Pulmonary perfusion assessment in HLHS patients after Glenn operation and left pulmonary artery stenosis treated with stent implantation - preliminary study

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Introduction: Left pulmonary artery (LPA) stenosis is a common complication in patients with hypoplastic left heart syndrome (HLHS) after Norwood operation. LPA stenting is a well-established treatment in those patients.

Aim: Assessment of pulmonary perfusion in patients with HLHS after bidirectional Glenn (BDG) operation and pulmonary artery stenting using planar lung perfusion scintigraphy.

Material and Methods: Planar pulmonary perfusion scintigraphy was performed in ten patients (6 boys and 4 girls) at the median age of 56 months (21-104 months), after BDG operation. Mean Nakata index was 239 (118-341). Eight patients underwent stent implantation of which 7 had LPA stenting and the remaining one had RPA treated. Mean time from the stent implantation to scintigraphy was 19 months (3 days - 7 years).

Results: Pulmonary perfusion asymmetry was noted in 9/10 (90%) patients. Hypoperfusion of the left lung was noted in 7 cases. Contribution of the left lung to the global lung perfusion ranged from 11 to 59% (mean 28%). Hypoperfusion of the left lung was diagnosed in two patients (age 21 months and 30 months) with no previous pulmonary artery interventions. After LPA stenting both patients presented with improved left lung perfusion; from 18 to 37% and from 28 to 34%, respectively. In one patient (8 years old) after previous LPA stent implantation, hypoperfusion of the left lung (25%) was noticed, which was related to restenosis due to stent fracture. Despite successful stent implantation with significant improvement in the diameter of the obstruction from 6.25mm to 12.5 mm, there was no improvement in the follow-up scintigraphy performed 3 days after the intervention.

Conclusions:
1. Patients with HLHS present with pulmonary perfusion abnormalities due to non-physiological pulmonary flow and pulmonary artery obstructions.
2. Hypoperfusion of the left lung is the most common finding even after successful LPA stenting.
3. Pulmonary artery stent implantation in young patients shortly after BDG operation brings the biggest improvement in pulmonary perfusion.
4. Planar lung perfusion scintigraphy may be a useful method for outcomes assessment of percutaneous interventions for pulmonary stenosis in HLHS.