Coronary Artery Fistulas: How to Manage Them

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Coronary artery fistulas are rare persisting vascular connections from a coronary artery to a cardiac chamber or major central blood vessel. The true incidence is difficult to discern because at least 75% may be asymptomatic and clinically undetectable until an echocardiogram or coronary arteriogram is performed. Small coronary artery fistulas that are not clinically detectable are not clearly associated with significant long term complications. Medium or large fistulas are associated with significant long term problems including angina, arrhythmias, myocardial infarction, endocarditis, and progressive dilation. Treatment options include surgical and catheter approaches to significant fistulas. The long term outlook after fistula closure is not well defined and deserves further study.

Key words: coronary artery fistula; coronary anomaly; congenital heart defect; catheter intervention

INTRODUCTION

Coronary artery fistulas are rare congenital malformations that can be defined as direct vascular connections from a coronary artery to a cardiac chamber or major central blood vessel without an intervening capillary bed. The true incidence of coronary artery fistulas is highly speculative since many may be small and never detected, or only detected incidentally with imaging for another indication. Coronary artery fistulas have been seen in ~0.3% of patients with congenital heart disease [1], 0.06% of children undergoing echocardiography [2], and 0.13–0.22% of adults undergoing coronary angiography [3–5]. At least 75% of coronary artery fistulas found incidentally are small and clinically silent.

There is controversy about the best management of coronary artery fistulas because both spontaneous regression and life threatening complications have been seen. Data about long term risks of clinically silent fistulas and the long term effects of interventions are particularly lacking [6]. Improved methods of detection, treatment, and follow-up have become available in the last 15 years, but there are no good large, complete, long-term comparative studies of a significant number of patients with coronary artery fistulas treated by specific strategies or left untreated. Our aim in this article is to attempt to review the limited available information along with our own experiences and discuss our current management strategies for patients with this rare malformation.

ANATOMY AND PHYSIOLOGY OF CORONARY FISTULAS

Coronary artery fistulas can originate from almost anywhere in the coronary artery system and terminate in any of the cardiac chambers, great veins, or pulmonary arteries. If one looks at older (preechocardiographic) series in which nearly all patients had large clinically detectable fistulas, ~60% originated from the right coronary artery, and there were multiple fistulas in only ~1% [7]. A more recent literature review that includes more small fistulas tabulated that ~55% arose from the left coronary artery system and 8% had multiple origins [8]. Small, asymptomatic fistulas arise much more commonly from the left coronary system (87%) [2]. In nearly all series, over 90% of fistulas drain to the right side of the circulation. Drainage to the left ventricle is least common.

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Coronary artery fistulas that drain to the right side of the circulation create a left-to-right shunt of oxygenated blood back to the pulmonary circulation. Those that drain to the systemic veins or right atrium have a circulatory physiology similar to an atrial septal defect. Those that drain to the pulmonary arteries are similar hemodynamically to a patent ductus arteriosus. A coronary fistula that drains to the left atrium results in no left to right shunt, but does cause a volume load similar to mitral regurgitation. Similarly, a coronary artery fistula that drains to the left ventricle produces hemodynamic changes similar to aortic insufficiency. In addition to shunts or volume loads on the heart, a coronary artery fistula can result in the coronary artery “steal” phenomenon with coronary blood preferentially passing through the fistula instead of into the more distal myocardial capillaries. Symptoms of angina have been reported in many patients with large fistulas, and may be aggravated by distal coronary artery disease. Anginal symptoms have disappeared with closure of the fistula in most patients with no distal disease [9].

TYPICAL CLINICAL PRESENTATIONS AND NATURAL HISTORY OF CORONARY FISTULAS

Patients with a coronary artery fistula can present in several common ways. Patients with the largest fistulas (relative to body size) may present with signs of congestive heart failure in infancy. Other neonates or infants with a fistula may be diagnosed during evaluation for separate congenital heart defects or because of a murmur. The typical murmur of a moderate or large coronary artery fistula is continuous with diastolic accentuation, but isolated systolic or diastolic murmurs are often described. Pediatric patients with moderate or large fistulas are most often brought to medical attention because of a murmur, but as many as 19% may have either a complication of the fistula or symptoms possibly related to the fistula [9]. Small, incidentally detected fistulas were not associated with symptoms or complications over a mean follow-up period of 9.3 years in the largest study of this patient subgroup [2]. Although symptoms and complications of medium or large sized coronary artery fistulas are not common in children, they become increasingly apparent in patients diagnosed after 20 years of age. In the literature review and analysis by Liberthson et al. in 1979, (which would likely include only moderate or larger fistulas) 63% of patients over age 20 years had either symptoms or a complication of the fistulas [9]. The most common symptoms and complications in adults include angina (especially if there is concomitant coronary artery disease), myocardial infarction, heart failure, arrhythmias, and endocarditis. A very small number of large fistulas have been documented to expand aneurysmally and even dissect or rupture [9–11].

FACTORS TO CONSIDER IN THE MANAGEMENT DECISION

The best management of a patient with a coronary artery fistula would provide the greatest probability of event-free survival over a lifetime. One must consider what is known about the risks of nonintervention versus the risks of any available intervention in the location and timeframe in which a decision is made. For any given patient there are many factors which may enter into the decision. Some of the more important ones in our opinion include the size of the fistula, presence of symptoms or complications likely related to the fistula, the age of the patient, the relative size of the fistula, the anatomy of the fistula, and whether the patient has other indications to be undergoing an invasive procedure.

Defining the relative size of coronary artery fistulas has not been systematically done, but it is clear that many of the symptoms and complications of the fistulas are greatly affected by size. Absolute size is a poor measure since a 3-mm fistula in a neonate may have far more detrimental effect than a similar diameter fistula in an adult. Left to right shunt ratios (QP/QS) are routinely measured at catheterization, but are not large (over 2:1) except in the most extreme cases. Aneurysmal dilation of portions of large, thin walled fistulas is common, but
the time-course of this dilation is not well understood. Fistulas are often irregularly shaped and tortuous. The smallest diameter of a fistula, frequently near the exit point or at a sharp bend of a tortuous vessel, is often considerably smaller than the largest portion. The excess flow through a fistula tends to be associated with dilation of the proximal portion of its associated coronary artery. Very distal coronary artery fistulas cause (or are associated with) dilation, and often tortuosity, of the entire length of the main coronary artery (Fig. 1). Those that arise near the origin of the coronary will generally have normal distal coronary artery anatomy (Fig. 2). In our practice we subjectively consider that trivial or small fistulas are ones that result in little or no dilation of the proximal coronary artery from which they arise, and are themselves no larger at any point than twice the normal expected proximate coronary artery diameter for the patient (Fig. 3). These small fistulas are usually, but not always, clinically silent and are generally detected incidentally. Fistulas that, at any point, are larger than two times but less than three times the expected proximate normal coronary artery diameter, or that are associated with similar ranges of dilation of the proximal associated coronary artery, are considered to be medium size fistulas. Fistulas with bigger dimensions are considered large.

Symptoms of coronary artery fistulas tend to vary with age and the size of the fistula. Heart failure related to a large volume shunt is most likely to occur in neonates or young infants with a large fistula. The neonatal symptoms from a large left-to-right shunt are often controllable with typical medical management and an initial trial of medications is indicated in very young patients. Those who do not respond to medical management need intervention in infancy. Intervention for asymptomatic patients may best be delayed for observation unless the fistula is large. Approximately half of symptomatic neonates are likely to become asymptomatic with the relative size of the fistula decreasing over time [12,13]. Spontaneous closure has been seen in some patients over time, but these cases appear to be in patients with small or medium size fistulas [8]. The most common symptoms in adults are angina, arrhythmias, or shortness of breath. These symptoms are most likely to occur in adult patients rather than children, but signs of coronary ischemia have been seen in some small children and should be sought. Heart failure related to myocardial ischemia is more likely to be seen in adults with concomitant coronary artery disease. Intervention is indicated in any patient with likely myocardial ischemia.

Endocarditis is a risk for any type of arterio-venous fistula and has been reported in ~3% of patients (both pediatric and adult) with a medium or large coronary artery fistula [9]. Endocarditis appears to be rare, but not unheard of, before teenage years. The risk of endocarditis in patients with clinically unapparent fistulas detected incidentally by echocardiography or angiography in modern times appears to be much less. Intervention
should be offered to any patient with a coronary artery fistula and a history of endocarditis or, at an appropriate age, if the patient has a large or medium sized fistula that is clinically detectable. Standard endocarditis prophylaxis precautions should probably be observed by any patient with any type of coronary artery fistula. We have not seen any reports of endocarditis in patients after successful closure of coronary artery fistulas. We also are unaware of any cases of endocarditis in patients with a “silent,” clinically undetectable, small coronary artery fistula. Details about the clinical findings in patients with endocarditis is lacking in most reports however. Therefore, it is not clear that an invasive procedure is warranted in this subgroup with the sole justification of endocarditis prevention, especially if the anatomy is difficult for catheter closure. We do conservatively recommend routine endocarditis prophylaxis for the presence of any small fistulas, even if they are not clinically apparent.

Age has a significant bearing on the decision to intervene in a particular patient at a particular time. There are numerous reports of spontaneous regression or even complete resolution of coronary artery fistulas in childhood. The size and developmental maturity of the patient may have some effect on the ease and safety of either surgical or catheter interventional procedures to close a fistula. Symptoms and complications of coronary artery fistulas are less common in children, but become more significant in adults [3,9]. Elective intervention for medium or large asymptomatic fistulas in childhood appears reasonable to prevent late complications. The exact age in childhood to intervene may depend on some of the other factors such as the anatomy of the fistula and whether the fistula appears to be growing or shrinking over time relative to the child’s size. Small, asymptomatic fistulas that are clinically silent may best be observed over time since the risks of complications or symptoms appear to be low and spontaneous closure is a possibility [14].

Approximately 10–30% of patients with a coronary artery fistula also have another congenital cardiovascular anomaly [3,8]. The most commonly seen defects include variations of Tetralogy of Fallot, patent ductus arteriosus, and atrial septal defect. If a pediatric patient is undergoing a surgical or catheter intervention for the additional defect, elective closure of even a small coronary artery fistula at the same procedure is justified if the fistula can be easily accessed. Similarly, intervention for even a small fistula may be justified in an adult who is undergoing a surgical or catheter procedure for an acquired cardiovascular problem such as coronary artery or valvular disease.

**SURGICAL CLOSURE OF CORONARY ARTERY FISTULAS**

The first reported successful attempt at surgical closure of a coronary artery fistula was by Fell et al. in 1958 [15]. Coronary artery fistulas may be closed by external pllication on the beating heart or by intracardiac closure using cardiopulmonary bypass with or without cardiac arrest. In recent series, bypass was utilized in...
Coronary artery fistulas should be rare (<1%) [17]. There have been occasional instances of periprocedural ST changes, myocardial infarctions, arrhythmias, and strokes. Recurrence or incomplete closure may occur in ~10% [3,18]. Most recurrent or residual shunts have been small and repeat surgical procedures have rarely been reported [19]. Surgical approaches for coronary artery fistula closure can be readily utilized in patients of all ages and sizes, and surgical closure is the preferred approach in patients who are undergoing operative repair of other cardiovascular problems [20]. The surgical approach can also allow for reduction in the size of very large aneurysmal dilations of either the fistula or the proximal coronary artery [21].

CATHETER DEVICE CLOSURE OF CORONARY ARTERY FISTULAS

Transcatheter occlusion of a coronary artery fistula was first reported in 1983 [22]. Since then there have been multiple reports of series including up to 33 patients [18,23–28]. Results have generally been comparable with reported surgical results with an expected mortality rate of <1%. There have been rare instances of periprocedural ST changes, myocardial infarctions, or arrhythmias. Occluder device embolizations have occurred in a number of reports, but the devices have been removed successfully in the catheterization lab in nearly all cases. Many different types of occluding devices have been utilized including detachable balloons, Gianturco and other types of coils, Rashkind and Amplatzer PDA and ASD devices, Grifka Bags, and Amplatzer Muscular VSD Device, and Vascular Plugs. Improved devices and delivery systems have made catheter closure applicable to over 90% of patients, including infants and some neonates, in centers with extensive experience [29–31]. Catheter techniques may be difficult or impossible in a small percentage of patients due to extreme vessel tortuosity and inability to deliver a catheter far enough distally, presence of multiple drainage sites, presence of normal coronary branches too close to the drainage site to allow selective occlusion, or very small size of the patient (neonate requiring treatment of large fistula). Small residual or recurrent leaks have been seen in ~10% of patients treated by catheter techniques [18,30]. Catheter interventions for residual small shunts after either surgical or catheter interventions are readily repeatable if necessary. Because of a much shorter recovery time and avoidance of a scar, transcatheter closure is considered the procedure of choice in applicable patients.

The basic technique of transcatheter closure is to advance a delivery catheter to the most distal portion of the fistula (past any branches that may feed normal myocardium) and place the occluding device in that location. In smaller fistulas, it may not be difficult to deliver small devices like coils directly through a small arterial catheter (Fig. 3). Controlled release coils or delivery techniques may widen the spectrum of applicable patients [32]. Small diameter coils can be delivered through 3F microcatheters coaxially placed through a coronary guide catheter [27]. In some large, high flow fistulas, we have found that it may be advantageous to first pass a small catheter and/or guidewire completely through the fistula. The arterial guidewire can then be snared and exteriorized on the venous side to provide a guidewire rail. This arterio-venous guidewire rail can be used to position even a large delivery system through the vein in preference to the retrograde arterial approach. An adequate size closure device such as an Amplatzer PDA device or Amplatzer Vascular Plug can be precisely deployed using this technique. If the fistula itself is large and lengthy with its origin proximally from a main coronary artery, we generally will try to fill the fistula with coils or also place a proximal occluding device. We feel that this may reduce the risk of clot propagation into normal structures or later dilation of the distally occluded and possibly thin-walled fistula (Fig. 2). A covered stent may be a good device to close some coronary artery fistulas [33]. The origin of the fistula from the normal coronary artery is excluded by placement of a covered coronary artery stent in the main coronary vessel. This is an especially useful technique in adult patients with coexisting coronary artery disease.

LONG TERM OUTCOMES AFTER TREATMENT OF CORONARY ARTERY FISTULAS

There is limited longitudinal information about the long term outlook for patients with coronary artery fistulas following surgical or transcatheter treatment. Many papers indicate that most patients remain clinically asymptomatic for a number of years after a closure procedure, but there are concerning findings from papers in which follow-up has been more detailed. Small residual leaks after either type of procedure appear to be present in ~10% of patients, but there is tremendous variability between series. As expected higher rates are seen if follow-up evaluations have included detailed echocardiographic or angiographic study in all the patients [3]. Other residua and sequelae of the fistulas and closure procedures include persistent dilation of the coronary artery, late stenosis in tortuous coronary arteries, arrhythmias, and late occlusion of dilated and tortuous coronary arteries with or without myocardial infarction.
In one of the most rigorous follow-up studies to date, Cheung et al. performed coronary arteriograms on 21 of 41 asymptomatic, mostly adult, patients after surgical fistula closure and found 20 of the 21 studies to show a persisting significant abnormality [3]. There was persisting proximal coronary dilation in 16/21. In six of these 16, the distal coronary artery was normal, but in 10, the distal vessel was threadlike or completely occluded. The remaining four patients had a small residual fistula with the exit at the original or a previously unseen exit location. McMahon et al. found persisting coronary artery dilation in all four of their pediatric patients 4 years after catheter closure of large fistulas performed at 1.5–3 years of age [26]. Hiraishi et al. found an absolute or relative decrease in coronary artery size in three children after surgical fistula closure. IVUS and angiography however showed areas of stenosis and intimal thickening in two of three who demonstrated an absolute reduction in coronary artery size. Inspite of normal exercise ECGs, two out of their three patients had abnormalities on thallium stress testing. At our institution, three of four patients who recently underwent follow-up evaluations 4–41 years after surgical fistula closure were found to have thrombosis of the entire parent coronary artery. Thus, even though most patients are asymptomatic after coronary artery fistula closure, they should not be dismissed from follow-up. It may be prudent to follow them with appropriate tests to evaluate myocardial perfusion. Long term anticoagulation with at least an antiplatelet agent appears to be advisable and we currently recommend warfarin for patients with more than moderate persisting aneurysmal dilation of the coronary arteries.

**SUMMARY OF RECOMMENDATIONS**

There appears to be good consensus that all symptomatic patients should undergo closure of medium or large coronary artery fistulas. In our opinion, trivial to small (not clinically discernable), asymptomatic fistulas do not require a specific procedure for closure simply because they are detected incidentally. However, closure of even small fistulas may be recommended (1) if the patient is considered to be at high risk for later endocarditis, or (2) if longitudinal follow-up is not feasible, or (3) if the patient is undergoing an invasive procedure for some other cardiac problem. The decision of whether, and when, to submit a patient with a clinically detectable, but asymptomatic, moderate or large fistula can be difficult. The risk of endocarditis in patients detected clinically in earlier studies is not insignificant (3%). The incidence of symptoms related to clinically apparent fistulas increases significantly with advancing age (especially after age 20). Surgical and catheter closure procedures are relatively safe and catheter procedures leave no residual scar and are well tolerated. In children, we generally recommend elective closure of any coronary artery fistula which remains clinically apparent past 3–5 years of age, even if the patient is asymptomatic.

The risk of closure may be less, and the risk of late giant aneurysm higher, in coronary artery fistulas that arise near the coronary artery origin (Fig. 2). Fistulas that arise proximally in a coronary artery lead to dilation of only a short segment of the proximal true coronary artery. These types of fistulas in our experience may be the most likely to become grossly dilated and may be at risk for rupture. We would close these types of fistulas at any age.

We are most uncertain about medium or large fistulas that arise very distally in the coronary system and are already associated with long segment coronary artery dilation at the time of detection. Longitudinal studies are lacking, but it seems reasonable that the longer abnormally high flow persists in a coronary artery with a distal fistula, the more likely for the coronary to become increasingly dilated and tortuous. We generally recommend closing such fistulas early if discovered in children. Recent studies indicate that late stenosis due to intimal hyperplasia may still leave these children at risk for later myocardial ischemia, but we hope that relatively early intervention may lessen the secondary changes. In older patients, if the proximal coronary has become grossly enlarged, closure of distal fistulas has been associated with a very high incidence of late thrombosis of the permanently dilated and tortuous proximal coronary artery. If the older patient is asymptomatic, the best course of action in this scenario is unclear to us. Closure of the fistula results in relative stasis in the giant coronary artery, and this is likely the etiology of late thrombosis. These types of fistulas can be difficult to close by catheter techniques. Since late thrombosis is likely and may be no better than late development of the coronary “steal” syndrome, nonintervention may have a similar outlook to surgical closure, without the thoracotomy. It is not known whether innovative approaches such as surgical reduction of the coronary artery would be beneficial or detrimental. A conclusive trial of intervention versus novel therapy or expectant medical management for these patients with the worst coronary artery fistulas will be nearly impossible because of the relative rarity of this specific anatomy.

Coronary artery fistulas are rare but fascinating anomalies. The long term outlook of specific types of coronary artery fistulas is sketchy at best. These fistulas can be closed, but our understanding of the long term outcomes of patients after fistula closure is woefully lacking. We feel that this lack of information warrants following patients with coronary artery fistulas, whether closed or...
not, indefinitely. Routine noninvasive, or even invasive, studies of the anatomy and function of the coronary system in these patients should be the standard of care.

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