

Recurrent second-trimester intrauterine fetal death due to undiagnosed atrioventricular block: A case report

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Keywords: Atrioventricular block, hydrops fetalis, intrauterine fetal death, heart failure

Abstract

Fetal cardiac abnormalities are one of the common causes of non-immune fetal hydrops. Early diagnosis and treatment may prevent the late consequences that can occur as heart failure and intrauterine fetal death. Herein we report the case of a 32-year-old patient with a history of recurrent second trimester intrauterine fetal death. She presented with fetal hydrops at 23 weeks. A detailed echocardiography revealed that the fetus had a third degree atrioventricular block and advanced hydropic changes due to heart failure. Corticosteroid therapy was started but the fetus died in utero after 2 weeks.

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Introduction

Hydrops fetalis is defined as abnormal fluid accumulation in fetal extravascular compartments and body cavities leading to edema, ascites, pleural and pericardial effusions. It is classified into

2 groups; immune and non-immune hydrops fetalis, with the latter representing more than 80% of the described causes of hydrops fetalis.¹ Perinatal mortality in hydrops fetalis is high unless the original cause is recognized and treated properly either intrauterine or extrauterine.²

The most common cause of non-immune hydrops (NIH) is fetal cardiac anomalies.³ Fetal bradyarrhythmia is one of the documented causes of NIH.⁴ It is a rare but serious disease that leads to fetal heart failure without appropriate management. An improvement in fetal echocardiographic equipment (M-mode echocardiogram and Doppler 5) has enabled accurate intrauterine identification of such arrhythmia.

Herein, we report the first case of recurrent intrauterine fetal death (IUFD) in the second trimester pregnant female due to heart failure secondary to fetal atrioventricular block.

Please cite this paper as: Abbas AM, Ali SEA, Michael A. Recurrent second-trimester intrauterine fetal death due to undiagnosed atrioventricular block: A case report. Proc Obstet Gynecol. 2017;7(2): Article 7 [7 p.]. Available from: <http://ir.uiowa.edu/pog/> Free full text article.

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Financial Disclosure: The authors report no conflict of interest.

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Case presentation

At 23 weeks of gestation, a 32-year-old female patient G5P4 (with no living children) was referred to our tertiary health care center because of hydrops fetalis. Her initial investigations revealed negative TORSCH screening tests,

negative Coomb's test and her blood group was AB positive Rh factor. Ultrasound evaluation performed at the fetal medicine unit revealed bi-parietal diameter and femur length of 23 weeks. Abdominal circumference measured 35 weeks due to marked fetal ascites (Figure 1).



Figure 1: Ultrasound showing marked fetal ascites

Detailed echocardiography performed by a level III sonographer revealed fetal cardiomegaly, bradyarrhythmia, ventricular contraction independent of atrium contraction (3rd degree heart

block) (video), and lastly, hypertrophy and dilatation of all heart chambers. A non-stress test confirmed the fetal bradycardia, which revealed a heart rate of approximately 60 bpm (figure 2).

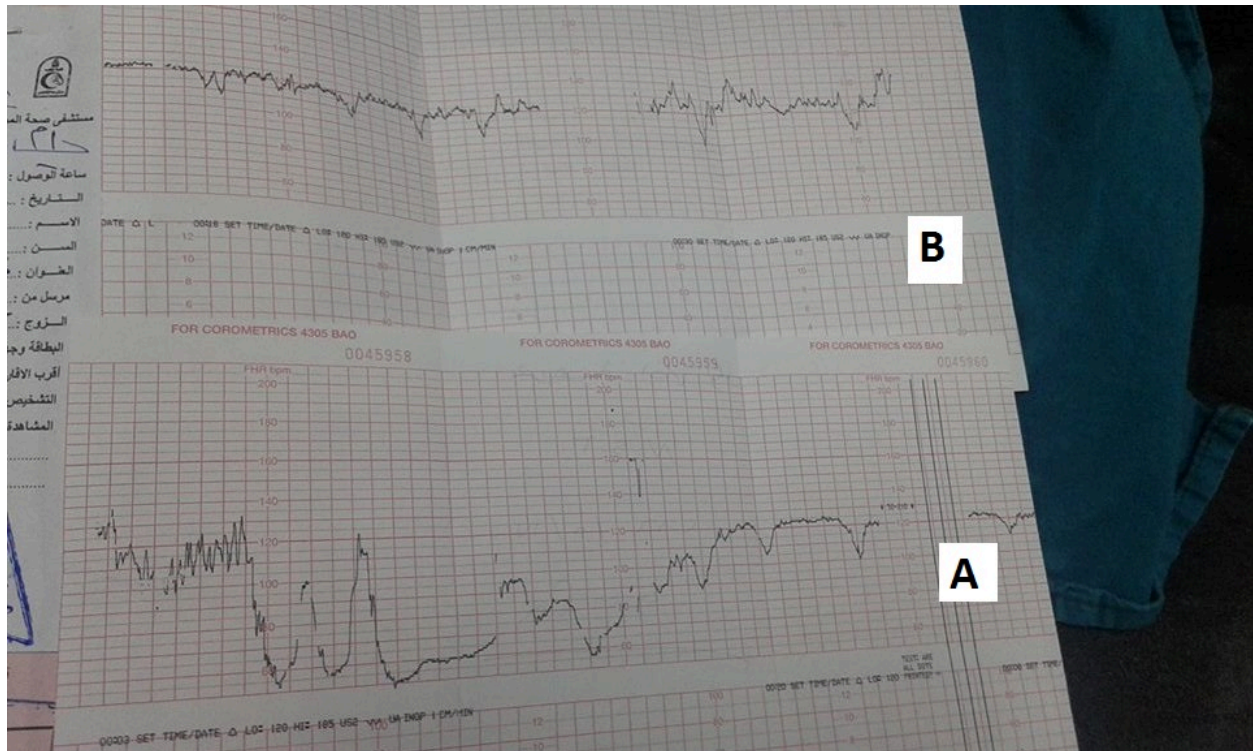


Figure 2: (A) Non stress test before treatment revealed fetal bradycardia – (B) Non stress test after treatment revealed normal fetal heart rate at 120 beats/minutes.

Although the patient had 4 previous pregnancies which were all complicated by hydrops fetalis and intrauterine fetal death, testing for SSA/Ro and SSB/La antibodies had not been performed. We requested these investigations and the results were positive.

The patient started corticosteroid therapy (5 mg prednisolone tablets, twice daily), and the fetal heart rate and arrhythmia improved (figure 2). However, the fetus died 2 weeks later, possibly due to heart failure.

Discussion

This case highlights that early diagnosis

and management of fetal atrioventricular block may be important. A woman with a past history of recurrent second-trimester fetal death may be a candidate for the screening of SSA/SSB-antibody.

There are two types of hydrops fetalis; the first is immune hydrops fetalis that represents about 12.7% of cases of hydrops fetalis and is associated with antigen-antibody mediated red blood cell hemolysis. The second more frequent type of hydrops fetalis is NIH which represents about 87.3% of cases.^{6,7} The incidence rate of NIH is estimated at 1 in every 3000 pregnancies.⁸ The main causes are infectious and metabolic diseases, chromosomal abnormalities, cardiovascular diseases, twin-twin transfusion syndrome, anemia and idiopathic NIH which has the worst prognosis.^{9,10}

There are different approaches for the

detection of fetal arrhythmias: the non-invasive methods like magnetocardiography (FMCG) and fetal echocardiography (M-mode, pulsed-Doppler, tissue-Doppler).¹¹ The invasive methods like scalp electrodes attached to electrocardiographic recordings are seldom employed; it can be used only in cases of ruptured membranes. The Doppler method is better than the M-mode for measurement of the fetal atrioventricular time intervals.¹²

Congenital atrioventricular block can be classified according to the etiology of it into 3 groups. The first is due to physical defects that structurally displace the distal conduction system. The second is due to the maternal antibodies SSA/Ro or SSB/La that yield into an inflammatory myocardial response, and the last is a block of unclear etiology. It is also can be classified depending on its severity as first-degree, second-degree (incomplete) or third-degree (complete) block.¹³

Isolated, complete atrioventricular block with negative SSA/Ro antibody appears to have the best long-term prognosis.^{13,14} The cause of this type of block remains unclear. Lopes et al. described that natural improvement of atrioventricular block in utero is possible in fetuses whose mothers are seronegative for antinuclear antibodies throughout pregnancy.¹⁵

Immune atrioventricular block occurs due to placental transfer of maternal autoantibodies against ribonucleoproteins of unknown function. SSA/Ro and SSB/La autoantibodies are labeled 'antinuclear antibodies', but ~70% of these ribonucleoproteins are actually situated in the cytoplasmic compartment.¹⁶

First-degree and second-degree atrioventricular block have short-lived period of reversibility, and early diagnosis is critical for treatment, but third-degree atrioventricular block is permanent.^{17,18} Complete improvement of second-degree block has been reported intrauterine.¹⁵ Friedman et al. documented cure of second-degree atrioventricular block in two of six fetuses treated with corticosteroids, but in spite of this treatment, three of the fetuses (50%) progressed to complete atrioventricular block (one intrauterine, two after delivery).¹⁹ The incidence of fetal atrioventricular block is about 2% of pregnancies in females with SSA/ Ro or SSB/La autoantibodies.¹⁸

Serial echocardiography screening is not helpful because there has been no evidence that this prevented the development of atrioventricular block. The risk of recurrent atrioventricular block when a previous fetus has been affected is about 19%.¹⁹ The most serious complication seen in females with positive antibodies is the occurrence of repolarization abnormalities and this may increase the incidence of sudden cardiac arrest documented in this group of patients.²⁰

In a females with SSA/Ro-related fetal cardiac complications, follow-up with FMCG is highly recommended to assess depolarization and repolarization anomalies, confirm the degree of block, and evaluate myocardial hypertrophy^{21,22} but, this method is still not readily accessible to doctors in most centers. Actually, it was unavailable in our tertiary fetal medicine center. If FMCG is not available, such females should be followed up with fetal echocardiography to measure mechanical PR interval and to detect

signs of endocardial fibroelastosis, tricuspid valve regurgitation, cardiac dysfunction, or atrioventricular block.¹⁹ Subsequently repolarization abnormalities cannot be documented without FECG or FMCG.

A number of progresses have been made in the treatment of fetal atrioventricular block. In utero, terbutaline has been used to increase the heart rate to >55 bpm, or in fetuses with signs of hydrops,²³ but it has not been proven to decrease the incidence of fetal or neonatal death. The use of steroids for treatment of fetal atrioventricular block is debated and at present no clear evidence of care is available. Due to the non-reversible condition of the third-degree atrioventricular block, steroids are infrequently used to treat it, unless myocardial dysfunction or hydrops fetalis are also present. Rein and colleagues²⁴ suggested that mothers who had previously had a fetus with atrioventricular block should receive steroid treatment in following pregnancies if fetal mechanical PR intervals ≥ 150 ms develop.

Due to the small number of cases in each treatment center, no randomized control trial is available to properly evaluate and compare the efficacy of drugs currently used. Available information was based on descriptive studies.

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