A 47-year-old patient visited our outpatient clinic. She was born with tricuspid atresia with normally connected great arteries without pulmonary stenosis (type Ic); a ventricular septal defect connected the pulmonary artery and hypoplastic right ventricle (RV) with the left ventricle.

When she was 14 years old, a Fontan-Bjork procedure was performed. The atrial and the ventricular septal defects were closed and a valved conduit (Hancock prosthesis) was placed between the right atrium (RA) and the hypoplastic RV, thus providing pulsatile flow to the pulmonary arteries. Ten years later, the conduit was severely obstructed and replaced by a 23-mm aorta homograft. At the age of 38 years, she had atrioventricular block for which a dual-chamber pacemaker system was implanted with placement of the ventricular lead through the homograft in the RV apex.

In the last 2 years, she had progressive heart failure and diarrhea. She was clinically diagnosed with protein-losing enteropathy (PLE), based on hypalbuminemia, edema, and diarrhea. Other causes of protein losses or decreased protein production were excluded. Within a few months, serum albumin levels decreased from 29 to 19 g/L. She was hospitalized, and despite increasing doses of diuretics and dietary protein adjustments, her clinical condition deteriorated (New York Heart Association functional class III/IV) with decrease of renal function.

Echocardiography and angiography demonstrated significant regurgitation of the RA-RV homograft and severe dilation of the RA. The originally hypoplastic RV had a remarkably good size and systolic function. The homograft appeared to be heavily calcified and attached to the dorsal side of the sternum. The risk of surgical conversion to a total cavopulmonary connection was considered too high because of her poor clinical condition.

It was decided to restore “tricuspid” valve function by placement of a Melody transcatheter valve (Medtronic, Minneapolis, MN) in the RA-RV homograft, as recently described by Eicken et al. Coronary angiography was performed during sizing of the conduit with a 22-mm balloon to exclude the potential risk of right coronary artery compression. The Melody valved stent, mounted on a 22-mm Ensemble delivery system (Medtronic), was positioned in the RA-RV homograft.
homograft over an Amplatzer superstiff wire. The Melody valve was successfully deployed in the most proximal part of the homograft, herewith securing the ventricular pacemaker lead between the valved stent and the homograft. Further dilation was performed with a 22-mm high-pressure Mullins balloon (Numed, Hopkinton, NY). Despite heavy calcification, the valved stent showed good circular deployment, with expansion to the dorsal side as demonstrated by “down-the-barrel” fluoroscopy. RV angiography showed a competent Melody valve in stable position and good systolic function of the RV (Figure 1). After valve implantation, RA pressure slightly decreased and pulmonary artery pressure (34/15 mm Hg) indicated good pulsatile flow (Table). The procedure was uneventful and the pacemaker lead function remained unchanged. The patient slowly recovered, and 4 weeks after valve placement she was discharged from the hospital. On follow-up, she showed no signs of fluid retention, New York Heart Association functional class improved to class II, and her exercise tolerance improved dramatically. Her renal function normalized and her serum albumin level gradually increased to 34 g/L, and PLE did not recur during 14-month follow-up (Figure 2).

Discussion

The prevalence of PLE after Fontan surgery is estimated 1% to 11%. PLE probably is caused by a combination of increased systemic venous pressure and a low cardiac output. PLE is a life-threatening complication of a failing Fontan circulation, with a 5-year survival rate of only 46% to 59%. Surgical or interventional solutions for treatment of PLE will involve conversion from the classical atriopulmonary connection to the modern cavopulmonary connection, fenestration of the Fontan baffle or atrial septum, and ultimately cardiac transplantation.

In the present case, PLE developed late after the so-called Fontan-Bjork operation. Because of severe degeneration of the RA-RV homograft, significant regurgitation led to congestive heart failure and PLE. This is the first report that demonstrates that transcatheter “tricuspid valve” implantation can improve hemodynamics and resolve PLE in the case of RA-RV regurgitation after Fontan-Bjork operation.

Disclosures

None.

References


Table. Invasive Pressure Recordings Under General Anesthesia Before and After Placement of the Melody Valve in Tricuspid Position

<table>
<thead>
<tr>
<th></th>
<th>RA</th>
<th>RV</th>
<th>PA</th>
<th>Ao</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before</td>
<td>15*</td>
<td>24/13</td>
<td>23/11/15*</td>
<td>84/55/64*</td>
</tr>
<tr>
<td>After</td>
<td>13*</td>
<td>32/12</td>
<td>34/15/22*</td>
<td>114/48/70*</td>
</tr>
</tbody>
</table>

PA indicates pulmonary artery; Ao, aorta.

*Systolic/diastolic and mean (all values in mm Hg).

Figure 2. Concentration of albumin (g/L) over time. From the moment of implantation of the valved stent (arrow), albumin levels normalize, indicating diminishing PLE.

Key Words: heart defects, congenital | Fontan procedure | regurgitation | valves | catheterization | balloon
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SUPPLEMENTAL MATERIAL

**Movie 1.** Angiogram before implantation. Note the regurgitation of the RA-RV conduit and the blood flow via the hypoplastic right ventricle.

**Movie 2.** Angiogram after implantation of the Melody valved stent providing competent tricuspid valve function.

**Movie 3.** “Down-the-barrel”-fluoroscopy showing good circular stent deployment

**Movie 4.** Adjusted parasternal short axis echocardiographic view of regurgitant homograft in tricuspid position