A case of heart block of unknown cause in fetus requiring a pacemaker insertion

Sajnach-Menke M. A., Abu-Harb M.
The Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, United Kingdom

Objective
Introduction: Congenital heart block is usually related to maternal anti-Ro/SSA and anti-La/SSB antibodies or abnormal conduction in congenital cardiac anomaly, such as the left isomerism, congenitally corrected transposition of the great arteries or double inlet left ventricle. Counselling should aim to give the prognosis and the neonatal outcome. This is challenging, particularly in rare cases with unknown etiology.

Methods
Methods: We present a fetus with normal heart and second-degree heart block, in which the mother had negative anti-Ro/SSA and anti-La/SSB antibodies. The baby required a pacemaker insertion at two days of life. Thirty-two years old G2 P2 woman at 28 w. g. was referred for a fetal echocardiogram with concerns regarding fetal arrhythmia. The ultrasound revealed a structurally normal heart, at an atrial rate of 136 bpm, and a ventricular rate of 48 bpm. The cardiac function was normal, and there was no evidence of hydrops. Differential diagnosis included complete or second-degree AV block with 3: 1 conduction. Maternal blood sample for anti-Ro/SSA and anti-La/SSB antibodies was obtained. The result was negative. During counselling, parents were informed that the cause of heart block is unknown, and the baby would likely require a pacemaker insertion immediately after birth. Besides, long QT syndrome had to be excluded. Subsequent scans showed a second-degree AV block with 2: 1 conduction, ventricular rate of 62bpm. We informed parents that the heart rate is acceptable, and that timing of a pacemaker insertion remained unpredictable. At 39 w. g., she underwent an elective caesarean section. A female baby was born in good condition weighing 3220 grams. An ECG confirmed diagnosis of second-degree AV block with 2: 1 conduction, ventricular rate of 62 bpm. On day two of life, the baby deteriorated, and an ECG revealed second degree AV block with 3: 1 conduction, ventricular rate of 39 bpm.

Results
Results: We proceeded with an epicardial ventricular permanent pacemaker insertion. The baby remains clinically stable. On the pacemaker check, the underlying rhythm remains at 50 bpm—no evidence of long QT syndrome in subsequent ECG's.

Conclusion
This is an atypical case of congenital heart block, which proved challenging. This will impact counselling as it was difficult to predict the need for pacemaker insertion.