

Sinoatrial nodal disease presenting with tachy-bradycardia syndrome in a fetus of anti-SSA/SSB-positive mother

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ABSTRACT

Tachy-bradycardia syndrome (TBS) has been reported rarely in the fetus. We present here an unusual dominant involvement of sinoatrial node in a fetus presenting with TBS of anti-Sjögren's syndrome-related antigen A (SS-A) and antigen B (SS-B) autoantibodies positive mother. Prenatal imaging findings, Doppler hemodynamics, and outcomes are described.

Keywords: Atrial arrhythmia, autoimmune associated tachy-bradycardia syndrome, fetus

INTRODUCTION

Tachy-bradycardia syndrome (TBS) is defined as an atrial arrhythmia formed by the alternation of various atrial tachycardias with sinus bradycardia or sinoatrial block.^[1] It has been reported in fetuses with the missense variant p.(Gly482Arg) in HCN4 gene.^[2] Atrial ectopics and junctional ectopic tachycardia were reported in fetuses with autoimmune atrioventricular block.^[3] An unusual dominant involvement of sinoatrial node in a fetus presenting with TBS of anti-SSA/SSB-positive mother is presented in this report.

CASE REPORT

A 22-year-old primigravida was referred for fetal echocardiography at 24 weeks' gestation given suspicion of fetal tachyarrhythmia on screening. The four-chamber view showed moderate pericardial effusion, hyperechoic endocardium, papillary muscles, and both atrioventricular valve annuli [Figure 1a and b]. The heart was otherwise structurally normal. There was ventricular diastolic dysfunction as evident in the left ventricle inflow-outflow Doppler (presystolic

flow in ascending aorta) [Figure 2a and b)] with no atrioventricular valve regurgitation.

Left ventricle inflow-outflow Doppler showed periods of fetal sinus bradycardia [Figure 3] alternating with frequent conducted atrial ectopics triggering episodic atrial tachyarrhythmia. Left brachiocephalic vein-aorta Doppler showed the beginning and termination of the tachyarrhythmia. It was a 1:1 long

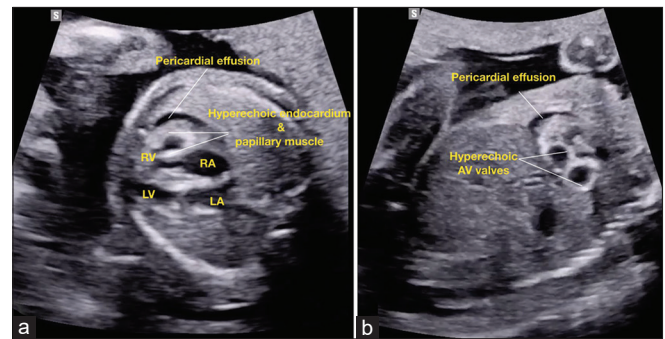


Figure 1: (a) Four-chamber view showing hyperechoic endocardium, papillary muscle, and pericardial effusion. (b) Sagittal view showing hyperechoic atrioventricular valves

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VA tachyarrhythmia (VA interval 600 ms, AV interval 267 ms) triggered by an atrial ectopic, and spontaneous termination of the tachyarrhythmia followed by an atrial ectopic and bradycardia [Figure 4]. Hence, a diagnosis of TBS was made. There was no evidence of atrioventricular block. Maternal serum SSA and SSB antibody levels were elevated 145 U (normal <20) and 181 U (normal <20), respectively. There was no family history of sinus node dysfunction.

In utero course

Transplacental therapy with oral dexamethasone at a dose of 4 mg/day was given initially for 7 days. Reassessment after 1 week showed absent pericardial effusion and improved ventricular diastolic function [Figure 5]. However, fetal bradycardia, conducted atrial ectopics, and episodic atrial tachycardia persisted.

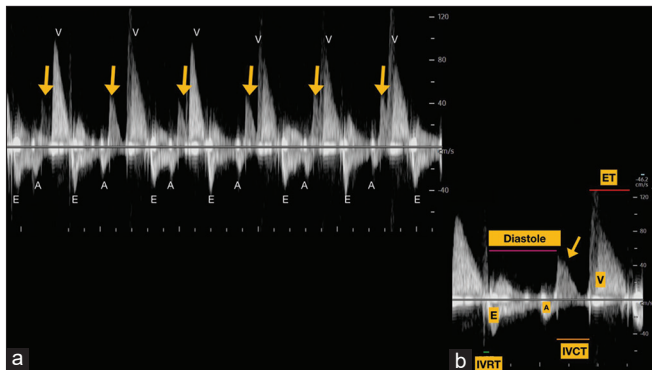


Figure 2: (a) Left ventricle inflow-outflow Doppler showing presystolic forward flow into the aorta (yellow arrow). This finding suggests increased stiffness of the left ventricle due to endocardial inflammation resulting in a significant increase in left ventricular pressure during left atrial contraction leading to presystolic forward flow into the aorta. (b) Left ventricle inflow-outflow Doppler showing forward flow into the aorta (yellow arrow) during the isovolumetric contraction time

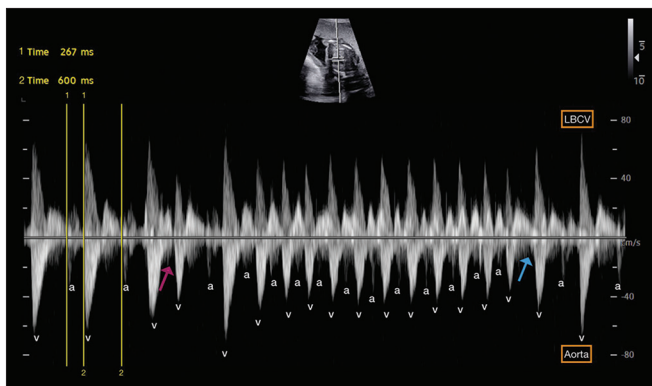


Figure 4: Left brachiocephalic vein-aorta Doppler demonstrating the beginning of the tachyarrhythmia triggered by an atrial ectopic (pink arrow) and termination of the tachyarrhythmia followed by an atrial ectopic (blue arrow) and bradycardia. It is a long ventriculoatrial tachyarrhythmia (atrioventricular interval 267 ms; ventriculoatrial interval 600 ms) with 1:1 atrioventricular conduction

Hence, oral dexamethasone dose was increased to 8 mg/day. Reassessment after a week showed no further improvement.

DISCUSSION

Fetal cardiac failure due to arrhythmias is associated with high fetal and neonatal mortality and affects the long-term neurodevelopmental outcome.^[4] Tachyarrhythmias has been reported in fetuses with complete heart block. The reported tachyarrhythmias in the fetal literature include junctional ectopic tachycardia, atrial flutter, and ventricular tachycardia.^[5-7] To the best of our knowledge, immune-mediated TBS is not reported in fetuses so far. Presystolic flow in ascending aorta has been reported in adults with ventricular diastolic dysfunction. It occurs due to impaired relaxation of the left ventricle resulting in decreased filling of left ventricle in early diastole which is compensated by

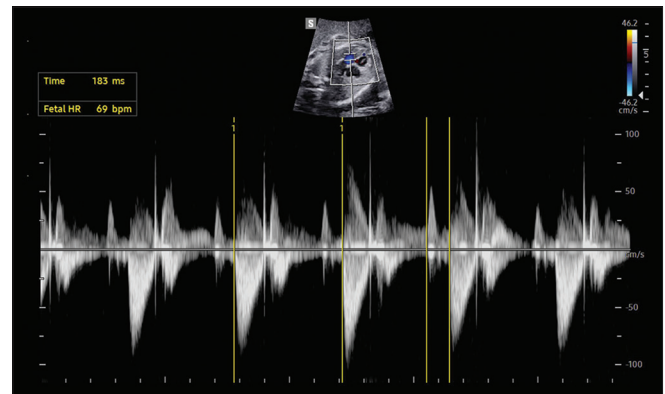


Figure 3: Left ventricle inflow-outflow Doppler showing sinus bradycardia with atrioventricular interval of 183 ms

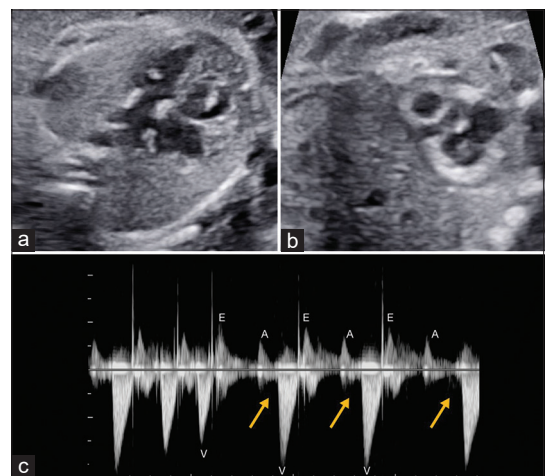


Figure 5: (a) Four chamber view and (b) Sagittal view of the fetal heart showing disappearance of pericardial effusion after steroid therapy. (c) Left ventricle inflow-outflow Doppler showing disappearance of presystolic forward flow into the aorta suggestive of improved diastolic function and persistence of tachycardia-bradycardia

forceful contraction of the left atrium to complete left ventricle filling.^[8] Immune-mediated SA node injury has discordances between the echocardiographic and pathologic findings.^[9]

Hemodynamic evaluation using Doppler echocardiography helps in understanding the electrophysiological mechanism and to make an accurate diagnosis of fetal arrhythmias.^[10] In this case, Doppler echocardiography helped in understanding both the arrhythmia mechanism and cardiac function. Although the cardiac function stabilized after transplacental therapy with oral steroids, the arrhythmia persistent probably due to immune-mediated fibrosis of the sinoatrial node and the atrium. No pharmacological therapy has yet resulted in permanent reversal of fetal third-degree heart block due to maternal lupus. However, transplacental therapy with fluorinated steroids has some efficacy in treating second-degree heart block and cardiac disease beyond the atrioventricular node and beta-agonists have been used to increase fetal heart rate *in utero*. No controlled experiments regarding the use of plasmapheresis have been performed, and due to the costly and time-consuming process, it does not play a significant role in its management. Intravenous immunoglobulin has shown promise in the treatment specifically associated with fetal cardiomyopathy/endocardial fibroelastosis.^[11] Hydroxychloroquine is a highly beneficial drug in the management of women with autoimmune disease and now has a new role for the prophylaxis of recurrent fetal heart block in which it halved the rate of development. The risk of heart block in a fetus exposed to maternal anti-SSA/Ro antibodies is approximately 2% if the mother has never had an affected child and 18% if she has. Hydroxychloroquine 400 mg daily initiated at or before 10 weeks gestation is associated with a recurrence rate less than one-half that of historical control subjects.^[12]

CONCLUSION

Analysis of the arrhythmia mechanism is very important to assess the degree of conduction tissue involvement by the disease process and will aid in prognostication and decision regarding maternal transplacental therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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