

Sudden death of a premature new-born with hypoplastic left heart syndrome. Morfological study of the heart

Florin Mihail Filipoiu¹, Mihaela Balgradean^{2,*}, Viorel Jinga³

Abstract: The Hypoplastic left heart syndrome (HLHS), also known as the Norwood Syndrome, is a complex clinical entity, a part of the cardiac congenital maladies, with a birth frequency of 1/15000. It is a severe disease with a huge mortality index after-birth. The lack of development of the left ventricle and the atresia with mitral or aortic stenosis mainly characterizes the syndrome.

We report a case of a premature male fetus which was diagnosed with HLHS at 6 months of age by cardiac echographic examination. The new-born died at 3 hours after birth by cardio-respiratory arrest due to ventricular fibrillation.

We noticed that the specialized literature does not include photographic iconographies, owing to the fact that all articles use only explaining schemes. This fact has determined us to achieve a suggestive dissection of the heart.

Key Words: hypoplastic left heart syndrome, HLHS, dissection.

The Norwood syndrome known as the hypoplastic left heart syndrome (HLHS) describes a group of cardiac malformations which consist in various underdevelopment stages of the left heart and of the aorta. Its consequences are related with the incapacity of the heart to sustain the systemic circulation and with a significant obstruction to blood flow.

Lev [1] in 1952 was the first to use the term hypoplasia of the aortic tract complexes.

The first to use the term of HLHS was Noonan and Nadas [2] in 1958 describing an atresia or stenosis of the left side of the heart, but in a general sense, the area of discussions ranging from the aortic atresia to an underdevelopment of the left ventricle.

In the 1980's the Norwood operation was developed and resulted in a synonymous use with

HLHS, because it implies the inability of the left heart to support systemic circulation [3].

The subject related to the HLHS was debated by the Heart Surgery Nomenclature and by the European Association for Cardiothoracic Surgery to establish a common nomenclature [4].

MATERIAL AND METHODS

The investigated heart has been harvested from a seven-month fetus, deceased in an obstetrics service, three hours after a premature birth. The fetus was previously diagnosed by fetal eco-cardiography with HLHS.

After the demise of the child, the mother gave her approval so that the corpse may be used in

1) Associate Professor, Department of Morphological Sciences, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

2) *Corresponding author: Associate Professor, Department of Pediatrics, "Carol Davila" University of Medicine and Pharmacy, Tel: 0722.280.110, E-mail: mbalgradean@gmail.com, Bucharest, Romania

3) Associate Professor, Department of General Surgery, "Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

scientific purposes.

The heart has been dissected and we made sagittal sections through both left and right part. The dissection stages were photographed using a digital camera.

RESULTS

The examined corpse presented generalized adenopathy. The thymus was hypertrophied. Between the thymus and the base of the heart a tumoral mass can be observed, having the aspect of a lymphoma (Figure 1).

The heart is generally hypertrophied, with a dominant right hypertrophy. After removing the pericardium the following aspects can be observed:

- on the sternocostal side the coronary vessels

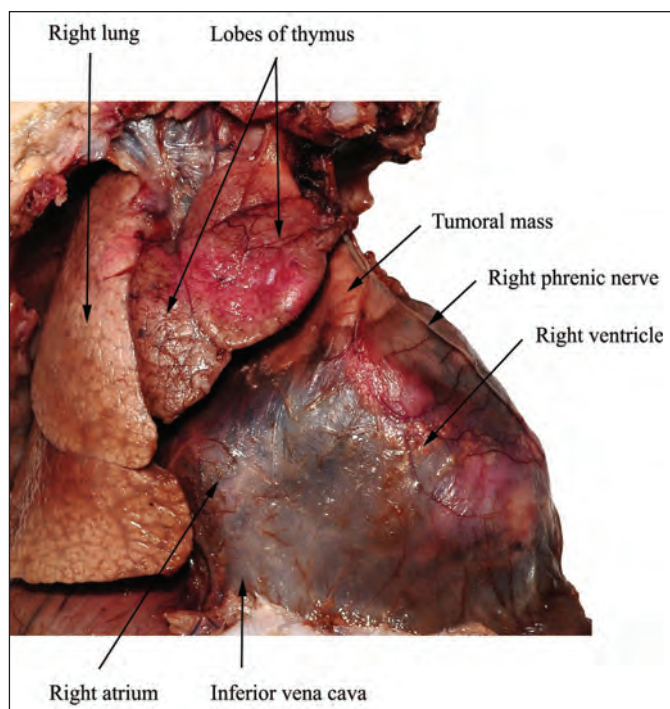


Figure 1. Heart-lungs-thymus ensemble, antero-lateral view.

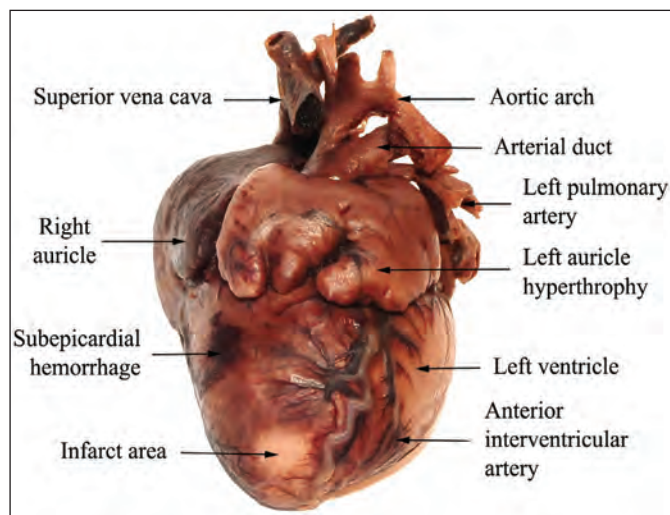


Figure 2. Heart-anterior view.

are turgid and sinuous, with a developed collateral circulation.

- the left auricle is very large in dimensions. It comes in contact and even covers the right auricle (Figure 2).

On the base of the heart the coronary sinus requires a special attention. It appears to have stopped evolving, without reducing its diameter size up to the usual. It still has the aspect of a horn of the venous sinus, in which opens the great cardiac vein. (Figure 3).

Proximal to the junction with the great cardiac vein, the oblique vein of left atrium (Marshall) can be observed.

Between the ascending aorta and the pulmonary artery we observe two embryological remains of the brachial arteries. At their origins they are united in a conjunctive mass and have no lumen. They have no orifices of origin in the aorta or in the pulmonary artery.

The ductus arteriosus is permeable and well represented, above the pulmonary artery origin. (Figure 4).

On sagittal section we can observe a hypoplastic left atrium with the endocardium being thickened and undifferentiated. Unlike the atrium, the left auricle is large, but atretic and almost without a lumen.

On the interatrial septum we can see that there is no interatrial communication and the oval fossa was closed by the overlapping of various successive structures (Figure 5).

The mitral valve is atresic, the cusps are small, thickened, inserted in the ventricle on small papillary muscles. There are no chordae tendinae.

The left ventricle is hypoplastic. Its endocardium is thickened. The left ventricular myocardium is very thick, has no regular structure and keeps a poorly differentiated appearance (Figure 6).

The aortic orifice is atresic and stenosed. The free edges of the semilunar valves are stuck together in a common mass. The left coronary sinus and the non

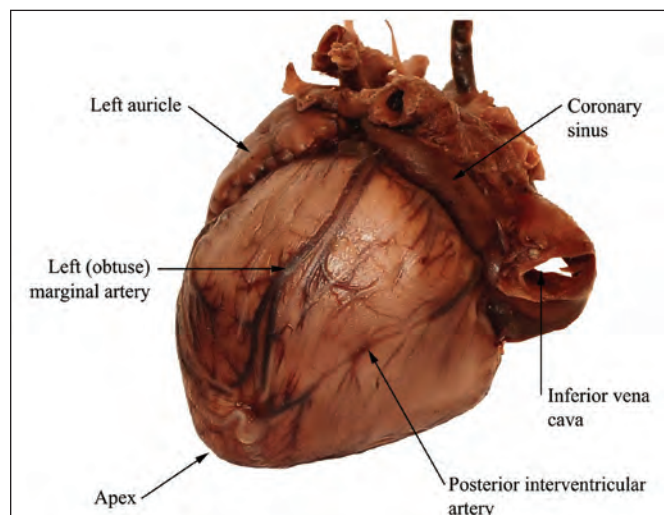


Figure 3. Heart-pulmonary and diaphragmatic surfaces.

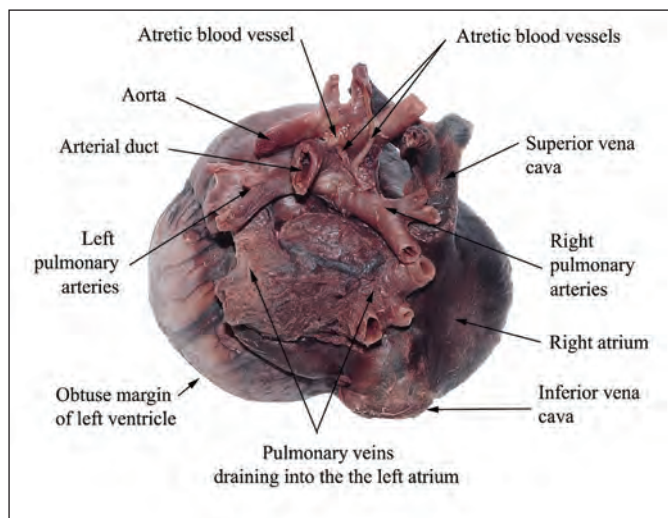


Figure 4. The base of the heart.

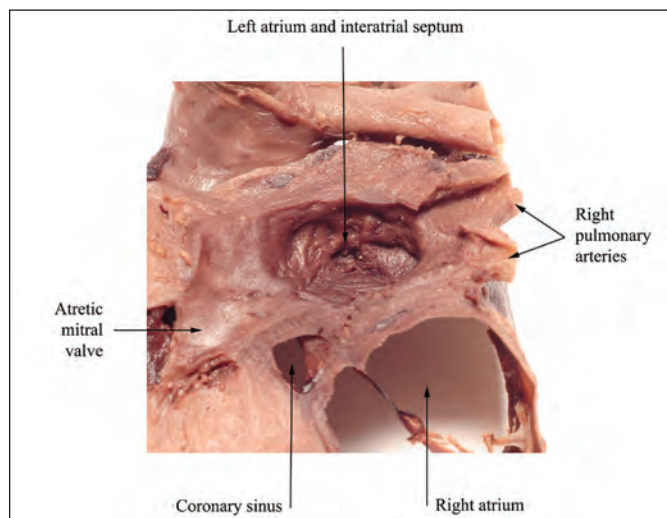


Figure 5. Sagittal cross-section through the left atrium. Left view of the interatrial septum, the left atrial cavity and of the mitral orifice.

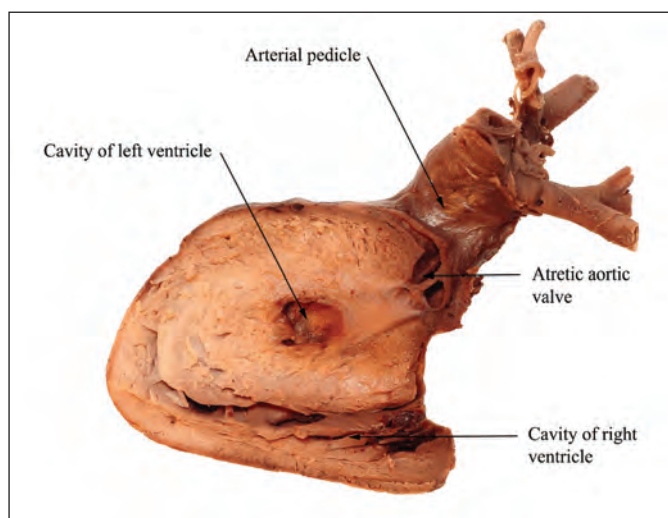


Figure 6. Sagittal cross-section through the left ventricle-left view.

coronary one are differentiated and have lumen.

The coronary right sinus is atretic and has no lumen. There is a coronary right permeable artery. The section includes the origin of the left coronary artery, which presents a lumen and thick walls. We mention that beyond the level of the coronary ostium the ascending aortic lumen is present and well formed.

DISCUSSION

The left ventricular hypoplasia refers to the absence of forming a normal-sized ventricular cavity [4-7]. But the left ventricle exists and has very thick walls, with a nearly embryonic aspect of the myocardium, which stopped its evolution. This issue might be due to the lack of blood flow through the ventricle.

The presence of the coronary, both sinuous and turgid, suggests that the left ventricular myocardial mass is not sufficiently permeable for the coronary flow.

The coronary ostium is located above the atretic area. The coronary perfusion is retrograde through the arterial duct.

- The atresia of the left auricle prevents the achievement of an efficient surgical communication with the right auricle. Because in the Norwood type operations the surgeons apply for an interauricular anastomosis, our observation becomes important for that moment when the surgical technique is to be chosen [9].

- The presence of the generalized adenopathy and thymus type of hypertrophy draws attention to the associated pathologies that may be involved in the pathogenesis or aggravation of the heart disease.

CONCLUSION

We consider that presenting and commenting the dissection of a heart with the Norwood syndrome, has several benefits:

- shows the clinician the seriousness of this pathology.
- accommodates the cardiac surgeon with anatomic details of the hypoplastic left heart syndrome.
- it represents a good reference for the effort of the imagistic diagnosis.
- the grim prognosis may be better explained to the patient's caregiver, using these images.

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