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Original

Lung perfusion scintigraphy in the assessment of pulmonary circulation after completion of surgical treatment of a hypoplastic left heart syndrome (HLHS)

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Abstract

INTRODUCTION: Hypoplastic left heart syndrome (HLHS) is an inborn complex heart malformation. A multi-stage treatment is initiated in a neonatal period with a Norwood surgery. The next step is Glenn surgery — a bidirectional superior cavo-pulmonary anastomosis. At the last stage anastomosis of inferior vena cava (IVC) with the right pulmonary artery (RPA) is formed as a result of a Fontan surgery. The aim of this study was to assess lung perfusion in patients with HLHS after completion of a surgical therapy, using a scintigraphic method.

MATERIAL AND METHODS: In 92 patients with HLHS a planar lung scintigraphy in anterior and posterior projections after administration of 99mTc-macroaggregates in activity 18-111MBq was carried out twice (in several day intervals). At first, a radiopharmaceutical was administered to the right extremity in order to assess the lung distribution of blood flowing through the anastomosis of superior vena cava (SVC) with RPA. In the next study, after administration of the tracer to the right lower extremity, the distribution of blood flowing through the anastomosis of IVC with RPA was assessed. The relative percentage of each lung in the total lung perfusion was calculated on a Xeleris workstation using the "Lung perfusion analysis" program. Lung perfusion was considered close to symmetrical when the proportion was in the range of 40–60%.

RESULTS: In spite of the fact that mean relative values of distribution of blood flowing through the anastomosis of SVC with RPA to the left lung (LL) and right lung (RL) in the entire study group did not differ significantly: LL_{mean} = 47%; RL_{mean} = 53%, p = 0.14, relative values of perfusion of both lungs were differentiated — in 26% of patients LL was better perfused, in 38% RL was better perfused and in 36% a perfusion of both lungs was similar. The analysis of blood distribution by anastomosis of IVC with RPA showed that the mean relative perfusion of RL was significantly higher than that of LL (70% vs. 30%, $p < 0.0000001$). No signs of pulmonary emboli were detected. In 13% of studied patients, uptake of the radiopharmaceutical in kidneys was shown indicating the presence of shunt "from right to left".

CONCLUSIONS: After completion of surgical treatment of patients with HLHS, differentiated blood supply of the lungs was observed through SVC with RPA anastomosis and a tendency to higher blood supply of RL than LL by anastomosis of IVC with RPA. No signs of pulmonary embolism were detected. The study revealed a "right to left" shunt in some patients.

KEY words: HLHS, Fontan surgery, lung perfusion scintigraphy

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Introduction

Hypoplastic left heart syndrome (HLHS) is the fourth most frequent congenital heart defect. HLHS is a complex malformation consisting of atresia or hypoplasia of left heart structures: mitral and aortic valve, left ventricle, ascending aorta and aortic arch. Palliative multistage operation is a widely approved treatment of this syndrome. Reconstruction of a fully normal anatomy is an inaccessible option in this anomaly. Surgical treatment is a three-stage procedure initiated in a neonatal period with a Norwood surgery which involves: atrial septectomy, reconstruction of ascending aorta and aortic arch through their anastomosis with a pulmonary trunk. Blood inflow to the lungs is provided by a systemic-pulmonary shunt (Blalock-Taussig shunt) — connection between right subclavian artery and right pulmonary artery in a classical Norwood procedure or right ventricle to pulmonary artery conduit in Sano modification. A second stage is a Glenn operation — a bidirectional anastomosis of superior vena cava with right pulmonary artery. In the third stage a total cavo-pulmonary anastomosis of full Fontan circulation is formed. The Fontan operation, in which a systemic arterial and pulmonary blood flow is completely separated, is the final stage of the palliative surgical therapy [1–3].

Lung perfusion scintigraphy is a valuable diagnostic tool in the evaluation of pulmonary circulation. It is increasingly used in the diagnosis of pulmonary circulation disorders accompanying congenital heart diseases and in the assessment of effectiveness of the surgical correction of these defects [4–8]. Due to the fact that one of the important goals of the three-stage surgical treatment of HLHS is the separation of systemic and pulmonary circulation, scintigraphy has also been used to assess the impact of surgery on both the distribution of blood to both lungs and the presentation of perfusion disorders in the lungs. So far, there are relatively few reports on the use of perfusion lung scintigraphy in patients with HLHS [9–14].

In this study scintigraphy was used to assess pulmonary circulation in patients with HLHS after completing multistage surgical treatment.

Material and methods

The study involved 92 patients (63 boys and 29 girls) aged 3–20 years (mean 9 years). Scintigraphy was performed between 6 months and 13 years (mean 3.4 years) after the Fontan surgery. In all patients, planar lung perfusion scintigraphy was performed using a dual-head scintillation camera in the anterior and posterior projections after intravenous injection of 99mTc-macroaggregates with activity from 18 to 111 MBq (depending on the age and body mass of a patient).

In each patient, the study was carried out twice (in several day intervals). In the first study, a radiopharmaceutical was administered to one of the veins of the right elbow flexure in order to assess the lung distribution of blood flowing through the anastomosis of superior vena cava (SVC) with right pulmonary artery (RPA). In the next study, after administration of the tracer to the right dorsal foot vein, the distribution of blood flowing through the anastomosis of inferior vena cava (IVC) with right pulmonary artery (RPA) was assessed.

The relative percentage of each lung in the total lung perfusion was calculated on a Xeleris workstation using the "Lung perfusion analysis" program, based on the geometric mean number of counts from both study projections. Lung perfusion was considered similar (close to symmetrical) when the proportion was in the range of 40–60% (Fig. 1).

Results

Mean relative values of a distribution of blood flowing through the anastomosis of SVC and RPA to the left lung (LL) and right lung (RL) in the whole patient group (n = 92) did not differ statistically significantly and amounted to: $LL_{mean} = 47\%$; $RL_{mean} = 53\%$, $p = 0.14$ (Tab. 1). However, it is noteworthy that only in 33 patients (36%) the perfusion of both lungs was similar (within 40–60%), and in 59 (64%) it was asymmetric: in 35 (38%) RL perfusion was better than LL, whereas in 24 (26%) better perfusion was observed in the LL (Tab. 2a.). The analysis of blood distribution by anastomosis of IVC with RPA showed that the mean relative RL perfusion was statistically significantly higher than that of LL (70% vs. 30%); $p < 0.0000001$ (Tab. 1). In 75 patients (82%), a relatively higher perfusion of RL was observed, and only in 17 (18%) of the blood distribution in both lungs was similar (Tab. 2b). Distribution of the radiopharmaceutical in each lung was uniform, no focal perfusion defects were observed that could correspond to pulmonary emboli.

In addition, 12 patients (13%) also showed uptake of the radiopharmaceutical in kidneys, indicating the presence of shunt "from right to left" (Fig. 2).

Discussion

In this study lung perfusion scintigraphy was performed at various time points after completion of surgical treatment, ranging from 6 months to 13 years (mean 3.4 ± 3.1 years), in the majority of patients (75%) the time after Fontan surgery did not exceed 5 years. Yin Z et al. examined patient group with HLHS with lung perfusion scintigraphy shortly after the Fontan surgery and 5 years later and showed that the distribution of blood to the lungs by both anastomoses (SVC with RPA and IVC with RPA) did not change with time [9]. It should be considered that the distribution of blood to the lungs remains the same for a long time after the surgical correction of the defect. Authors of the work applied a slightly different methodology of scintigraphy than assumed in this work. The radiopharmaceutical was administered first in the right upper extremity and just after completion of the study acquisition another portion of activity (the same as previously) was injected into the right lower extremity to obtain an image summing the uptake of the radiotracer in the lungs after administration from the "top" and from the "bottom". Blood distribution through the IVC-RPA anastomosis was calculated by subtraction of counts obtained after administration of the radiopharmaceutical to the upper extremity from the total number of counts, for each lung separately. The advantage of the method described above is limiting the study to a one-day procedure. However, analysis of results obtained with this method is more complicated and in our opinion may generate additional errors.

The adopted criteria of asymmetric lung perfusion (< 40% or > 60%) are similar to those used by Tamir et al. [4]. In our work, the distribution of blood to the lungs is presented as a percentage of the individual lung in the summed lung perfusion, although one can also use the ratio of perfusion of one lung to another [9].

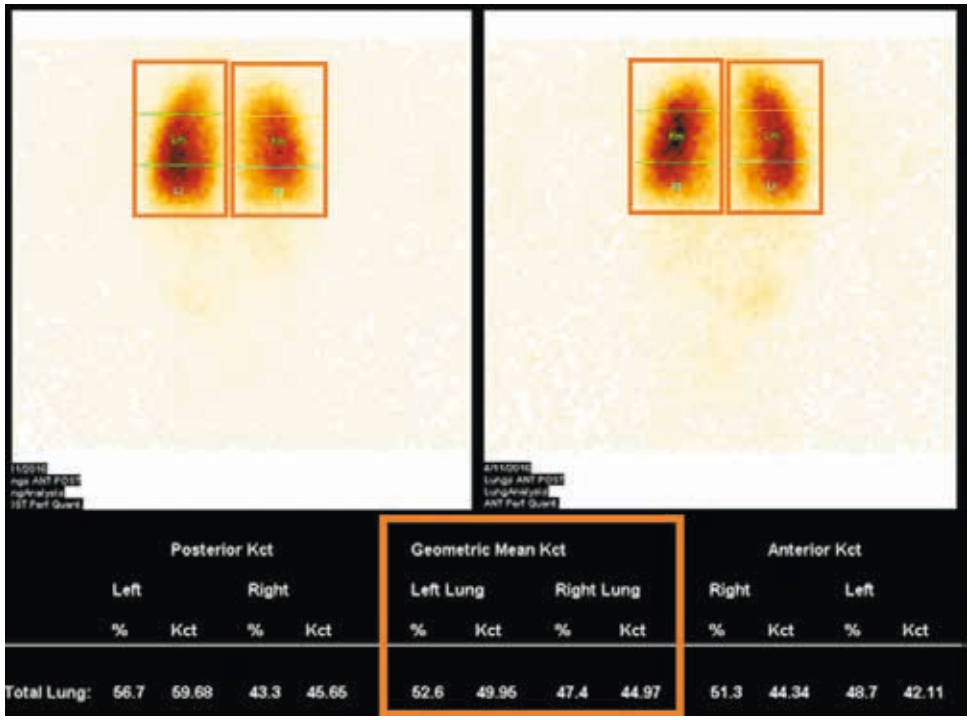

Figure 1. Example of the application of "Lung perfusion analysis " program on the Xeleris workstation to calculate the percentage of each lung (thicker frame) in the overall lung perfusion, based on the geometric mean number of counts (Kct) in both projections of the study (posterior and anterior). Relative lung perfusion: LL = 53%, RL = 47%

Table 1. Mean relative values of a distribution of blood flowing into the right lung and left lung by the SVC-RPA and IVC-RPA anastomoses

N = 92 patients		Mean relative percentage	SD standard deviation	P
SVC-RPA anastomosis	RL	53	19	0.14
	LL	47	19	
IVC-RPA anastomosis	RL	70	11	< 0.0000001
	LL	30	11	

RL — right lung; LL — left lung; SVC — superior vena cava, IVC — inferior vena cava, RPA — right pulmonary artery

Table 2. Differentiation of blood distribution to lungs: **a)** through SVC-RPA anastomosis; **b)** through IVC-RPA anastomosis

a) Blood distribution through SVC-RPA anastomosis	Number of patients	Percentage of patients [%]
Uniform 40–60%	33	36
Higher to LL	24	26
Higher to RL	35	38
b) Blood distribution through IVC-RPA anastomosis	**Number of patients**	**Percentage of patients [%]**
Uniform 40–60%	17	18
Higher to LL	0	0
Higher to RL	75	82

Authors of several papers point out that, after the completion of HLHS surgery, blood distribution through the SVC-RPA anastomosis to the LL is generally higher than to the RL, whereas by the IVC-RPA anastomosis the distribution of blood to the lungs takes place with the right side preference. [4–6, 8, 9] Our results indicate differentiated distribution of blood through the SVC-RPA anastomosis (only in about 1/3 of patients higher perfusion was related to LL, about 1/3 to RL, and in the remaining 1/3 blood supply to both lungs was similar). We confirmed, however, a definite predominance of blood distribution to RL by IVC-RPA anastomosis (in 82% of patients, with similar perfusion of both lungs in the remaining 18%).

In patients with HLHS treated surgically, attention is paid to the risk of embolic complications, including pulmonary embolism (sometimes asymptomatic), especially in the early postoperative period (up to about 1 year) [15–17]. In light of this, it seems surprising that in our clinical material no lung perfusion defects were found that could correspond to embolic changes. This may be related to the fact that the vast majority of children were examined later than one year after the completion of surgical treatment, and scintigraphic changes after earlier, especially less extensive emboli, show a tendency to disappear.

In patients after surgery, the inflow of blood into the lungs occurs directly from the systemic veins; in addition, the lack of ventricular ejection of blood to the pulmonary circulation is followed by the absence of a typical, pulsatile blood flow in the lungs. This induces such abnormalities as arteriovenous malformations causing blood shunt from right to left. The intensity of such shunts can be assessed by comparing the number of counts in the lungs with counts throughout the body [5, 6, 18, 19]. In this study such a shunt was detected in 13% of patients, based only on the uptake of the tracer in kidneys, which indicated that its intensity was not lower than 10% [20].

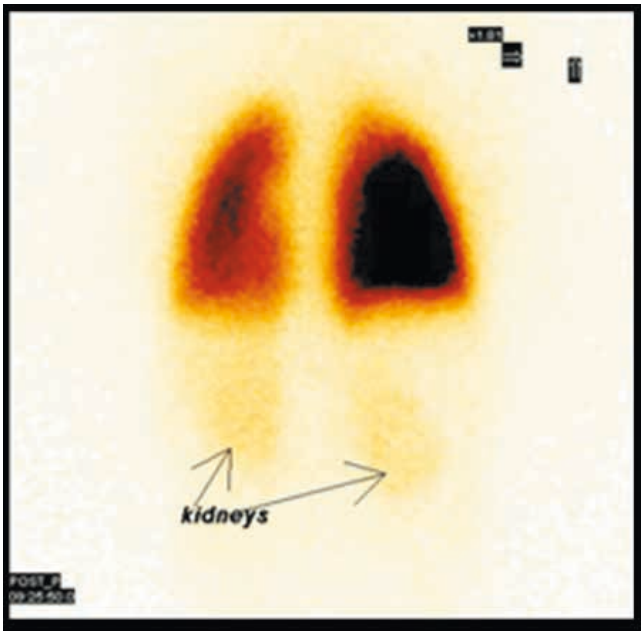

Figure 2. Planar perfusion lung scintigraphy in posterior projection. The presence of 99mTc-macroalbumin uptake in the kidneys indicates at least a 10% shunt from right to left

Conclusions

In patients after completed surgical treatment of HLHS, a differentiated distribution of blood to the lungs through the SVC-RPA anastomosis and a tendency to higher perfusion of RL than LL by IVC-RPA anastomosis were revealed with lung perfusion scintigraphy.

No signs of pulmonary emboli were detected in the lungs, while the features of shunt from right to left were demonstrated in over a dozen of patients.

Planar perfusion scintigraphy, showing lung perfusion after a separate administration of a radiopharmaceutical to the upper and lower extremity, is a useful diagnostic tool in the assessment of pulmonary blood supply in patients after the completion of surgical treatment.

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