A 72 year old woman with ALCAPA

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Abstract

ALCAPA syndrome (anomalous origin of the left coronary artery from the pulmo-

nary artery), which causes the left coronary artery to grow with an anomalous or-

igin from the pulmonary artery, is a rare disease which may result in myocardial

infarction, congestive heart failure, and sometimes death during the early infantile

period. A 72 year old woman with ALCAPA syndrome is presented. The asympto-
matic patient presented with a cardiac murmur which was discovered during a

routine check up for a gynaecological intervention. Coronary cineangiography

established the diagnosis. Although surgical correction is the usual treatment for

such cases, medical treatment was preferred for this patient because she was

asymptomatic without clinical signs of heart failure.

Keywords: coronary vessel anomalies; ALCAPA syndrome

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare

congenital cardiovascular defect.1–6 An extended left ventricle infarction can be fatal if

the defect is not corrected surgically in the first years of life.2 5–9 Patients with ALCAPA syn-
drome who survive past childhood often have varying symptoms of myocardial ischaemia or

heart failure, depending on the development of collateral circulation, and up to 90% die

suddenly at a mean age of 35 years.2 7 9 There are few patients older than 50 who have ALCAPA

syndrome that has not been surgically treated.

Until now only 26 patients have been described.4 We report a 72 year old woman

with ALCAPA, who was almost asymptomatic in spite of a significant left to right shunt.

Case report

A 72 year old woman was referred to our hospital to determine the clinical importance of a

continuous heart murmur which had been discovered during a routine check up before a

gynaecological procedure. She had no cardiac antecedents but at presentation she mentioned

mild shortness of breath (New York Heart Association functional class I) and palpitations
during heavy exercise.

Physical examination revealed a continuous murmur in the 3rd and 4th parasternal

intercostal spaces (grade 3/6) and a mild holo-
systolic murmur at the apex. There were no

clinical signs of left or right heart failure. ECG

showed lateral repolarisation abnormalities (fig

1). Chest x ray revealed cardiomegaly and

prominent vascular trauma (fig 2), and trans-

thoracic echocardiography showed a normal

right ventricle and a mildly dilated left ventricle

with a minimal hypertrophic and hypokinetic

terventricular septum. Left ventricular ejec-
tion fraction, measured in M mode, was 74%.

No septal defects were found and colour Dop-
pler imaging revealed only mild mitral and tri-
cuspid regurgitation. Estimated systolic right

ventricular pressure was 40 mm Hg, and we

observed a dilated blood vessel with an

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Figure 1 Electrocardiogram showing lateral repolarisation abnormalities.

Figure 2 Chest x ray showing cardiomegaly and prominent vascular trauma.
approximate diameter of 1 cm. It seemed to originate from the ventral side of the aorta, with prominent laminar diastolic flow compatible with coronary artery flow. A referring cardiologist suggested a single coronary artery or an important coronary fistula.

A transoesophageal echocardiogram showed a severely dilated right coronary artery originating from the right coronary cusp and connecting distally to the pulmonary trunk 2 cm above the pulmonary valve. The origin of the left coronary artery in the left coronary cusp could not be found.

The diagnosis of ALCAPA was made by coronary angiography, which revealed an extremely dilated and tortuous right coronary artery with strong collateral filling of the left coronary artery, shunting into the pulmonary trunk (fig 3). Saturation sampling during right heart catheterisation showed a mild left to right shunt (Qp/Qs = 1.5/1) with a clear saturation gap of 10% between the lower and the higher section of the pulmonary trunk (table 1). The suspected pulmonary hypertension which had been indicated by echocardiography was confirmed (table 2). Nuclear magnetic resonance was also able to locate the origin of this vessel (fig 4).

Conservative treatment with an angiotensin converting enzyme inhibitor was proposed, due to the age of the patient and the fact that she

![Figure 3](image_url) 
Coronary angiography showing an extremely dilated and tortuous right coronary artery (RCA) with strong collateral filling of the left coronary artery (LCA) shunting into the pulmonary trunk (PT).

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Saturation sampling during right heart catheterisation showing a mild left to right shunt (Qp/Qs = 1.5/1) with a clear saturation gap of 10% between the lower and the higher section of the pulmonary trunk</th>
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<tbody>
<tr>
<td>Oxygen saturation (%)</td>
<td></td>
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<tr>
<td>Vena cava inferior</td>
<td>65.6</td>
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<tr>
<td>Vena cava superior</td>
<td>62.1</td>
</tr>
<tr>
<td>Right atrium high</td>
<td>61.5</td>
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<tr>
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<td>63.1</td>
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<tr>
<td>Right atrium low</td>
<td>53.3</td>
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<tr>
<td>Right ventricle high</td>
<td>62.2</td>
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<tr>
<td>Right ventricle mid</td>
<td>61.6</td>
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<tr>
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<td>59.2</td>
</tr>
<tr>
<td>Arteria pulmonalis trunk low</td>
<td>68.7</td>
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<tr>
<td>Arteria pulmonalis trunk high</td>
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<tr>
<td>Arteria pulmonalis left</td>
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<tr>
<td>Arterial</td>
<td>95.9</td>
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<table>
<thead>
<tr>
<th>Table 2</th>
<th>Pressures measured during right heart catheterisation showing pulmonary hypertension (mean 30 mm Hg)</th>
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<tbody>
<tr>
<td>Pressure</td>
<td>Systolic (mm Hg)</td>
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<tr>
<td>Right atrium</td>
<td>–</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>40</td>
</tr>
<tr>
<td>Arteria pulmonalis</td>
<td>40</td>
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<tr>
<td>Pulmonary capillary wedge pressure</td>
<td>23</td>
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<tr>
<td>Aorta</td>
<td>170</td>
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<tr>
<td>Left ventricle</td>
<td>176</td>
</tr>
</tbody>
</table>

![Figure 4](image_url) 
Nuclear magnetic resonance scan showing a dilated left coronary artery (LCA) merging with the pulmonary trunk (PT). (A) T1 weighted longitudinal section; (B) T2 weighted cross section.
was asymptomatic without clinical signs of heart failure.

**Discussion**

ALCAPA was first mentioned in 1908, in a 60 year old woman. The syndrome was also described by Bland et al in 1933 after the study of the clinical and necropsy findings from an infant with symptoms of dyspnoea and myocardial infarction.

The estimated incidence of ALCAPA is 1/300 000 live births (between 0.24% and 0.46% of all congenital cardiac anomalies). Some believe that this is a significant underestimation of the true incidence, as many patients may be asymptomatic until their death and therefore remain undiagnosed. This case confirms the possibility that many cases may remain undetected.

ALCAPA develops before birth when the systemic and pulmonary arterial pressures are equal and there is antegrade flow in both the left and right coronary. In the neonatal period this gradually changes as the pulmonary blood pressure diminishes, the ductus arteriosus closes, and the flow in the left coronary artery reverses. The development of collateral circulation between the right and left coronary artery during closure of the duct and lowering of the pulmonary pressure will determine the extent of myocardial ischaemia.

If not treated surgically, up to 85% of patients with ALCAPA die in infancy or childhood, and untreated ALCAPA in the elderly is extremely rare. In 1995, Alexi-Meskishvili and colleagues found only 26 patients over 50 years old. The clinical presentation of older ALCAPA patients is usually non-specific: syncope, cardiac arrhythmias, exertion fatigue, nocturnal dyspnoea and less commonly, angina pectoris. The defect is sometimes discovered during necropsy after sudden death.

Clinical examination reveals a systolic murmur in almost 70% of cases. As in our patient, the ECG usually shows lateral ischaemia or sequel of a (anterior) myocardial infarction. Bicycle testing, myocardial scintigraphy, positive emission tomography, and dobutamine stress echocardiography can all reveal chronic ischaemia, which can be improved through surgery. Diagnosis is usually established by a coronary angiogram, which shows a dilated and tortuous right coronary artery with collateral filling of the left coronary artery system; variable degrees of shunting to the pulmonary artery may be present. Right heart catheterisation may show an increase in oxygen saturation at the point where the blood of the left coronary artery merges with the blood from the pulmonary artery. Surgical treatment usually recreates a dual coronary perfusion, regardless of the presence of symptoms or state of myocardial function.

However, in our patient, who may be the oldest to be reported with this anomaly, the risk of cardiac surgery seemed to outweigh the natural prognosis. Therefore we chose not to intervene surgically.

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