for at least 24 hours. Careful clinical and electrocardiographic monitoring were performed, and serial levels of creatinine kinase were measured in 3 patients. Three patients were submitted to further coronary angiography 6, 7 and 12 months, respectively, after the procedure. One patient (case 3) died in a car accident 7 months after the procedure while in the other patient (case 4), only clinical follow up is available.

Results

Immediate results

Occlusion was successful in all patients, with immediate disappearance of the continuous murmur. In case 4, transient inversion of the T wave was observed during the stay in the intensive care unit, and this was associated with only a slight increase in the level of creatinine kinase. Both abnormalities had reverted to normal 2 days later. In the remaining patients, no electrocardiographic abnormalities or elevation of serum enzyme levels were noted. No patient showed evidence of arterial occlusion. The hospital stay ranged from 2 to 3 days.

Follow up results

Three patients (case 1, 2 and 5) underwent repeat coronary angiography 6, 7 and 12 months respectively after the procedure. They were asymptomatic, and showed no evidence of recanalization. In addition, there were no signs of deflation of the balloons. Although coronary angiography was not repeated in case 4, the patient was asymptomatic, and the size of the heart has returned to normal one year after the procedure.

Discussion

A congenital coronary arterial fistula is an uncommon malformation having similar incidences in males and females. Up to two-thirds of the fistulas arise from the right coronary artery, and rarely may take origin from both coronary arteries. About nine-tenths drain into the low pressures chambers or vessels, most commonly into the right ventricle, followed by the right atrium, the pulmonary trunk or arteries, the coronary sinus, and the superior caval vein. The pattern observed in our patients is in accordance with these observations, although it is somewhat unusual to have three of five draining to the pulmonary trunk. Generally, the fistula is a single tortuous vessel, but multiple fistulas have also been described. These fistulas are occasionally associated with other congenital heart defect. Due to increasing medical awareness, the fistulas are becoming readily recognized in adults, and some authors no longer consider them as a rare malformation.

This condition is rarely symptomatic in infancy or childhood. Most commonly, it presents as a heart murmur in a symptom-free child. After the second decade, the frequency of symptoms and complications increases. Fatigue, exertional dyspnea, atrial fibrillation, angina, myocardial infarction, endarteritis, aneurysmal dilatation with eventual rupture, and sudden death have all been reported. The cardinal physical finding is a continuous murmur usually heard best at the middle left or right sternal border. In our series, all patients were symptomatic and all had continuous murmurs. Small fistulas may cause no murmur, and are considered to have a relatively benign natural history. Electrocardiographic and chest X-ray findings are non-specific, and depend on the extent of the left-to-right shunt. Standard echocardiography with Doppler studies, including color flow mapping, is a useful noninvasive tool with which to establish the diagnosis. Transesophageal echocardiography can provide better imaging of the coronary arteries and the origin, course and drainage of the fistula. Magnetic resonance imaging has also successfully been employed to establish the diagnosis. Nuclear radioisotopic evaluation is usually helpful.
in demonstrating reversible myocardial ischaemia in adults.\textsuperscript{28-30} Definitive diagnosis is provided by cardiac catheterization with selective coronary cineangiography. Opacification of the involved coronary artery, which is often tortuous and markedly dilated, with the contrast material spurring into the chamber of drainage, are the main angiographic features.\textsuperscript{5,6,23} To date, surgery has been the standard method of treatment.\textsuperscript{2,9-11} Surgical closure can be recommended in adults, either on the basis of symptoms or to prevent future complications, and in symptomatic children.\textsuperscript{2,7,9-11} Due to low mortality rates, most authors have advocated early prophylactic surgical obliteration, even in the asymptomatic child.\textsuperscript{2,7,11,22} Although myocardial ischaemia and infarction have been reported after surgery,\textsuperscript{2,7,10} these complications are uncommon in the modern area.\textsuperscript{11} Surgical ligation, nonetheless, carries a significant risk of recurrence.\textsuperscript{2,7,10} Rates of mortality and morbidity are higher when surgery is performed at an older age.\textsuperscript{2,7,10} The operative procedure also involves a median sternotomy and, in about half the cases, requires cardiopulmonary bypass with its attendant morbidity.\textsuperscript{2,7,9-11,17}

With the development of interventional techniques, several reports have appeared of percutaneous therapeutic embolization.\textsuperscript{12-22} Different materials were employed with good results: Ivalon and micro particles,\textsuperscript{12,13} Gianturco coils,\textsuperscript{16,18} detachable coils,\textsuperscript{23} umbrella\textsuperscript{19} and detachable balloons.\textsuperscript{14-17,19-21} Because our group had been working with detachable balloons for other purposes, we were more familiar with this technique, which has some features that make it particularly useful in closing coronary fistulas.\textsuperscript{17,19,21} The release of the balloon is controlled, so an improperly placed balloon can be repositioned. "Test occlusions", with simultaneous inflation of the balloon, coronary cineangiography and electrocardiographic monitoring, can be performed prior to the final release. The balloon can usually be positioned smoothly because it is flow-directed. This property also helps to avoid dangerous manipulations with large catheters in distal coronary arterial branches. In addition, the balloon can be constructed to suit the size of the fistula, and, theoretically, a multichannel fistula can be occluded with one balloon since it assumes an elongated shape as it expands. There are some disadvantages, including the need for a large guiding catheter, which is less worrisome in the adult population, the risk of premature deflation of the balloon due to damage in the valve with consequent embolization to other sites,\textsuperscript{17} and cost, if compared to Gianturco coils.\textsuperscript{19} Coronary occlusion has also been reported after failure of the procedure.\textsuperscript{15} Detachable balloons have recently been implanted through 5 French sheaths,\textsuperscript{19} making the technique suitable for small children.

Although we do not have any experience with interlocking detachable coils, this technique has also been used with excellent results.\textsuperscript{22} Its main advantage is that the platinum coils can be introduced via 3 French catheters passing coaxially through a 5 French guiding catheter. Such low profile catheters allow the operator to advance them easily through the course of the fistula over a floppy guide wire to the required location.\textsuperscript{22} Complex fistulas can thus be addressed in this manner. In addition, coils can usually be retrieved if they embolize, a feature which is virtually impossible with deflated balloons.

After detailed selective coronary angiography, some specific technical requirements must be stressed.\textsuperscript{12,17,19,22} The artery leading to the fistula must be large enough to accept the guiding catheter and the delivery system without causing ischaemia, especially when using balloons. There must be a stenotic segment in which to anchor safely the device. The point of occlusion must be as distal as possible in the feeding artery, so that the risk of occluding normal coronary arterial branches is minimal. In fistulas with high flow, however, normal coronary arterial branches may not be clearly visualized due to a "steal" effect.\textsuperscript{17,21,32} Careful electrocardiographic monitoring prior to release of the device helps to minimize this problem.\textsuperscript{17,19} Temporary balloon occlusion of the proximal vessel with a Berman catheter can help to reduce flow across the fistula in this setting.\textsuperscript{22} A combination of techniques, utilizing detachable balloon and Gianturco coils in the same patient, may occasionally be needed to achieve complete occlusion of the fistula, especially if there are multiple feeding branches.\textsuperscript{16,17} Sometimes, arterial-venous guidewire circuits are required for occlusion, with antegrade release of steel coils and retrograde release of platinum coils.\textsuperscript{22} Rashkind umbrellas have also been implanted by a retrograde approach.\textsuperscript{18} Finally, when using detachable balloons, it is important to fill them with isotonic contrast medium to prevent early deflation or rupture due to osmotic mechanisms.\textsuperscript{17} There are several advantages for embolization as opposed to surgery, including short hospital stays, short recovery times, elimination of a thoracotomy, and reduced morbidity.\textsuperscript{16,22} Embolization probably results in less myocardial damage, and makes recurrence of the fistula less likely.\textsuperscript{17} General anesthesia was also avoided in the adults treated in our series. If the anatomy is complex, however, necessitating a versatile and flexible technique that is
not available for the operator at the time of the procedure, surgery remains an excellent option with very good long-term results.11,33

Despite the relatively small number of patients in our series, we conclude that percutaneous embolization with detachable balloons is a safe and effective alternative to surgical ligation in those adults with single coronary artery fistulas. With technical refinements, such as the use of interlocking detachable coils, transcatheater occlusion of coronary fistulas can even be employed in the treatment of the asymptomatic small child with either simple or complex fistulas.22

References


