Sinus Node Dysfunction in a Patient with Left Atrial Isomerism

Sírio Hassem Sobrinho, Airton Camacho Moscardini, Ulisses Alexandre Croti, Carlos Henrique de Marchi, Moacir Fernandes de Godoy

Faculdade de Medicina de São José do Rio Preto - São José do Rio Preto, SP - Brazil

We report the case of an asymptomatic six-year-old child with left atrial isomerism and sinus venosus atrial septal defect. The physical examination revealed several periods of bradycardia. During a 24-hour electrocardiographic monitoring the patient presented a significant sinus node dysfunction with sinus pauses of up to 2.4 seconds. A permanent pacemaker was implanted, with a satisfactory outcome.

Isomerism of the atrial appendages is a rare congenital anomaly characterized by symmetry of organs which are normally asymmetrical. There are two types of atrial isomerism: the right atrial isomerism – also known as asplenia syndrome or Ivemark syndrome, and the left atrial isomerism or polysplenia syndrome.

Polysplenia was first described by Abernethy in 1793[1], and only in 1967 did Moller et al[2] make a full description of the syndrome. In left atrial isomerism the most prevalent alterations are: polysplenia, presence of two morphologically left lungs, two morphologically left atrial appendages, absence of the hepatic segment of the inferior vena cava, midline liver, and high incidence of congenital heart defects.

The congenital heart defects most frequently associated with isomerism of the left atrial appendages are: double right ventricular outflow tract, single atrium, atrioventricular septal defect, pulmonary valve stenosis, and persistence of the left superior vena cava[2-4]. Heart rhythm disturbances are also described.

In Momma et al's[5] review, all patients with left atrial isomerism presented with a wandering pacemaker. Another common finding is the decrease in heart ratealong the years. At fifteen years of age, 70% of the patients present with sinus bradycardia. Atrioventricular block may be present in 50% of the patients in some phase of their lives, and may be progressive[6-8], with a higher prevalence of atrioventricular block in the fetus[9].

Histological studies show the absence of the sinus node in the junction of the superior vena cava with the right atrium[10-12]. A hypoplastic and abnormally located sinus node has been demonstrated in the upper lateral atrial wall[10], in the medial lateral atrial wall[11], and in the lower lateral wall, close to the coronary sinus ostium[12]. In most instances the sinus node cannot be found[11].

Key words

Congenital heart defects, left atrial isomerism, sinus node dysfunction.

Case Report

Six-year-old Caucasian child from the city of Mirassol do Oeste (State of Mato Grosso, Brazil), with a history of heart murmur and absence of symptoms. Her physical examination revealed an acyanotic, eupneic, hydrated child in good general conditions. Palpation of the chest revealed a mild precordial bulge, with no thrills. Cardiac auscultation showed a grade 2/6 systolic murmur in the pulmonic area, constant and fixed splitting of the second heart sound, and mild accentuation of the pulmonic component. Several periods of bradycardia could be observed.

The electrocardiogram (ECG) showed a sinus rhythm with a 70 bpm heart rate, right bundle branch block, electrical axis deviated to the right, and short sinus pauses.

Chest radiography revealed a mild enlargement of the heart area at the expense of the right cavities, mild increase of the pulmonary vasculature, prominent main pulmonary artery, and two morphologically left main bronchi.

The echocardiogram showed the presence of a moderate-sized sinus venosus atrial septal defect, two morphologically left atria, absence of the hepatic segment of the inferior vena cava, and presence of a dilated azygos vein following the descending aorta posteriorly.

The Holter test showed a predominant sinus rhythm with a mean 65 bpm heart rate (HR), alternated with a junctional rhythm with a minimum 31 bpm HR. Sinus pauses of up to 2.4s occurred (Fig. 1), followed by junctional escapes, 6,114 isolated supraventricular extrasystoles and 512 supraventricular extrasystoles in couplets. No symptoms were reported.

Discussion

Polysplenia is a complex syndrome with a high variability of anatomical findings.

Three types of heart rhythm disturbances may occur in the isomerism of the left atrial appendages: sinus node dysfunction[5], atrioventricular block[6-8], and dual atrioventricular nodal pathways[5].

The patient reported was a child diagnosed with left atrial isomerism associated with a sinus venosus atrial septal defect and sinus node dysfunction with sinus bradycardia, sinus pauses and junctional rhythm. Although the patient was asymptomatic,

Mailing address: Sírio Hassem Sobrinho •
Rua Siqueira Campos, 2597/13 - 15025-055 • São José do Rio Preto, SP, Brazil
E-mail: siriohassem@cardiol.br
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we decided to implant a pacemaker considering the duration of pauses and the unfavorable progression of sinus node dysfunctions in the polysplenia syndrome[5].

In patients with sinus node dysfunction the outcome is better when the AAI C/R and DDD C/R pacemakers are used for electrical stimulation because of the lower prevalence of atrial fibrillation, thromboembolic events, and heart failure[13]. However, because of the presence of atrial septal defect, pacemaker implantation exclusively in the ventricle was found to be convenient. Pacemaker implantation in most of the patients with sinus node dysfunction does not provide an increased survival[13], but rather a better quality of life. For this reason, pacemakers are not indicated for asymptomatic patients.

Although few studies on sinus node disease in atrial isomerism are available, one cannot ensure that the outcome will be favorable without pacemaker implantation, because the disease is progressive in this type of patient, even in the absence of symptoms[5]. After a four-year follow-up, the patient's outcome remains satisfactory.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

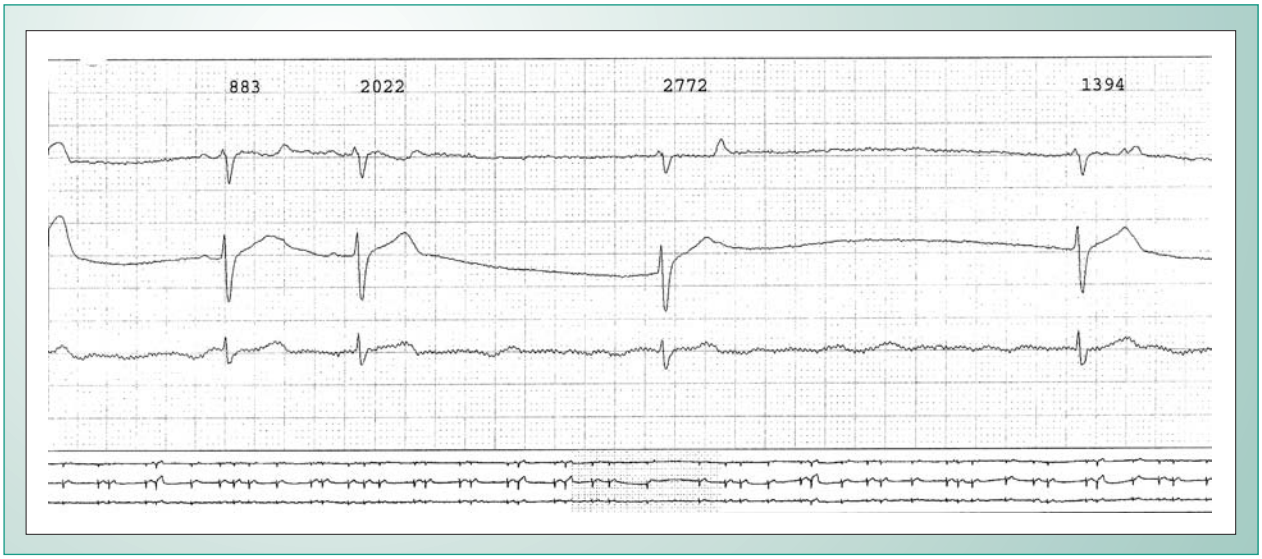

Fig. 1 - *Sinus beats followed by 1.6 and 2.4s pauses, respectively, with junctional escapes.*

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