

## CLINICAL AND THERAPEUTIC ASPECTS IN THE SEVERE AORTIC COARCTATION WITH DUCT DEPENDENT SYSTEMIC CIRCULATION IN THE NEWBORN

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### ABSTRACT

The aortic coarctation has an incidence of 0.6-0.8/1,000 newborn, with clinical manifestation starting with the neonatal period. In the newborn, the heart congenital malformations associated to heart failure and clinical response early in the first hours of life have a guarded prognosis, needing an early diagnostic and an adequate therapeutic conduct. We present a clinical case of severe aortic coarctation with duct dependent systemic circulation, diagnosed early in the neonatal period. The early treatment with Prostaglandin E1 allowed the maintenance of the haemodynamic balance, the newborn benefiting from surgical correction in the first month of life.

**Keywords:** newborn, duct dependent severe aortic coarctation, heart failure, prostaglandin E1, surgical treatment

### CASE PRESENTATION

Patient A.D., age – 10 days, is admitted through clinical transfer for generalized cyanosis, tachypnea, dyspnea on nursing effort and the investigation of a systolic murmur detected in the maternity. The newborn is the first child of an apparently healthy couple (mother-18 years old, father-22 years old), born through natural birth, at a gestational age of 40 weeks, having the birth weight of 2,700 g and the APGAR score: 8 in 1 minute and 9 in 5 minutes. Upon admission, the child presents a serious general condition, size -1.52DS, weight -2.2DS, cranial perimeter -2.9DS, pale, elastic tegmina, generalized hypotonia, perioronasal and extremity cyanosis, severe respiratory functional syndrome: tachypnea, intercostal and subcostal retraction, RF: 35 respirations/minute, rhythmic

The aortic coarctation in children represents almost 6-8% of the total number of heart congenital malformations, being the fourth most frequent lesion which requires surgical intervention or catheterization during childhood (1). The most frequent deficiencies associated to the aortic coarctation are the arterial channel persistence, VSD and the aortic stenosis. Therefore, the aortic bicuspidy can be noticed in almost two thirds of the children with aortic coarctation (2). According to Abbott's studies, the patients survive, on an average, up to 33.5 years (3). If the surgical intervention is performed in an early stage (up to the age of 14), the survival up to 20 years following the intervention is of 91% and in case of the tardy abnormality correction, the survival decreases to 79% (4,5). The treatment options include surgical interventions, balloon angioplasty and endovascular stent graft.

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tachycardic heart sounds, with the heart rate: 140 beats per minute, systolic murmur grade IV/6 right i.c III and left parasternal space, SaO<sub>2</sub>: 65-70%, bilateral non-palpable femoral pulse, cold lower extremities, blood pressure at the level of the right upper limb: 70/50 mmHg, blood pressure at the level of the right lower limb: 50/30 mmHg, depressible abdomen, liver at 3.5 cm below the costal margin, intestinal tract and physiological urinations, fed through gavage, FA:2/2 cm normotensive.

At paraclinical level, we noticed a deficiency anemia and thrombocytosis.

The thoracic radiography shows an increased heart, through the crowning of the left lower arch, cardio-thoracic index of 0.61, elevated heart edge, charged pulmonary circulation.

The electrocardiogram shows a sinus rhythm of 160/min, the electrical physiological right axle (+120 grade), flattened T waves in the frontal derivations, slightly elongated QT: 0.28 sec.

The 2D echocardiography, the parasternal section of the short axle highlights a concentric hypertrophy of the ventricle walls and the aorta which presents 2 cusps; the suprasternal section, long axle, allows the assessment of the aortic arch, the coarctation area, the post stenotic dilatation. The color Doppler superposition at coarctation level

and in the trunk of the pulmonary artery shows a turbulent flow and the continuous Doppler superposition shows an envelope with increased velocity in the systole.

We instituted the treatment with Prostaglandin E1, in order to maintain the permeability of the arterial and diuretic channel, for the heart failure therapy.

Considering the clinical condition of the patient upon admission in the hospital, the easy-going favorable evolution under the instituted medication and the echocardiograph aspect, after 20 days of hospitalization, we performed the transfer to a specialized center of pediatric cardio-vascular surgery, where the surgical correction is performed.

After anesthesia induction, two arterial lines were performed: one at the level of the right radial artery and another at the level of the left femoral artery, in order to measure the blood pressure at the level of the upper and lower limbs. The systolic blood pressure at radial level was 55 mmHg higher than the systolic blood pressure at femoral level. After a left posterolateral thoracotomy at the level of the i.c. III space, we performed the resection of the segment which comprises the aortic coarctation, followed by termino-terminal anastomosis and the arterial channel ligation. The postoperative

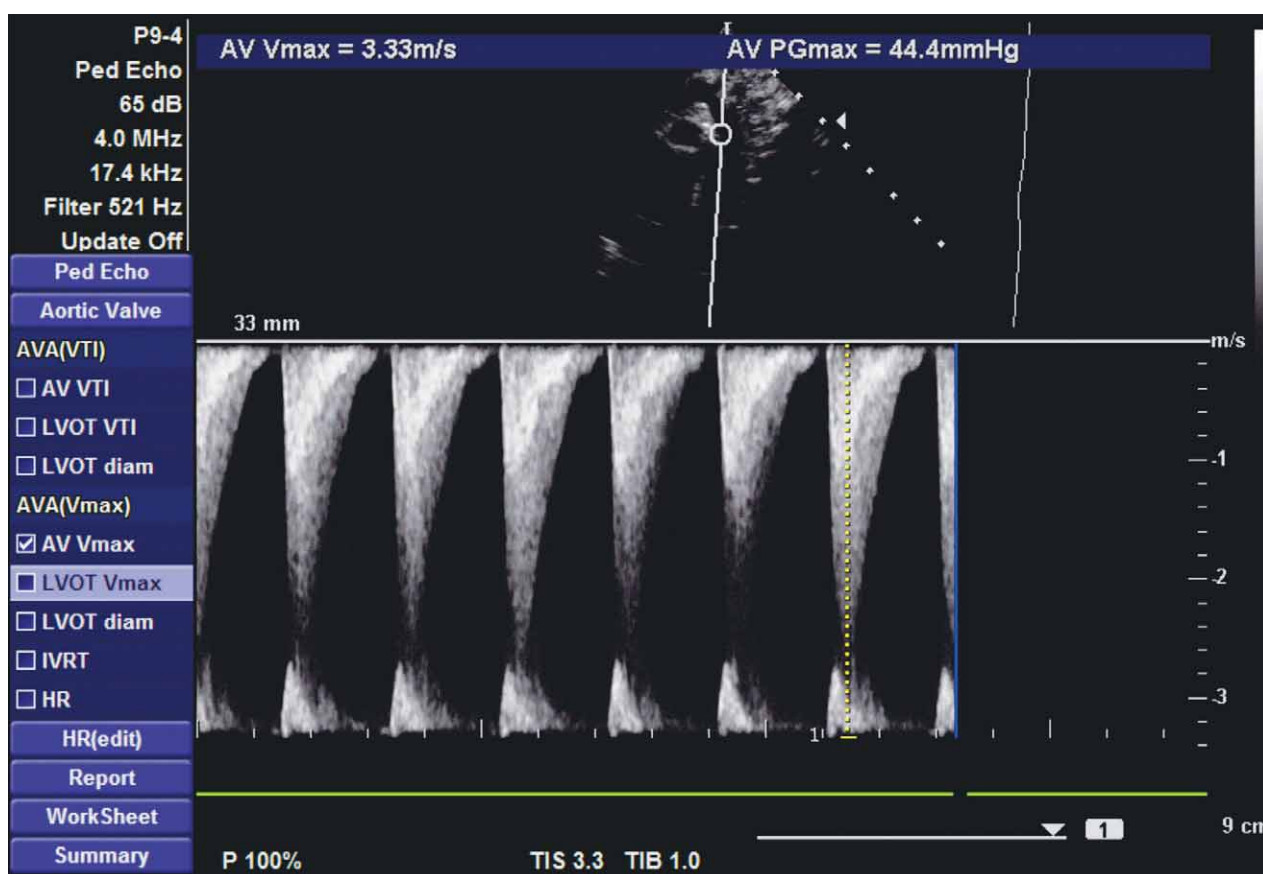


FIGURE 1. Patient A.D. – Aortic coarctation - transthoracic echocardiography, suprasternal section

evolution was favorable, but with difficult haemodynamic adaptation, needing a prolonged inotropic support and vasodilatation treatment in increased doses for pressure values control. As after the surgery, we noticed the presence of the anemic syndrome (Hb 8.8 g/dl), the patient needed the administration of erythrocytic mass, without side reactions.

The echocardiograph proved a normal flow in the descending aorta, without the existence of a gradient at anastomosis level. The pulmonary artery gradient was of 50 mm Hg (residual pulmonary high blood pressure), being considered as satisfactory.

8 months following the surgery, the patient presents a good general condition, normally colored tegmina, postoperative scar with normal appearance, systolic murmur grade II/6 in the right i.c. II space, AV: 120 bpm, bilateral arterial pulse at extremities level, blood pressure at the level of the upper limbs: 102/61 mmHg, at the level of the lower limbs: 110/65 mmHg, liver edge at the costal margin.

The patient receives treatment with captopril (3 mg/day), furosemide (1 mg/day), spironolactone (3.125 mg/day), aspacardin, aspenter, the infectious endocarditis prophylaxis and shall be periodically examined in order to monitor the blood pressure, the echocardiograph tracking of the associated abnormalities and of a new eventual stent narrowing.

## DISCUSSIONS

The isthmus aortic coarctation represents aorta narrowing at the level of the joint between the distal aortic arch and the descending aorta, immediately below the origin of the left subclavian artery. Coarctation presence determines pressure increase at the level of the proximal aorta and of the left ventricle post charge, and at distal level, the pressure is low. When the coarctation is associated to the arterial channel persistence, takes place the blood vein-arterial return through the arterial channel persistence with right ventricle solicitation.

Because of the severe form of the aortic coarctation, the clinical signs have started early in the first day of life through signs of brutally installed congestive heart failure, translated from a clinical point of view through tachypnea, cyanosis, cold lower extremities with absent pulse at the level of the femoral and pedis arteries.

The systemic flow of the lower extremities of the body being duct dependent, Prostaglandin E1 0.1 mcg/kg was administered in order to maintain the arterial channel permeability, by paying attention to side effects occurrence.

The surgical correction through CoAo resection with termino-terminal extended anastomosis was the election treatment in this case, because of the transcoarctation gradient of maximum > 20 mmHg, the presence of the trailing symptoms and of the haemodynamic consequences (severe heart failure). CoAo correction in the early period allows the vascular elasticity maintenance and the prevention of the residual high blood pressure development.

But, in spite of the favorable postoperative early results, a new stent narrowing at the coarctation level was frequently reported in several clinics (20-86%) (6,7,8), mainly in the patients of less than one year old; but the use of the resorbable sutures instead of those of silk determined a significant decrease of the new stent narrowing.

Several studies show that almost 68% of the patients suffering a surgical intervention for the aortic coarctation correction in the first year of life shall present high blood pressure in their evolution. The physiopathology mechanism is not fully known, but it seems that the postoperative modification of the aortic arch architecture and the affection of mechanoreceptors sensitivity can be favorable factors in high blood pressure development (10).

Therefore, the periodic monitoring of the patient through a detailed anamnesis, a full clinical examination, with pulse palpation at the level of the femoral and pedis arteries, pressure values measurement and the echocardiograph assessment allow the identification of the eventual complications.

## CONCLUSIONS

The severe aortic coarctation with duct dependent systemic circulation presents a suggestive neonatal beginning, needing a diagnostic as early as possible. The initiation of the perfusions with prostaglandin E1, in order to maintain the arterial channel permeability, to avoid the heart oversteering, the pulmonary circulation overloading and the surgical treatment are essential stages in the management of these cases.

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