Sinoatrial nodal disease in a Fetus presenting with tachy-bradycardia syndrome of Anti-SSA/SSB-Positive Mother.

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Abstract

Atrial, sinoatrial node and atrioventricular node disease with preserved conductivity results in tachy-bradycardia syndrome.(1) Tachy-bradycardia syndrome has been reported in fetuses with the missense variant p.(Gly482Arg) in HCN4 gene.(2) Atrial ectopics and junction ectopic tachycardia were reported in fetuses with autoimmune atrioventricular block.(3,4) An unusual dominant involvement of sinoatrial node in a fetus presenting with tachy-bradycardia syndrome of Anti-SSA/SSB-Positive mother.

Case report :

A 22-year-old primigravida was referred for fetal echocardiography at 24 weeks gestation in view of suspicion of Fetal tachyarrhymia on screening. The 4-chamber view showed moderate pericardial effusion, hyperechoic endocardium, papillary muscles and both atrioventricular valve annuli. **(Figure 1A&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&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 **(Figure 4)** triggering episodic atrial tachyarrhythmia.**(Figure 5)** . Left brachicephalic vein-aorta Doppler showed the beginning **(Figure 6)** and termination of the tachyarrhythmia**(Figure 7).** It was a 1:1 long VA tachyarrhythmia (VA interval 175ms ; AV interval 92ms) triggered by an atrial ectopic and spontaneously terminating with V. **(Figure 8)** Hence a diagnosis of tachy-bradycardia syndrome secondary to atrial, sinoatrial node and atrioventricular node disease with preserved conductivity was made. There was no evidence of atrioventricular block. Maternal serum SSA and SSB antibody levels were 145 U (normal <20) and 181 U (normal <20) respectively. In-utero course : Transplacental therapy with oral dexamethasone at a dose of 4mg/day was given initially for 7 days. Reassessment after one week showed absent pericardial effusion and improved ventricular diastolic function. However fetal bradycardia, conducted atrial ectopics and episodic atrial tachycardia persisted. Hence oral dexamethasone dose was increased to 8mg/day. Reassessment after a week showed no improvement. The parents decided for termination of pregnancy.

Discussion :

Tachyarrhythmias has been reported in fetuses with complete heart block. The reported tachyarrhythmias were junctional ectopic tachycardia, atrial flutter and ventricular tachycardia.(3,5,6) Immune-mediated atrial inflammation and atrial dilatation secondary to myocardial dysfunction can result in atrial flutter. Ventricular tachycardia occur as a result of abnormal ventricular repolarization. Immune-mediated focal necrosis and fibrosis in the AV node can result in junctional ectopic tachycardia.(7) To our knowledge, immune-mediated tachy-bradycardia syndrome is not reported in fetus so far. Presystolic flow in ascending aorta has been

reported in adults with ventricular diastolic dysfunction. It occurs due to impaired relaxation of left ventricle resulting in decreased filling of left ventricle in early diastole which is compensated by forceful contraction of 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 to understand both the arrhythmia mechanism and cardiac function. Though the cardiac function stabilized after transplacental with oral steroids, the arrhythmia persistent probably due to immune-mediated fibrosis of the sinoatrial node and the atrium.