

Congenital Complete Atrioventricular Block with Hydrops Fetalis in a Pregnant Woman with Sjögren's Syndrome

A Case Report
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Background

Anti-Ro/SSA and Anti-La/SSB antibodies may cause congenital heart block.

Sjögren’s syndrome is an autoimmune disorder frequently associated with maternal anti-Ro/SSA and anti-La/SSB antibodies. These antibodies can cross the placenta and cause immune-mediated injury to the fetal cardiac conduction system, leading to congenital heart block (CHB). Complete atrioventricular (AV) block is usually irreversible and can be complicated by fetal hydrops and intrauterine fetal demise.

Case

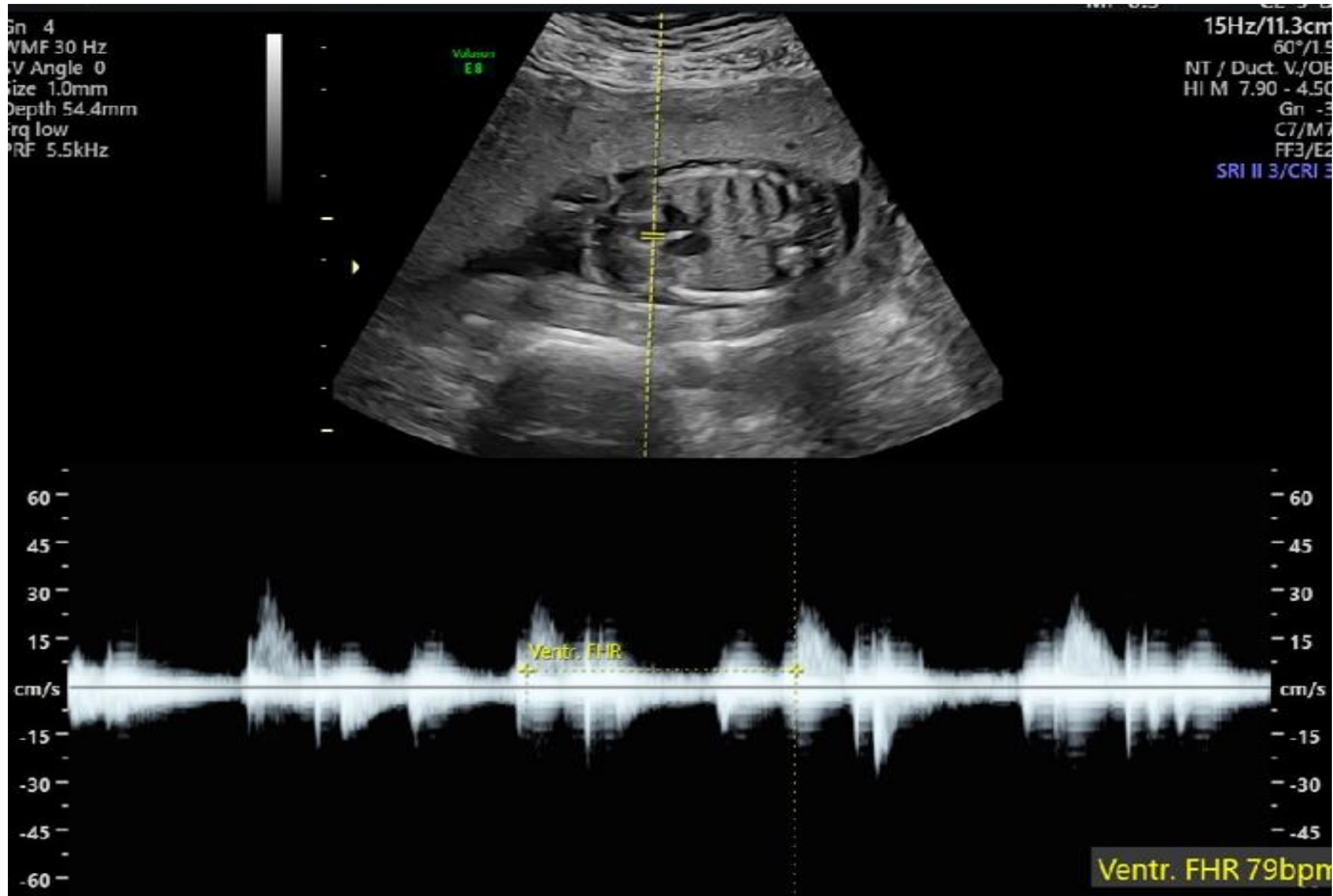
A 35-year-old primigravida woman with known Sjögren’s syndrome, positive for anti-Ro and anti-La antibodies, conceived spontaneously with an estimated due date of January 5, 2026. She had discontinued her rheumatologic medications after confirming pregnancy due to concerns about fetal safety. Until 16+4 weeks of gestation(July 25, 2025), routine ultrasound showed a structurally normal fetus with normal cardiac rhythm.

At 18+2 weeks of gestation (August 6, 2025), follow-up ultrasound revealed severe fetal bradycardia (FHR 79 bpm), complete AV block on Doppler, fetal ascites, and generalized hydrops. These findings were consistent with immune-mediated congenital complete heart block complicated by hydrops fetalis.

Discussion

This case illustrates the abrupt onset of complete AV block between 16–18 weeks of gestation in the setting of maternal Sjögren’s syndrome. Once complete block develops, it is typically irreversible despite maternal corticosteroid or immunomodulatory therapy. The presence of hydrops indicates poor prognosis. Early and continuous monitoring with fetal echocardiography is essential in anti-Ro/SSA and anti-La/SSB positive pregnancies, ideally starting from 16 weeks of gestation.

Ultrasound Image



Conclusions

1. Anti-Ro/SSA and anti-La/SSB positive pregnancies require close fetal cardiac surveillance.
2. Rapid progression to complete AV block can occur during the second trimester.
3. Early fetal echocardiography is critical for timely diagnosis and multidisciplinary management.

Key Message

Weekly fetal echocardiographic surveillance should be considered in anti-Ro/SSA positive pregnancies.