THE LEFT TO RIGHT SHUNT CONGENITAL CARDIOPATHY
Part II

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***Abstract**: Heart diseases with left to right shunting are the most frequent congenital heart diseases. All of them have in common the increase of pulmonary blood flow. Because of the abnormal communication between the heart cavities and vessels, the passage of the oxygenated blood from the systemic circulation into the pulmonary circulation increases. The evolution of congenital heart diseases is dominated by the irreversible alteration of the pulmonary blood vessels resulting in time, in fixed pulmonary hypertension. Medical discoveries, especially echocardiography combined with Doppler examination, and surgical progressions, that made possible corrective heart surgeries in younger ages, brought about better results in pediatric cardiology.*

***Keywords**: congenital heart diseases, congenital cardiac shunts*

***Rezumat**: Cardiopatiile responsabile de un shunt stânga-dreapta sunt cel mai frecvent cardiopatiile congenitale. Ele au toate în comun creşterea debitului sanguin pulmonar. Datorită prezenţei comunicării anormale între cavităţi sau vase se antrenează pasajul sângelui oxigenat din marea circulaţie în mica circulaţie. Evoluţia lor este dominată de riscul alterării ireversibile a vaselor circulaţiei pulmonare, ducând spre hipertensiune arterială pulmonară fixă. Progresele medicale, graţie echocardiografiei cuplate cu examinarea Doppler şi a progreselor chirurgicale, care au permis operarea copiilor la vârste tot mai mici prin cura completă, au adus foarte bune rezultate în cardiologia pediatrică.*

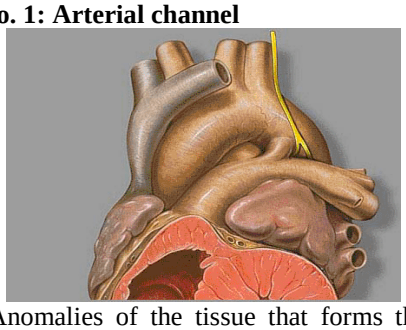
***Cuvinte cheie**: cardiopatii congenitale, cardiopatii cu shunt*

INTRODUCTION

A left to right shunt is a quantity of saturated blood that passes from left to right and overcharges the pulmonary blood flow. It is recognized by pulmonary and cardiologic clinical signs, pulmonary hypervascularity on radiography and echographic signs.

The arterial channel is a fetal structure that makes the connection between the pulmonary artery and the aorta isthmus. It plays a major part during the fetal period of time and is closed immediately after the birth.

Picture no. 1: Arterial channel



Anomalies of the tissue that forms the arterial channel wall may render difficult its closing up, leading to the persistence of the arterial channel, a quite frequent congenital cardiopathy (10-15% of the congenital cardiopathies). It is a cardiopathy easy to be diagnosticated and treated.

Cardiopathy etiology is less known. The persistence of the arterial channel may be found in a polimalformative context – it belongs to the malformations encountered in the rubeolic embryofetopathy – or it may be encountered in an isolated form in premature babies. It was observed that the frequency is raised in those who are living at higher altitudes.

Evolution

The arterial channel may increase in sizes, in the babies born at term, or it may close spontaneously in the first three weeks of life.

Risks

Before 6 month old:

- The cardiorespiratory insufficiency is essential

After 6 months old:

- Pulmonary arterial resistances may be fixed;
- At any age:
- Rarely, a hypokinetic myocardiopathy was emphasised, occurring after many months or years of evolution with diastolic overcharge of the left ventricle.
- The prophylaxis of the bacterial endocarditis is indicated;
- Aortic and mitral insufficiency is supervised, due to the dilatation of the valvular rings.

Catheterization

Catheterization is not indispensable, it becomes exceptional and it is indicated only when the quality of the echographic image is not accurate, or if there is doubts regarding the presence of a fixed pulmonary hypertension.

Treatment

The only treatment in the case of arterial channel is its closing up. The closing may be spontaneous, medical or surgical. If waiting, in case the clinical condition is good, the cariorespiratory insufficiency may be treated as in all badly tolerated left-rights shunts.

Medical closing

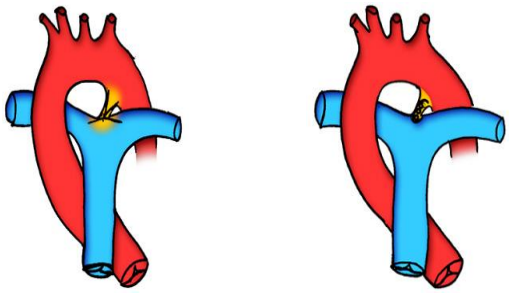
The administration of drugs that inhibit the synthesis of prostaglandins (more frequently in the pre-term new-born babies and rarely among those at term) may obtain the closing of the arterial channel. The drug used is Indometacin (80% efficacious); the dose is of 0,1mg/kg/day, for 7 days.

Closing through interventional catheterization

This method consists in obliteration of the arterial channel on the endovascular duct, with an "umbrella" prosthesis, inserted through vascular puncture, prosthesis that will be left in the channel lumen. In the case of a large channel, the prosthesis will let a residual shunt in 10-20% of the cases.

Using "Coil serpentine" for closing the smaller channels is much safer, even if there were few cases of repermeabilization of the arterial channel. There is a minimal risk of vascular obstruction, that is why it is indicated in children above 1 year old.

Picture no. 2: "Umbrella" prosthesis" - "Coil serpentine".



Surgical closing

Surgical closing is made through left lateral thoracotomy. This method registers a null percentage of mortality and the hospitalization period of time is of around 8 days. Complications are rare: pneumothorax, or the paralysis of the phrenic or recurrent nerve. The simple suture may be repermeabilised.

The channel closing is made by placing two "clips" with the help of the video-surgery.

The channel obliteration is made by using two titan clips with the help of endoscopy, avoiding scars forming. The hospitalization period of time is of about three days, mortality is null, the complication encountered is the paralysis of the left recurrent nerve, but in rare cases.

Indications:

- Closing the arterial channel which is badly tolerated in pre-term babies, in the absence of counter indications (renal insufficiency, digestive problems) may be closed with Indometacin, and in case of failure, the surgical cure is proposed.
- Arterial channel that is badly tolerated and associates hypertension: the surgical cure (video surgery) is suggested immediately after the new-born period of time.
- Arterial channel with large flow but well tolerated and without important arterial hypertension: surgical cure (video surgery) around the age of 1 or in case, IAo or IM occur.
- Small arterial channel: closing through catheterization: between 1 and 2 years old.

Further evolution

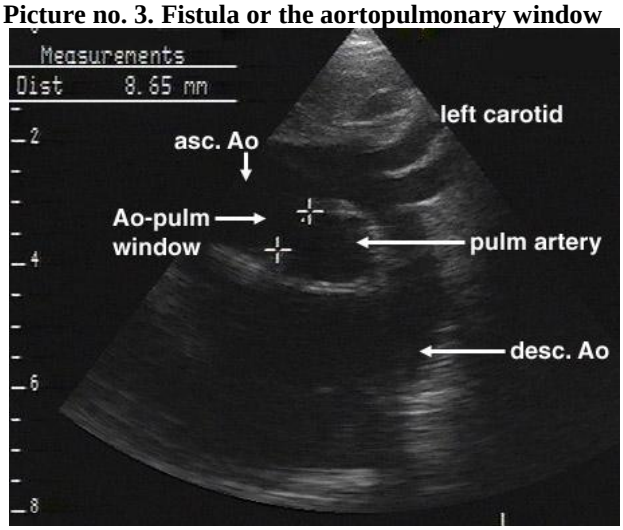
With the exception of the closing through endovascular prosthesis, which should be subsequently supervised and submitted to the antioslerien prophylaxis, arterial channel closing is considered definitive.

AORTOPULMONARY FISTULA

Fistula or the aortopulmonary window represents an abnormal communication between the ascendant aorta and the trunk of the pulmonary artery. It is a rare affection, representing 1% of the congenital cardiopathies and is more frequent in boys than in girls (3/1). The associated malformations are: anomalies of the sigmoids and anomalies of coronaries position.

It accomplishes an important arterial left-right shunt, associating the clinical signs of the shunt – a strong peripheral pulse.

Picture no. 3. Fistula or the aortopulmonary window



Spontaneous evolution

As long as the pulmonary arteriolar resistances are low, the evolutive risk is that of an important left-right shunt: cariorespiratory insufficiency.

After the age of 6 moths, the pulmonary resistances evolve towards a pulmonary obstructive malady with clinical improvement, secondarily to the decrease of the shunt. Cyanosis occurs subsequently, by inversing the shunt. The closing is made surgically through median sternotomy and by using a Dacron patch. The relation between the Ao-pulm window and the origin of the coronary arteries must be observed. The prognostic is good and these patients are considered cured.

BIBLIOGRAPHY

1. Batisse A. Cardiologie Pédiatrique Pratique, Dion Editeurs, Groupe Liaison SA, 2002 ;
2. Sidi D. "Classification des malformations, physiopathologie et hémodynamique ". In : Association Saint-Vincent-de-Paul prénatal, ed. Diagnostic et prise en charge des affections foetales . Paris : Vigot 2001;
3. Nelson J. Textbook of pediatrics – 17th edition. Editura: Saunders 2004 ; 1499 – 1554;
4. Simoneau G et Coll. Clinical classification of pulmonary hypertension. J Am Coll Cardiol 2004; 43 : S5-12;
5. European Society of Cardiology. Guidelines on diagnosis and treatment of pulmonary arterial hypertension. Eur Heart J 2004; 25 : 2243-2279,
6. Philippe L., Classifications couramment employées en pathologie cardiovasculaire, 2000.