Case report

The role of echocardiography in fetal tachyarrhythmia diagnosis. A burden for the pediatric cardiologist and a review of the literature

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Abstract

Supraventricular tachyarrhythmia represents the most frequent fetal dysrhythmia. In the lack of diagnosis and treatment these fetuses may develop hydrops and even death. For the therapeutic approach it is important to establish the diagnosis of the type of supraventricular tachycardia. In this paper we report 29 cases with different types of supraventricular tachycardia in which the diagnosis was established using our own protocol, which allowed us to make the difference between the types of tachycardia (atrioventricular reentry tachycardia due to the accessory pathway, atrial ectopic tachycardia and permanent junctional reciprocal tachycardia). We acquired the data by a series of recordings in M mode and pulsed Doppler by simultaneous recording of an artery and a vein flow. First of all, we diagnosed the supraventricular tachycardia type, with short or long ventriculoarterial interval, and afterwards, we made the difference between atrial ectopic tachycardia and permanent junctional reciprocal tachycardia using methods to decrease the atrioventricular conduction.

Keywords: fetal tachyarrhythmia; echocardiography; pediatrics

Introduction

Arrhythmia represents an important aspect of neonatal pathology with onset in the fetal period. Certain types of this pathology, premature atrial and ventricular beats and sinus tachycardia, have a natural history with complete resolution without treatment, but fetal and neonatal tachycardia can lead to fetal hydrops and heart failure, respectively and therefore, the therapeutic approach is mandatory [1].

Supraventricular tachyarrhythmia (SVT) and atrial flutter (AF) represent the most frequent fetal dysrhythmia with onset during the fetal period [2]. The therapeutic approaches in the prenatal and postnatal period, the immediate and long-term prognosis are different depending on the type of tachyarrhythmia. Considering this, the tachyarrhythmia diagnosis has to be as close as possible to electrophysiological mechanism. After birth the diagnosis of this pathology is made by interpreting the electrocardiogram, while it is much more difficult to determine an accurate diagnosis during the intrauterine period, and therefore, echocardiography is considered to be the most useful method for this phase [2,3].

Case reports

From February 2007 to April 2016, 29 cases of fetuses with electrical disorders were diagnosed in the Pediatric Cardiology Clinic and Obstetrics Gynecology Clinic of Targu Mures – Romania: 10 cases with different types of SVT, 2 cases with AF, 5 cases with atrioventricular block, and 12 cases with premature atrial beats. Any congenital heart disease was excluded. Using echocardiographic criteria, the following types of SVT were diagnosed in the fetuses group with SVT: atrioventricular reentry tachycardia (AVRT) due to the accessory pathway, atrial ectopic tachycardia (AET) and permanent junctional reciprocal tachycardia (PJRT).
We established the diagnosis of SVT based on the following echocardiographic elements: elevated heart rate, higher than 180-200 beats per minute and changes of relation between atrial and ventricular activity. We acquired the data by a series of recordings in M mode and Pulsed Doppler (PW). The M mode recording was performed in four chambers view, including the atrial and ventricular wall (fig 1a), in order to analyze the following elements: atrial and ventricular rate (fig 1b), regular and irregular aspect of tachyarrhythmia and also, the relation between atrial and ventricular activity. Ventriculoarterial (VA) interval was established by pulsed Doppler, based on simultaneous recording of an artery and a vein flow, the aorta and the superior vena cava respectively (fig 2). Using these methods, we established the following types of SVT: AF, SVT with short VA interval and SVT with long VA interval.

**Fig 1.** a) M mode recording of a fetus with SVT – The M mode cursor line intersects the right ventricle and right atrium. The atrial contractions are in the superior part of the image and the ventricular contractions are in the inferior part of the image. The heart rate is over 200 bpm; b) M mode recording of a fetus with SVT – M mode recording of a SVT. The atrial contractions are in the inferior part of the image and the atrial rate is 225 bpm. The ventricular contractions are in the superior part of the image and the rate is 222 bpm.

**Fig 2.** a) The view with the aorta and the superior vena cava which allows simultaneous recording of an artery and a vein; b) Pulsed Doppler aspect in the aorta and superior vena cava during sinus rhythm. Below the baseline the atrial activity is recorded (atrial revers in the superior vena cava) and ventricular activity (aortic flow velocity).

**Fig 3.** a) M – mode recording of a fetus with AET. The heart rate decreases progressively (“the cooling phenomenon”); b) Pulsed Doppler of the aorta and superior vena cava in a fetus with AET. Few atrial contractions are blocked.

**Fig 4.** a) Pulsed Doppler of the aorta and superior vena cava in a fetus with regularSVT, with a long VA interval, more precisely PJRT; b) Pulsed Doppler of the aorta and superior vena cava in a fetus with irregular SVT, with a long VA interval, respective AET; c) Pulsed Doppler of venous ductus in a fetus with SVT. The waveforms are monophasic with reversal flow.
Table I. The fetus group with supraventricular tachycardia

<table>
<thead>
<tr>
<th>Type of SVT</th>
<th>Number of cases</th>
<th>Age of gestation (weeks)</th>
<th>Answer to therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVRT</td>
<td>6</td>
<td>22–24</td>
<td>5 cases – intrauterine therapy with favorable answer; 34</td>
</tr>
<tr>
<td>AET</td>
<td>2</td>
<td>24</td>
<td>1 case – intrauterine therapy with favorable answer; 34</td>
</tr>
<tr>
<td>PJRT</td>
<td>2</td>
<td>29, 32</td>
<td>27, 30</td>
</tr>
</tbody>
</table>

SVT, supraventricular tachycardia; AVRT, atrioventricular reentry tachycardia due to the accessory pathway; AET, atrial ectopic tachycardia; PJRT, permanent junctional reciprocal tachycardia.

Finally, the difference between the two types of SVT with long VA interval, AET and PJRT respectively, was realized by influencing the atrioventricular conduction of the fetal heart. The atrioventricular conduction can be influenced through a vagal maneuver by increasing the abdominal pressure during echocardiography. Similar effect on atrioventricular conduction could be obtained by digoxin administration. In AET this maneuvers decreased the heart rate progressively (fig 3a) or blocked the atrioventricular conduction with appearance of uncontacted P waves (fig. 3b), while in PJRT no modification was detected.

In order to establish the most accurate diagnosis, we followed the next steps: determine the heart anatomy, define the atrial and ventricular rate, establish the permanent or intermittent aspect, establish the regular or irregular character (fig 4b), define the VA interval and determine the answer to the decrease of the atrioventricular conduction (Table I.). Also, it is very important to analyze the hemodynamic status of the fetus by checking the flow in venous ductus (fig 4c).

Discussions

Tachycardia with an elevated heart rate of about 200-400 beats/min, with regular aspect and short VA interval is well-known as AVRT [4,5]. The diagnosis of flutter was established in the case of a very high atrial rate, of about 300-400 beats/min, a low ventricular rate, and a relatively regular blocking of atrioventricular conduction (2:1, 3:1, 4:1). Meanwhile, in the case of a tachycardia with a long VA interval, the possible diagnosis is PJRT or AET [6,7]. The presence of an irregular aspect, “warming and cooling” phenomena or blocked atrial contraction confirms the diagnosis of AET, while a continuous tachycardia with a regular aspect confirms the diagnosis of PJRT [7]. Very often, the differential diagnosis of these two types of SVT is difficult to be made [8]. Because of the different evolution and prognosis of these two types of SVT in the postnatal period, we consider that it is important to establish an adequate diagnosis as soon as possible.

The method used for establishing the diagnosis of SVT is different during the intrauterine period in comparison to the postnatal period, being also more difficult during the intrauterine life. The first report on SVT diagnosed using M mode was made by Kleinman et al in 1980 and the measurement of VA interval in M mode was published eight years later by Jaeggi et al [3,9]. Later, in 2000 it was described the simultaneous PW recording of superior vena cava and aorta in sheep fetus [10]. In the same year, Fournon et al proved the superiority of the Doppler tracing in comparison with M mode regarding the diagnosing of SVT in fetus [11]. The prenatal diagnosis difficulty of the SVT is augmented by the decreased contractility, velocity changing at the vessels level which, leads to an increased difficulty in interpreting the images both in M mode and Doppler. Therefore, a series of recordings are most required in order to obtain an accurate diagnosis.

In the following years, multiple studies were focused on this topic. They described prenatal therapeutic protocol depending on the tachycardia type. Thus, the diagnosis of the type of SVT was established based on the VA interval. Most of the studies agreed on a SVT with a short, respectively long VA interval [8,11,12]. SVT with a short VA interval responds very well to antiarrhythmic therapy and offers a long standing favorable prognosis. Regarding SVT with a long VA interval, the therapeutic answer is variable and the prognosis is different depending on the type of SVT with long VA interval: AET or PJRT. The distinction between these two SVT types during the intrauterine life is difficult, both types representing a tachycardia with a continuous aspect and a risk of developing dilated cardiomyopathy. Slightly irregular aspect and blocked atrial contractions are diagnostic elements for AET. Although the PJRT diagnosis is difficult to be established before birth [13], there are a few suggestive elements for this type of SVT: a continuous, regular tachycardia with a long VA interval and with dilated cardiomyopathy aspect.
SVT has side effects on fetal heart and blood flow causing decreased contractility, atrioventricular valve regurgitation, increased central venous pressure followed by hydrops [14]. Central venous pressure, the main cause of hydrops, can be established by the PW interrogation of venous duct, more precisely by demonstrating the presence of a holodiastolic flow reversal [15,16]. PW of venous duct is important for both the estimation of the risk of hydrops and the therapy efficiency.

These aspects of fetal pathology continues to be an important subject worth studying. A recent published study shows a new method for establishing the electrophysiological diagnosis of fetal tachyarrhythmia, by magnetocardiography, proving how complex a fetal tachyarrhythmia can be [17].

Although, many studies have been published in literature regarding the management of fetal tachyarrhythmia, a therapeutic guideline for fetal tachyarrhythmia remains unknown [18]. In this situation an accurate diagnosis of the type of SVT can help us to choose the proper therapy guided by our experience in the treatment of postnatal arrhythmia.

Despite the small number of cases reported, we were able to show that using a protocol of examination with M mode, pulsed Doppler in the aorta and superior vena cava and completing these well-known methods by adding the decrease of the atrioventricular conduction, it is possible to obtain an accurate diagnosis.

In conclusion we can say that the influence of vagal stimulation, digoxin administration, or both on the atrioventricular conduction of the fetuses with SVT can provide important information in order to establish an accurate diagnosis of the type of SVT.

Conflict of interest: none

References