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Fetal atrioventricular heart block as a first manifestation of maternal connective tissue disease: a case report

Milhano JC⁽¹⁾, Ladislau F⁽²⁾, Rato JP⁽³⁾, Noronha N⁽³⁾, Teixeira AM⁽³⁾, Landim E⁽²⁾, Matos T⁽²⁾.

⁽¹⁾ Obstetrics Department, Unidade Local de Saúde Lisboa Ocidental; ⁽²⁾ Obstetrics Department, Unidade Local de Saúde Amadora/Sintra; ⁽³⁾ Pediatric Cardiology, Unidade Local de Saúde Lisboa Ocidental.

Objective: Most cases of congenital atrioventricular heart block (AVHB) without major structural abnormalities diagnosed in utero are associated with maternal anti-Ro/SSA antibodies. We present a case of AVHB as a first manifestation of maternal connective tissue disease.

CLINICAL CASE



A 31-year-old nullipara, with no previous significant medical history, was referred to an obstetrics appointment at her local hospital due to 1st trimester gestational diabetes. A **fetal echocardiogram** was then requested in our centre.



21 weeks: Normal FHR + frequent supraventricular extrasystoles ? + suspected aortic subvalvular membrane



2 weeks



Fetal bradycardia: 2:1 atrioventricular heart block

Ventricular rate 71 bpm + atrial rate 150 bpm
Mild pericardial effusion

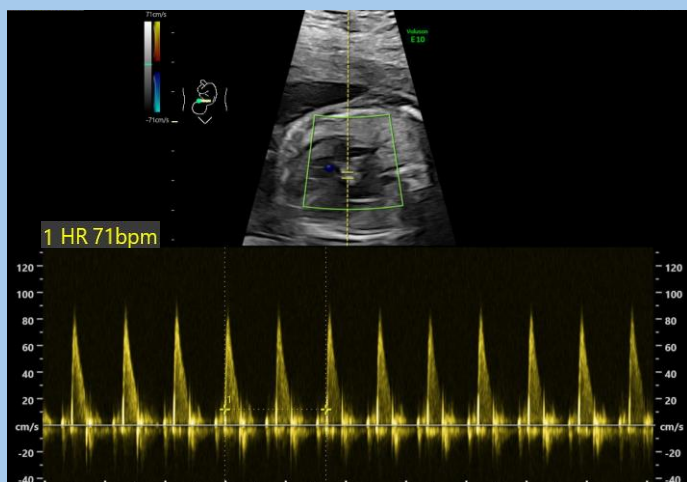


Image 1 – Ventricular heart rate at GA 23w4d.

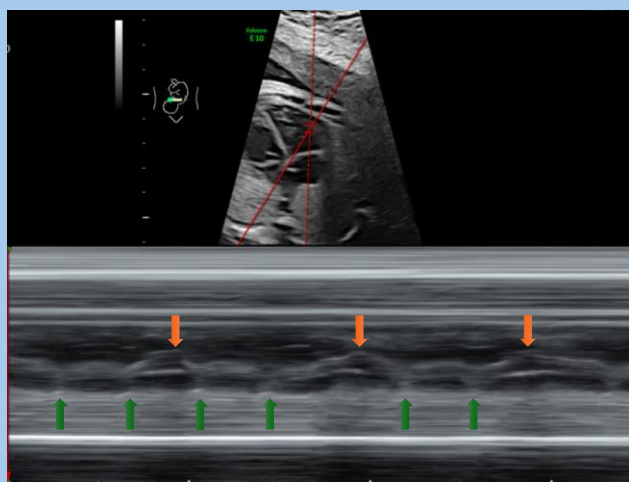


Image 2 – M-mode demonstrating 2:1 AV heart block.
Green arrows: atrial systole. Orange arrows: ventricular systole.



Autoimmune disease panel:
positivity for **anti-Ro/SSA** and **anti-La/SSB** antibodies



Dexamethasone 6 mg id *per os*
at 26 weeks



36-weeks pregnancy: stable FHR with no progression of the heart block and a reduction in pericardial effusion volume. Insulin therapy due to poor metabolic control.

Conclusion: The cardiac manifestations of neonatal lupus are very rare and usually detected in pregnant women with previously diagnosed anti-Ro/SSA and/or anti-La/SSB antibodies. It is highly unusual for the diagnosis of maternal connective tissue disease to be made with fetal AVHB as a first manifestation. The detection of heart block at earlier stages prompts early intervention, which is thought to improve outcomes.