Long-term effects of combined percutaneous atrial septal defect occlusion and pulmonary valvuloplasty in children below 2.5 years old

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Long-term effects of combined percutaneous atrial septal defect occlusion and pulmonary valvuloplasty in children < 2.5 years old

**Brief title:** ASD and PS treatment in children < 2.5 years

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**INTRODUCTION**

The combination of atrial septal defect (ASD) type II and pulmonary valve stenosis (PS) is a relatively uncommon congenital heart disease.

When this condition is present, usually significant left-to-right shunt is prevented by right outflow obstruction, but right ventricular hypertrophy progresses. Transcatheter pulmonary balloon valvuloplasty (PBV) and closure of the ASD are the treatment of choice [1,2]. Combined percutaneous ASD closure and PBV is an infrequent but feasible method of treatment, mostly described in adults [3,4] but also in children [5,6]. We present a unique group of young children, in whom the main indication for percutaneous intervention was significant PS and simultaneously important clinical ASD. The long-term results of this procedure are also presented.

**METHODS**

Between 1999 and 2014 in our centre, 157 cases of ASD in children under 3 years old were closed
percutaneously [7]. In this group, there were 8 patients aged under 2.5 years (3 girls) with PS treated simultaneously by PBV. In all, the indication for such therapy was a transpulmonary gradient ≥ 40 mmHg. The clinical data of these patients are presented in Table 1. The mean age of the patients was 1.7 years (0.5–2.5 years) and mean weight was 12 kg (8.7–16.7 kg). In all patients (apart from one), overload of right cardiac cavities was found. In seven patients, left-to-right shunt through the ASD was present. In a two-year-old child with borderline hypoplastic RV and critical PS in the neonatal period, PBV was performed with subsequent surgical left Blalock-Taussig anastomosis (Table 1, patient 7). In this child, bidirectional shunting through the ASD was observed with an oxygen saturation of 86%. In another child in the neonatal period, pulmonary valve atresia with intact ventricular septal and ASD were diagnosed and treated by perforation of the atretic valve with stiff part of guidewire, with subsequent PBV (patient 5). So, in these two patients, a second PBV was performed because of restenosis. In six patients, a single ASD was present, and in two patients, multiple ASDs were present (there were small additional fenestrations located close to the main defect (patients 4 and 8). In all cases, the extent of ASD and PS were estimated before the procedure by transthoracic echocardiography (TTE), confirming the diagnosis during catheterisation by transoesophageal echocardiography (TEE). The decision for treatment (PBV and simultaneous ASD closure) was made by a paediatric surgeon and the paediatric cardiology team.

All the transcatheter interventions were performed in a haemodynamic laboratory under general anaesthesia with intubation and antibiotic coverage. Standard application of heparin 100 IU/kg before the procedure were used. Diagnostic right heart catheterisation was done. Thereafter, PBV as performed in a standard fashion with a Tyshak balloon as the first procedure in all patients (to avoid catheter manipulation after ASD device implantation). The balloon diameter/valvular annulus ratio was 1.1–1.4. The transpulmonary gradient was measured again immediately after PBV. ASD was closed with an Amplatzer Septal Occluder (ASO, St. Jude Medical now Abbott Co.). The ASD closure was performed under fluoroscopic control (mean time 11 min) and TEE guidance. A necessary condition for transcatheter ASD closure was the presence of atrial septal rims (at least 5 mm) in order to avoid embolisation of the device. The exception was the aortic rim, which could be deficient. The implant size was selected either on the basis of the defect stretch diameter (patients 1, 2, 3, 6, 8) or with reference to the defect diameter defined by TEE (it was equal to 1 mm or bigger than the biggest measurement in this examination (patients 4, 5, 7). So, catheter balloon calibration was omitted in 3 children with centrally located defects with good surrounding rims. Each ASO was implanted in a standard fashion [8], and before its release, a precise evaluation of the implant position was made using TEE. After the procedure, aspirin in a dosage of 3–5 mg/kg daily during the first 6 months was prescribed. The results of the interventions were evaluated after the first, third, sixth, and 12th months, as well as when good results were present - every year.
RESULTS AND DISCUSSION

All the procedures were successful, without any intraoperative or postoperative complications. Development of all treated children was excellent in follow-up over 10.3 years (7.4–15.7 years). The mean transpulmonary gradient of 56 mmHg (35–81 mmHg) decreased to 20.4 mmHg (15–31 mmHg) after PBV. In two patients, slight PS with moderate pulmonary valve insufficiency (PI) was observed immediately after the procedure and in follow-up (patients 5 and 7). The probable reason for PI in both cases was a dysplastic pulmonary valve. Both patients had complex defects treated by PBV in the neonatal period. The development of the RV in the first child was adequate (patient 5). In the second child (patient 7), normalisation of oxygen saturation was observed, with some degree of persistent RV hypoplasia. The mean ASD diameter on TEE was 10 mm (4–13 mm), and the mean size of applied ASO was 12.1 mm (5–18 mm). The mean implant size and body weight ratio in all of the groups was 1.0 (ranging from 0.45 to 1.38). In the two-year-old child with a relatively small ASD (5 mm) with bidirectional shunting, saturation after the procedure permanently increased from 86% to 96% (patient 5). Residual shunting through the ASD was present in this child (from the beginning of follow-up). In this patient, an initially double ASD was present and the second ASD was distal to the applied device. In all other patients, residual leaks were not observed.

In patient no. 2 (Table 1) with a 4-mm ASD estimated on TEE (stretch diameter of 8mm) and with enlarged RV, the decision for ASD closure during pulmonary valvuloplasty procedure was made at a firm request of the parents.

The optimal time for transcatheter ASD closure in children is still an open issue. It is believed that small defects tend to close spontaneously, while the bigger ones tend to increase in size [9]. It is also believed that significant ASDs should be closed around the age of 4 to 5 years. Our experience indicates that percutaneous closure of ASD is also feasible in infants and children below 3 years of age [7, 8], but we have to remember that spontaneous closure of even large ASDs can occur [10].

The data presented here document that another indication for early transcatheter ASD closure is the coexistence of important PS.

The youngest documented patient treated with these techniques was a neonate with critical PS and ASD [6]. In our study, the youngest patient was six months old, and the mean age of the treated patients was 1.7 years. In our practice, we performed PBV as the first procedure to avoid catheter manipulation after the placement of the ASD device. Other authors preferred to perform the ASD closure first (before PBV) [4] or as a staged procedure (postponing PBV a few days or
months) [3]. Chinese authors compared 35 patients who underwent percutaneous correction of both
defects with 43 patients treated surgically. All patients had simultaneous complete correction, and
no serious complications occurred in either group. They stated that the transcatheter procedures of
ASD and PBV were valuable alternatives to cardiac surgery correction [11]. Our experience of the
long-term efficacy of such percutaneous procedures confirm their observations.

In conclusion, interventional technology addressing multiple simultaneous congenital
cardiac defects (ASD and PS) as a combined procedure in young children (< 2.5 years old) is safe
and effective in short- and long-term observations.

Conflict of interest: none declared

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**Table 1.** Clinical data of children (< 2.5 years old) in whom percutaneous closure of atrial septal defect (ASD) and pulmonary balloon valvuloplasty (PBV) were simultaneously performed.

<table>
<thead>
<tr>
<th>No.</th>
<th>Sex</th>
<th>Age (y)</th>
<th>Weight (kg)</th>
<th>Date of procedure</th>
<th>ASD diameter (mm)</th>
<th>PS gradient (mmHg)</th>
<th>Fluor (min)</th>
<th>Additional info</th>
<th>Follow-up years</th>
<th>Observations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>2.3</td>
<td>13.0</td>
<td>07.2000</td>
<td>12 17 18</td>
<td>40 20</td>
<td>21</td>
<td></td>
<td>15.7</td>
<td>RV-; PS-; PI-</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>0.5</td>
<td>8.7</td>
<td>04.2002</td>
<td>4 8 7</td>
<td>70 15</td>
<td>1.7</td>
<td></td>
<td>14.5</td>
<td>RV-; PS-; PI+</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>1.1</td>
<td>12.0</td>
<td>04.2004</td>
<td>13 18 16</td>
<td>51 17</td>
<td>15</td>
<td></td>
<td>10.9</td>
<td>RV-; PS 16; PI+</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>1.5</td>
<td>11.0</td>
<td>04.2007</td>
<td>13 (-) 14</td>
<td>80 18</td>
<td>8</td>
<td>Doubling</td>
<td>9.5</td>
<td>RV-; PS-; PI+</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>2.2</td>
<td>16.7</td>
<td>12.2007</td>
<td>8 (-) 9</td>
<td>50 21</td>
<td>6</td>
<td>*</td>
<td>8.8</td>
<td>RV-; PS 26; PI++</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>2.5</td>
<td>12.0</td>
<td>02.2008</td>
<td>12 16 14</td>
<td>41 23</td>
<td>13</td>
<td></td>
<td>7.5</td>
<td>RV-; PS-; PI+</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>2.0</td>
<td>11.5</td>
<td>07.2008</td>
<td>5 (-) 5</td>
<td>35 18</td>
<td>20</td>
<td>**</td>
<td>8</td>
<td>HRV; PS 25; PI +/++ , rest ASD</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>1.6</td>
<td>11.0</td>
<td>06.2009</td>
<td>13 16 14</td>
<td>81 31</td>
<td>3.5</td>
<td>Doubling</td>
<td>7.4</td>
<td>RV-; PS-; PI+</td>
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<tr>
<td></td>
<td></td>
<td>1.7</td>
<td>12</td>
<td></td>
<td>56 24.4</td>
<td></td>
<td>11</td>
<td></td>
<td>10.3</td>
<td></td>
</tr>
</tbody>
</table>

* M - male, F - female, y - years of age, PS gradient - transpulmonic gradient in direct measurement during catheterisation, RV - right ventricular enlargement, PS - pulmonary valvular stenosis (max gradient in echocardiography - mmHg), PI - pulmonary insufficiency, ASD - diameter of atrial
septal defect, TEE – transoesophageal echocardiography, Stretch - diameter of ASD measured by balloon “stop flow method”, ASO - applied Amplatzer Atrial Septal Occluder, Fluoro - time of fluoroscopy during catheterisation, Double - double ASD closely localized, rest ASD - residual shunt through atrial septal defect, *Pulmonary atresia perforated with the stiff end of guidewire in the neonatal period with subsequent PBV, **After neonatal PBV and left Blalock-Taussig surgical procedure with borderline hypoplastic RV (HRV), during actual procedures bidirectional shunting through the ASD (sat O 86%). Additionally, ASD closure and PBV balloon angioplasty of stenosed left pulmonary artery branch.