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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 10^9/l. Electrocardiography showed a normal sinus rhythm. Chest X-ray revealed no abnor-
A 15-year-old boy presented with decreased effort tolerance. In retrospect, the patient had had slight cyanosis at rest for years. On physical examination, a cyanotic boy was seen with a height of 170 cm and weighing 50 kg. He had finger clubbing, but no signs of  

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 cardiac abnormalities.
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 syndrome and family history was also negative for this disorder. On auscultation of the heart, a soft systolic and diastolic murmur were both best heard parasternally 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 $171 \times 10^9$/l. Chest X-ray revealed a prominent right heart and a shadow at the side of the left atrium. Echocardiography and cardiac catheterization, 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 arterial saturation at rest was 90%. On 4 June 1981, a median sternotomy was performed. The LA was somewhat 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 asymptomatic 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 important, 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).](image-url)
but the fistula causes huge volume load of the left heart only if there is a very large shunt. TTE and TEE sometimes reveals LVH with poorly filled pul-
monary arteries, an abnormal systolic mitral regurgitation, and a dilated atrial septum. Cardiac catheterization with cardiac MRI may also be performed to better assess the lesion clearly. At cardiac catheterization, oxygen saturation of the LA and/or involved pulmonary vein is markedly reduced and does not respond to the admin-
istration of extra oxygen. Selective angiography of the involved pulmonary artery reveals the anomaly with-
out contrast injection. By contrast, when not treated, the lung, corresponding to the involved pulmonary artery, may lead to lung edema with irreversible pul-
monary hypertension and microorganisms coming

Fig. 2. Aortography of patient B which revealed markedly dilated ascending aorta.

Fig. 3. Intraoperative view of the double-ligated fistula between the right main pulmonary artery and the left atrium in patient B. SVC, right superior vena cava; RAA, right atrial appendage; AO, aorta (embolized with teflon.)
directly into the systemic circulation, thereby bypassing pulmonary filter function, may lead to cerebral complications. Transient ischemic attacks, cerebral infarctions and abscesses occur more often with these fistulas [19,34,37]. Other complications are: endocarditis, infective endarteritis and aneurysmatic growth of the fistula, with the risk of fatal rupture.

To prevent these complications, elective surgery is recommended [8,15,19,37]. Absolute indications for surgical correction are severe cyanosis with a significantly decreased systemic oxygen saturation or severe polycythemia. It is possible to perform embolization of the fistula without surgery [10]. However, direct communication between the RPA and LA exposes the patient to a high risk of major complications. Surgical correction is therefore preferable. In general, the fistula can easily be ligated and divided. Procedures are also described where the fistula is only ligated (without division) or where an intracardiac repair is performed with the use of ECC. A lack of information about the consistency of the fistulous tissue during extracardiac procedures may be a reason to use ECC.

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.

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