

# Left main coronary artery compression in a young woman with Eisenmenger syndrome

## INTRODUCTION

Secondary pulmonary hypertension is a common finding in patients with congenital cardiac defects. Eisenmenger syndrome, a severe manifestation of pulmonary hypertension, affects quality of life, morbidity, and mortality. Patients usually present with cyanosis and reduced physical ability due to dyspnoea or sometimes chest pain on exertion. Nevertheless, coronary artery compression is an uncommon and probably underestimated cause for angina-like chest pain and ventricular dysfunction in patients with severe pulmonary artery hypertension. We report on an adolescent woman with unoperated ventricular septal defect and Eisenmenger syndrome who experienced myocardial infarction owing to a compression of the left main coronary artery (LMCA) by a distended pulmonary artery.

## CASE

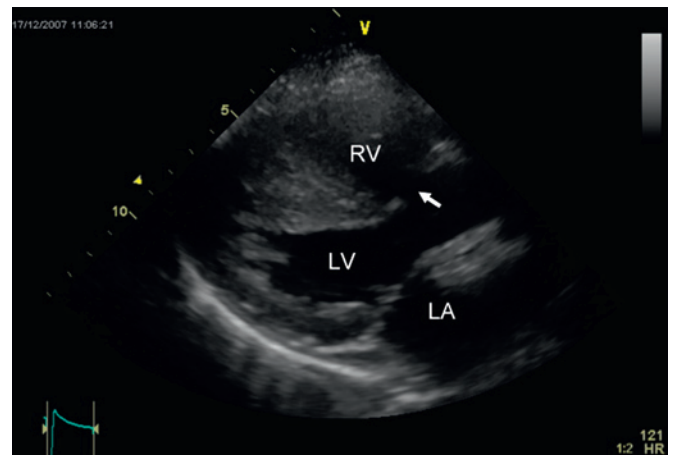
A 16-year-old cyanotic woman was admitted to hospital because of severe chest pain during minimal exertion. On admission, she was in functional class III (New York Heart Association).

She had a history of unoperated unrestrictive ventricular septal defect with a bidirectional shunt leading to Eisenmenger reaction.

Clinical examination revealed a holosystolic heart murmur with fixed narrow splitting of the second heart sound. There were neither signs of acute heart failure nor arrhythmia. Electrocardiogram indicated regular sinus rhythm with delayed conduction, signs of biventricular hypertrophy as well as ST segment and T wave changes in the left precordial leads.

Echocardiography showed a perimembranous malalignment-ventricular septal defect with a bidirectional shunt, biventricular hypertrophy and dilation with reduced biventricular systolic function, but no regional wall motion abnormalities (figure 1). Mild mitral regurgitation was assessed. Moreover, the pulmonary valve had a gradient of 28 mm Hg, and the pulmonary artery trunk was dilated by approximately 40 mm in diameter.

Laboratory tests revealed an elevated Troponin I level of 10.7 ng/ml (upper reference limit 0.1 ng/ml) as well as a creatinine kinase level of 221 U/l (upper reference limit 171 U/l) with an elevated CK-MB/CK fraction. Haematological markers showed secondary erythrocytosis with haemoglobin of 17.6 g/dl and haematocrit fraction of 50% as well as a normal platelet count. The oxygen saturation was 82% on pulse oximetry at rest. A cardiac MR scan presented a late enhancement in the left ventricular myocardium of the interventricular septum. Coronary angiogram revealed a severe stenosis of the LMCA and the proximal left anterior descending (LAD) without additional stenosis of the coronary arteries (figure 2). A percutaneous coronary intervention was successfully performed implanting a 3.5 mm/18 mm sirolimus-eluting coronary stent (Ciper, Cordis, J&J) (figure 3). A larger balloon and high pressure of 20 atm were used for postdilatation. Antiplatelet therapy consisted of pretreatment with 600 mg of clopidogrel and 500 mg of acetylsalicylic acid prior to intervention and 100 IU/kg of body weight unfractionated heparin. Maintenance antiplatelet therapy consisted of daily 200 mg of acetylsalicylic acid and



**Figure 1** Perimembranous ventricular septal defect, showing a parasternal long axis view from a two-dimensional echocardiogram: left ventricle (LV) and right ventricle (RV) with biventricular hypertrophy and perimembranous ventricular septal defect (arrow).

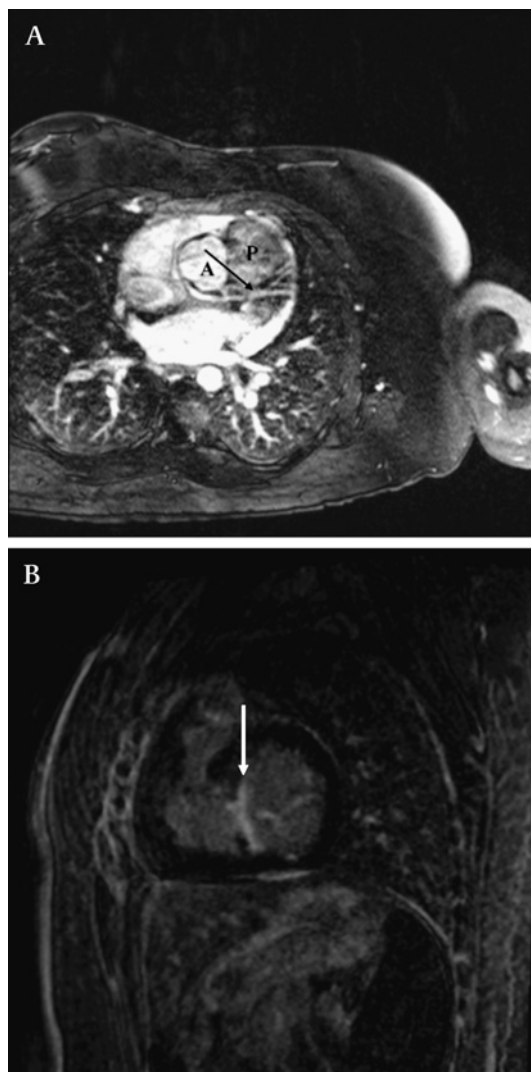
75 mg of clopidogrel. Haemodynamics showed severe pulmonary hypertension with a pulmonary vascular resistance of 25 Wood units with vasodilatory response to inhaled nitric oxide (19 Wood units). Pulmonary hypertension was treated with bosentan, an unselective endothelin receptor blocker. During the 5-day hospital stay, no complications occurred. On clinical follow-up 6 months after the initial procedure, the patient was in functional class II with marked improvement in the patient's regular activities, and no recurrence of angina was reported.

## DISCUSSION

Angina-like chest pain might be common in adolescents with congenital cardiac defects but is usually not associated with cardiovascular disease, particularly not in cyanotic patients.<sup>1</sup> The extramural coronary arteries in cyanotic congenital heart disease are typically dilated, sometimes aneurysmatic, because of vascular remodelling because of the endothelial vasodilative substances released.<sup>2</sup>

There might be several reasons for chest pain in context with congenital heart anomalies—for example, aortic pathology, pulmonary embolism and pericarditis or other non-cardiac reasons such as musculoskeletal disorders. The underlying mechanisms of angina pectoris-like chest pain in patients with severe pulmonary hypertension still remain controversial.<sup>3</sup> A painful dilation of the pulmonary artery during transient increases in pulmonary artery pressure as well as an increased myocardial oxygen demand secondary to right ventricular overload and right ventricular subendocardial ischaemia have been suggested as possible mechanisms.<sup>4–6</sup>

Our patient represents a rare example of coronary artery obstruction caused by external mechanical compression by an enlarged pulmonary artery trunk which could be treated successfully by coronary artery stenting. However, to our knowledge, this is the first case described of this entity in an adolescent. In adults with idiopathic or secondary pulmonary hypertension, a few cases of extrinsic LMCA compression have been suggested to cause left ventricular dysfunction.<sup>7–11</sup> Although left ventricular dysfunction is commonly attributed to be a consequence of ventricular interdependence in patients with pulmonary hypertension, physicians should be alert about an external compression of the LMCA by a distended pulmonary trunk in these patients.<sup>12</sup> In most case reports on LMCA

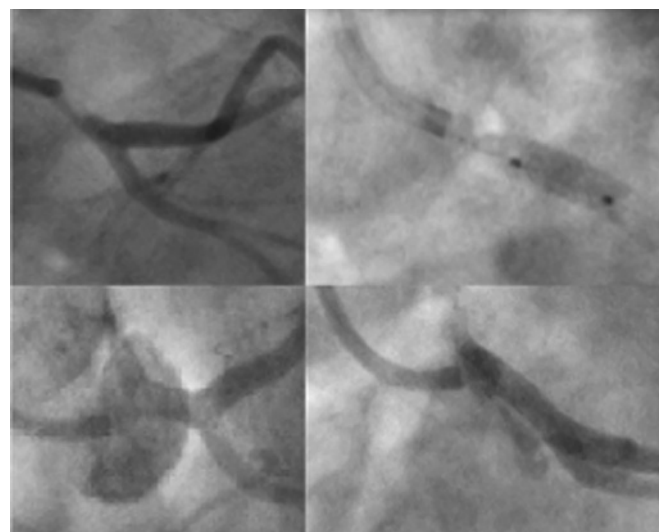


**Figure 2** (A) MRI scan showing a dilated pulmonary artery trunk (P) and compressed left main coronary artery. P, pulmonary artery trunk; the arrow shows narrowing of the coronary artery. (B) MRI scan showing late enhancement of the interventricular septum (arrow).

compression in cyanotic adults, diagnostic imaging was primarily performed by selective coronary angiography, which was the method of choice because of the patients' clinical presentation, cardiovascular risk factors and age.<sup>13 14</sup>

Because chest pain and left ventricular dysfunction are induced by coronary artery disease only very rarely in younger cyanotic patients, they can easily be overlooked. However, as timely diagnosis and treatment may be lifesaving, clinicians need to be aware of the presence of coronary artery disease in younger cyanotic patients. Markers of myocardial ischaemia such as Troponin T and CK/CK-MB can be helpful in forming a diagnosis, whereas non-invasive studies, such as MRI, may also be valuable but delay time to revascularisation in an acute myocardial ischaemia. Coronary angiography remains crucial in defining the diagnosis and treatment.

With the advent of drug-eluting stents, percutaneous coronary intervention for LMCA disease has become a widely accepted technique.<sup>11 15</sup> Several registries and randomised studies have shown no differences in survival with bypass surgery or coronary stenting for unprotected LMCA disease or multivessel coronary artery disease.<sup>16 17</sup> The higher rate of



**Figure 3** Coronary angiography demonstrating a severe stenosis of the left main coronary artery (left upper). Successful stenting with a drug-eluting stent (right upper). Following stenting, injection into the left aortic sinus, the left main coronary artery has no obstruction any longer (bottom left and right).

repeat revascularisation procedures observed with bare metal stenting as compared with bypass surgery has been reduced with the use of drug-eluting stents.<sup>18 19</sup> However, optimal treatment for Eisenmenger patients with LMCA compression remains unknown. Sivakumar *et al* reported on stenting of the LMCA in a 58-year-old patient with diabetes with secondary pulmonary hypertension using a bare-metal stent, whereas Dubois *et al* treated a 51-year-old smoker with pulmonary hypertension in a similar presentation with a zotarolimus eluting stent.<sup>13 14</sup> Caldera *et al* performed a literature review of the few reported cases revealing the currently heterogeneous strategies for this condition mainly depending on interventional techniques.<sup>20</sup> Further information has to be gathered to develop helpful guidelines for clinicians. Still, particularly in patients with a high risk for surgery, for example patients with congenital cardiac defects, drug-eluting stenting of the unprotected LMCA represents a favourable option of revascularisation in an acute condition.

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