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Anomalous left Coronary Artery from the Pulmonary Artery

Anomalous origin of the left coronary artery arising from the pulmonary artery (ALCAPA) is a rare and isolated anomaly. ALCAPA is rarely associated with tetralogy of Fallot, patent ductus arteriousus, and coarctation of the aorta.

21.1 Pathophysiology

Shortly after birth, as the poulmonary vascular resistance and pulmonary artery pressure decrease as does the oxygen content of pulmonary blood, the left ventricular myocardium is perfused by a relatively desaturated blood under low PA (pulmonary artery) pressure, leading to myocardial ischemia.

Though myocardial ischemia is transient initially that occurs when the infant is feeding and crying, further increases in myocardial oxygen demand lead to anterolateral left ventricular free wall myocardial infarction. The myocardial infarction may result in papillary muscle dysfunction and a variable degree of mitral insufficiency.

As myocardial ischemia progresses, the collaterals develop between the right and left coronary systems. Both the right and left coronary artery flow reverses and enters the pulmonary trunk due to the low pulmonary vascular resistance, producing coronary steal phenomena. As a result, left ventricular myocardium remains underperfused. Consequently, the entire left ventricular myocardium remains underperfused resulting in left ventricular dysfunction and significant mitral valve insufficiency, which leads to congestive heart failure.

21.2 Clinical Symptoms

In young infants, the following symptoms predominate: Tachypnea, poor feeding, irritability, and diaphoresis leading to poor weight gain. Myocardial ischemia causes significant chest pain and these symptoms may be misinterpreted as infantile colic. Some infants may outgrow these symptoms and may gradually

become asymptomatic. During later childhood or in adults, periods of dyspnea, chest pain, syncope, or sudden death may occur.

21.3 Diagnosis

In a suspected patient with ALCAPA, an anterolateral infarct pattern on EKG, a characteristic finding of basal and anterolateral subendocardial myocardial fibrosis on cardiac magnetic resonance (MR) imaging, and reversible ischemia on stress adenosine MR imaging is highly suggestive of ALCAPA.

Two dimensional echocardiogram may identify abnormal origin of the left coronary artery from the main pulmonary artery. The color flow doppler demonstrates retrograde flow from the anomalous left coronary into the pulmonary trunk if collaterals between the left and right coronary artery systems exist.

Oximetry may show a small left-to-right shunt into the pulmonary arteries.

Aortography or selective right coronary arteriography demonstrates an enlarged right coronary artery system with collateralization to the left coronary artery and reflux of contrast into the main pulmonary artery (see Figure 21.1).

Angiogram with a large bolus of contrast under high pressure or with a balloon occlusion of the distal main pulmonary artery, retrogradely fills the anomalous left coronary artery.

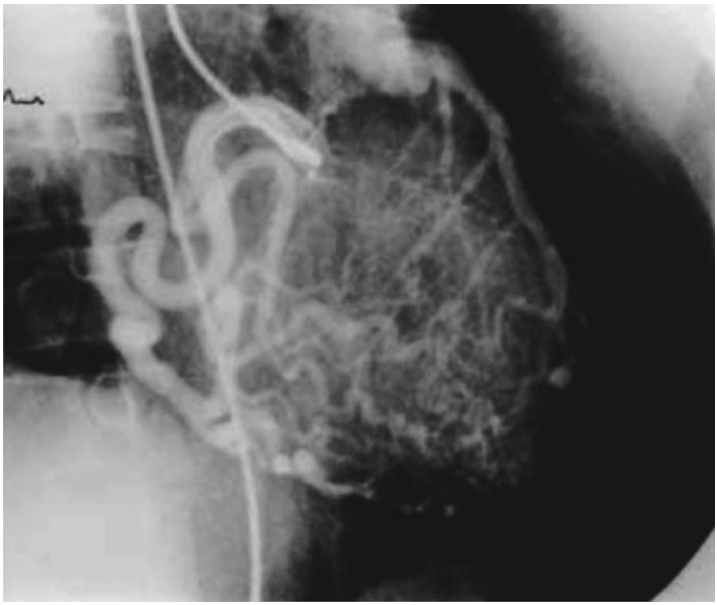

Figure 21.1 *Right coronary artery angiogram. Note the right coronary artery is tortuous and dilated. The left coronary artery is filled through extensive collaterals and the contrast is seen draining into the main pulmonary arery. Reproduced from Takemi Kawara et al. Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery. Ann Thoac Cardiovasc Surg Vol 9. 2003.*

21.4 Surgical Treatment

1. Ligation of the left coronary artery: Performed at its origin from the main pulmonary artery without the use of CPB (cardiopulmonary bypass).

The technique is not optimal since myocardial perfusion remained solely dependent on extensive collateralization from the right coronary artery, and the risk of sudden death exists due to continuing ischemic episodes.

2. Myocardial revascularization by creating two coronary artery systems:

A: Left subclavian artery- left coronary artery anastomosis.

B: Aorto - left coronary (left anterior descending coronary artery) saphenous vein bypass graft.

C: Takeuchi procedure (creation of an aortopulmonary window and an intrapulmonary tunnel by a Gore-Tex baffle extending from the anomalous ostium to the window).

3. Direct reimplantation of left coronary artery into aorta.

Once perfusion through two coronary arteries is accomplished, most patients demonstrate improved left ventricular systolic function, decreased mitral valve insufficiency, and resolution of CHF (congestive heart failure) symptoms.

The need for a simultaneous mitral valve reconstruction, in the presence of severe insufficiency, is controversial because spontaneous improvement of mitral valve function occurs following surgical revascularization. Rarely, refractory mitral regurgitation will necessitate delayed mitral valve repair or a replacement.

21.5 Perioperative Management

The usual postoperative course lasts 7 days to 14 days.

Prior to surgical correction, the management is primarily focused on treatment of congestive heart failure, improvement of myocardial performance, systemic perfusion, and oxygen transport to tissues and myocardium. Agents commonly used are diuretics, afterload reducing medications, and cardiac inotropic drugs (see Section I Chapters 4 & 16). The same medications are used postoperatively to treat ventricular dysfunction and to ensure smooth postoperative course.

Optimal oxygen transport is achieved with adequate replenishment of intravascular blood volume and O_2 carrying capacity. The O_2 saturations are maintained at > 98% with adequate PaO_2 (see Section I Chapter 5). Care is taken to not to use 100% oxygen for any lengthy periods in order to improve pulmonary gas exchange as it may result in deleterious effects on alveolar capillary membranes, and may further reduce the pulmonary vascular resistance

and increases coronary steal from the right coronary artery into the pulmonary arteries, aggravating myocardial ischemia.

Afterload reduction is done judiciously to improve myocardial performance (see Section I Chapter 4 & 16). A similar phenomenon occurs with aggressive afterload reduction which reduces coronary flow through right coronary artery collaterals leading to myocardial ischemia.

Non-judicious inotropic support may significantly increase myocardial oxygen consumption, which in the presence of reduced myocardial blood flow may result in worsening ischemia. In addition to inotropes, afterload reducing drugs may worsen myocardial ischemia, decrease cardiac output, and create a potential for onset of ventricular arrhythmias.

However, all the above medications may be used more aggressively, subsequent to surgical correction for the continued treatment of CHF, left ventricular dysfunction, and mitral valve insufficiency.

21.5.1 Hemodynamic Management

Aimed at maintenance of adequate systemic perfusion pressure for the age, with maintenance of optimal tissue perfusion and maintenance of adequate cardiac output.

21.5.2 Invasive Monitors

Arterial line, central venous catheter, and LA (left atrial) catheters (in severe mitral insufficiency).

An oximetry catheter may be placed in the pulmonary artery (PA) to monitor PA pressure (in presence of severe mitral insufficiency) and cardiac output.

21.5.3 Vasoactive Drug Infusions

Dopamine or dobutamine, epinephrine, milrinone, nitroprusside, and phenoxybenzamine (see Section I, Chapter 4 & 16).

21.5.4 Postoperative Bleeding

Rarely occur from the suture lines.

21.5.5 Atrial and Ventricular Arrhythmias

These may occur due to myocardial ischemia or reperfusion injury subsequent to myocardial revascularization. Treat with antiarrhythmic drugs (see Section I Chapter 16).