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Surgical treatment of a fistula between the right pulmonary artery and the left atrium: presentation of two cases and review of literature

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Abstract

Objective: A direct communication between the pulmonary artery and the left atrium is a rare anomaly. On the basis of two cases of our own and a literature review of 49 cases, we focus on clinical presentation, anatomy, diagnosis, and the role of surgery.

Methods: Two cases of a fistula between the right pulmonary artery and the left atrium are described in a girl of 4 years and a boy of 15 years. Both presented with unexplained cyanosis. Diagnosis was made on echocardiography and angiography. The fistula was ligated using extracorporeal circulation in the first case and not in the second case. Results: The surgical results were successful with resolution of the cyanosis. Conclusions: In newborns, urgent surgery may be necessary. In other patients, early elective surgical correction should be performed to prevent complications, especially systemic and cerebral emboli, cerebral abscesses, and rupture of aneurysmal fistulas. Complete cure can be achieved by ligation and possible division or by intracardiac repair. © 1997 Elsevier Science B.V.

Keywords: Cerebral abscess; Cyanosis; Extracorporeal circulation; Right pulmonary artery–left atrium fistula

1. Introduction

A direct communication between the right pulmonary artery (RPA) and the left atrium (LA) is a rare anomaly. Central cyanosis with clubbing of fingers and toes, exertional dyspnea and decreased arterial oxygen saturation usually accompany the lesion. Complications are cerebral and systemic emboli, cerebral abscesses and rupture of the fistula. On the basis of our two cases and a literature review, we focus on clinical presentation, anatomy, diagnosis and the role of surgical correction, as complete cure can be achieved by ligation and possible division of the fistula or by intracardiac repair.

2. Case reports

2.1. Case A

A 4-year-and-3-months-old girl was admitted to hospital because of sleeping problems. She had severe cyanosis and, in retrospect, her mother stated, that for many years she had had a cyanotic color. On examination, a cyanotic girl was seen, 110 cm tall and weighing 17.5 kg. Her blood pressure was 95/65 mm Hg. She had cyanosis of the lips and limbs with clubbing of the fingers. No tachypnea, dyspnea, or murmurs were noted. Liver and spleen were not palpable. Laboratory examination showed a hemoglobin concentration (Hb) of 11.7 mmol/l, a hematocrit (Ht) of 0.57 and a platelet count of 244 x 10⁹/l. Electrocardiography showed a normal sinus rhythm. Chest X-ray revealed no abnor-
pulmonary vein with a continuous flow pattern at its entrance into the LA. Perfusion scintigraphy of the lungs clearly demonstrated a right-to-left shunt (R-L shunt) of 34%. On cardiac catheterization, the systolic pressure in the right ventricle was 20 mm Hg. Angiography revealed a large central arteriovenous fistula (AVF), between the RPA and the LA (Fig. 1). On 17 October 1994, a median sternotomy was performed. A large arteriovenous communication between the right main pulmonary artery and the roof of the LA, with a diameter of 5 mm, was dissected free. The fistula was doubly ligated and divided with the child on extracorporeal circulation (ECC). Arterial oxygen saturation, being about 80% before ECC, increased to 100% immediately after ECC. The postoperative course was uneventful. Until now, 1 1/2 years after surgery, the patient has been in good health. Digital clubbing has disappeared and Hb and Ht have normalized.

### 3. Discussion

Direct communication between the right pulmonary artery and the left atrium is not as rare as recently suggested by Sawant and Nair [48]. The first case was operated by Blalock and described by Friedlich et al. in 1950 [18]. Since then, 51 cases (including our two cases) have been reported in the literature (Table 1). The male:female ratio was approximately 3:1. Only one fistula has been reported between the left pulmonary artery and the LA [26].

Eight patients were diagnosed at birth. All of them needed urgent surgical intervention. Three newborn patients deteriorated before surgical intervention could be undertaken. Only three newborn patients survived. Causes of death were probably related to additional
congestive heart failure. Genetic examination excluded
dolichostenomelia, wrist/thumb sign and Steinberg sign.
The metacarpal index was 7.25 and pedes caves were
present. Ophthalmologic examination excluded lens
luxation. The urine amino-acid chromatogram was
normal. There were no definitive signs of Marfan's syn-
drome and family history was also negative for this
disorder. On auscultation of the heart, a soft systolic
and diastolic murmur were both best heard parastern-
ally in the fourth left intercostal space. Laboratory
tests showed a Hb of 10.7 mmol/l, a Ht of 0.52 and a
platelet count of $1 \times 10^9$/l. Chest X-ray revealed a
prominent right heart and a shadow at the side of the
left atrium. Echocardiography and cardiac catheteriza-
tion, including selective angiography, showed a wide
main pulmonary artery with wide branches. A direct
communication was seen between the RPA and the LA.
The ascending aorta was dilated (Fig. 2) and there was
minor regurgitation of the aortic valve. Systemic arte-
rtrial saturation at rest was 90%. On 4 June 1981, a
median sternotomy was performed. The LA was some-
what dilated at the site of connection of the right
pulmonary veins. The aortic root and ascending aorta
were dilated. The main pulmonary artery was also
markedly ectasic. Because of the suspected friability of
the aortic wall, it was elected to attempt ligation of the
fistula without ECC. Since the patient was asym-
tomatic of aortic disease, it was also decided not to
perform reconstructive surgery at this point. The large

lesions, which mostly included patent ductus arteriosus
and/or patent foramen ovale [15,17,24,43,47]. Therefore,
patency of the ductus arteriosus seemed im-
portant, causing a huge volume load of the left ventricle
and leading very rapidly to heart failure. Older patients
had a milder form of the disease with an important

Fig. 1. Angiogram of patient A showing clearly the anomalous fistula
between the right main pulmonary artery and the left atrium (white
arrow).
ventricles hypertrophy (LVH), with left axis deviation. 

Figure 1: ECG may reveal left atrial dilatation, and left

ventricular hypertrophy (LVH), with left axis deviation.

The image below shows atrial hypertrophy and poststenotic
capillary dilatation. The right atrium and the left atrium in patien \textit{B} and \textit{C}.

The 2-L approach may occur. Characteristics of the lesion are

Dependent site of atrial location of the R-1.

Figure 2: The outcome of valve replacement is

some annular thinning, valvular tissue, and leaflet

damage in diastole, as we age, at dissection range form

3 months to 10 years, with an average of 4 years

known.

The image above of patient \textit{B} which reveals markedly thickened
In conclusion, apart from mild cyanosis and finger clubbing, direct communication between the right pulmonary artery and the left atrium may give few symptoms, causing a significant delay in diagnosis. Electrocardiography and chest X-ray may be completely normal. Echocardiography and cardiac catheterization, including selective angiography, provide the necessary information. To prevent complications, especially systemic and cerebral emboli, early surgical intervention should be performed. Complete cure can be achieved by ligation and possible division or by intracardiac repair.

**References**


