

## Transcatheter Coil Embolization in Coronary Artery Fistulae

**Budi Y. Setianto<sup>1,2</sup>, Pudya L. Arshant<sup>2</sup>**

<sup>1</sup> Department of Cardiology and Vascular Medicine, Gadjah Mada University - dr. Sardjito Hospital. Jl. Kesehatan Sekip no. 1, Yogyakarta, Indonesia.

<sup>2</sup> Department of Internal Medicine, Gadjah Mada University - dr. Sardjito Hospital, Yogyakarta, Indonesia. Correspondence mail: budyuls@yahoo.com.

### ABSTRAK

*Fistula arteri koroner atau coronary artery fistulae (CAF) merupakan kelainan yang jarang terjadi, sebagian besar bersifat kongenital dengan insidens sebesar 0,1% - 0,2% dan insidens CAF yang didapat sebesar 0,2%, sebagaimana ditunjukkan oleh angiografi koroner. CAF dengan pintas aliran tinggi (high flow shunting) atau yang bersifat simtomatik harus ditangani dengan pembedahan atau teknik tertentu tanpa pembedahan seperti embolisasi dengan kumparan lintas kateter atau transcatheter coil embolization (TCCE).*

*Artikel ini menyajikan suatu kasus, yakni seorang wanita berusia 63 tahun dengan keluhan utama nyeri dada sejak satu bulan yang lalu. Pasien telah menjalani angiografi koroner yang menunjukkan adanya 95% penyempitan sisi proksimal arteri desenden anterior bawah atau LAD (left anterior descendens) dan CAF dari LAD ke atrium kanan. Untuk penyempitan LAD sebelah proksimal telah ditangani dengan PCI (percutaneous coronary intervention) dan implantasi stent bersalut obat (drug eluting stents), tetapi pasien tetap mengeluhkan nyeri dada yang dialaminya. Para dokter memutuskan untuk melakukan TCEE untuk menangani CAF. Setelah dilakukan TCEE, pasien tidak lagi mengeluhkan nyeri dada dan evaluasi dengan EKG menunjukkan hasil yang normal.*

**Kata kunci:** fistula arteri koroner, simtomatik, transcatheter coil embolization.

### ABSTRACT

*Coronary artery fistulae (CAF) is a rare anomaly, mostly with the incidence of congenital CAF 0.1% - 0.2% and the incidence of acquired CAF 0.2% at coronary angiography. Symptomatic or high flow shunting CAF must be treated with surgery or non surgery approach such as transcatheter coil embolization (TCCE).*

*A women 63 years old with chief complain of chest pain since one month ago. The patient had undergone coronary angiography with conclusions as follows 95% narrowing of proximal LAD (left anterior descendens) artery and CAF from LAD into the right atrium. Whereas narrowing of proximal LAD was performed PCI (percutaneous coronary intervention) with the implantation of drug eluting stents, but patients still complained chest pain. CAF decided to do the TCCE. Post TCCE chest pain and ECG evaluation were within normal limits.*

**Key words:** coronary artery fistulae, symptomatic, transcatheter coil embolization.

### INTRODUCTION

Most coronary artery fistulae (CAF) are asymptomatic, but some are symptomatic because of manifestation of coronary steal such as angina pectoris, myocardial infarction, heart failure, endocarditis, and aneurysm formation. Triad diagnostic of CAF is a location of abnormal

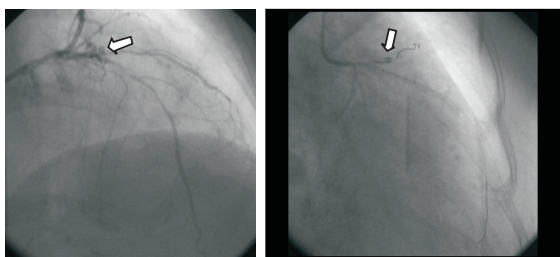
murmur 'to and fro' corresponding persistent ductus arteriosus (PDA), a left to right shunt in atrium or ventricle and tortuosity of coronary.<sup>1-3</sup> The gold standard diagnostic of CAF is coronary angiography.<sup>4,5</sup>

Indications therapy of CAF are asymptomatic patients with high - flow shunts and the presence

of symptoms. The goal of therapy to prevent symptom or complication.<sup>2,5-7</sup> Clinical signs related to increased morbidity and mortality. Fistulae-related complications especially in elderly and mortality increases with age.<sup>8</sup> Therapy strategies of CAF are divided into conservative therapy, surgical and transcatheter coil embolization (TCCE). The success of surgery is quite high but the risk of cardiac arrest.<sup>1,9</sup> TCCE technique is choice of treatment because of the cost-effectiveness, and less risk compared than surgery.<sup>7,9</sup>

### CASE ILLUSTRATION

A 63-year-old female patient that was treated with chief complaint of chest pain came and went since one month ago. The patient, non-obese diabetes mellitus (DM2NO), hypertension and coronary artery disease, one vessel disease (CAD-1VD) post stenting dated January 15, 2010 with the conclusion in the following, proximal LAD 95% stenosis, found the coronary artery fistulae from LAD into the right atrium. Other vessels are within normal limits. On physical examination there is no murmur or others abnormality. ECG finding shows ventricle extra systole (VES). The working diagnosis is observation of chest pain may caused by CAF, CAD-1VD post stenting, VES, hypertension and DM2NO. The patient is administered by clopidogrel 75 mg twice a day, bisoprolol 5 mg once a day, valsartan 160 mg once a day, and Insulin gargline 26 units subcutaneous a day. The patient underwent transcatheter coil embolization (TCCE) on February 17, 2010 in the catheterization laboratory of Dr. Sardjito Hospital, Yogyakarta. The result had been installed two coil units with no residual fistulae, no chest pain anymore and no significant complications. Another problem has not been controlled was hypertension, so that given additional therapy with amlodipine 5 mg once a day and patients discharged after controlled blood pressure.



**Figure 1.** CAF from the LAD into the right atrium pre and post TCCE

### DISCUSSION

Coronary artery fistulae (CAF) is a relationship between one or more coronary arteries with a heart space or large vessel through myocardial basis, was first reported by Krauss in 1865.<sup>1-3</sup> In embryology, CAF divided into congenital and acquired. Congenital coronary artery fistulae caused by failure of obliteration of the sinusoids intramyocardial is due to defects in embryonic development of coronary arterial branches between the ages of 6-8 weeks of life.<sup>1,10</sup> Postulate pathophysiology of congenital CAF originating from the two forms of embryologic growth disorders. Based on fetal growth retardation, Steinberg, Dotter and Baldwin divided CAF into type 1 due to coronary capillary element differentiation defect and type 2 are anomalous origin of coronary arteries with concomitant inter-coronary relationship due to defect formation of a primitive septum bulb.<sup>11</sup> Approximately 0.25 to 0.4% CAF associated with other congenital heart diseases such as atrial septal defect (ASD), tetralogy of Fallot (TOF), persistent ductus arteriosus (PDA), ventricular septal defect (VSD) and pulmonary atresia.<sup>8</sup> Acquired coronary artery fistulae can result from blunt or penetrating trauma, post-cardiac surgery (valve replacement, transplant-coronary artery bypass, coronary angioplasty, re-post myocardial biopsy in heart transplant) and the omission of CABG through the coronary veins or complications of myocardial infarction (rare).<sup>1-3</sup> Normal anatomy of the main coronary artery has two branches of right coronary artery (RCA) and left main coronary artery (LMCA), which branched into left circumflex (LCX) and left anterior descendens (LAD).<sup>13</sup> Normally, the two major coronary arteries from aortic sinus occur consecutively, but one or more branches of the fistulae connects directly with a heart chamber or pulmonary trunk, coronary sinus, superior vena cava or pulmonary veins showed a shortcut left-to-right. Great shunts happen if CAF ended up in the atrium space.<sup>14</sup> Coronary artery fistulae can arise from several parts of the three major coronary arteries. CAF generally grow from the RCA or LAD, LCX is rarely involved. Most of the CAF originated from the RCA or the branches (55%), 30% of the LAD and 5% bilateral.<sup>2,5,15</sup> Ninety percent of the CAF ended on the right side of the heart, usually on the single surface. Location drainage is the most

low-pressure structure, including the right of heart chamber, pulmonary artery, superior vena cava and coronary sinus. Most end up in the right ventricle (45%), then the right atrium (25%), pulmonary trunk (15%), coronary sinus (7%), left atrium (5%) and left ventricle (3%).<sup>2,7,8,16</sup>

Coronary artery fistulae in these patients was likely congenital because there was no history of or risk factor for acquired CAF. LAD coronary fistulae originated from and ended up in right atrium. LAD artery as a place of origin of CAF found on 30% of cases, and CAF in the right atrial endings in 25% of cases. Forty percent up to 55% patients with CAF are asymptomatic. Patients can be asymptomatic in several years, then experienced symptoms of cardiovascular disorders such as growth and development in childhood, fatigue, dyspnea, orthopnea, palpitations, angina pectoris, myocardial ischemia or infarction, stroke, congestive heart failure, chest pain atypical, subacute bacterial endocarditis or arrhythmias (such as premature ventricular contractions, atrial tachycardia, ventricular tachycardia, bundle branch block and 1st degree atrioventricular block) or sudden death.<sup>4,5,7,17</sup> Clinical presentation of the CAF is influenced by age, severity of left-to-right shunt, the size and structure of relationships. In theory, CAF 'steal' of coronary blood flow (coronary steal) from the origin of coronary artery fistulae, resulting in a decrease in coronary blood flow resulting in ischemia of the distal CAF or myocardial infarction which gives the manifestation of chest pain.<sup>5,10</sup> These alerts can be worsened by other cardiac conditions such as coronary artery disease, valve disease and myocardial hypertrophy.<sup>1</sup> Physical examination revealed no sign if there was no ischemia or myocardial infarction. The first clinical signs may include sudden death in young athletes and military personnel.<sup>18,19</sup> Most asymptomatic patients referred for unexplained angina, congestive heart failure or continuous murmur that sounded most loudly in the precordial. Murmur tends to crescendo-decrescendo systolic and diastolic, but even harder on the diastolic. Conversely, most continuous murmur reaches peak intensity in the second heart sound. Location of continuous murmur will be heard at the entrance to the heart fistulae. If the CAF entrance on the right atrium, the loudest murmur heard at the bottom along the sternal border. If CAF

entered the pulmonary artery or right ventricle, the loudest murmur heard at the left sternal spatium intercostal 2-3 and if it went into the left ventricle most obvious murmur near the apex. If CAF is connected with the left ventricle, probably just sounded early-diastolic murmur.<sup>1,2,5,21</sup> Haller- and Little describe the triad of diagnostic CAF. An abnormal location of murmur "to and from" in line with the persistent ductus arteriosus, a the left to right shunt on atrial or ventricular levels and coronary artery winding (turtousity).<sup>5</sup> Chest radiography is generally within normal limits or cardiomegaly with increased vascularities, especially if there is a great left-to-right shunt.<sup>1,2,15</sup> Electrocardiography (ECG) is generally normal or ischemic changes that show the effects of ventricular volume overload or left ventricular hypertrophy left, right or both.<sup>1,2</sup> Trans thoracal echocardiography (TTE) - two dimensions demonstrated the large coronary artery dilatation. Turbulence pattern of continuous systolic and diastolic flow describe the location of the beginning of the shunt, dilated nutrient arteries or twisting with abnormal flow patterns. Shunt size are classified based on the ratio of pulmonary to systemic flow ratio (Qp/Qs). High-flow shunts if the ratio Qp/Qs >1.5. The ratio of Qp/Qs in the CAF typically <1.5 and rarely >2.1,<sup>17</sup> Transesophageal echocardiography (TEE) with descriptions include dilatation of the heart chamber, color flow doppler jet narrowest >4 mm or descending aortic back flow.<sup>24</sup> Resolution of TEE is higher because the transducer is placed in the esophagus, just behind and very close to the left atrium and thoracic aorta.<sup>4,22</sup> Treadmill test results can be positive ischemic response.<sup>16</sup> ECG changes on the treadmill test can occur if there are other cardiovascular conditions.<sup>1</sup> Coronary angiography is the gold standard diagnosis of CAF because it can accurately describe the origin, size, number and flow fistulae and presence of aneurysm. Coronary angiography in congenital heart disease is the classification of the recommendations of ACC/AHA class I, level of evidence C.<sup>21</sup> Angiography aimed to find influence of coronary lesions on coronary hemodynamic and knowing abnormality.<sup>2,21</sup> Coronary magnetic resonance angiography (CMRA), more and more used to know the anatomy of the coronary anomaly due to non-invasive, able to identify the main causes of CAF and thus their minimizing radiation exposure is

an ideal procedure.<sup>19</sup> This patient complained recurrent chest pain though the patients were diabetic patients and post stenting one month before. Chest pain was suspected because of ischemia or infarction due to coronary steal phenomenon. Chest pain in patients with DM is very meaningful, because it possible existence of neuropathy. Physical examination found no noisy heart. Results of laboratory and others have hyperglycemia within normal limits. Results of chest X-ray within normal limits and ECG demonstrated VES. Therapy indication of CAF is that there are clinical signs (symptomatic), mainly heart failure and asymptomatic myocardial ischemia as well as CAF with high-flow shunt. The goal of therapy is to prevent the signs or complications, especially in childhood. Closure of fistulae absolutely be done immediately to prevent complications.<sup>2,5-7</sup> There are three options CAF governance namely conservative, surgical and transcatheter coil embolization (TCCE). Conservative therapy is performed for small and asymptomatic CAF. Antiplatelet therapy is recommended especially at the distal CAF and abnormal coronary arteries are dilated. Suggested prophylactic sub-acute bacterial endocarditis due to a complication of coronary fistulae.<sup>7</sup> Definitive therapy of CAF is usually needed cardiac surgery. CAF surgical therapy was first performed Bjork and Crafoord in 1947.<sup>20</sup> Small or asymptomatic CAF surgery remains controversial because of losses due to surgery.<sup>13</sup> Indications of surgery include shunt ratio >30%, signs of ischemia, pulmonary hypertension, congestive heart failure, aneurysm formation, large CAF with high-flow shunt, relationships and multiple endings, the need for coronary bypass-simultaneous and social reasons.<sup>3,4,7,23</sup> Complete occlusion of fistulae after surgery achieving >95%. Morbidity and mortality of surgery 0-6%, myocardial infarction <5% and risk-recurrent fistulae.<sup>1-3,7,8,14</sup> TCCE technique was first performed in 1982. This therapy is effective, safe and complete occlusion was achieved >95%. TCCE successful therapy is similar to surgical therapy in patients without cardiac surgery-related morbidity due to avoid medial sternotomy and cardiopulmonary bypass. TCCE purpose is occlusion of the arteries and fistulae as distal as and as close as possible to the end point to prevent the possibility of occlusion of the branches of myocardial normal. Blockage of arteries that were treated at the first level of

branches and reduction of shunts left-to-right so-closure of post myocardial perfusion became normal. That is too distal embolization, and if there is no significant stenosis in blood vessels, embolization may occur migration tools out fistulae and into the pulmonary circulation.<sup>2,5,6,9,24</sup> TCCE therapy is recommended during secure access to the nutrient artery, single fistulae, there are no multiple large branches, no vessels are extremely winding, narrow single-stream location and the little branches of blood vessels. Despite extensive fistulae with rapid blood flow, TCCE still can be done because at least supplementing, the existence of an optimal location for the main coil and attached to flow into the aneurysm location is relatively narrow and restrictive.<sup>7,9</sup>

The choice of techniques influenced the age, size of catheters that can be used, size of blood vessels that will be occluded and bend the catheter to reach the point of occlusion fistulae, and nutrient artery morphology as well as price considerations. Morphology fistulae include a bend, there are at least high-flow fistulae, aneurysm dilatation of nutrient arteries and location of fistulae.<sup>2,24,25</sup> TCCE tools such as amplatzer duct occluder, microcoil and hydrocoil. Tool such as the double-umbrella device amplatzer duct occluder is used on a larger CAF with branches close to the location of coronary occlusion and more accurate placement. Orifice occlusion in a heart chamber with the surrounding location, then elected a specific coil or double-umbrella-sized two times the diameter of orifice.<sup>6,24,25</sup>

Issenberg microcoil first introduced in 1990, initially used on a smaller CAF. Those advantages and the catheter sheath size smaller and cheaper prices.<sup>24,25</sup> Selected microcoil diameter 20-50% larger than the largest diameter of the fistulae to prevent coil repositioning or migration. Gianturco coil, independent silicon, latex balloon or a combination of coils and balloons used as embolization material. TCCE tool most frequently used is the coil and is the largest type of platinum coil.<sup>7</sup> Coil delivered 2.5 Fr micro-catheter and the coil is released near the meandering arteries. One or more straight 0.018-inch coil is used until there is complete obstruction and after micro-coil fixed to prevent distal migration. The advantage is to reduce the amount of coil needed to close the CAF, reduce costs and time.<sup>17</sup> Microplex coil that can



be controlled from larger and has a stronger shape memory than conventional coil can be like an anchor, with an additional coil to close the shunt. During the procedure, the absolute placement of the micro-catheter is as deep as possible into the fistulae and the meticulous care during coil placement.<sup>7,9</sup> Hydrocoil is a kind of hybrid-endovascular the coil material, used in large fistulae due to high-volume, low pressure due to expansion of the outer layer of a hydrophilic acrylic polymer gel which widened slowly and deployment capacity of up to nine times the volume since the first initial contact with the blood thus reducing the number of coils which needed to close fistulae or aneurysms of the aorta. For example using expandable hydrogel-coated platinum coils.<sup>17</sup> Indication of therapy in these patients because of symptomatic fistulae and VES on ECG. Consideration of selection of TCCE treatment modality is the age and morphology fistulae. Fistulae of the LAD into the right atrium with a single drainage and there is no involvement of major blood vessels. Moreover, the patient is a geriatric patient with a complication of hypertension and DM2NO. TCCE procedure was performed on February 17, 2010. Time required in this patient TCCE 73 minutes. Time required similar reports Kabbani (2008) average 35±15 minutes.<sup>17</sup> Evaluation post coil installation, chest pain disappeared even though remains VES on ECG. Complications in the post-TCCE include transient ischemia on ECG, whereas atrial arrhythmia, embolization coils outside and pulmonary artery dissection post fistulae.<sup>6</sup> Re-occlusion angiography is required to view the nutrient arteries that should be closed during TCCE.<sup>24</sup> Complications of CAF include congestive heart failure, pulmonary hypertension, ischemia, myocardial infarction. A rare complication of ruptured aneurysms and infective endocarditis.<sup>5,7</sup>

## CONCLUSION

TCCE technique is the treatment choice in selected symptomatic CAF patients with a high success rate, saving cost and time compared to surgery. The goal of therapy is to prevent symptoms and complications especially in elderly. Clinical signs and increasing age are related to increased morbidity and mortality.

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