

## Isolated anti-Ro/La antibody-negative fetal complete atrioventricular block: a case report

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*To the Editor:* A 28-year-old woman (P1G0 at 30 weeks of gestation) was referred to our diagnostic center for an irregular fetal cardiac rhythm. No risk factors were noted and the tests for detecting maternal auto-antibodies (anti-Ro/La, anti-dsDNA, and anti-nuclear antibodies) were negative. The fetal systematic ultrasonic examination was normal. The fetal echocardiogram revealed that the fetal cardiac structure and function were normal; neither hydrops nor any other malformations were detected.

However, a fetal echocardiogram conducted by passing the sampling line through the right atrial wall, ventricular septum, and left ventricular wall (one after another), revealed a complete atrioventricular block (CAVB). The atrial rate, at 130 to 140 beats/min, was regular and faster, while the ventricular rate was 68 beats/min.

Subsequent follow-up revealed that she underwent a cesarean section and delivered a healthy female baby at 38 weeks. The amniotic fluid was contaminated (III degree). No premature rupture of fetal membranes, nuchal cord, or any stifling was observed. The Apgar scores were 9 and 10 at the first and the fifth minute, respectively. An electrocardiogram, taken by the bedside after birth, confirmed the CAVB diagnosis, as the atrial and ventricular rates were 136 and 68 beats/min, respectively [Figure 1]. Two-dimensional ultrasound reconfirmed the normal anatomical structure of the heart; only the ductus arteriosus and the foramen ovale were not closed. The infant and her mother were discharged after 3 days of observation.

Since then, the child has developed well, can play normally, and has a good academic performance. She continues to perform well without any medications and pacemaker. Now aged 9 years, she revisited our center, where echocardiography revealed a normally sized and structured heart; the ventricular rate was 58 beats/min. A 24-h ambulatory electrocardiogram revealed that the average heart rate was 54 beats/min, the total heart rate was 77,631 beats/24 h, the slowest heart rate was 43 beats/min, and the fastest heart rate was 80 beats/min.

Fetal CAVB is a serious and rare form of bradycardia. Presently, around 50% of CAVB cases are caused by congenital heart disease (CHD) and nearly 40% are mediated by immunity. About 10% of CAVB is found no definite etiology.

In CHD patients, the prognosis of CAVB is strongly associated with the type of CHD, depending upon isomerism, corrected transposition of the great arteries (cTGA), and critical pulmonary stenosis. Fetuses with cTGA often have a good prognosis, while a poor prognosis is observed in left isomerism. Pacemaker implantation was needed in 89% cases. Recent studies from Japan reported that a ventricular rate of less than 55 beats/min has significant effects on fetal myocardial dysfunction and fetal hydrops, resulting in high mortality.<sup>[1,2]</sup>

In a retrospective, multi-center study of 175 fetuses with isolated CAVB and maternal auto-antibodies, the risk factors associated with death included a gestational age of less than 20 weeks, a ventricular rate of less than 50 beats/min, fetal hydrops, and an impaired left ventricular function at diagnosis.<sup>[3]</sup> In this study, the presence of more than one of these variables was associated with a ten-fold increase in pre-birth mortality and an independent six-fold increase in neonatal mortality. Two-thirds of the survivors had pacemakers at the age of 1 year and eight children developed cardiomyopathy.

Due to the low incidence of auto-antibody negative fetal CAVB, associated studies are few. The natural prognosis and risk factors remain unelucidated. A multi-center study reported that the fetal and neonatal mortalities did not differ significantly between antibody-positive and -negative pregnancies in those with a known outcome at 1 month of age.<sup>[3]</sup> However, another multi-center study reported favorable long-term outcomes of congenital, non-immune, isolated atrioventricular (AV) block; no patient died or developed dilated cardiomyopathy and pacemaker-related complications were few.<sup>[4]</sup> In this case, the child

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10.1097/CM9.0000000000000581

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Chinese Medical Journal 2020;133(1)

Received: 31-08-2019 Edited by: Ning-Ning Wang

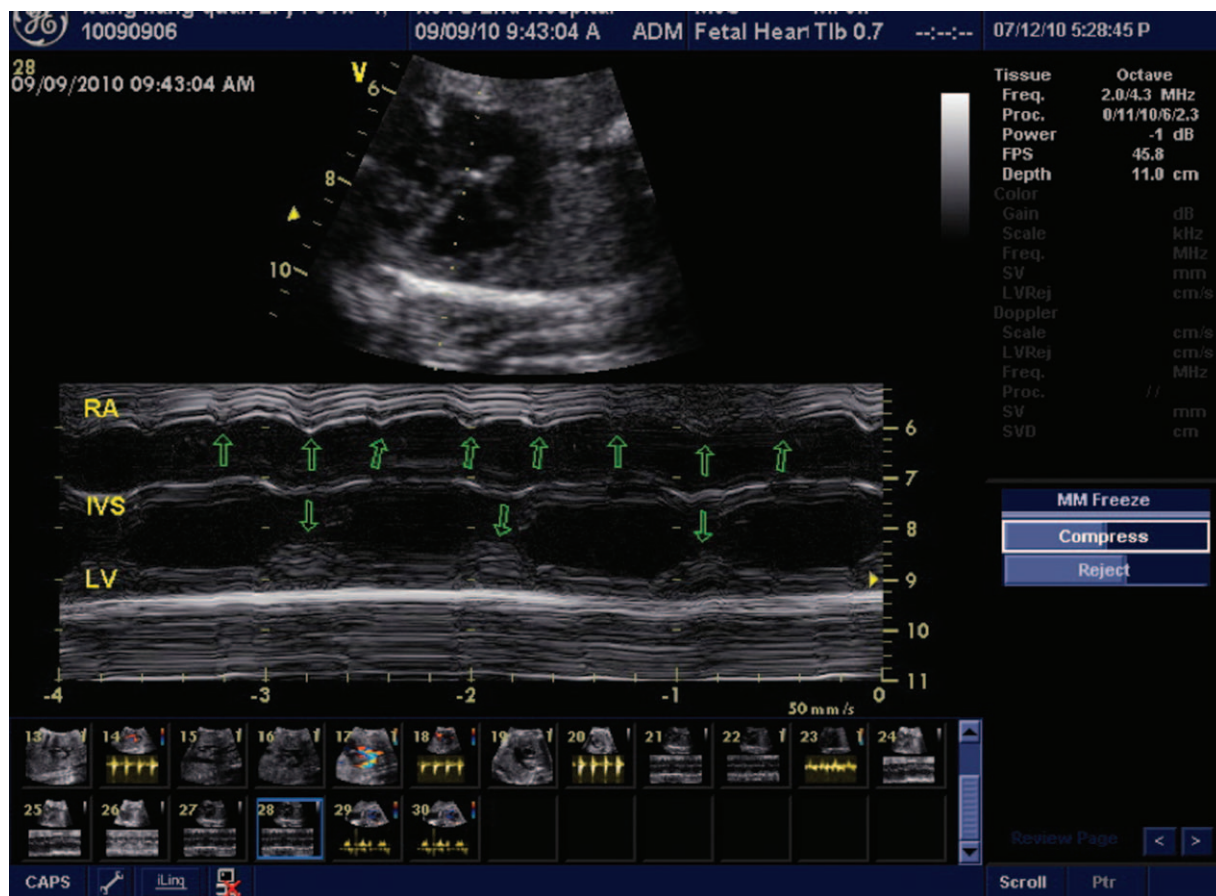


Figure 1: Fetal echocardiogram showed a complete atrioventricular block.

with isolated, auto-antibody negative CAVB is alive, with no symptoms including syncope, fatigue, and heart failure as of yet. It is noteworthy that the child performed well without any medication or pacemaker. Her heart rate ranged around 68 beats/min at the last follow-up. The exact reason for this phenomenon remains unclear; it may be related to the relatively higher ventricular rate (55 beats/min or greater), which we inferred was associated with its high pacing site. In an observational study of nine anti-Ro/La-negative cases (similar to this case), three cases with a heart rate of less than 50 beats/min died, regardless of timely pacemaker implantation.<sup>[5]</sup> Thus, based on our findings, if the fetus presents with seronegative CAVB and the heart rate is more than 55 beats/min, the prognosis may be favorable.

In conclusion, unlike previously reported studies on pediatric auto-antibody positive cases, our study, which focuses on an auto-antibody negative pediatric case, is rare. Furthermore, we observed a relatively longer follow-up as well as a better outcome. Existing literature has revealed that risk factors influencing the prognosis of isolated CAVB include a gestational age of less than 20 weeks, ventricular rate lower than 50 beats/min, fetal hydrops, and impaired left ventricular function. We speculate that the heart rate is a key factor in deciding the prognosis of fetal auto-antibody negative CAVB, which we believe is related to the relatively high pacing site of the heart.

### Conflicts of interest

None.

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How to cite this article: Huo HH, Wei YJ, Qi YH, Liu BM. Isolated anti-Ro/La antibody-negative fetal complete atrioventricular block: a case report. *Chin Med J* 2020;133:97–98. doi: 10.1097/CM9.0000000000000581