Early and Late Outcomes of Coil Embolization of Pulmonary Sequestration in Children

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Background: Pulmonary sequestration (PS) is characterized by non-functioning lung tissue fed from 1 or several aberrant systemic arteries. The classical therapeutic approach is surgical resection. Several case reports have shown that coil embolization is feasible, but this technique has not been evaluated in a larger series of consecutively treated patients. The purpose of our study was to assess the early and long-term outcomes of coil embolization of PS in children and to determine the risk factors of early and late major adverse cardiovascular and pulmonary events.

Methods and Results: Between March 1999 and December 2004, 6 patients (2 boys, 4 girls, mean age 4.7±3.8 years) with PS were treated by coil embolization of the feeding systemic artery. Four patients were considered to have been cured and 2 patients required a second coil embolization 6 months later because of residual systemic flow seen on computed tomography. Transient ischemic change of the lower limb occurred in the youngest patient. None of the other patients had any late complications or recurrent pneumonia.

Conclusions: Coil embolization of PS is safe and feasible, with a good late outcome. (Circ J 2009; 73: 938–942)

Key Words: Coil embolization; Extralobar sequestration; Intralobar sequestration; Pulmonary sequestration
Results

All patients had left PS: intralobar in 1 patient and extralobar in the 5. Only the left lower lobes were involved. CT angiography or magnetic resonance angiography (MRA) of the chest after bolus infusion of contrast medium revealed the aberrant systemic artery in all patients. The main source of arterial supply was the thoracic aorta in 5 patients, and in the other the aberrant artery arose from the upper abdominal aorta at the level of the diaphragmatic artery.

The aberrant artery was single in 5 patients and double in 1. The venous drainage differed between the types of PS. The veins drained into the azygos system in the 5 cases of extralobar PS and into the inferior pulmonary vein in the 1 case of intralobar PS. After coil embolization, angiography showed complete occlusion in 4 patients (Figure 2) and partial occlusion in 2 cases (Figure 3), for whom a second coil embolization was performed 6–13 months later. Angiography showed complete occlusion in these 2 patients after the secondary coil embolization. Patient characteristics, the type of PS and embolization coil size are given in Table.

Complications occurred in 1 patient who had a transient
ischemic change of the lower limb after cardiac catheterization, but this resolved after 2 days of medical treatment with heparin. No other complications of fever, arrhythmia, pleural effusion or cardiopulmonary disorder were noted. None of the other patients had any complications and they were discharged from hospital after 3 days.

Patients were followed for a mean of 13 months (range 6–18 months) after the coil embolization. Follow-up examinations, including echocardiography and CT angiography or MRA, were scheduled at 6 or 18 months after the cardiac catheterization. One year later, there was complete involution of the lung parenchyma of the sequestration on CT scan and MRA in 3 patients (Figure 4). In the other 3 patients, involution was partial (Figure 5). No cardiopulmonary complications had occurred 6 or 18 months later. There was no infection of the sequestrated lung after the coil embolization, although 1 patient developed pneumonia 8 months after the cardiac catheterization.

Table. Patient Background and Characteristics of the Type of PS, Complications and Long-Term Follow-up

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Aberrant artery no.</th>
<th>Result</th>
<th>Complication</th>
<th>Long-term follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1.4</td>
<td>M</td>
<td>Shortness of breath</td>
<td>Extralobar PS &amp; PAPVR, ASD</td>
<td>1</td>
<td>Complete occlusion</td>
<td>No</td>
<td>No infection or complications</td>
</tr>
<tr>
<td>2</td>
<td>2.9</td>
<td>F</td>
<td>Recurrent infection</td>
<td>Extralobar PS &amp; severe MR, MVP s/p mitral valve replacement</td>
<td>1</td>
<td>Complete occlusion</td>
<td>No</td>
<td>Pneumonia</td>
</tr>
<tr>
<td>3</td>
<td>13.8</td>
<td>F</td>
<td>Recurrent infection</td>
<td>Intralobar PS</td>
<td>1</td>
<td>Complete occlusion</td>
<td>No</td>
<td>No infection or complications</td>
</tr>
<tr>
<td>4</td>
<td>0.3</td>
<td>F</td>
<td>Heart murmur, shortness of breath</td>
<td>Extralobar PS</td>
<td>1</td>
<td>Complete occlusion</td>
<td>Ischemic change in the lower leg</td>
<td>No infection or complications</td>
</tr>
<tr>
<td>5</td>
<td>1.3</td>
<td>F</td>
<td>Shortness of breath</td>
<td>Extralobar PS</td>
<td>1</td>
<td>Partial occlusion &amp; re-embolization</td>
<td>No</td>
<td>No infection or complications</td>
</tr>
<tr>
<td>6</td>
<td>8.3</td>
<td>M</td>
<td>Recurrent infection</td>
<td>Extralobar PS</td>
<td>2</td>
<td>Partial occlusion &amp; re-embolization</td>
<td>No</td>
<td>No infection or complications</td>
</tr>
</tbody>
</table>

PS, pulmonary sequestration; PAPVR, partial anomalous pulmonary venous return; ASD, atrial septal defect; MR, mitral valve regurgitation; MVP, mitral valve prolapse.

Figure 4. A 4-month-old infant with pulmonary sequestration suffered from shortness of breath. (A) Pre-embolization contrast-enhanced computed tomography (CT) scan demonstrates a soft-tissue density mass in the left lower chest (black arrow) and a feeding vessel originating from the aorta. (B) Post-embolization contrast-enhanced CT scan shows complete occlusion of the feeding vessel by embolization coils (black arrow) and complete involution of the sequestration.

Figure 5. A 1.3-year-old girl with a large aberrant artery from the descending aorta suffered from shortness of breath. (A) Pre-embolization magnetic resonance angiography T1-weighted view of the chest shows a hypervascular mass with cystic changes in the left lower lung. (B) Post-embolization contrast-enhanced computed tomography scan shows coil embolization of the aberrant feeding artery (black arrow) and partial involution of the lung parenchyma of the sequestration.
Discussion

PS is a rare clinical entity, fewer than 2% of congenital abnormalities, and the term “sequestration”, which was first used in the medical literature by Pryce in 1946, originates from the Latin word sequastare, which means “to separate”. PS is the term given to a region of lung parenchyma that is partially or completely separated from the bronchopulmonary tree of the lung proper. It may present in children or adults as recurrent pneumonia in a persistent location. The sequestration has an aberrant systemic, rather than pulmonic, arterial supply. Multiple aberrant arteries are seen in 15–20% of cases.

Multiple theories of the pathogenesis of PS have been proposed and were summarized by Corbett and Humphrey. The recent hypothesis is that it results from the formation of an accessory lung bud inferior to the normal lung buds during development. During embryogenesis this “accessory” lung bud develops an independent vascular supply, usually from the aorta, and remains independent of the normally developing tracheobronchial tree.

Two forms of PS are recognized: extralobar, which is separated from the lung tissue by a separate lining of pleura, and intralobar, which is embedded in the normal lung and shares it with a common pleural investment. Extralobar sequestration is a disease confined to neonates or children and almost uniformly occurs in the left lower lobe. More than 60% of patients with extralobar sequestration have coexistent congenital anomalies. Intralobar sequestrations comprise 75% of all sequestrations, and are typically diagnosed in adulthood. In contrast to extralobar sequestration, they are usually isolated anomalies. In our study, there were 5 patients with extralobar PS and 1 with intralobar sequestration; 2 of the 5 patients with extralobar sequestration had associated congenital heart disease: 1 had coexistent severe mitral valve regurgitation and valve prolapse, and the other had coexistent partial anomaly of pulmonary venous return and atrial septal defect.

Angiography is considered the conventional standard method for identifying the aberrant feeding artery of PS. In recent years, less invasive imaging techniques have proved to be equally effective and safer alternatives to angiography, including CT angiography, Doppler ultrasound, and MRA. Color-enhanced, 3-dimensional MRA and 3-dimensional CT angiography can identify both arterial and venous aberrations and define the soft tissue abnormalities associated with sequestration. In our series of patients, PS was verified by CT angiography and cardiac angiography.

Surgery is the conventional treatment of PS in order to prevent possible infection. But in asymptomatic patients, surgical treatment is debatable. Endovascular embolization and coil embolization are less invasive alternatives to surgery. These methods can be used to protect the lungs from excessive intraoperative bronchial blood flow. We treated PS with embolization coil in order to prevent recurrent infection or shortness of breath in principle and report our results to answer the question whether asymptomatic patients should be treated by embolization coil.

In our study, 4 of 6 patients were cured after the first embolization and the remaining 2 patients were cured by partial embolization of the aberrant artery. In 1 of those patients, who had a large aberrant artery, partial embolization did not prevent infection and re-embolization was performed 6 months later to block the aberrant artery completely. In the other patient, who had 2 aberrant arteries, coil embolization of 1 artery was performed at the first embolization and re-embolization was done 14 months later because of the recurrence of infection.

After embolization, complications occurred in 1 patient, who had a transient ischemic change of the lower limb, which resolved after 2 days of medical treatment with heparin. No complications of fever, chest pain, or pleural effusion were noted in the remaining 5 patients.

All patients were followed up for 6 months or 18 months after embolization. All patients had a normal chest X-ray during follow-up. CT angiogram and MRA best demonstrate the pulmonary parenchymal abnormalities and seem to be more sensitive than chest X-ray for following these patients. In our study, 5 patients with stainless steel coils were followed up by CT angiography and the remaining patient with a platinum coil was followed up by MRA. In our study, at 1-year follow-up, 2 patients had complete involution of the PS parenchyma on CT angiography, 1 patient had complete involution of the sequestration parenchyma on MRA and 1 had partial involution. In the 2 cases in which the aberrant artery was partially occluded at the first embolization, CT angiography was performed 6 months after re-embolization and on CT angiography there was partial involution of the sequestration parenchyma in both patients. Four months later, infection of sequestration occurred in the patient whose large aberrant artery had been partially embolized and re-embolization was performed 2 months after infection had resolved. After re-embolization, there was no further infection of the sequestration for 8 months. Infection of the sequestration frequently occurred in the other patient whose 2 aberrant arteries were partially occluded at the first embolization, re-embolization was performed 14 months after the first. There was no infection of the sequestration during 6 months of outpatient dept (OPD) follow-up of this patient. None of the other 4 patients had infection of the sequestration.

Long-term follow-up studies of children treated surgically for PS have been reported and complications include pneumonia, asthma, gastroesophageal reflux, peptic ulcer, pyloric stenosis, and intralobar sequestration. Garcia-Pena et al reported spontaneous shrinkage of PS without complete involution in 1 patient. In their study, cases of complete involution of the lung parenchyma of sequestration had no recurrent pulmonary infection. It is not known whether the risk of infection remains the same for partial involution, and this question remains controversial. In our study, no cardiopulmonary complications occurred in any of the patients during OPD follow-up. One patient developed pneumonia 8 months after embolization, but improved after 10 days of antibiotic treatment.

Because spontaneous regression of PS has been reported, we suggest that patients less than 3 months of age should not be treated by coil embolization, because femoral arterial puncture in young infants is more difficult and ischemic change of the lower leg of young infants is more likely. In our study, the only complication occurred in a 4-month-old infant with ischemic change of the lower leg.

Conclusion

We suggest the following algorithm for the non-surgical management of children with PS.

1. PS in children can be treated by coil embolization.
2. OPD follow-up 6-18 months after embolization to
include chest X-ray, CT angiography, MRA and cardiac angiography. Coil embolization seems to be a safe and effective alternative to surgery in children with PS.

3. Long-term morbidity seen after surgery for PS, including pneumonia, gastroesophageal reflux, asthma, pyloric stenosis and musculoskeletal deformity, does not occur after coil embolization.

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References