Anomalous origin of the left coronary artery from the pulmonary artery with patent ductus arteriosus: a must to recognize entity

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Anomalous left coronary artery from the pulmonary trunk (ALCAPA) presents in early infancy with a clinical picture of congestive heart failure with left ventricular (LV) dysfunction and mitral insufficiency. These manifestations of myocardial ischaemia may be masked in the presence of an associated patent ductus arteriosus (PDA) or ventricular septal defect (VSD) which prevents the fall of pulmonary artery pressures and allows perfusion of the anomalous coronary artery. We present a case of a patient with large PDA-associated ALCAPA and preserved LV function. The importance of such a finding lies in the fact that VSD closure or PDA ligation in such cases would unmask the ALCAPA.

Keywords
ALCAPA • PDA

Anomalous left coronary artery from the pulmonary trunk (ALCAPA) most commonly occurs in isolation. However, it has been known to be associated with other cardiac lesions, including patent ductus arteriosus (PDA).1–3 In a case of ALCAPA, clinical manifestations are due to left coronary artery steal, from the pulmonary circulation.4 The associated lesions become significant when the associated lesions increase the pulmonary artery pressures, thus maintaining the coronary artery perfusion. Previous reports have described a fatal outcome after ligation of a PDA in patients with undiagnosed ALCAPA.5,6 We present the case of an infant with pulmonary hypertension secondary to a large PDA which resulted in normal flow in the aberrant coronary artery preserving the myocardial contractility and function.

Case report
A 4-month-old male infant was referred to our institution after murmur was detected by paediatrician. He was the 2.92 kg product of a 37-week normal pregnancy and delivery. The physical examination showed that heart rate was 140 bpm and regular. Respiratory distress and tachypnoea were not noted. The cardiac examination revealed a hyperactive pericardium. The first heart sound was normal and the pulmonary component of the second heart sound was increased in intensity. A 2/6 holosystolic murmur was heard at the apex with radiation to the left axilla. A mid-diastolic rumble was appreciated at the apex. Chest X-ray showed mild cardiomegaly, with features of increased pulmonary blood flow. Electrocardiography showed biventricular hypertrophy with sinus rhythm. Transthoracic echocardiogram showed a large arterial duct with left to right flow (PDA max PG of 14 mmHg), mild mitral regurgitation, and pulmonary artery hypertension. Anterior papillary muscle appeared hyperechoic (Figure 1). Further evaluation showed anomalously originating left main coronary artery (LMCA). It originated from the rightward portion of the pulmonary sinus (Figure 2). Antegrade flow was maintained and seen on colour Doppler interrogation (Figure 3). Child underwent successful translocation of the LMCA to the aorta and ligation of arterial duct.

Discussion
ALCAPA or Bland–White–Garland syndrome is a rare congenital cardiac anomaly occurring in one of 300 000 live births and generally occurring in isolation.1 Clinical manifestations of ALCAPA were due to myocardial ischaemia secondary to low-pressure coronary perfusion with poorly oxygenated blood and insufficient collateral flow from the right coronary artery and also from the coronary steal from the pulmonary circulation and reversed flow in the left coronary vessels to the pulmonary artery. Dramatic changes occur in the coronary circulation in infants with

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ALCAPA within the first weeks of life. Although the foetus remains in utero, the heart develops quite normally. Initially after birth, there still relatively high pulmonary vascular resistance and pulmonary artery pressure allow the myocardium supplied by the anomalous artery to remain well perfused. In first few days after birth, pulmonary vascular resistance falls so does the pressure in the pulmonary trunk. Subsequent compromise of left ventricular (LV) subendocardial perfusion develops because of both reduced antegrade flow and to frank retrograde flow in the LMCA and resultant coronary steal. In addition, all antegrade left coronary artery flow is deoxygenated venous blood. Severe LV dysfunction and mitral insufficiency generally develop rapidly. The age of onset of symptoms varies depending on the rate of fall of pulmonary artery pressure and the development of collateral connections with the right coronary artery. Left untreated, 90% of infants will not survive the first year of life. In a small proportion, the LV gradually improves over a period of 4–6 weeks as collaterals develop. The presence of a significant post-tricuspid shunt prevents the fall in the pulmonary artery pressure and also provides oxygenated blood to the left coronary artery system, thus preventing the dramatic changes seen in a child with associated ALCAPA.

Post-operative LV dysfunction is a common finding in patients who have been operated for large post-tricuspid left to right shunts and it may not be possible to delineate coronary artery blood flow due to inadequacy of echo windows. However, the presence of poorly contracting LV with hyperechoic papillary muscles and variable degree mitral valve incompetence may be the only markers for an ALCAPA that has been unmasked by surgical repair. A cardiac catheterization with aortic root angiogram will help in diagnosing the condition.

In our patient, the presence of a large PDA prevented fall in pulmonary artery pressure and maintained antegrade delivery of oxygenated blood to the LMCA. However, the presence of hyperechoic papillary muscle was suggestive of ongoing myocardial ischemia. Ligation of arterial duct alone would have been met with poor outcome. Hence, it is imperative to demonstrate coronary artery anatomy on two-dimensional echocardiography and colour Doppler in all patients before surgical repair of post-tricuspid shunt lesions.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References


