

PULMONARY HYPERTENSION ASSOCIATED TO CONGENITAL HEART DISEASES

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Abstract: Congenital heart diseases are among the most common congenital malformations at birth with an incidence of 8/1000 newborn. These defects are characterized by a heterogeneous group of abnormal defects and connections between the cardiac chambers and vessels with different haemodynamic consequences and hence, varying need for follow-up and interventions. The most common forms are congenital cardiac shunts (i.e.: ventricular septal defects, atrial septal defects, patent ductus arteriosus) that account for almost 60% of the malformations.

Keywords: pulmonary hypertension, congenital heart diseases

Rezumat: Cardiopatiile congenitale sunt malformațiile congenitale cele mai frecvente, cu o incidență în jur de 8 pentru 1000 născuți. 60% dintre aceste anomalii sunt caracterizate printr-un shunt stânga dreapta. Acestea din urmă asociază frecvent și hipertensiune arterială pulmonară (HTAP).

Cuvinte cheie: hipertensiune arterială pulmonară, cardiopatii congenitale

Pulmonary hypertension (PAH) is frequently encountered in malformative or acute heart diseases, but it may occur even on a healthy heart by the reversible or irreversible accentuation of the pulmonary arteriolar resistances.

Assessment of the pulmonary arterial pressure (PAP)

Physiopathology

Pulmonary pressure is assessed according to the blood flow and pulmonary resistances ($P = Q \times R$).

More than the pressure, pulmonary resistances are the key element to be known. Their assessment requires the preliminary evaluation of the pressure and of the pulmonary flow.

PHYSIOPATHOLOGIC CLASSIFICATION

GRO UP	BLOOD FLOW	PULMONARY PRESSURE	PULMONARY RESISTANCES
I	Low	Normal	Normal
II a	Important	< 2/3 P Systemic	Normal
II b	Important	> 2/3 P Systemic	Normal (or slightly increased)
III	Low	= P systemic	Increase ++
IV	Moderated	Normal (but there is gradient VD-AP)	Normal

As a result of knowing the pulmonary arterial resistances, pulmonary arterial hypertension may be classified into:

- PAH with low resistances
- PAH with increased resistances
 - Postcapillary blocking
 - Precapillary blocking
 - Reversible
 - Fixed

The increase of the pulmonary resistances to the flow of the pulmonary blood may be secondary to an obstacle situated after the pulmonary arterioles (hypocontractility of the left ventricle, mitral stenosis etc.). It is about a postcapillary obstacle or about an increase of the pulmonary arterioles resistances, increase that may be reversible or irreversible (fixed pulmonary arterial hypertension).

Pressures are expressed in mmHg and resistances in Wood/m². The difficulty in assessing a correct pulmonary flow in the moment of left to right shunt makes the pulmonary resistances be only related to the systemic resistances: (RP/RS) relation is normally inferior to 0,25.

The increase of the pulmonary pressure, irrespective of the reason, will have as a consequence the adaptation of the right ventricle. The higher pressure is the more the right ventricle will be hypertrophied and the septal curve will be flat in systole and then concave.

Above a certain value of the pressure, the right ventricle will be dilated and become hypocontractile. Stasis signs will be manifested through hepatomegaly and edema.

Auscultatoric

Pulmonary hypertension is responsible for an accentuation of the II noise and it is rather late, in case of an important PAH. Pulmonary insufficiency is perceived as a diastolic murmur.

Radiological

Thorax radiography is less representative; it shows us a convex medium arch and a variable vascularization according to the PAH cause.

EKG

EKG aspect is proportional with the pressure existing in the right ventricle, but no exact appreciation of PAH can be made.

Doppler echography

It is a non invasive exam and very viable. In practice, five signs are easily to be obtained with the help of echography, and two signs are more sensitive, but unfortunately inconstant.

Septal curvature

The aspect of the septal curvature is brought about the action of the systolic pressure in each ventricle. Normally, in systole, the septal curvature is concave towards the left ventricle cavity, becoming rectilinear in case of pressures equality, or convex if the right pressure is superior to the pressure of the left ventricle.

Muscle hypertrophy of the right ventricle

Hypertrophy is important when the intracavitary pressure is very high. This sign is less sensitive and of low interest in the neonatal period of time, because at this age, the right ventricle is hypertrophied normally.

Pulmonary flow

The aspect of the pulmonary ejection flow changes according to the pressure. The pressure is higher if flow acceleration time is increased.

In practice, the appreciation of the pulmonary flow is less sensitive, because in infants, the cardiac frequency is more rapid.

Tricuspidian insufficiency

The assessment of velocity of a possible tricuspidian insufficiency allow the accurate appreciation of the pressure gradient between the right ventricle and the right atrium, so the systolic pressure in the right ventricle may be deduced.

If there is no obstacle between the right ventricle and the pulmonary artery, accurate appreciation of the pulmonary systolic pressure can be made.

Pulmonary valvar insufficiency

If pulmonary insufficiency is important in order to give us a good "wave" in continuous Doppler, the assessment of the diastolic gradient between the right ventricle and the pulmonary artery is relevant.

Gradient between the two ventricles or between the two vessels.

If there is an arterial channel or an interventricular communication shunt velocity may be frequently measured between the two circulations and deduce the pressure gradient. Knowing the arterial pressure, the pulmonary systolic pressure may be assessed as well.

Catheterization

This manoeuvre is the only one that may measure the pressure. The pulmonary resistances may be assessed, previously knowing the pulmonary flow. Catheterization allows the localization of the secondary precapillary obstacles upon the accentuation of the pulmonary arteriolar resistances. Catheterization is the most important investigation which allows finding out whether the increase of the pulmonary resistances increase is reversible or definitively fixed.

The pharmacodynamic test is made with:

- Oxygenation or even ventilation under O₂,
- Prostacyclin perfusion (Flolan: 5-15ng/kg/min),
- Inhalation of NO (5 – 10ppm),

Spectacular results may be obtained by the occurrence of a shunt of the pulmonary resistances, or more frequently (unfortunately) a minimum shunt without significant values is emphasised.

Capillary angiography completes the examination through catheterization, showing in case of PAH, a poor arteriole bed, rigid with the disappearance of the blood flow.

Pulmonary biopsy

After a certain period of time and in certain patients, pulmonary arterial hypertension brings about histologic lesions of the pulmonary arterioles, classified by taking into account their importance level, with good predictive value for the fixed or irreversible character of the resistances.

Pulmonary arterial hypertension – consequence of a cardiopathy

Left to right shunt

In infants, PAH occurs in the presence of a ventricular or arterial left to right shunt. In the case of an important auricular shunt, PAH has a low level (large ASD or RVPAT).

This pulmonary hypertension is secondary to the shunt and to the increased pulmonary resistances, but practically reversible until the age of 6 months years old. The evolution towards a pulmonary obstructive disease is variable, from one cardiopathy to another and from one patient to another.

Fixed pulmonary hypertension is a pulmonary obstructive vascular disease (Eisenmenger's syndrome), and frequently, it is the consequence of a shunt cardiopathy and PAH left to evolve without any treatment.

The pulmonary obstructive vascular disease may evolve variably. There are forms that evolve rapidly, in few years reaching suprasystemic resistances badly

tolerated, with cyanosis and/or cardiac insufficiency. Other forms are well tolerated many years and the patients live a cvasinormal life.

Doppler echography confirms and appreciates the arterial hypertension and appreciates the ventricular or arterial tolerance.

During evolution, a terminal right cardiac insufficiency may be generally observed, with hepatomegaly, murmur of tricuspidian insufficiency and a dilated right ventricle, hypocontractile in echography. Death usually occurs within this context due to an infection or pulmonary bleeding.

Symptomatic treatment consists in the fight against polyglobuly by correcting a possible microcytosis. The gravity of the pulmonary bleeding should make us attentive to prescribing anti-inflammatory medication non steroidian and anticoagulants.

Pulmonary or cord-pulmon transplant remains the unique alternative.

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