

CONGENITAL CORONARY ARTERY – PULMONARY ARTERY FISTULA CAUSING STEAL PHENOMENON: CASE REPORT AND REVIEW OF THE LITERATURE

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CONGENITAL coronary artery fistulas terminating in cardiac chambers, pulmonary artery, or systemic veins are the most common hemodynamically significant coronary congenital anomaly found in up to 0.2% of the adult population.¹ It was first described by Krause in 1865.² The first surgically treated patient was reported by Bijork and Crafoord in 1947³ at thoracotomy in a patient diagnosed as a case of patent ductus arteriosus.

Case Report

A 54-year-old man was admitted to the coronary care unit in another hospital following an attack of chest pain that was associated with sweating and dizziness. His electrocardiogram (ECG) showed raised ST segment in leads III and VF and depressed ST segments I, aVL, V₄ and V₆. The enzymes were normal on admission but showed an early rise after 8 h and then gradually declined by the end of 24 h. The patient responded well to antianginal therapy. A continuous murmur was noticed in the second and third left parasternal spaces. The patient was referred to King Khalid University Hospital for further management.

On admission, his heart rate was 65 beats/min and regular; the blood pressure was 140/100 mm Hg. The patient was afebrile, had no edema of the lower limbs, and the jugular venous pressure was not raised. The apex beat was in the left fifth intercostal space midclavicular line. Auscultation

revealed a continuous murmur in the second and third left parasternal spaces. The ECG was normal, apart from a leftward axis with inverted T-waves in leads III and AVF. Serum, electrolytes, blood urea nitrogen, creatinine, triglycerides, and complete blood count were normal. The chest x-ray was unremarkable.

Echocardiographic study was within normal limits. Coronary angiography showed bilateral coronary artery fistulas arising from the left anterior descending (LAD) and intermediate branches of the left coronary artery, the dye opacifying the pulmonary artery and poor filling of the LAD due to stealing of coronary flow (Figure 1). Right coronary injection demonstrated a critical lesion in the proximal right coronary artery (RCA) and two proximal branches arising from the RCA and feeding the fistula (Figure 2). The Q_p/Q_s was estimated to be 1.2:1. In view of the subjective and objective findings, surgery was offered.

Through median sternotomy, the creation of a pericardial cradle revealed angiomatous plexus with a palpable thrill, overlying the outflow tract of the right ventricle arising from the LAD and intermediate branches of the left coronary artery leading to a vascular channel that ended in the pulmonary artery about 1 cm above the level of the pulmonary valve. A branch from the RCA was passing over the left anterolateral aspect of the pulmonary artery to join the vascular channel.

Cardiopulmonary bypass (CPB) was conducted by aortic bicaval cannulation with systemic cooling to 30°C. After cross clamping the aorta, cold crystalloid cardioplegia was injected in the aortic root with topical cooling. During delivery of cardioplegia, digital pressure was applied to the fistula to achieve proper myocardial protection.

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Venting was achieved by suction applied to the aortic root. Reversed saphenous vein graft was anastomosed end-to-side to the RCA with continuous 7-0 prolene.

After declamping the aorta, the pulmonary artery was opened longitudinally. A continuous flow of arterial blood was seen coming from the orifice of the fistula (Figure 3). It was sutured by pledgetted 4-0 prolene. The pulmonary artery was closed by continuous 5-0 prolene, including the branch of the RCA in the suture line. After defibrillating the heart, the top end of the vein graft was anastomosed to the ascending aorta with continuous 5-0 prolene.

The thrill was noticed to disappear completely over the fistula. The postoperative course was uneventful, the continuous murmur was no longer audible, and the patient was discharged from the hospital on the 10th postoperative day after repeat coronary angiography.

In Figure 4, postoperative coronary angiography shows patent graft to the RCA. Figure 5 illustrates proper filling of the LAD coronary artery after closure of the fistula. This is a clear demonstration of the stealing effect of the fistula on the coronary flow. Figure 6 shows attenuation of the feeding vessels that arise from the RCA with no opacification of the pulmonary artery. There is residual insignificant jet from the vascular maze draining into the outflow tract of the right ventricle.

Discussion

Most coronary artery fistulas are congenital in origin. Myocardial vessels in the embryo arise from branches of the coronary arteries and veins and also from endothelial protrusions into the intertrabecular spaces.⁴ During normal development the outermost intertrabecular spaces become narrow and join the coronary vessels to form a capillary network. The intertrabecular vessels retain their ventricular communications to form the thebesian vessels of the adult heart. Failure of obliteration of embryonic intertrabecular vascular network leads to coronary artery fistula formation. Recently, other categories of patients have been recruited due to expanded invasive measures of investigation and therapy.

Iatrogenic coronary artery fistulas following endomyocardial biopsy in heart transplant patients have been reported.⁵⁻⁷ In coronary bypass surgery, inadvertent vein grafting was documented.⁸

Left coronary artery fistula-to-left ventricle is a reported complication following percutaneous transluminal coronary artery angioplasty.⁹ Permanent endomyocardial pacemaker electrode is a potentially serious cause of coronary artery fistula formation.¹⁰ Penetrating injury of the heart may result in coronary artery fistula.¹¹

With regards to the involved coronary artery and the site of fistulous connection, the RCA is involved in about 55% of cases.¹² The left coronary artery is involved in 35% and both coronary arteries in about 5%.¹³

More than 90% of the fistulas open into the right heart chambers or its connecting vessels. About 40% drain to the right ventricle, 25% to the right atrium, 15% to 20% to the pulmonary artery, 7% to the coronary sinus, and only 1% to the superior vena cava.^{12,13} This leads to left-to-right shunt with the Q_p/Q_s rarely above 1.8. Large shunts lead to congestive heart failure after the second or third decade of life.¹⁴ Less than 10% of the fistulas end in the left side of the heart, mainly in the left atrium (less frequently in the left ventricle and rarely in the pulmonary veins).

Hemodynamically, fistulas draining into the left heart are arteriosystemic, and when the volume overload is significant, they could be similar to aortic valve insufficiency.¹⁵

Multiple fistulas from all three major coronary arteries to the left ventricle are extremely rare, with less than 20 reported cases.^{16,17} The salient presentation of this type of fistula is coronary insufficiency¹⁷ or even myocardial infarction.¹⁸ Occasionally, the involved coronary artery undergoes aneurysmal dilatation¹⁹ but rupture is rare.

The clinical presentation is usually late in life. Patients under the age of 20 years are usually asymptomatic.^{11,13,20-23} However, in a review of 286 cases, 55% were symptomatic.¹²

A continuous murmur that is maximal to the left or right side of the sternum is the main clinical feature which may resemble patent ductus arteriosus, ruptured sinus of valsalva aneurysm, ventricular septal defect with aortic insufficiency, pulmonary arteriovenous fistula, and fistulas of systemic arteries to veins of the chest wall or to the lung. Exertional dyspnea and fatigue are fairly common symptoms. Other symptoms include congestive heart failure, palpitations, respiratory infection, and hemoptysis.





Figure 1 (pre-op). Left coronary artery injection, right anterior oblique view, showing opacification of the pulmonary artery (small arrow), faint filling of the left anterior descending coronary artery (medium arrow), and angiomatous plexus overlying the right ventricular outflow tract (big arrow).

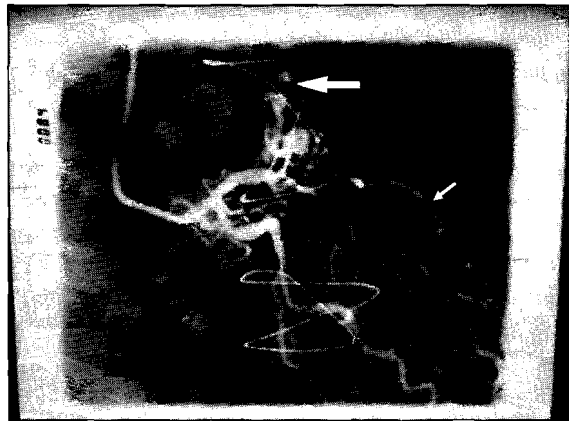


Figure 4 (post-op). Left coronary artery injection showing proper filling of left anterior descending after fistula closure (small arrow) and arrest of the dye with no filling of the pulmonary artery (big arrow).

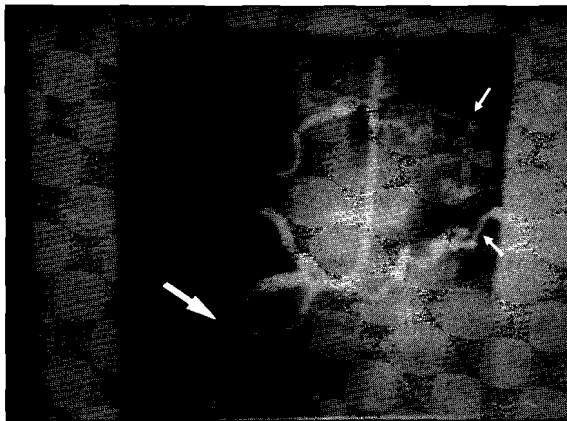


Figure 2 (pre-op). Right coronary artery injection, left anterior oblique view, showing branches from the right coronary artery opacifying the pulmonary artery (small arrow) and critical lesion in the proximal part of right coronary artery (big arrow).

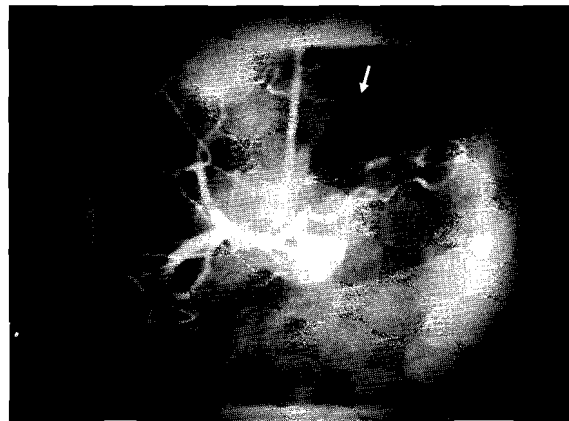


Figure 6 (post-op). Showing attenuation of the branches of the right coronary artery with no opacification of the pulmonary artery after fistula closure (small arrow).



Figure 3. The pulmonary artery is opened (big arrow) with a jet of arterial blood coming from the fistula orifice (small arrow).

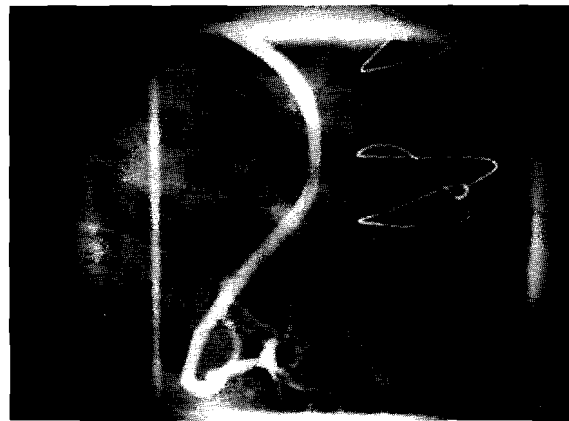


Figure 5. Patent graft to the right coronary artery.



Infective endocarditis was reported in about 5% of patients with coronary artery fistulas.^{22,24} In another series, the risk of endocarditis in coronary artery fistula was 0.004 per patient-year.²⁵ Coronary angiography is the standard method to reach an accurate diagnosis and to determine the site and degree of the shunt. Two-dimensional echocardiography and doppler color-flow mapping is one of the diagnostic tools.^{26,27} However, diagnostic errors²⁸ have shown that coronary angiography is indispensable for arriving at the precise diagnosis. Cine magnetic resonance imaging has been introduced as a valuable noninvasive technique in the diagnosis of coronary artery fistula.²⁹

Indications for Operation

Spontaneous closure of the coronary artery fistula has been reported,³⁰ although exceedingly rare. In the review of the literature, most authors believe that surgical treatment is advised for symptomatic patients with large shunts. Asymptomatic patients with small shunts represent the grey area where the opinion splits. Jaffe et al³¹ believe that the expectant approach is advised, based on long-term follow-up. On the other hand,^{12,14,21,23,32} many authors advise surgical intervention in the asymptomatic group of patients in the young age to reduce the likelihood of complications such as congestive heart failure, myocardial ischemia and infarction, infective endocarditis, aneurysm formation, dissection and rupture of the fistula,³³ and pulmonary hypertension.

Surgical Techniques

The aim of surgical treatment is to interrupt the fistulous connection and to restore the blood supply to the myocardium through the coronary artery. Several techniques have been employed such as ligation of the coronary artery above and below the fistula; this technique, employed by Abott,³⁴ was associated with a high incidence of myocardial ischemia and infarction.^{19,35} Ligation or division of the fistula is the operative technique of choice.¹² Horiuchi and associates³⁶ mentioned the hazard of ligating a RCA proximal to the fistula draining into the superior vena cava or right atrium, possibly due to ischemia of the sinoatrial node. Tangential arteriography was described by Cooley, Hallman, and associates for multiple fistulas.³⁷⁻³⁹

When the coronary artery is aneurysmal, the

fistula could be closed from within the artery that is tailored to near normal vessel. If the aneurysm is very large, it should be excised with fistula obliteration¹⁹; and to restore continuity, vein graft or internal mammary artery is anastomosed distally if the receiving vessel is of reasonable caliber.³⁹ Fistulas draining into a cardiac chamber or its large vessels are obliterated by opening the receiving chamber. Fistula ligation can be accomplished without the aid of CPB. However, the use of CPB is advised if the recipient chamber is opened, in inaccessible site of the fistula, e.g., posteriorly in the distribution of the circumflex coronary artery, hugely dilated torturous vessel to avoid hemorrhage, aneurysm resection, coronary artery bypass graft, or repair of concomitant cardiac lesion.

Surgical Results

Early in the surgical experience, Libberthson et al¹⁴ reported 4% hospital mortality, 4% recurrence, 3% ischemia, and 3% myocardial infarction.

Lowe and associates¹² published the surgical results of 22 patients without early or late death after 10 years of follow-up. Only one of their patients had small residual shunt. In another series Urrutia et al³⁹ presented their experience with 56 patients over 25 years. They divided their patients into two groups. In group 1 (37 patients) with isolated coronary artery fistula, there was no operative death. Long-term follow-up (6 months to 18 years) in 89% of patients showed that all but one were asymptomatic. In group 2 (21 patients) with associated cardiac defects, 93% survived the operation; 67% of the patients were followed up to 9 years with 10 patients asymptomatic, one improved, and one died.

In the University of Alabama (Birmingham, Alabama, USA) and Green Lane Hospital (Auckland, New Zealand) experiences, the hospital mortality approaches zero in the absence of a giant aneurysm such as in the 25 patients who were operated upon; there were excellent long-term results where 24 of these 25 patients were in New York Heart Association (NYHA) class I. Bogers et al²⁵ reported 22 operative survival among 23 patients.

Bilateral coronary artery fistula terminating in the main pulmonary artery is a unique subgroup of patients with congenital coronary artery fistula. Failure of obliteration of embryonic intramyocardial sinusoids leads to congenital coronary artery



fistula.⁴ Accordingly, bilateral involvement requires two separate developmental errors.¹³ One can speculate that this defect of coronary implantation may be related to that which causes anomalous origin of the right or left coronary from the pulmonary artery.⁴⁰ The incidence of bilateral coronary artery-pulmonary artery fistula is 3% in Rittenhous collective review²¹ and about 1.7% in Horiuchi's report.³⁶ Levin et al⁴¹ mentioned that bilateral fistulas occurred only in 5% of 363 cases reviewed. More than half the cases drained into the pulmonary artery.

The mechanism of myocardial ischemia is attributed to coronary steal where coronary flow is preferentially directed to the lower resistance path.^{42,43} This could be accentuated by coronary atherosclerosis distal to the fistula as in our case and others.⁴³ This was demonstrated in our case where coronary angiography showed poor filling of the LAD that opacified clearly after fistula closure (Figures 1-5).

Coronary artery fistula has been incriminated as a contributing factor in the development of premature atherosclerosis due to shear induced intimal damage.^{44,45} Since shear stress is inversely related to the third power of the vessel diameter, under conditions of laminar flow, the grossly dilated coronary artery makes the shear induced intimal damage insignificant. This was not the case in our patient where the RCA diameter was about 2 mm. However, smoking was a documented risk factor.

The use of detachable balloon embolization is evolving as an interventional cardiology technique for treatment of coronary artery fistula.⁴⁶ It is too early to consider this technique as an established line of treatment to replace surgical treatment because of (1) the small number of cases that have been reported, (2) the presence of concomitant cardiac defects requiring surgical intervention, (3) that long-term results are still to be reported, and (4) the procedure is not without complications.⁴⁷ With expanding experience it could be complementary to the surgical approach.

In conclusion, congenital coronary artery fistula is a condition amenable to surgical treatment with satisfactory early and late results.

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References

1. Gilbert C, Van Hoof R, Van de Werf F, et al. Coronary artery fistulas in an adult population. *Eur Heart J* 1986;7:437-47.
2. Krause W. Über den Ursprung einer akzessorischen Coronaria aus der a. pulmonalis. *Z Ratl Med* 1865;24:225-36.
3. Bijorck G, Grafoord C. Arteriovenous aneurysm on the pulmonary artery simulating patent ductus arteriosus botalli. *Thorax* 1947;2:65-75.
4. Grant RT, Regnier M. The comparative anatomy of the cardiac coronary vessels. *Heart* 1926;13:285-317.
5. Yeoh JK, Anderson ST, Federmen J, Esmore D. Coronary artery to middle cardiac vein fistula following endomyocardial biopsy in heart transplant patient. *Cathet Cardiovasc Diagn* 1991;24(2):108-10.
6. Somers JM, Verney GI. Coronary cameral fistulae following heart transplantation. *Clin Radiol* 1991;44(6):419-21.
7. Henz Lova MJ, Nath H, Bucy RP, et al. Coronary artery to right ventricle fistula in heart transplant recipients, a complication of endomyocardial biopsy. *J Am Coll Cardiol* 1989;14(1):258-61.
8. Marin Nito JA, Simoes MV, Vicent WV. Acquired aortocoronary vein fistula after bypass graft surgery: report of 2 cases with long-term follow-up. *Int J Cardiol* 1991;30(1):121-4.
9. Iannone LA, Iannone DP. Iatrogenic left coronary artery fistula-to-left ventricle following PTCA: a previously unreported complication with non-surgical management. *Am Heart J* 1990;120(5):1215-7.
10. Sacian K, Vellinga T, Troup P, Weatherbee J. Coronary artery fistula formation secondary to permanent pacemaker placement. *Chest* 1991;99(3):780-1.
11. Juca ER, Carvallo W Jr. Right coronary artery-right atrium fistula resulting from a penetrating injury of the heart. *J Thorac Cardiovasc Surg* 1989;98(4):638-9.
12. Lowe JE, Oldham HN, Sabiston DC. Surgical management of congenital coronary artery fistulas. *Ann Surg* 1981;194:373-80.
13. Baim DS, Klin H, Silverman JF. Bilateral coronary artery-pulmonary artery fistulas. Report of 5 cases and review of the literature. *Circulation* 1982;65:810-5.
14. Liberthson RR, Sagar K, Berkoben JP, et al. Congenital coronary arteriovenous fistula. Report of 13 patients. review of the literature and delineation of management. *Circulation* 1979;59:849-54.
15. Dobell ARC, Long RW. Right coronary left ventricular fistula mimicking aortic valve insufficiency in infancy. *J Thorac Cardiovasc Surg* 1981;82:785-9.
16. Sheikhzadeh A, Stierle U, Langbehn AF, et al. Generalized coronary arterio-systemic (left ventricular) fistula: case report and review of the literature. *Jpn Heart J* 1986;27:533-44.
17. Black IW, Loo CKC, Allan RM. Multiple coronary



- artery-left ventricular fistulae: clinical, angiographic and pathologic findings. *Cathet Cardiovasc Diagn* 1991;23:133-5.
18. McLellan BA, Pelickan PCD. Myocardial infarction due to multiple coronary-ventricular fistulas. *Cathet Cardiovasc Diagn* 1989;16:247-9.
 19. Meyer MH, Stephenson HE Jr, Keato TE, Martt JM. Coronary artery resection for giant aneurysmal enlargement and arteriovenous fistula. *Am Heart J* 1967;74:603-13.
 20. Kirklin J, Barratt-Boyes B. *Cardiac surgery*. New York:John Wiley, 1986.
 21. Rittenhouse EA, Doty DB, Ehrenhaft JL. Congenital coronary artery-cardiac chamber fistula. *Ann Thorac Surg* 1975;20(4):468-85.
 22. McNamara JD, Gross RE. Congenital coronary artery fistula. *Ann Surg* 1969;65(1):59-69.
 23. de Nef JJE, Varghese PJ, Losekoot G. Congenital coronary artery fistula: analysis of 17 cases. *Br Heart J* 1971;33:857-62.
 24. Symbas PN, Schlant RC, Hatcher CR Jr, Lindsay J. Congenital fistula of right coronary artery to right ventricle complicated by *Actinobacillus actinomycete* concomitant endarteritis. *J Thorac Cardiovasc Surg* 1967;53:379-84.
 25. Bogers AJJC, Quaegebeur, Huysmans HA. Early and late results of surgical treatment of congenital coronary artery fistula. *Thorax* 1987;42:369-73.
 26. Wen MS, Lin FC, Yeh ST, Wu DL. Doppler color flow imaging in coronary artery fistula. *Am Heart J* 1991;122(4-1):1180-3.
 27. Nishiguchi T, Matsouk Y, Sennari E, et al. Congenital coronary artery fistula: diagnosis by two-dimensional Doppler echocardiography. *Am Heart J* 1990;120(5):1244-8.
 28. Lloyd TR. Failure of colour flow mapping to identify coronary artery to pulmonary artery fistula. *Pediatr Cardiol* 1991;12(4):227-8.
 29. Kubota S, Suzuki T, Murata K. Cine magnetic resonance imaging for diagnosis of right coronary arterial ventricular fistula. *Chest* 1991;100(3):735-7.
 30. Muthusamy R, Gupta G, Ahmed RA, et al. Fistula between a branch of left anterior descending coronary artery and pulmonary artery with spontaneous closure. *Eur Heart J* 1990;11(10):954-6.
 31. Jaffe RB, Glancy L, Epstein SE, et al. Coronary arterial-right heart fistulae long-term observation in seven patients. *Circulation* 1973;48:133-43.
 32. Hudspeth AS, Linder JH, Salem W. Congenital coronary arteriovenous fistula. *Arch Surg* 1968;96:832-5.
 33. Habermann JH, Howard ML, Johnson ES. Rupture of the coronary sinus with hemopericardium: a rare complication of coronary arteriovenous fistulas. *Circulation* 1963;28:1143-7.
 34. Abbott OA, Rivarola CH, Longue RB. Surgical correction of coronary arteriovenous fistula. *J Thorac Cardiovasc Surg* 1961;42:6670-8.
 35. Cooley DA, Ellis PR. Surgical considerations of coronary arterial fistula. *Am J Cardiol* 1962;10:467-77.
 36. Horiuchi T, Abe T, Tanaka S, Koyamada K. Congenital coronary arteriovenous fistulas. *Ann Thorac Surg* 1971;11(2):102-12.
 37. Hallman G, Cooley DA, Singer DB. Congenital anomalies of the coronary arteries. Anatomy, pathology and surgical treatment. *Ann Surg* 1966;59:133-47.
 38. Liotta D, Hallman G, Hall RJ, Cooley DA. Surgical treatment of congenital coronary artery fistula. *Ann Surg* 1971;70:856-67.
 39. Urrutias CO, Falaschi G, Ott D, Cooley DA. Surgical management of 56 patients with congenital coronary artery fistulas. *Ann Thorac Surg* 1983;35:300-7.
 40. Lerberg DB, Ogden JA, Zuberbuhler JR, Bahnsen HT. Anomalous origin of the right coronary artery from the pulmonary artery. *Ann Thorac Surg* 1979;27:87-96.
 41. Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries. *Circulation* 1978;58:25-37.
 42. Cason BA, Gordon HJ. Coronary steal caused by a coronary artery fistula. *J Cardiothoracic Vascular Anaesth* 1992;6(1):65-7.
 43. Swan KM, Koepke DC. Coronary artery to left atrium fistula requiring revascularization: case report and literature review. *Thorax* 1982;37:376-80.
 44. Fry DL. Acute vascular endotracheal changes associated with increased blood velocity gradients. *Circ Res* 1968;22:165-72.
 45. Fry DL. Certain histological and chemical responses of the vascular interface to acutely induced mechanical stress in the aorta of the dog. *Circ Res* 1969;24:93-104.
 46. Doorey AJ, Sullivan KL, Levin DC. Successful percutaneous closure of a complex coronary-to-pulmonary artery fistula using a detachable balloon: benefits of intraprocedural physiologic and angiographic assessment. *Cathet Cardiovasc Diagn* 1991;23:23-7.
 47. Meier B. Coronary occlusion after failed closure of coronary-to-pulmonary fistula with detachable balloon. *Cathet Cardiovasc Diagn* 1989;18:237-9.

