Haemodynamic impact of pulmonary vasodilators on Fontan circulation

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Introduction
The Fontan circulation is the current palliation for patients with a single ventricle physiology. The absence of a sub-pulmonary ventricle makes low pulmonary vascular resistance and optimal systemic ventricular function the essential elements of a successful Fontan circulation. The aim of this retrospective study was to investigate the potential effect of pulmonary vasodilators on pulmonary vasculature in Fontan patients.

Methods
Eighteen single ventricle patients in therapy with pulmonary vasodilators from 6 months or more were enrolled. Nine of these patients were in therapy after Glenn procedure or just after the Fontan completion (Group A) and nine more than 5 years after Fontan completion (Group B). Ten patients without pulmonary vasodilators who underwent two catheterization at a maximal distance of two years (Group C).

Data from two different right catheterizations (before starting therapy and 6 to 24 months after) were collected: mean pulmonary pressure (mPAP), arteriolar pulmonary vascular resistances (PVR), pulmonary capillary wedge pressure (PCW), transpulmonary pressure gradient (dP), Cardiac output (CO), pulmonary blood flow (PBF), right pulmonary artery (RPA) and left pulmonary artery (LPA) diameter, Nakata index and pulmonary arterial compliance (PAC).

Results
After starting therapy, PBF (p=0.04), PVR (p<0.001), RPA and LPA diameters (p<0.001) and Nakata index (p=0.001) improved after starting therapy. These results were confirmed analyzing the subgroups A and B independently.

At baseline Group A had a smaller Nakata Index (p=0.047) and a tendency to higher mPAP and higher PVR than Group C. At the second catheterization the PCW decreased only in Group C (p=0.039), dP (p=0.025) and PVR (p=0.009) decreased in group A. Nakata Index (p=0.003) and PAC (p=0.001) increased in group A.

Conclusions
Pulmonary vasodilators reduce PVR and increase PAC, Nakata index and CO. Pulmonary vasodilators could be used before Fontan completion in selected patients as “bridge to Fontainability”. In adults the best effect was seen in patients with higher dP and lower PAC. In patients with low dP and high PCW the treatment increased cardiac output but also wedge pressure raised. For these reasons, given also the costs of the therapy, we suggest to perform the heart catheterization before starting this therapy.