SEVERE JUNCTIONAL BRADYCARDIA BY DESTRUCTION OF THE SINUS NODE AT A PATENT WITH COMPLEX SURGICAL CORRECTION OF A CYANOGENIC CONGENITAL HEART DEFECT – THE FONTAN PROCEDURE

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Abstract

The Fontan procedure, in the case of the unique ventricle, can affect the specialized circulation system, starting with the sinus node. The incidence of the arrhythmias increases with the passing of time from the surgical event. The authors present the case of a 12-year-old patient, with severe junctional bradycardia by destruction of sinus node, following a the Fontan procedure performed for the surgical correction of a cyanogenic congenital heart defect.
Key words: Fontan procedure, jonctional bradycardia, desctruction of the sinus node.

Case presentation

We shall present the case of 12-year-old patient, B.R., with a known complex cyanogenic congenital heart disease. During his first 5 years of life, his evolution manifested the progressive intensification of the cyanosis, staturoponderal retard, and after this age were additionally present crises of hypoxia, of medium severity, with a mild initial evolution, followed by a progressive aggravation.

In the 6th year of life he is explored by the Institute of Cardiac Diseases: the physical objective exam shows generalized cyanosis, more obvious around the nose and in the nailbed, Hippocratic fingers, presternal extension, apexial shock in the 6th intercostal space, left, on the anterior axial line, intensive presternal murmur, rough intensive murmur with a maximum of intensity in the 2nd-3rd intercostal space, left parasternal.

Paraclinic investigation:

Electrocardiography: Sinus rhythm, AV = 90 beats/ min, electric axis with left deviation, left ventricular hypertrophy (fig. 1).

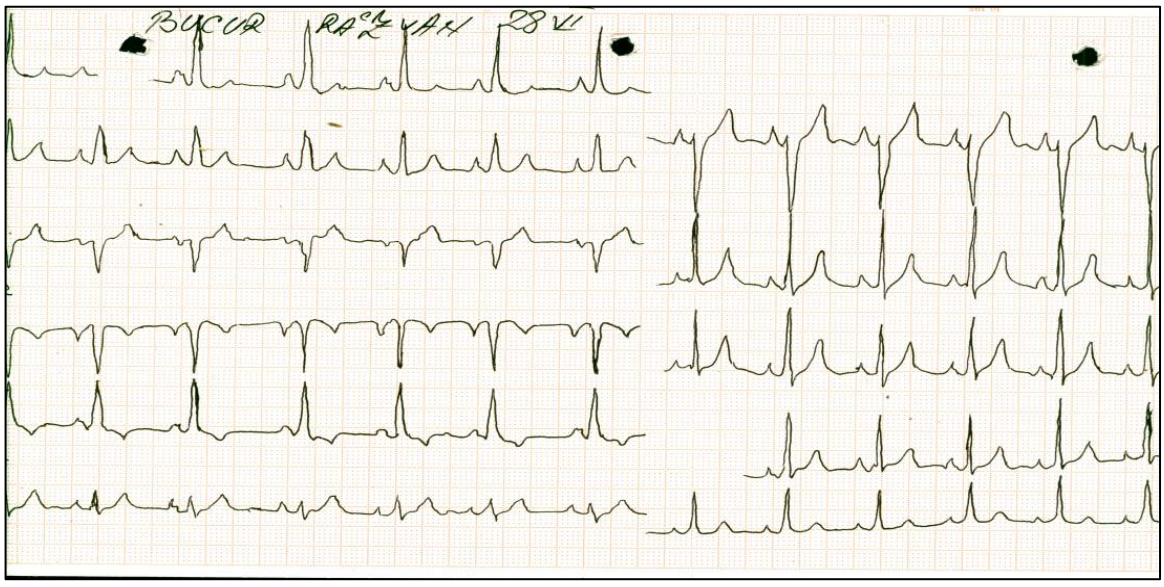

Fig: 1 EKG

Heart and lung radiography: slight cardiomegaly, cardiothoracic index = 0.52 slightly congestive lung hiluses, with a limitation of the peripheral lung circulation, prominence of the left inferior arch, with high cardiac apex lifted from the diaphragm plane, prominence of lung arch.

Echocardiography (fig. 2): atriums in situ solitus, normal atrioventricular connections, tricuspid valve atresia (fibrous chord), malpositioned vessels with parallel trajectory, atrium septal defect, ventricular septal defect, perimembranous, slightly right oriented aorta, rudimentary right ventricle, pulmonary, valvular and supervalvular artery stenosis, systemic venous blood passing through the atrium septal defect in the left atrium, where it mixes with the venous pulmonary blood, both passing though the mitral valve (highly hyperkinetic); then, the left ventricle contributes to the performing of both circulations: systemic, though the aorta, and pulmonary, through the pulmonary artery, the right ventricle being hypoplasic and reduced only to the ejection tract.

After applying the cardiac catheter (fig. 3), we have observed an interatrial and interventricular communication, proven by the probe trajectory. Bidirectional shunt, causing systemic desaturation of 88,3%. Systolic gradient VD-AP=47 MMHg. It was selectively catheterized, AP retrograde, (Ao-VS-DSV-VD-AP). Efficient ventricles, RPT=1.6 HRU, RPT/RST=0.1; normally positioned VCS. The contrast passes in turns into AD-AS-VS-Ao and though the ventricular septal defect, right ventricle and pulmonary artery.

Ventricular septal defect, largely perimembranous, though which it is distinguished a rudimentary right ventricle, of small dimensions. Valvular pulmonary artery stenosis with small ring, trunk and branches developed. Adequate VAo, normal coronaries.

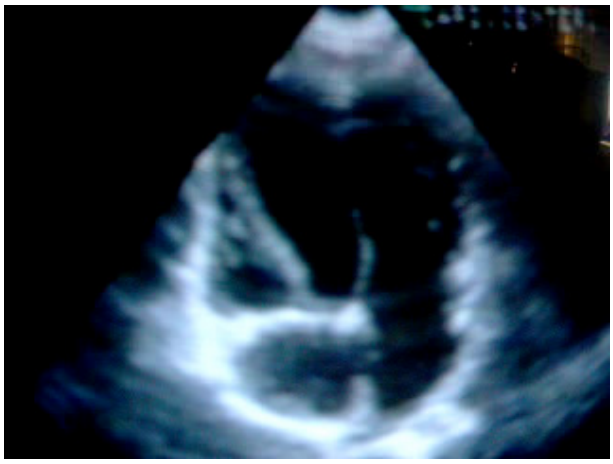

Fig. 2: Echocardiography preoperatory.

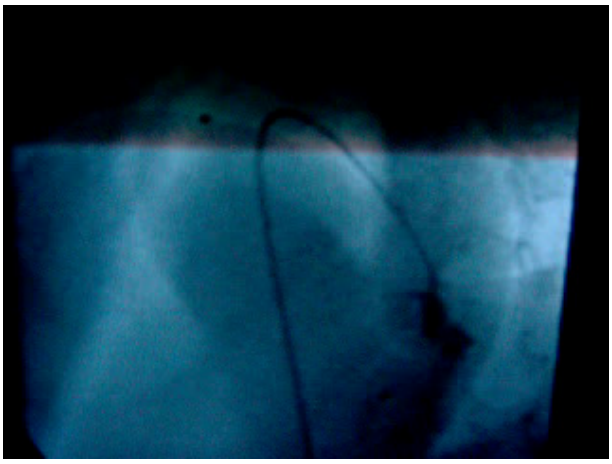

Fig. 3: cardiac catheter.

Following these explorations, we have issued the diagnosis of pulmonary valve atresia Type II B, with transposed vessels and pulmonary stenosis and we have decided to perform the complete correction - the Fontan procedure (fig. 4). (1) There are some criteria for the selection of the patients candidates to the Fontan procedure; our patient can be included in these criteria, lowering significantly the immediate post-procedure risk and ensuring a good evolution in the long term; these criteria are the following:

- total pulmonary resistances under 6 U WOOD
- average pressure in the pulmonary artery under 18 mmHg
- pulmonary branches of normal dimensions
- efficient left ventricular function
- tele-diasystolic pressure VS < 12 mmHg
- FE > 60%
- Absence of backflow of the atrioventricular valve
- Age between 4-10 years

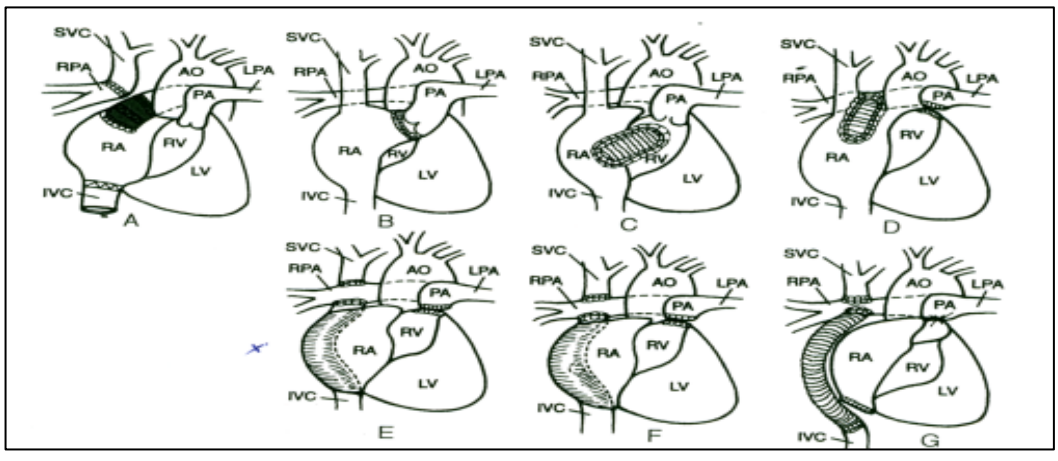

Fig 4: The Fontan procedure.

Immediately after the procedure, the patient's electrocardiography modified (fig. 5): the negative P wave, preceding the QRS complex, FC=65-80 beats/ min, aspect of junctional rhythm with the efficient adaptation to effort.

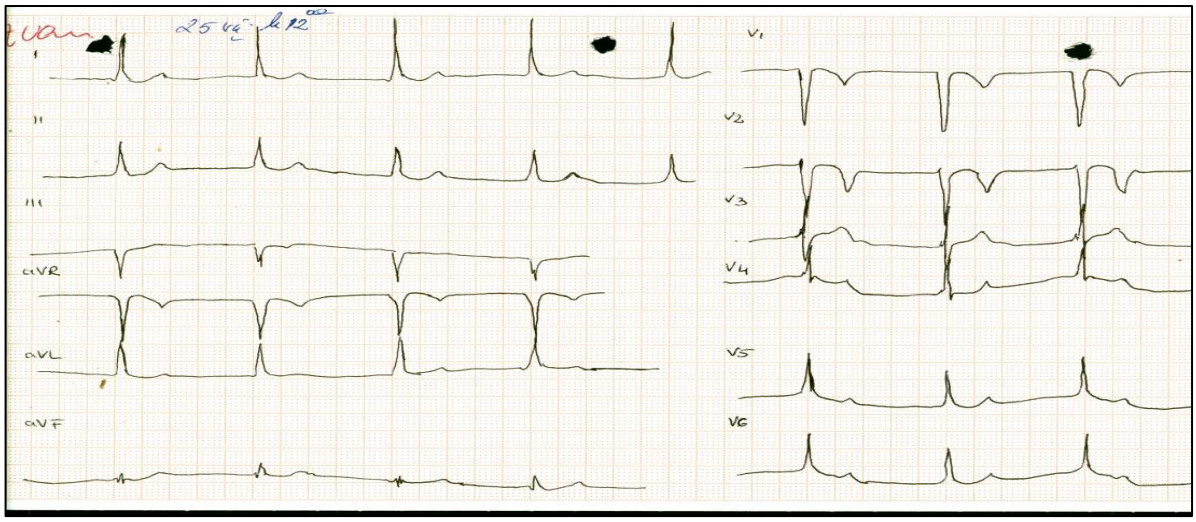

Fig. 5: EKG after the procedure.

In time, the patient gradually lost the cyanotic effect, the perioperatory recovery was good; the patient was released from hospital 9 days after the procedure, having the surgical plaque already healed; as unique medication, it was recommended the anticoagulant treatment, under the supervision of INR. During the periodic medical examinations performed, a marked tendency to bradycardia was observed, with FC rest = 55-60 beats/min in the first 4 years, then FC rest tends to lower below 50 beats/min in the last 2 years, this being interpreted as a significant bradycardia and determining the exploration with a view to determine the suitability of permanent electro-stimulation. (2,3)

On the electrocardiography appears junctional rhythm, FC=43 beats/min, electrical axis QRS left deviated, wave P negative in DII, DIII, AVF, wave QS in V1, wave T negative in V1, V2; rare supraventricular extrasystoles.

Holter-EKG: average FC = 40 beats/min, minimum FC = 26 beats/min, maximum FC = 86 beats/min, without TPSV or TV episodes, with 306 sinus pauses > 2.5 seconds, the longest pause being of 3.2 seconds, inferior atrial rhythm (wave P negative in DII, DIII, aVF), short periods of sinus rhythm (wave P positive in DII, DIII, aVF).

The effort test: TA start = 120/70mmHg, FC = 47 beats/min, inferior atrial rhythm (wave P negative in DII, DIII, AVF) at 100w, TA = 150/80 mmHg, FC = 125 beats/ min at 75w, FC = 90 beats/ min, aspect of sinus rhythm (wave P positive in DII, DIII, AVF – fig. 6) with the reappearance of the inferior atrial rhythm during rest, at the same time with the lowering of the cardiac frequency.

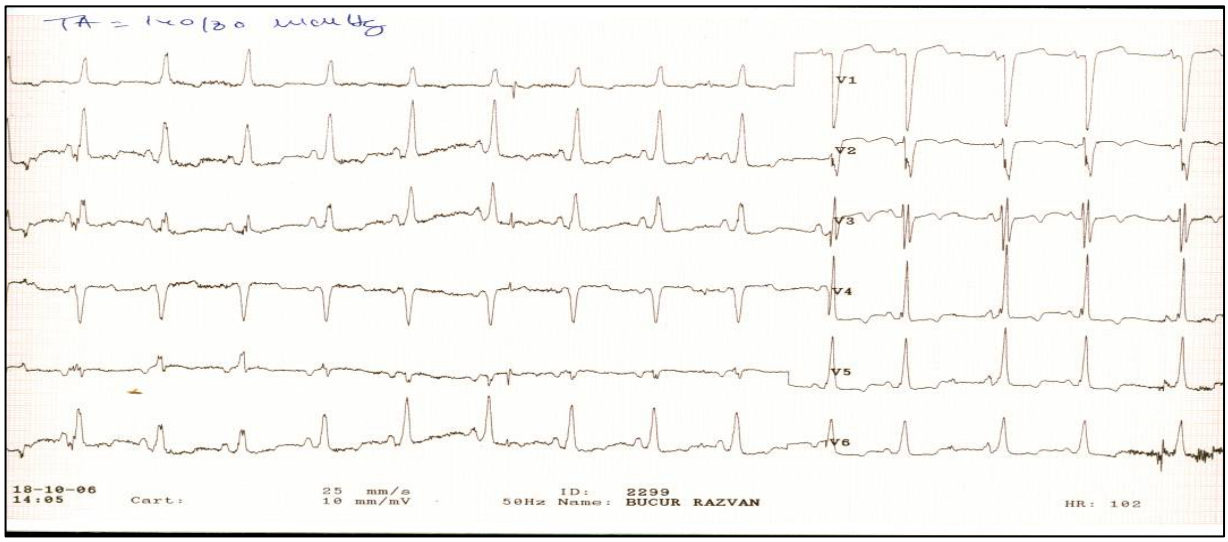

Fig 6: Wave P positive in the effort test.

The patient can be included in the 2nd indication class of diffuse electro-stimulation, asymptomatic sinus bradycardia for a child with complex heart disease and heart frequency during rest under 40 beats/min or ventricular pauses > 3seconds. In this situation, were considered the type and way of electrostimulation. The only logical procedure in this case is the permanent electrostimulation.

The ideal stimulation would have been an atrioventricular bicameral stimulation, as the patient had a good atrioventricular circulation; the preservation of the atrial pump would have been beneficial.

Due to the complex morphological aspect resulted after the Fontan procedure, the accession of the endovascular probe in the right atrium is extremely difficult from the technical point of view and, for this reason, the patient was explored morphologically.

The status after the Fontan procedure for tricuspid atresia, subaortal septal defect – 1.3 cm; right ventricle small, hypoplasic, without communication between the left and right atriums; without communication between right and left atriums, separated by a fibrous chord; Ao at valvular level = 2.12 cm, Ao ascending = 2.9 cm, DTDVS = 47 mm, PPVS = SIV = 1cm, FE = 64% (fig. 7).

The x-ray venography and the right cardiac catheterization were performed by injecting the contrast substance though the right femoral vein at the level of the right atrium; this showed the communication between the atrium and the inferior caval vein and with the right pulmonary artery though a duct; the injection of the contrast substance through the brachial vein at the level of the superior caval vein shows its communication with the pulmonary artery, without communication between the right atrium and the superior caval vein.

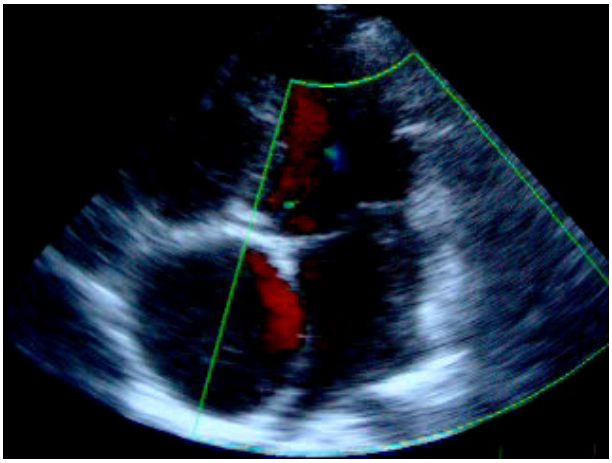

Fig 7: ECO after the Fontan procedure.

This patient's case is extremely interesting, both morphologically and from the point of view of the arrhythmia evolution of post-surgical origin. The more complex the congenital heart defect is, the greater the possibility of a complex arrhythmia; this makes necessary the interdisciplinary collaboration.

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