



Fetal Atrioventricular Block Leading to the Diagnosis of Maternal Sjögren’s Syndrome: A Case Report



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Introduction

Fetal bradycardia may be the first clinical sign of an underlying maternal autoimmune condition. Anti-Ro/SSA and anti-La/SSB antibodies can cross the placenta and damage the fetal atrioventricular (AV) node, resulting in congenital heart block (CHB). This is the most severe manifestation of neonatal lupus syndrome, occurring in 1–5% of fetuses exposed to maternal anti-Ro/SSA antibodies. CHB may present in isolation or be associated with fetal cardiac dysfunction or hydrops¹.

The pathogenesis involves passive transplacental transfer of maternal IgG autoantibodies that bind to fetal cardiac conduction tissue, triggering an inflammatory response. This can lead to fibrosis and irreversible disruption of AV conduction. The spectrum ranges from first-degree block to complete atrioventricular block (CAVB)¹.

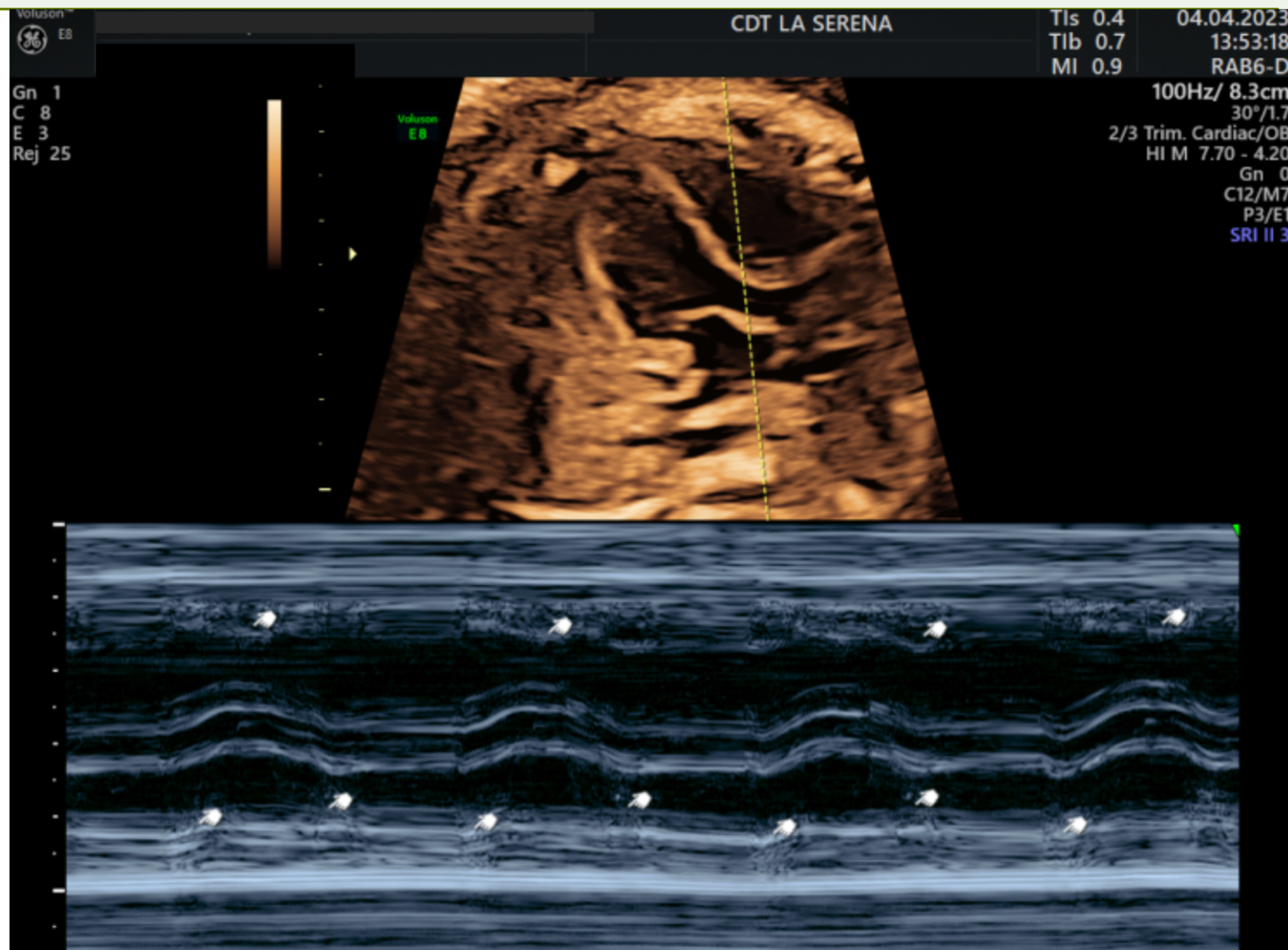
There is no universally accepted treatment protocol. However, several observational studies^{2,3} suggest that early corticosteroid therapy (e.g., dexamethasone) may reduce the risk of progression to complete atrioventricular block in selected cases.

Case presentation

- Patient: 28-year-old woman in her first pregnancy (G1P0)
- Medical history: Hashimoto’s thyroiditis with primary hypothyroidism, on levothyroxine
- Finding: Routine mid-trimester ultrasound revealed fetal bradycardia (around 70 bpm)
- Referral: Transferred to a tertiary care center for evaluation

On Evaluation

- Persistent fetal bradycardia
- No signs of hydrops or heart failure
- Fetal echocardiogram: Second-degree AV block (2:1)
- Maternal labs: Positive anti-Ro/SSA 180 U/mL and anti-La/SSB 30.9 U/mL.
- Further testing: Positive Schirmer’s test and bilateral parotid chronic parotitis: **Sjögren’s syndrome diagnosed**



Delivery and neonatal outcome

- Gestational age: 36 weeks (spontaneous preterm labor)
- Delivery: Cesarean section due to inability to ensure adequate fetal monitoring
- Newborn:
 - Male, 2230 g
 - Apgar scores: 8 and 9 at 1 and 5 minutes
 - Postnatal Holter: Persistent bradycardia ~70 bpm, no episodes <60 bpm
 - Pacemaker was not required.

Follow up

The child has been followed regularly by pediatric cardiology. At 2 years of age, he remains clinically stable with persistent second-degree AV block and bradycardia around 70 bpm. There have been no signs of heart failure, and pacemaker implantation has not been indicated to date. The mother remains clinically stable, however, she has not attended follow-up with a rheumatologist since delivery, underscoring a gap in long-term autoimmune care.

Discussion

- Congenital heart block due to maternal anti-Ro/SSA and anti-La/SSB antibodies occurs in approximately 1–5% of exposed fetuses. Complete AV block is associated with significant morbidity and mortality, with perinatal mortality rates of up to 20%. Nearly 50% of surviving infants require permanent pacemaker implantation within the first year of life³.
- Although evidence is limited, corticosteroid therapy may be beneficial when initiated early in second-degree AV block to prevent progression. Early detection through fetal echocardiography and timely maternal autoimmune screening are essential to optimize outcomes.
- Persistent fetal bradycardia should prompt evaluation for maternal anti-Ro/SSA and anti-La/SSB antibodies. If positive, a comprehensive fetal cardiac assessment is warranted. Early diagnosis and individualized treatment may stabilize conduction and avoid irreversible damage.
- Long-term pediatric cardiology follow-up is critical, as delayed complications may develop even in initially stable infants.

Bibliography

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