Coronary Artery Fistulas; in Adult Period

Erişkin Yaş Grubunda Koroner Arter Fistülleri

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Geliş Tarihi/*Received:* 02.07.2008
Kabul Tarihi/*Accepted:* 02.09.2008

This study was presented 57th ESCVS International Congress, Interactive Cardio Vascular and Thoracic Surgery, Barcelona, Spain Vol 7(1) S115 (2008).

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ABSTRACT Objective: Coronary artery fistulae are rare anomalies. These are rarely seen in the adult, particularly with additional cardiac pathologies. Therefore we present our surgical experience and results carried out in adult patients with additional cardiac pathologies. **Material and Methods:** Between March 1998 and November 2007, five patients were incidentally diagnosed as having coronary artery fistulae by cardiac catheterization which was performed for investigation of ischemic or valvular heart disease. Coronary angiography and 2D echocardiography were used to assess coronary artery and fistula anatomy, before determining eligibility for surgical treatment. All of the patients were operated under cardiopulmonary bypass and median sternotomy. With the help of cardioplegia, the fistula's orifices were located. Initially, coronary fistulae were ligated. Associated cardiac disease was corrected by surgical procedure. **Results:** The median patient age was 40.6 years (range: 27 - 52 years). Two of the patients were female. The median patient follow-up time after surgical closure of fistulae was 13 months (range 4 - 37 months). Four patients had no residual shunts, multislice CT was performed to confirm this. In patient 4, we observed that two of the multiple fistulae from RCA to the PA persisted 6 months later for which coil occlusion was performed then with success. Subsequent echocardiography shows a trivial persisting fistula communication. No further intervention was planned. **Conclusion:** Surgical closure of CAF is technically safe, and indicated especially in the presence of additional cardiac pathology in adult period. The percutaneous closure for CAF is unsuccessful; surgery is the reasonable next option.

Key Words: Coronary vessels; vascular fistula; adult

ÖZET Amaç: Koroner arter fistülleri çok az rastlanan bir anomalidir. Erişkin yaş grubunda, özellikle kardiyak ek bir patoloji ile çok nadir görülen bir anomalidir. Bu nedenle erişkin yaş grubunda görülen ve eşlik eden ek kardiyak patoloji ile beraber sonuçlarımızı irdelemeyi ve cerrahi deneyimlerimizi sunmayı uygun bulduk. **Gereç ve Yöntemler:** Mart 1998-Nisan 2008 tarihleri arasında iskemik veya kapak hastalığı nedeni ile koroner anjiyografi yapılan ve koroner arter fistülü saptanan 5 hasta çalışmaya alındı. Cerrahi tedavi stratejisi için, sağ-sol koroner anjiyografi ve 2D ekokardiyografi ile koroner arterler, koroner fistüllerin anatomisi ortaya konuldu. Tüm hastalar kardiyopulmoner bypass altında ve median sternotomi ile opere edildiler. Fistülün köken aldığı yer antegrad kan kardiyopleji ile saptandı. Önce koroner arter fistül ligasyonu ardından ek patoloji giderildi. **Bulgular:** Hastaların ortalama yaşları 40.6 (yaş aralığı: 27-52 yıl), hastaların 3'ü erkek 2'si kadın hasta idi. Hastaların ortalama takip süresi 13 ay idi (4-37 ay). Çok kesitli bilgisayarlı tomografi ile olguların 4'ünde fistüllerin kapandığı izlendi. Dördüncü hastada sağ koroner arter ile pulmoner arter arasında 2 adet koroner fistül saptandı. Ameliyat sonrası 6. ayında koil embolizasyon ile fistüller kapatıldı. 12. ay ekokardiyografik takibinde hemodinamik olarak önemsiz rezistans fistül saptandı. Ancak ek tedavi düşünülmedi. **Sonuç:** Koroner arter fistülü cerrahi olarak güvenle kapatılabilir. Perkütan olarak kapatılamayan veya eşlik eden başka kardiyak cerrahi problemi olan olgular cerrahi olarak güvenle kapatılabilir inancındayız.

Anahtar Kelimeler: Koroner arter; fistül; erişkin

Turkiye Klinikleri J Med Sci 2008;28(6):886-91

Coronary artery fistulae (CAF) are anomalous connections between coronary vessels and the cardiac chambers and/or the great vessels. They are usually congenital, but also may be acquired.[1] CAF is an infrequent abnormality. The incidence of CAF in the overall population is estimated to be 0.002%; coexistence with other cardiac pathology is even less common.[2,3]

Occasionally, clinical diagnosis of CAF can be made by detecting a continuous heart murmur in the upper precordial area.[4] Fistulae are usually detected by means of angiography. CAF tend to get larger with age, so early electively closure is usually recommended in patients who have continuous murmurs and/or systolic murmurs with an early diastolic component.[5]

The mean age at diagnosis is 7.2 years, and a male predominance has been well established. Although the majority of patients remain asymptomatic for life, some do have fistulae that progress to sympomatic state or they are usually found incidentally at the time of coronary angiograpy for other reasons like atherosclerotic heart disease. Although most patients suffering from CAF remain asymptomatic during childhood and adolescence, many of them develop complications in adulthood due to volume overload, endocarditis or ischemia. The clinical presentations include fatigue, dyspnea, orthopnea, angina, endocarditis, arrhythmias, stroke, myocardial ischemia or myocardial infarction.[6,7]

Surgical closure has been described as the gold standard for closure of CAF.[8] The main indications for closure are clinical symptoms especially of heart failure and myocardial ischemia, and in asymptomatic patients with high-flow shunting, to prevent occurrence of symptoms or complications.[6]

We present five patients with CAF who were in relatively older ages. One patient with isolated CAF and 4 patients with CAF with other cardiac diseases were included in this study.

MATERIAL AND METHODS

Between March 1998 and November 2007, 5 patients incidentally diagnosed as having CAF by cardiac catheterization. Medical histories of these patients are detailed in Table 1. The origin and the drainage of CAF are shown in Table 2. The median patient age was 40.6 years (range: 27-52 years) and two of them were female (Table 3). One patient had severe mitral valve disease, one patient had severe aortic valve disease; other two patients had coronary artery disease (Table 3). Patient 4 had isolated CAF with documented ischemia on Thallium stress test with no significant lesions on the coronary angiogram and a high pulmonary artery pressure. Informed consent was obtained from all patients before intervention. Selective right and left coronary angiography and 2D echocardiography were used to assess coronary artery and fistula anatomy, before determining eligibility for surgical treatment.

All of the patients were operated under cardiopulmonary bypass. Surgery was performed through a median sternotomy. Extracorporeal circulation was instituted using bicaval cannulation or two stage venous cannulation and arterial cannulation of the ascending aorta; initial antegrade isothermic blood cardioplegia maintained by retrograde blood cardioplegia and moderate hypothermia was applied. The draining orifices of the fistulae were located in the right atrium and the pulmonary artery with the help of cardioplegia via

TABLE 1: Past medical history of our patients (n= 5).

Characteristic	No of pts.
Hypertension	3
Diabetes Mellitus	2
Current smoker	1
Former smoker	1
Hypercholesterolemia	3
Cardiac history	
Coronary artery disease	2
Prior myocardial infarction	1
Prior coronary bypass	N
Mitral valve disease	1
Aortic valve disease	1
Arrhythmia	1
Chronic obstructive pulmonary disease	N
Chronic renal insufficiency	N
Peripheral vascular disease	1

TABLE 2: Locations of origins and drainage sites of CAF and surgical procedures performed on 5 patients.

Patient	CAF (Origin)	CAF (Drainage)	Surgical procedure
1	RCA	RA	MVR + Lg and Tr.
2	LAD	RA	CABG + Lg and Tr.
3	Cx	RA	CABG + Lg and Tr.
4	RCA	PA	Lg and Tr.
5	LAD	PA	AVR + Lg and Tr.

CAF: Coronary artery fistulae, RCA: Right coronary artery, LAD: Left anterior descending, Cx: Circumflex artery, PA: Pulmonary artery, MVR: Mitral valve replacement, AVR: Aortic valve replacement, CABG: Coronary artery bypass grafting, Lg and Tr: Ligation and Transfixion.

the aortic root. Initially, coronary fistulae were ligated by 5/0 polypropylene and transfixion sutures by 5-0 prolene in the outlow portion. After ligation of CAF, associated cardiac disease was corrected by surgical procedure (mitral valve replacement in 1, aortic valve replacement in 1, coronary artery bypass grafting in 2 patients).

Additionally, radiofrequency ablation was performed in one patient with mitral valve disease. Radiofrequency ablation was carried out with a Cardioblate BP (bipolar) surgical ablation device (Medtronic 60821; Medtronic, Mineapolis, MN, USA).

Follow-up was done by 2D echocardiography in all patients and angiographic control was made in the fourth patient.

RESULTS

Table 2 summarizes the origin and drainage of CAF and associated surgical procedure together with the closure of CAF. The median patient follow-up from the time of surgical fistula closure was 13 months (range 4-37 months). At last follow-up, 4 patients had no residual shunts. In patient 4 who had multiple complex fistulae to the pulmonary arterial tree, cardiac catheterization was performed which showed residual shunt. In this patient, two of the multiple fistulae from RCA to the pulmonary artery persisted 6 months later for which coil occlusion were performed with success. The only late complication was the development of a femoral artery aneurysm following coronary angiography in the fifth patient, which required surgical repair. No myocardial infarction due to the procedure or wound infection was observed during follow-up.

DISCUSSION

Recent advances in the techniques of selective coronary arteriography and echocardiography have led to the increased detection of many CAF. CAF's are rare anomalies and account for 0.27-0.4% of all congenital heart defects. The diagnosis can be achieved by catheterisation and coronary angiography. When the diagnosis achieved, transcatheter or surgical closure is indicated to prevent the early and late complications due to fistula.[9] The inci-

TABLE 3: Clinical data of patients undergoing cardiac surgery for coronary artery fistulation.

Patients	1.	2.	3.	4.	5.
Age (years)	27	42	44	38	52
Sex	Male	Female	Male	Male	Female
Body Surface Area (m^2)	1.36	1.40	1.52	1.38	1.63
BP (mmHg)	130/70	140/80	110/60	100/50	120/50
ECHOCARDIOGRAM					
LVDd (mm)	58	50	56	40	44
LVDs (mm)	47	34	46	30	33
LVEF (%)	40%	50%	45%	50%	50%
IVSd (mm)	0.9	1.1	1.3	1.0	0.9
PAP (mmHg)	55	35	60	35	40
Qp/Os	1.3	1.2	1.3	2.0	1.4

BP: Blood pressure, LVDd: Left ventricular end-diastolic diameter, LVDs: Left ventricular end-systolic diameter, IVSd: Interventricular septum end diastolic diameter, PAP: Pulmonary arterial systolic pressure.

dence is reported to be around 0.08% in our country.[10]

Persistence of large intertrabecular spaces of the embryonic development is involved in the origin of this abnormality.[11] The sinusoidal intertrabecular network closes with myocardial thickening and forms a capillary network during normal embryonic development. The retention of this network in the normal heart forms small communication from the coronary arteries through the myocardium as well as Thebesian veins.[12,13]

The incidence ranges from 0.3% to 0.8% in patients who undergo coronary angiography.[14] Much of the blood flow to the capillaries is bypassed via the arterio-sinusoidal vessels because the normal coronary circulation usually has a greater resistance than the fistulae. As a consequence, a substantial amount of the arterial blood drains by way of these anomalous pre-capillary vessels directly into the left ventricle, resulting in left ventricular diastolic volume overload.[15] Therefore the natural course is the enlargement of the fistulae over time; although a few cases of spontaneus closure have been reported.

Patients usually become symptomatic during the 5th or 6th decade of life and may present with sudden cardiac death, myocardial ischemia, systolic hypertension, heart failure, arrythmia, rupture or endocarditis.[7,9,16] Kimura et al reported that ratio of symptomatic patients was 30%. The same study showed that the ratio of symptomatic patients was 9% in patients younger than 20 years and 55% in patients older than 20 years.[17] The higher the flow rate through the fistula is the higher is the risk of bacterial endocarditis. Thus, antibiotic prophylaxis may be necessary.[6,18]

The most frequent sign of CAF is continuous murmur at the left parasternal region extending to the apex which was present in three of our patients. But in the presence of coexisting valvular pathology, murmur due to fistula may be inconspicuous.

Enç et al reported nonspecific ECG findings in 54% of the cases. Cardiomegaly and increased pulmonary vascularity are present in 5% cases. Cardiac catheterizarion and coronary angiography are still widely accepted diagnostic methods because they define the precise anatomy and reveal additional anomalies.[18- 21]

Clinical presentations are variable depending on the fistula, shunt volume, site of the shunt, and presence of other cardiac conditions.[22] The Hobbs et al series included the greatest number of patients, whose fistulae were classified according to their origin.[18,23] In this series, 50% of fistulae originated from RCA, 42% from left coronary artery (LCA), and 19% from both RCA and LCA. Moreover, the arrangement of the fistulae according to their drainage were as follows: 41% to RV, 26% to RA, 17% to PA, 5% to LA, 3% to LV, 7% to coronary sinus, and 1% to SVC.

The first case of a CAF was reported by Krause in 1865.[24] The priority in surgical treatment of CAF belongs to Bjork and Crafoord.[25] Percutaneous closure also has been described as an alternative and less invasive procedure. Various percutaneous catheteter devices have been used to close such fistulae, including coils, detachable balloon, alcohol foam, and double umbrellas.[7,26,27]

CAF occlusion is rarely reported in the literature. The spontaneous occlusion rate for CAF hovers between 1% and 2%. The most effective factors for spontaneous occlusion are the diameter of the fistula and flow rate.[7]

If there is a moderate or high flow of shunt or the patient is symptomatic, regardless of the age of the patient, surgery is recommended.[28] Surgery can be performed without CPB if the chamber into which the fistula drains will not be exposed. Some surgeons recommend CPB for all cases in order to avoid ligation from the wrong artery and to definitely occlude the fistula.[29] We operated on all of our patients under CPB since four of them were already going to be operated due to other cardiac pathologies. Our patient with isolated CAF was not technically eligible for off-pump closure, as well.

Recently, new methods have been developed to occlude the CAF by transcatheter intervention.[30] These methods include coil embolization, intravascular embolization with chemical embolization, and balloon occlusions. There are no

reported studies that cover the long-term results obtained through transcatheter occlusion interventions in detail. Although transcatheter occlusion interventions are less invasive, they cannot be used in all cases. Coil embolization of the largest fistula was not found to be an appropriate technique for management.[31] Transcatheter closure of CAF with coils is well described. The use of novel devices may offer advantages such as improved control of device placement, the use of a single device instead of multiple devices, and high rates of occlusion.[31]

Patients with CAF should undergo surgical operation if one of the following indications is present; I) ischemia on myocardial SPECT, II) Symtomps or signs of heart failure, III) Qp/Qs>1.5. IV) Another indication of cardiac surgery.[4] Our patients were not appropriate for percutaneous closure of CAF. Four patients required cardiac surgery since they had additional cardiac pathology and one patient with isolated CAF had previously had unsuccessful percutaneous closure attempt for CAF. Interestingly, we referred one patient for coil embolization because of having residual shunts after surgical procedure.

In this study, surgical closure of CAF was proved to be technically safe, especially in the presence of additional cardiac pathology. If the percutaneous closure for CAF is unsuccessful, surgery is the reasonable next option.

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