Creation of a secondary fenestration in the inferior vena cava stump with Occlutech Atrial Flow Regulator implantation in a Fontan patient

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The final step of palliation for complex heart defects of single ventricle physiology is the Fontan circulation. Contemporary surgical techniques utilize artificial material (Gore-Tex®) to create a tunnel and direct the flow from the inferior vena cava to pulmonary arteries, establishing the total cavo-pulmonary connection. In some patients, the surgeon creates a connection between the tunnel and atrium — primary fenestration — to improve outcomes [1].

Chronically increased systemic venous pressure, together with decreased cardiac output and venous congestion, result in the ultimate failure of Fontan circulation with clinical symptoms of Fontan-associated liver disease, plastic bronchitis, and protein-losing enteropathy [2]. Treatment of Fontan circulation failure is complex. The main goal is to address all potential sources of flow disturbances – conduit or vessel narrowing, heart dysfunction, and arrhythmias. If none is present, creating the connection between the pulmonary circulation and atrium (secondary fenestration) may alleviate symptoms. There are different potential routes for communication conduit to the atrium: from the thoracic inferior vena cava stump to the atrium or one of the pulmonary arteries to the atrium. To maintain the patency of created communication stents, self-made fenestrated device or special implants are used [3–5].

A 15-year-old boy with hypoplastic left heart syndrome, after the Norwood, bidirectional Glenn and Fontan operation — extracardiac conduit with interventionally closed primary fenestration was diagnosed with protein-losing enteropathy. After evaluation, we found no treatable cause, and we scheduled the patient for interventional creation of secondary fenestration.

Under general anesthesia, we punctured the right femoral and right jugular vein. Hemodynamic measurements revealed elevated Fontan circulation pressures (mean 20–22 mm Hg) with wedge pulmonary artery pressure of 11–12 mm Hg. Systemic saturation was 92%–94%. Based on the pre-procedure imaging (Figure 1A), we decided to puncture the inferior vena cava stump in proximity to the right atrium wall (Figure 1B). We introduced an 8 Fr Swartz SL0 sheath (Abbott, Chicago, IL, US) to the inferior vena cava and positioned it against the right atrium wall under transesophageal echocardiography guidance (Figure 1C). The sharp end of the 0.018" wire easily perforated the walls of the inferior vena cava and right atrium. The guidewire was exchanged through a Progreat 2.7 F catheter (Terumo, Japan) for a 0.014 coronary wire and crossed through the tricuspid valve to the right ventricle and aorta for a stable position. Serial dilatations with 4 × 20 mm, 8 × 20 mm, 12 × 20 mm, and 14 × 20 mm high-pressure balloons (Figure 1D) were performed to ensure adequate communication. In the next step, we introduced a 10 F delivery sheath over the 0.035" wire to the right atrium and implanted the Atrial Flow Regulator 65AFR006 (Occlutech, Sweden) into the fenestration (Figure 1E–F). After the procedure, the mean Fontan circulation pressure was 18 mm Hg, right atrium pressure was 13 mm Hg, and systemic saturation was 86%. During follow-up, the fenestration remained patent, and we observed a clinical improvement (increase in serum albumin and protein).

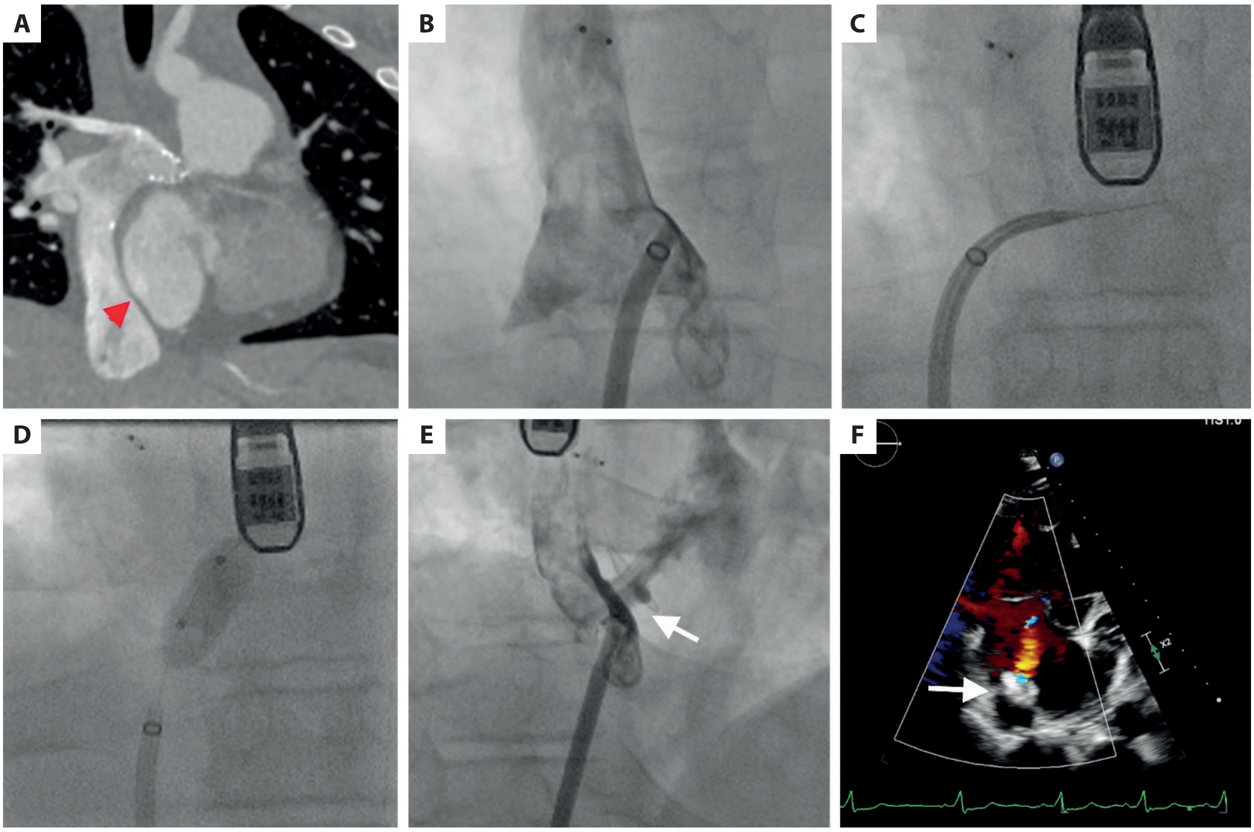

Figure 1. Secondary fenestration in the inferior vena cava stump and Atrial Flow Regulator implantation in a Fontan patient. **A.** Computed tomography — multi-plane reconstruction image. Potential place for the secondary fenestration marked with red arrow. **B.** The 8 Fr Swartz SL0 sheath placed at the place considered for the secondary fenestration in the angiography image. **C.** Puncture of the right atrium and inferior vena cava walls with the wire. **D.** Ballon dilatation of the secondary fenestration. **E.** Atrial Flow Regulator (white arrow) implanted in the secondary fenestration (angiography image). **F.** Atrial Flow Regulator (white arrow) implanted in the secondary fenestration (echocardiography image)

This is the first description of secondary fenestration in the inferior vena cava stump with Atrial Flow Regulator implantation. Our approach provided easily created, efficient, and safe palliation. Native tissue with specifically designed implants should support long-term patency of created communication. The Atrial Flow Regulator implanted in the inferior vena cava stump does not disrupt the flow through the extracardiac conduit.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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