CASE REPORT

Supraventricular microreentry in a newborn due to a giant atrial septum aneurysm

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Key Clinical Message
Supraventricular arrhythmias in neonates are rare and mostly not related to structural heart defects. We present the first case of a newborn with a supraventricular microreentrant tachycardia possibly associated with an atrial septum aneurysm and emphasize the importance of a thorough diagnostic workup allowing a fast and adequate therapy.

Keywords
Aneurysmatic atrial septum, atrial flutter, ectopic atrial tachycardia, microreentry tachycardia, newborn, supraventricular arrhythmia.

Introduction
Supraventricular tachycardias (SVT) occur in 13/100,000 children under 19 years of age [1]. They can be subdivided according to the substrates of the tachycardia, like atrioventricular node-dependent tachycardias (e.g., atrioventricular reentrant tachycardia (AVRT), AV node reentrant tachycardia (AVNRT)) or cavotricuspid isthmus-dependent tachycardias (e.g., atrial flutter [Aflu]). Another subdivision is, according to the underlying mechanism, roughly divided into reentry versus automatic foci (typically ectopic atrial tachycardia [EAT]) [2].

The most prevalent form of SVT in children is a pathway-mediated AVRT. Atrial tachycardia in which the primary electric disturbance is restricted to the atrial tissue, as in EAT or atrial flutter, occurs in only 14% of SVTs and is mostly not correlated to congenital heart disease [3]. The underlying basis for all reentrant tachycardias is a reentry circuit mediated via different anatomic substrates. In microreentries, these are areas of functional conduction block, as occurring in adjacent tissues with differing refractory periods. Rather than from reentry, primary atrial tachycardias can be due to localized automatic foci. A subdivision in foci due to abnormal automaticity and microreentry foci is complex but in children the tachycardia is typically automatic in nature and a focal microreentry tachycardia (FMRT) is very rare [1]. Atrial fibrillation is an exception in children, especially neonates, and therefore is disregarded here. The symptoms range from dyspnea, pallor, refusal of enteral feeding, sweating and agitation to cardiac dysfunction, and heart failure, but it can also be asymptomatic. The treatment depends on the mechanism of the arrhythmia and the clinical state of the patient. It varies from pharmacological therapy to transesophageal overdrive pacing to electric cardioversion or catheter ablation and cardiac arrhythmia surgery. The choice of drug varies significantly between centers, and as there are many effective options available today, the choice of drug is often determined by the physician’s experience [1]. In acute settings with a hemodynamic instable patient, electric cardioversion with 0.25–2 J/kg remains as an option of rescue therapy to end the SVT to avoid progressive ventricular dysfunction [4]. In general, SVT’s in young children with a structurally normal heart are often self-limited and do not need any other treatment despite the initial conversion. If relapses occur, an
electrophysiological study and catheter ablation or drug prophylaxis could be necessary.

There are few cases of adults with an atrial septum defect and atrial flutter as a form of atrial reentrant tachycardia but to the author’s knowledge this is the first report of a newborn with a FMRT possibly associated with a giant atrial septum aneurysm [5].

Case Presentation

A 13-days-old male patient was referred to our center from a primary care hospital due to recurrent tachycardia (240 bpm) in a stable clinical condition. The antenatal history was uneventful and primary cesarean section was performed after 36 + 4 weeks of gestation due to a pathologic cardiotocography result showing fetal tachycardia. Postnatal adaptation was unremarkable.

In our center, supraventricular reentrant tachycardia with a heart rate of 240 bpm was diagnosed after application of adenosine (0.2 mg/kg, Fig. 1A). Echocardiographically an atrial septal defect (ASD) with a giant atrial septum aneurysm, that took up about 1/2 to 2/3 of the right atrium, prolapsing into the tricuspid valve, and a very small muscular ventricular septum defect (VSD) were diagnosed; cardiac function was normal (Fig. 1B). As the SVT could not be terminated by the use of transesophageal overdrive pacing, we opted for an electric cardioversion with 5 J (2 J/kg), which was successful. After 3 h, the patient showed a relapse, which again could be terminated with an electric cardioversion (6 J). Therefore, we started the substitution of digoxin (blood serum level: 0.6 ng/mL)

Figure 1. (A) ECG showing the initial SVT with heart rate of 240 bpm and the FMRT after using adenosine. (B) Echocardiogram showing the ASD and atrial septum aneurysm.
and sotalol (112 mg/m²). With this therapy the patient remained in a stable sinus rhythm and free from further relapses. The patient was discharged in good clinical condition after 4 days.

In a follow-up after 6 months and 1 year, the patient was in good clinical condition without the history of any relapses. After 6 months, the medication has been discontinued. The giant septum aneurysm showed distinct regression in its dimensions.

**Discussion**

Supraventricular tachycardias occur in less than one percent of children and have various pathophysiologic mechanisms [1, 6]. To identify the underlying mechanism of the SVT, it is advisable to proceed step by step (Fig. 2).

Identification of the underlying tachycardia mechanism succeeds with adenosine that can help distinguish between intraatrial reentrant tachycardia and AV node-dependent tachycardias (AVRT/AVNRT), as the former will not be terminated by the use of adenosine. In the presented case, the differential diagnoses of a SVT not terminating after the use of adenosine are therefore Aflu, Intraatrial reentrant tachycardia (IART), microreentrant tachycardia, and EAT. The typical atrial flutter is a stable macroreentry capturing the atria in a clockwise or counterclockwise manner in between the normal anatomic boundaries [4]. In the case of an underlying congenital heart disease, the boundaries of the SVT are more unspecific and therefore many reentrant patterns, than called IART, may occur. The constant depolarization of the myocardium leads to the typical saw-tooth pattern. In contrast, a microreentrant circuit leads to simple depolarization of the atrium with isoelectric phases in between. Atrial flutter could be ruled out in our patient by the use of adenosine that revealed isolates p-waves, instead of the typical saw-tooth pattern. In contrast to an EAT which typically shows changes in tachycardia cycle length, a FMRT has a strictly fixed cycle length. Additionally, it presented as a benign condition and could easily be treated with cardioversion, showing that it is not an ectopic atrial tachycardia, as an ectopic focus normally does not respond to cardioversion (Fig. 2). A FMRT is a rare phenomenon and there has not been a lot of research concerning the pathomechanism of this arrhythmia in children. Studying the electrocardiogram (ECG) of our patient, we assume that there is

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**Figure 2.** Diagnostic approach of differentiating SVTs. The red dotted lines indicate a termination of the SVT via adenosine; the green boxes and dotted lines refer to a termination of the SVT via electric cardioversion. AVNRT=atrioventricular nodal reentry tachycardia, AVRT=atrioventricular reentry tachycardia, Aflu=atrial flutter, IART= intraatrial reentry tachycardia, EAT=ectopic atrial tachycardia, * some EAT’s are terminable via electric cardioversion.
a correlation between the giant atrial septum aneurysm and the focal microreentrant tachycardia, as positive p-waves can be seen in lead II, III, aVF, and V1-4, making a septal origin of the focal ectopic reentry likely. In addition, the tissue of the giant aneurysm will possibly represent an area with an altered refractory period compared to the adjacent atrial tissue. A similar, although not the same, phenomenon can be seen in cases of atrial fibrillation at the transition of the pulmonary veins to the left atrium. This anatomic structure may therefore represent a zone of slow conduction as a substrate for the FMRT. This theory may also explain the disappearance of the arrhythmia after regression of the atrial aneurysm. Yet, it has to be mentioned, that the disappearance of the FMRT after discontinuation of the antiarrhythmic drugs may also be a spontaneous resolution, as is frequently seen in other supraventricular arrhythmias in infants.

Atrial septum aneurysms occur in 4.9% of children and are a rare malformation that may occur isolated or in combination with other congenital heart defects with the potential of spontaneous regression [7]. Calderón Colmenero et al. described two cases of neonates with atrial septum aneurysms and SVTs: one with supraventricular extrasystoles with aberrant conduction and one with atrial flutter but the relationship of the arrhythmia and the aneurysm is discussed controversial.

In our case, a relationship between the two can be assumed, regarding the position of the atrial septum aneurysm, the p-wave morphology and the suspected mechanism described above. This is the first case describing a possible correlation between an atrial microreentry tachycardia and an atrial septum aneurysm in a neonate.

In neonates with SVTs, an echocardiography is crucial to rule out underlying congenital heart diseases and to determine the heart function prior to antiarrhythmic drug treatment, which is often negative inotropic. Especially, if the SVT is atypical, one should become alert. Particularly, if children are relatively asymptomatic, it can delay the diagnosis and therefore may lead to severe cardiac dysfunction.

**Conclusion**

This is the first report of a newborn with a focal atrial microreentrant tachycardia, assumingly related to a giant atrial septum aneurysm. The tachycardia was effectively treated by cardioversion and medicinal prophylaxis with digoxin and sotalol.

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**Authorship**

FW: resident assigned to patient during hospitalization and participated in review of the literature, drafting and editing of case report. RG: head of rhythmologic department performed critical revision approval. RW: performed critical revision and approval. CP: senior physician participated in critical revision of case report and final approval.

**Conflict of Interest**

None declared.

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