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Original

Lung perfusion scintigraphy in the diagnosis of peripheral pulmonary stenosis in patients after repair of Fallot tetralogy

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Abstract

BACKGROUND: The frequency of peripheral pulmonary artery stenosis in patients after surgical repair of tetralogy of Fallot (TOF) ranges from 20 to 40%. This can be either primary or secondary to the surgical intervention. The influence of resulting lung perfusion alterations on the life quality of patients is difficult to predict. The aim of this study was to compare the utility of the diagnostic procedures in this group of patients, with particular focus on lung perfusion scintigraphy.

MATERIAL AND METHODS: This study comprised 104 patients who underwent repair of TOF at ages from 5 months to 25 years. The patients have been followed up for from 4.2 to 25 years. On the basis of chest x-ray peripheral pulmonary artery stenosis was suspected in 11 patients, in 12 on the basis of echocardiography examination.

RESULTS: Lung perfusion scintigraphy has been performed on 87 patients. The disturbances in lung perfusion (mostly in the left lung) were show by means of lung perfusion scintigraphy in 43 (49%) of patients. In 27 of them heart catheterisation has been performed. Angiography revealed stenosis of the lung artery branch in 15/43 (34.9%) patients with abnormal perfusion lung scan and in 4/44 (9%) in patients with normal perfusion lung scan. Intervention procedures were carried out on 10 patients.

CONCLUSIONS: Lung perfusion scintigraphy may prove a valuable, non-invasive screening tool in the assessment of patients after TOF repair, although both false-negative and false-positive results may happen. Therefore, it should play an auxiliary role together with other diagnostic modalities.

Key words: peripheral pulmonary stenosis, perfusion scintigraphy, congenital heart disease, tetralogy of Fallot

Introduction

Peripheral pulmonary arterial stenosis is quite a common problem in patients with tetralogy of Fallot (TOF). The frequency is about 20–40% of all patients after surgical repair of TOF. The stenosis may be primarily due to congenital abnormalities in the structure and development of pulmonary vessels, or secondarily to palliative surgical procedures (systemic to pulmonary shunts) or total correction — reconstruction of right ventricle outflow tract.

It is difficult to estimate the influence of pulmonary artery stenosis on the haemodynamics and development of the lung circulatory system. This abnormality increases right ventricle pressure and pressure overload. The decreased blood flow in the pulmonary artery or one of its branches may affect the development of pulmonary bed in children.

The assessment of pulmonary stenosis is based on physical examination and the analysis of preoperative examinations, surgery protocol, x-ray, echocardiographic examination, pulmonary scintigraphy and angiographic examination. Lung perfusion scintigraphy and angiocardiography are the most valuable tools in the diagnosis of pulmonary artery stenosis. Pulmonary scintigraphy is a simple, non-invasive and inexpensive procedure, which gives the most precise information of the assessment of the severity of peripheral pulmonary stenosis. On the other hand its usefulness is somewhat limited, and its interpretation requires a comparison with other diagnostic procedures.

The aim of this study was to assess the value of pulmonary scintigraphy in the diagnosis of peripheral pulmonary stenosis in patients after the correction of TOF.

Material and methods

We studied 104 patients after total surgical repair of TOF. The mean age at the time of our study was 15 years (range 5–40). The time interval between our study and the surgery was mean 11.1 years (range 4.2–15 years). In 28 patients the surgical procedure was preceded by the systemic-pulmonary shunt — in 18 case right- and in 11 left-sided. In one case it was a central shunt, and in 2 patients two shunts were required. In 87 cases (83% of all patients) lung perfusion scintigraphy was performed 20 minutes following the intravenous infusion of albumin macroaggregates (CIS, Gif-sur-Yvette, France). Scanning was performed using a double-headed gamma camera MULTISPECT 2 (Siemens, Erlangen, Germany) in anterior, posterior, lateral and posterior-oblique projections. The semiquantitative data analysis was performed by ICON — Siemens Data Analysis Program. The percentage distribution of radioactive marker between both lungs and also between the lower, middle and upper parts of lungs was assessed. The normal right lung perfusion was assumed as 50–60% of total lung perfusion, and 40–50% for the left lung. Our cohort was divided into two groups according to lung perfusion study results.

The first group comprised patients with impaired lung perfusion, the second had normal lung perfusion. The results of pulmonary scintigraphy were compared with other clinical diagnostic procedures: chest x-ray, echocardiography and angiography in some cases. All the data were analysed in order to find the factors responsible for peripheral pulmonary stenosis.

Results

The mean right/left lung perfusion ratio between right and left lung was 60% to 40%. The right lung perfusion was between 23––89% (mean 59.4%) compared to 11–77% (mean 40.4%) in the left lung. Impaired lung perfusion was found in 43 out of 87 patients (49%). 44 patients had normal lung perfusion (group II). 34 out of 43 patients from the first group had impaired left lung perfusion, in 9 right lung perfusion was impaired. No correlation was found between perfusion disturbances and previous surgery. Increased lung perfusions on the same side as previous systemic to pulmonary shunt were found in 9 patients. Only in 5 cases out of 34 with decreased left lung perfusion was left-sided shunt performed. The other 10 patients with left lung perfusion disturbances had right-sided shunt. In 2 patients out of 9 with right pulmonary artery stenosis right-sided shunt was performed.

The chest x-ray examination detected the disproportion of pulmonary vascularity in 11 out of 104 patients. Doppler study suggested left branch stenosis in 9 cases and right branch stenosis in 2 patients. In one patient bilateral pulmonary stenosis was suspected on echocardiographic examination. Comparison between patients with impaired lung perfusion (group I) and subjects without any abnormality (group II) is shown in Table 1. The rate of asymmetry in chest x-ray was 10 times higher in the first group. Patients from group I were also more often qualified for cardiac catheterisation, which revealed stenosis.

Table 1. Comparison of groups

	Group I	Group II
Number of patients	43	44
Systemic to pulmonary shunt	14 (32.6%)	11 (25%)
Chest X-ray asymmetry	10 (23.9%)	1 (2.3%)
Cardiac catheterisation	19	78
Peripheral pulmonary stenosis in angiography	15 (34.9%)	4 (9%)
Therapeutic intervention	10 (23.3%)	1 (2.3%)
Mean age at the time of surgery (years)	5.05	4.7
Mean age at the time of study (years)	16.4	15.3
Postoperative follow-up (years)	14.5	14.7

Catheterisation was performed on 27 patients (19 from group I, and 8 in group II). Left pulmonary artery stenosis was found in 13 patients and right pulmonary artery stenosis in 3 patients. In 2 cases both-sided pulmonary artery stenosis was diagnosed. In 10 patients cardiac intervention procedure (stent implantation or balloon-angioplasty or stent implantation) was performed, 3 patients were qualified for surgery.

Discussion

Following the wider accessibility of surgical procedures and the improvement of their results, the number of patients after correction of TOF is constantly increasing. The increased number of patients and prolonged follow-up results in the increased detection of residual or acquired haemodynamic and anatomic abnormalities [1–3]. One of those abnormalities is peripheral pulmonary stenosis, primary or secondary to surgery. The systemic to pulmonary shunt is said to be one of the pathogenic factors in secondary pulmonary stenosis [8]. We did not confirm this finding in our study. A similar number of shunts was performed in two groups: with or without lung perfusion abnormalities. The influence of pulmonary artery stenosis on exercise capacity, right ventricle and lung function or sudden cardiac death was previously analysed in many publications [4–6, 9–11]. Thanks to new therapeutic options (balloon-angioplasty and stent implantation) the problem of peripheral pulmonary stenosis is being more widely discussed [12, 13].

Diagnostic imaging following TOF repair. There are many different diagnostic procedures following TOF repair. Although chest x-ray gives information about lung vascularisation, abnormalities must be severe to get visualised. In our group of patients pulmonary artery stenosis was suspected on the basis of x-ray in 11 cases. In 8 cases diagnosis was confirmed by catheterisation and in 3 intervention was performed. Echocardiography enables the visualisation of pulmonary trunk and short arterial pulmonary branches proximal both right and left. In our cohort pulmonary artery stenosis following echocardiography was suspected in 12 cases and in 9 patients it was confirmed by angiography. The disproportion in the diameter of the pulmonary artery and increased flow in Doppler study suggest the stenosis of the branch of the pulmonary artery. Echocardiography does not give information about the severity of lung perfusion disturbances. Echocardiography is also useless in distal pulmonary stenosis localised out of echo wave range.

Pulmonary scintigraphy is a very sensitive method in the diagnosis of pulmonary blood flow disproportion [7, 14, 15]. Hypoperfusion of one lung may suggest pulmonary artery stenosis. In our study disturbances in lung perfusion scintigraphy were shown in 49% of patients. Jonsson found perfusion disturbances in 86% of all patients, although he used different, more restrictive, criteria [7]. Normal scintigraphy does not exclude pulmonary artery stenosis. False negative results of scintigraphy examination can be shown in both-sided pulmonary branches stenosis — like one patient in our series. In this situation lung perfusion can be balanced and results of lung perfusion scintigraphy are false-negative [7, 14, 15]. In contrast, peripheral, diffuse or multi-artery stenosis or any other increase of vascular resistance in a lung gives abnormal results of lung perfusion scintigraphy. Systemic to pulmonary vascular connection or lung disease can also influence lung perfusion scintigraphy results [14, 15]. Patients with disturbances in scintigraphic image require the introduction of other diagnostic procedures. A well-planned diagnostic process enhances the probability of the detection of peripheral pulmonary stenosis and can help to calculate the severity and haemodynamics of stenosis. Any patient with peripheral pulmonary stenosis requires angiographic verification. Angiographic examination visualises the location and estimates the degree of stenosis. Angiography is necessary before therapeutic qualification — surgery or intervention procedure.

Conclusions

1. Disturbances in lung perfusion are quite common in patients after repair of tetralogy of Fallot, but only patients with severe abnormalities seem to require further therapy.
2. Lung perfusion scintigraphy is a sensitive method for detection of lung perfusion disturbances and semiquantitative analysis and seems to be a reliable non-invasive tool in this group of patients, but it seems to play an important, but only an auxiliary, role together with other diagnostic modalities.
3. A disproportion in lung perfusion scintigraphy suggests peripheral pulmonary artery stenosis. Additional diagnostic procedures are required for confirmation and localisation of stenosis.

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