Pulmonary perfusion in patients with HLHS after Fontan operation and pulmonary artery stenting.

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Introduction: Left pulmonary artery (LPA) stenosis is a common complication during multistage palliation of patients with hypoplastic left heart syndrome (HLHS). Fontan operation results in diminished pulsatility of pulmonary blood flow which affects blood distribution in the lungs.

Aim: Lung perfusion assessment in patients with HLHS after Fontan operation using planar lung perfusion scintigraphy.

Material and Methods: 73 patients (52 boys) at a median age of 7,5 years (3,3-19,5 years) took part in the study. 57 patients (78%) had LPA stenting. All patients underwent planar lung perfusion scintigraphy. Two injections of 99mTc – MAA into the right upper and then into the right lower extremity were performed. Webster's formula was used to calculate the activity and Hawkeye hybrid gamma camera to perform lung perfusion studies, processed on Xeleris workstation. Percentage share of every lobe and lung in a global lung perfusion was calculated after averaging of counts in anterior and posterior projection for inflow from superior and then from inferior cavopulmonary anastomosis.

Results: Mean left and right lung contribution to global lung perfusion was: 38% (upper lobe 15%; lower lobe 23%) and 62% (upper lobe 8%; middle lobe 29%, lower lobe 25%) respectively. Severe left lung hypoperfusion, defined as percentage ≤ 35% of global lung perfusion, was noted in 30 patients (41%). In two patients without prior LPA stenting we found severe left lung hypoperfusion – 12% and 9% respectively. Both patients were asymptomatic. We diagnosed severe LPA hypoplasia, first patient had also left diaphragmatic paralysis. After successful LPA stenting, left lung perfusion increased up to 24% and 30% respectively. Postoperative diaphragmatic paralysis causing decrease of one of the lungs was noted in 8 patients (11%). Radionuclide activity in kidneys and brain suggested pulmonary arterio-venous fistulas (PAVF) in 5 patients (7%).

Conclusions:
1. Patients with HLHS after Fontan operation have pulmonary perfusion abnormalities due to: non-physiological pulmonary blood flow, pulmonary artery obstruction and postoperative diaphragmatic paralysis.
2. Planar lung perfusion scintigraphy is an useful method in postoperative evaluation of patients after Fontan completion.
3. Hypoperfusion of the left lung is the most common finding in spite of successful LPA stenting.