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

C.J.A.M. Zeebregts a,*, A. Nijveld a, J. Lam b, A.M. van Oort c, L.K. Lacquet a

 a Department of Thoracic and Cardiac Surgery, Children’s Heart Centre, University Hospital Nijmegen St. Radboud, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands
 b Department of Pediatric Cardiology, University Hospital Amsterdam AMC, Amsterdam, The Netherlands
 c Department of Pediatric Cardiology, Children’s Heart Centre, University Hospital Nijmegen St. Radboud, P.O. Box 9101, 6500 HB Nijmegen, The Netherlands

Received 9 September 1996; received in revised form 22 January 1997; accepted 22 January 1997

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 × 10^9/l. Electrocardiography showed a normal sinus rhythm. Chest X-ray revealed no abnor-
An echocardiogram demonstrated a wide right 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, 1½ years after surgery, the patient has been in good health. Digital clubbing has disappeared and Hb and Ht have normalized.

2.2. Case B

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 communication between the RPA and the LA was doubly ligated (Fig. 3). Arterial oxygen saturation rose from 91 to 99%. The postoperative course was uneventful. He has been seen regularly for assessment of his aortic disease. In 1990 he underwent a Bentall procedure for aortic valve incompetence and ascending aorta aneurysm. In 1996, he underwent a reoperation because of dissection of the remaining native ascending aorta. His present status is good.

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 paras-
ternally 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 × 10⁹/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-
rial 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]. There-
fore, patency of the ductus arteriosus seemed impor-
tant, 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).
ventriculo-pulmonary (TAP) with left atrial deviation. [15] ECG may reveal left atrial deviation and left atrio-ventricular anulation usually shows atrial hypoxia and polycythemia. Clinical and radiologic examination Laboratory examination

copies and derivatives of this paper cannot be the reason for symptoms may occur. Characteristic of the lesion are

Dependent on size and location of the R-L shunt, the size of the LA can be located high or low in the

the lesion. Unilateral pulmonary artery hypertrophy may occur. Secondary cause. Echocardiography is a

ventriculo-pulmonary (TAP) with right atrial deviation. Echocardiography is a sensitive and specific diagnostic tool for the detection and follow-up of this anomaly. RA, RV, LA, LV, SFC, SVC. Heart only if there is a very large shunt (RA-LV channel

shows atrial hypoxia and polycythemia. Clinical and radiologic examination Laboratory examination

The lesion is a developmental anomaly in which the atrial septum is defective, allowing communication between the right and left atria. The diagnosis is usually made by echocardiography. RA, RV, LA, LV, SFC, SVC. Heart only if there is a very large shunt (RA-LV channel

ventriculo-pulmonary (TAP) with left atrial deviation. [15] ECG may reveal left atrial deviation and left atrio-ventricular anulation usually shows atrial hypoxia and polycythemia. Clinical and radiologic examination Laboratory examination

The lesion is a developmental anomaly in which the atrial septum is defective, allowing communication between the right and left atria. The diagnosis is usually made by echocardiography. RA, RV, LA, LV, SFC, SVC. Heart only if there is a very large shunt (RA-LV channel

shows atrial hypoxia and polycythemia. Clinical and radiologic examination Laboratory examination

Some additional variations exist in these patients, such as

Some additional variations exist in these patients, such as
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.

References


<table>
<thead>
<tr>
<th>Gender</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>37</td>
<td>73</td>
</tr>
<tr>
<td>Female</td>
<td>13</td>
<td>25</td>
</tr>
<tr>
<td>Unspecified</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Age</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-1 month</td>
<td>8</td>
<td>16</td>
</tr>
<tr>
<td>1-12 months</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>1-10 years</td>
<td>18</td>
<td>35</td>
</tr>
<tr>
<td>11-20 years</td>
<td>13</td>
<td>25</td>
</tr>
<tr>
<td>21-30 years</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>31-40 years</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>41-50 years</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>51-60 years</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Unspecified</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Approach</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right thoracotomy (RT)</td>
<td>28</td>
<td>55</td>
</tr>
<tr>
<td>Median sternotomy (MS)</td>
<td>12</td>
<td>24</td>
</tr>
<tr>
<td>RT followed by MS</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>No surgery</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Unspecified</td>
<td>4</td>
<td>8</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Type of operation</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligation</td>
<td>27</td>
<td>53</td>
</tr>
<tr>
<td>Division</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Excision</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Intracardiac repair</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Pneumonecctomy</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Exploration</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>No surgery</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Unspecified</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Use of ECC</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Used</td>
<td>29</td>
<td>57</td>
</tr>
<tr>
<td>Not used</td>
<td>11</td>
<td>22</td>
</tr>
<tr>
<td>No surgery</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Unspecified</td>
<td>5</td>
<td>10</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Outcome</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Died</td>
<td>11</td>
<td>22</td>
</tr>
<tr>
<td>Survived</td>
<td>38</td>
<td>75</td>
</tr>
<tr>
<td>Unspecified</td>
<td>2</td>
<td>4</td>
</tr>
</tbody>
</table>


