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Short title: Creation of the secondary fenestration 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 a surgeon creates a connection between the pulmonary circulation and atrium — primary fenestration — to improve outcome [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-loosing enteropathy [2]. Treatment of Fontan circulation failure is complex. The main goal is to address all potential sources of flow disturbances – conduits or vessels 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 places for communication: the conduit to the atrium, the thoracic inferior vena cava stump to
the atrium, or one of the pulmonary artery to the atrium. To maintain the patency of created communication stents, self-fenestrated devices 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 the 8Fr Swartz SL0 sheath (Abbott, Chicago, IL, US) to the inferior vena cava and positioned against the right atrium wall under the transesophageal echocardiography guide (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 the Progreat 2.7 F catheter (Terumo, Japan) to the 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 the 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 clinical improvement (increase in serum albumin and protein).

This is the first description of secondary fenestration in the inferior vena cava stump with the Atrial Flow Regulator implantation. Our approach provided easily created, efficient, and safe palliation. Native tissue with specifically designed implant should support long-term patency of created communication. Atrial Flow Regulator implanted in 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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Figure 1. Secondary fenestration in the inferior vena cava stump and the 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 potential place for the secondary fenestration in angiography image. C. Puncture of the right atrium and inferior vena cava walls with 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)