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PEDIATRIC CARDIOLOGY

Repair of Anomalous Origin of the Left Coronary Artery in the Infant and Small Child

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Anomalous origin of the left coronary artery from the pulmonary artery is associated with myocardial infarction, left ventricular dysfunction, mitral valve dysfunction and, occasionally, intracardiac congenital abnormalities. A technique that utilizes a flap of the anterior wall of the pulmonary artery to serve as a neocoronary artery to direct aortic flow from a created aortopulmonary window to the pulmonary artery orifice of the anomalous left coronary artery was used in five patients aged 2.5 months to 4.75 years. Two patients were less than 4 months of age at operation. There was one death 2 days after operation and one late death. The two youngest patients required mitral valve replacement. Two of the three surviving patients are well at follow-up at 7 to 44 months. One patient has been lost to follow-up study. One patient had postoperative catheterization which showed an intact repair. The pulmonary artery neocoronary procedure is applicable to infants and small patients with anomalous origin of the left coronary artery from the pulmonary artery.

Anomalous left coronary artery originating from the pulmonary artery is a rare defect with a high mortality rate during infancy. Since its first clinical definition in 1933 by Bland et al. (1), fewer than 250 cases have been reported (2). The mortality rate in the first year of life of untreated patients has been estimated to be as high as 90% (3,4). The patient who is symptomatic and refractory to medical treatment requires aggressive surgical treatment to alter an otherwise lethal course. The anomaly represents a challenge to the pediatric cardiovascular surgeon because it is associated with myocardial infarction, left ventricular dysfunction, mitral valve dysfunction and, occasionally, other intracardiac abnormalities.

From 1978 to 1983, ten patients with anomalous origin of the left coronary artery from the pulmonary artery were operated on at Children's Hospital National Medical Center. Five patients were treated by a variety of procedures including ligation of the anomalous left coronary artery in two patients, saphenous vein graft in one, Goretex graft interposition in one and subclavian coronary anatomosis in one. This report describes five patients who received a new type of surgical procedure described by Takeuchi et al. (5) whereby an intrapulmonary baffle directs aortic flow to the anomalous orifice of the left coronary artery.

Methods

Patients. All five patients were female; their ages ranged from 2.5 to 57 months (median 12) (Table 1). Three were 12 months or younger. Weights ranged from 3.7 to 20 kg (median 7.2). Four patients presented with severe congestive heart failure and anterolateral myocardial infarction on the electrocardiogram. All required prolonged periods of intensive care before surgery. One patient with elevated pulmonary artery pressures secondary to hypoplasia of the branch pulmonary arteries had clinical evidence of cardiomyopathy without infarction. The definitive diagnosis of anomalous left coronary artery was established by cardiac catheterization and cineangiography. In two patients, the timing of operation was based on failure to improve with maximal cardiorespiratory support. The remaining three patients had chronic myocardial failure.

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Table 1. Takeuchi Procedures (5) in Five Patients

Patient	Age at Op (mo)	Wt at Op (kg)	CPB Time (min)	Aortic Cross-Clamp Time (min)	Discharge Days Postop	Mortality		Follow-up (mo)
						Op	Late	
1	57	20	118	43	8	—	—	44
2	43	12.2	120	50	13	—	NA	13
3	12	7.2	80	42	—	+	—	0
4	2.5	3.7	139	75	64	—	+	10
5	3.5	4.7	148	106	146	—	—	7

CPB = cardiopulmonary bypass; NA = not available, lost during follow-up; Op = operation or operative; Postop = after operation; Wt = weight; + = present; — = absent.

Surgical technique. By means of a midline sternotomy using cardiopulmonary bypass and moderate systemic hypothermia (27°C), the aorta and pulmonary arteries were cross-clamped and cold potassium cardioplegia solution was infused in both vessels until the myocardial temperature reached 9°C. A pulmonary artery incision was made and the orifice of the left coronary artery was inspected (Fig. 1). The anterior pulmonary artery incision was lengthened to create a peninsula-like flap of 15 × 30 mm and the base remained attached to the pulmonary artery adjacent to the aorta. An aortopulmonary window with a 6 mm diameter was created in the region of the left coronary sinus between the aorta and the proximal main pulmonary artery. The flap of the anterior pulmonary artery wall was sutured to the medial and posterior aspects of the main pulmonary artery to produce a tunnel directing flow from the aortopulmonary window to the orifice of the anomalous left coronary artery, thus creating a "neocoronary" artery. The defect in the

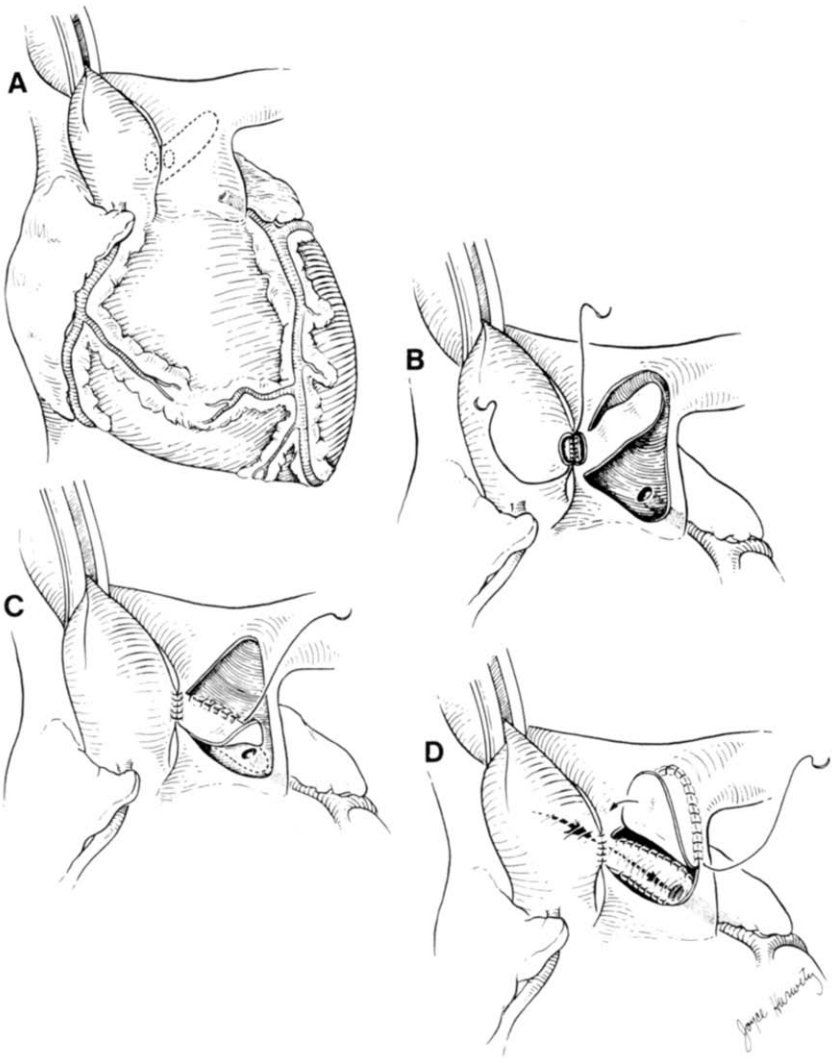


Figure 1. Surgical technique. **A, Dotted lines** indicate sites selected for aortopulmonary window (**circles**) and peninsula flap (U-shaped) of anterior pulmonary artery wall. **B** and **C**, An aortopulmonary window has been created and the pulmonary artery flap is applied to the posterior wall. **D**, A neocoronary tunnel has been constructed and the anterior pulmonary artery is being reconstructed with pericardium.

anterior wall of the main pulmonary artery was repaired with a patch of pericardium. Associated lesions were repaired in a routine manner.

Results

Table 1 lists the age at operation and other data relative to our five cases of surgical treatment of anomalous left coronary artery by the Takeuchi procedure.

Patient 1 had no additional abnormalities and is well at follow-up at 44 months.

Patient 2 had concurrent dilation of the branch pulmonary arteries and was doing well until being lost to follow-up after 13 months.

Patient 3 had concurrent closure of patent foramen ovale and died on the second postoperative day from persistent progressive arrhythmias and progressive biventricular failure. Autopsy showed transmural fibrosis of the anterolateral ventricular wall, endocardial sclerosis of the left ventricle and left atrium and a thin-walled, dilated right ventricle. The coronary artery repair was intact.

Patient 4, 2.5 months old, underwent mitral anuloplasty and closure of a large atrial defect. Because of persistent severe mitral regurgitation, she underwent mitral valve replacement 1 month later with a 17 mm Björk-Shiley valve. Pathologic examination revealed myxomatous degeneration of the mitral valve and fibrosis and calcific disease of the anterolateral papillary muscle. A third cardiopulmonary bypass procedure was necessary 1 month later for repair of a neocoronary baffle leak and a periprosthetic mitral valve leak. A fourth procedure was required 10 months later at the age of 14 months for chronic pulmonary venous hypertension and low cardiac output. Preoperative cineangiography demonstrated good function of the neocoronary pathway and prosthetic valve disc (Fig. 2), although a restrictive fibrous supramitral membrane was found at surgery. The prosthetic mitral valve was replaced with a 21 mm St. Jude valve, but the patient could not be weaned from cardiopulmonary support. Postmortem examination was not obtained.

Patient 5, 3.5 months old, had concurrent atrial septal defect repair and mitral valve replacement with a 17 mm Björk-Shiley prosthetic valve using a superior left atrial approach. She remains well 7 months after operation. There was primary disease of the mitral valve (myxomatous degeneration) as well as dysfunction of the mitral valve apparatus secondary to abnormalities of the papillary muscles, both grossly and microscopically. On pathologic examination, fibrocalcific degeneration of the papillary muscle was present.

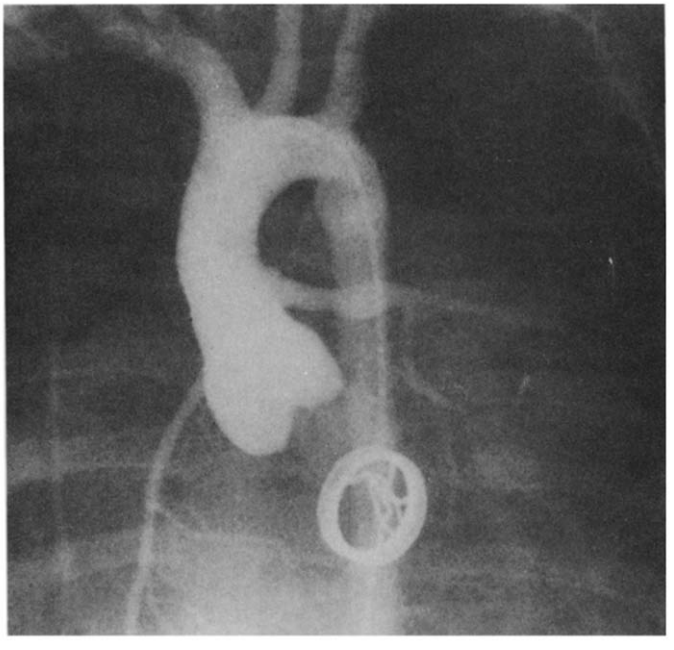
Discussion

Previous surgical procedures. Over the last 30 years, numerous surgical procedures have been proposed for palliation or total correction of anomalous left coronary artery (5–27). Among these are anastomosis with the left common carotid artery (10), the creation of an aortopulmonary fistula (11), pericardial poudrage with removal of the pericardium (12), pulmonary artery banding (13) and ligation of the pulmonary artery orifice of the anomalous left coronary artery (14). Saphenous vein graft interposition, anastomosis of the left coronary artery to the left subclavian artery (16–18), reimplantation (19–21), utilization of a free subclavian segment both within and without the pulmonary artery (22), the Vineberg procedure utilizing the internal mammary artery (23) and the pulmonary artery baffle procedure (5) also have been described.

We assume that the myocardium is best served on a short- and long-term basis by a two coronary artery system, and favor procedures with this result. Saphenous vein interposition, subclavian artery anastomosis and reimplantation are attractive procedures, yet are not without disadvantage. Long-term degeneration of saphenous vein bypass grafts has been described (24). Subclavian artery anatomy is not always favorable because of vessel size or kinking at the site of origin from the aortic arch. Reimplantation, while readily performed in many series, was not possible in any of our patients. In our patients, not only did the anomalous coronary artery orifice originate deep in the posterior lateral pulmonary sinus, 180° away from the aorta, but also the left main coronary artery was short.

Present procedure. The most favorable procedure applicable to the small infant would be a technique that has growth potential and obviates the need to manipulate directly

Figure 2. Patient 4. Cineangiogram demonstrating the neocoronary tunnel with normal left coronary branching. The disc of the prosthetic valve is open.



the left coronary artery system. Such an operative procedure was utilized in our five patients. The aortopulmonary window, neocoronary procedure had its initial stages of evolution in the proposal by Gasul and Loeffler (25) for creating an arteriovenous fistula between the pulmonary artery and the aorta to provide a higher pressure and higher oxygen-saturated blood flow to the anomalous left coronary artery. Creation of an aortopulmonary fistula for anomalous left coronary artery was reported by Potts (11) in 1955, but because it also increased left ventricular work it was uniformly unsuccessful. In 1975, Stern et al. (26) applied the arteriovenous fistula principle and added an intrapulmonary tunnel to direct flow from the fistula to the anomalous left coronary artery orifice and reported one success. Hamilton et al. (27) reported in 1979 the successful use of a pericardial intrapulmonary artery conduit with Dacron augmentation of the anterior wall of the main pulmonary artery in a 4 year old girl. In 1979, Takeuchi et al. (5) reported a modified technique using a flap of the anterior main pulmonary artery as a neocoronary tunnel in a 2 year old patient.

Advantages of technique. The technique as described by Takeuchi et al. has the advantage of constructing the neocoronary vessel from viable normal pulmonary artery wall which is able to function as a high pressure conduit without undergoing degenerative changes and is potentially capable of enlargement. Approximately one-third of the circumference of the tunnel is normal, undisplaced pulmonary artery wall. Although Takeuchi's procedure is complex and subject to technical failure, our experience demonstrates that the procedure can be performed in the infant and small child. No prosthetic materials are used. Manipulation of the left coronary artery is avoided and growth potential for the left coronary artery is preserved. The neocoronary artery establishes a two coronary artery system. Neocoronary construction can be performed in conjunction with repair of other major cardiovascular defects. The technique of aortopulmonary window, neocoronary construction should be given serious consideration as surgical treatment for anomalous left coronary artery, especially if reimplantation and subclavian artery procedures are not possible.

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