Clinical Case Report

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Dextrocardia with complete AV block and the implantation of a temporary pacemaker before cesarean section

A case report

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Abstract

Rationale: Patients with situs inversus totalis (SIT) and complete atrioventricular (AV) block are extremely rare, and only few cases have been reported up to now. Due to the rarity of such condition and its anatomical complexity, we reported this case as a reference for obstetricians, providing valuable insights into potential clinical treatment.

Patient concerns: We reported a case of 30-year-old patient with complete AV block, and her heart rate was only about 45 beats per minute.

Diagnoses: The patient was diagnosed with term delivery with SIT and complete AV block.

Interventions: A temporary pacemaker (TPM) was implanted before cesarean section (CS) because of complete AV block.

Outcomes: CS was performed successfully after the implantation of a TPM. The heart rate remained stable, and the 24-hour blood loss was limited.

Lessons: This study presents a complex heart disease case which needed more frequent antenatal examination and restriction of physical activity. To reduce the risk of pregnancy, basic disease needed to be cured before pregnancy. Our findings could provide guidance for future clinical studies.

Abbreviations: AV = atrioventricular, CHD = congenital heart disease, CS = cesarean section, DSA = digital subtraction angiography, ECG = electrocardiograph, SIT = situs inversus totalis, SVT = supraventricular tachycardia, TOF = tetralogy of fallot, TPM = temporary pacemaker.

Keywords: cesarean section, dextrocardia, temporary pacemaker, tetralogy of Fallot

1. Introduction

As a rare autosomal recessive condition, situs inversus totalis (SIT) is found in about 0.005% to 0.02% of the whole population.[1] SIT with dextrocardia is an uncommon congenital positional anomaly, which is characterized by a symmetrical "mirror-image" orientation of all organs in relation to the midline.[2] There is a 5% to 10% prevalence of congenital heart disease (CHD) in individuals with SIT. There are many types of CHD, such as Tetralogy of Fallot (TOF). Moreover, a higher risk of CHD has been observed during pregnancy due to the increased blood volume that exacerbates the burden on the heart. Therefore, CHD patients require more frequent antenatal examinations and present more challenges to obstetric care providers.

In the study, we reported a 30-year-old pregnant female with CHD who had corrective surgery when she was 6 years old. After then she led a peaceful life. She was examined 3rd atrioventricular (AV) block before pregnancy but not cured. Temporary pacemaker (TPM) was implanted before cesarean section (CS). Informed written consent was obtained from the patient for publication of this case report and accompanying images. This study was approved by Suzhou University on January 5, 2018. The number was (2018) science No. 002.

2. Case report

Our patient was born with SIT and TOF, and the TOF corrective surgery was performed when she was 6 years old. After the operation, she led a peaceful and asymptomatic life. She was pregnant when she was 30 years old, and then she was referred to

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CFY and WFY contributed equally to this work.

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the Department of Obstetrics in our hospital on December 9, 2017. The patient had a blood pressure of 112/65 mmHg, a pulse rate of 50 beats per minute (bpm), a body temperature of 36.5° C, and a respiratory rate of 18 bpm. At the time of admission, she was pregnant at 38 weeks and 1 day for CS. She had no symptoms of AV block during pregnancy with a heart rate of about 45 bpm. She had chest tightness occasionally when she climbed 3 staircases. Her echocardiogram (ECG) was abnormal, showing atrial fibrillation and 3rd degree AV block (Fig. 1). The AV block was diagnosed only before 3 days of delivery. The average heart rate was 49 bpm, ranging from 39 to 65 bpm. Maternal and fetal B-ultrasound examinations demonstrated that all maternal organs were reversed, while the fetal anatomy was normal (Fig. 2A, B). Maternal echocardiography (Fig. 2C, D) demonstrated no cardiac abnormalities. Her heart function was classified as level I. Obstetric examinations and clinical tests were in the normal ranges. Due to her ECG findings, cardiologist advised us to implant a TPM before delivery via CS. Therefore, a TPM was implanted before delivery on December 15, 2017. She underwent TPM implantation via her left subclavian vein with digital subtraction angiography (DSA) guidance (Fig. 3). The operation was successfully performed in 15 minutes. The pacing threshold was measured at 0.4 V, and the heart rate was at 60 bpm. After successful implantation, CS was performed by continuous epidural anesthesia. A 3410-g healthy female infant was born with Apgar scores of 9 and 9 at 1 and 5 minutes, respectively. After the CS, the TPM was removed. Moreover, 24-hour blood loss was 400 mL after birth. The patient was discharged 4 days postoperatively without any complications. She preferred breastfeeding, and the newborn developed well without any currently known congenital anomalies. She did not treat her 3rd AV block after the delivery. Both mother and child were in good conditions as of now according to the follow-up by telephone.

3. Discussion

3.1. The mechanism and diagnosis of SIT

SIT, also called mirror-image dextrocardia, is characterized by the presence of the heart and stomach on the right side of the midline, while the liver and gallbladder are present on the left side.[3] The pathogenesis of SIT remains unknown. In vitro and animal studies have found a possibly increased risk of dextrocardia following exposure to Fingolimod.[4] SIT may be caused

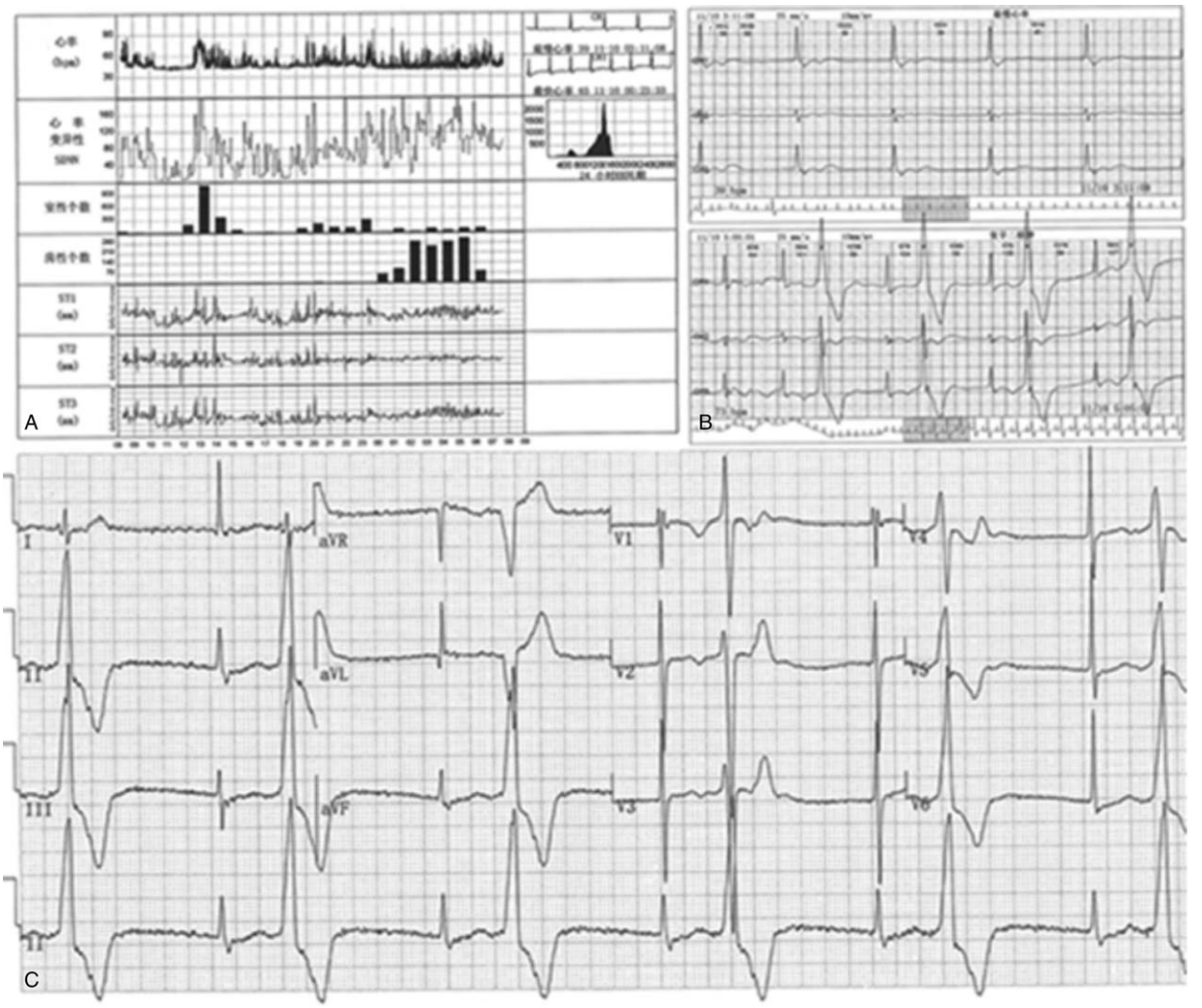

Figure 1. ECG findings. (A) 24-hour data; (B) ventricular premature beat; (C) 3rd degree AV block. AV = atrioventricular.

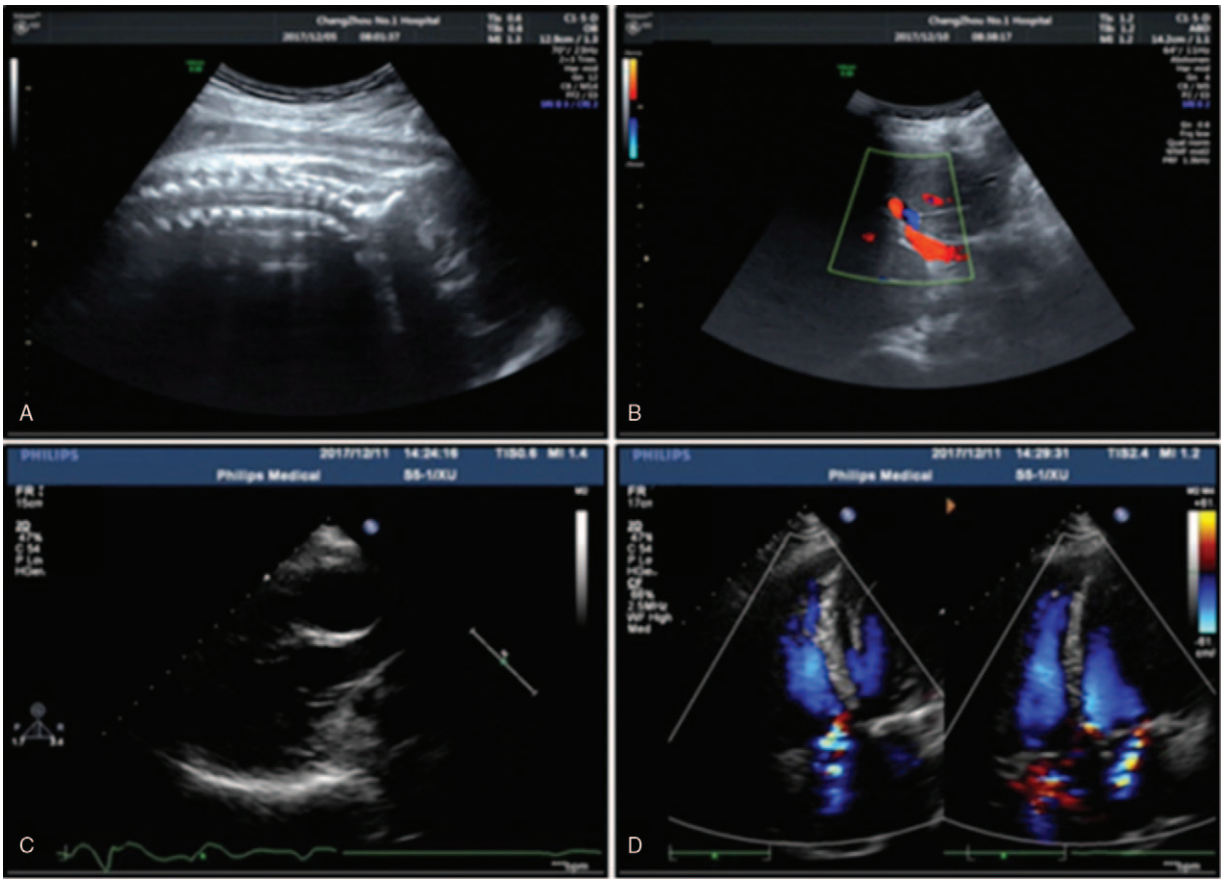

Figure 2. Maternal and fetal B-ultrasound and ECG. (A) fetal ultrasound showing the fetal spine; (B) maternal ultrasound showing the mother's liver; (C) maternal ECG apical 4 chamber view; (D) maternal ECG parasternal long axis view. ECG = electrocardiograph.

by a disorder in visceral rotation during embryonic development. As a recessive mutation, the probability of inheritance is very low. There are multiple modalities to diagnose SIT although patients are typically asymptomatic. SIT can be diagnosed using common radiographic imaging techniques, such as heart ultrasonography, chest X-ray, CT, and MRI. Furthermore, a properly interpreted electrocardiogram can be useful in suspecting the diagnosis of dextrocardia with SIT.[5] Our patient was diagnosed using X-ray and ultrasonography when she was 5 years old. She had corrective surgery when she was 6 years old. After the operation, her heart function was classified as level II.

3.2. TOF and pregnancy

TOF is one of the most common right-to-left shunt congenital heart defects, which forms during embryologic development. TOF is found in about 5% to 8% of CHD patients. Surgical correction, especially in early life, is the best way to treat such defect. Heart block has not been significantly correlated with post-operative outcomes.[6] Applications of new surgical techniques have led to improvements in treating TOF in early childhood.[7] Cardiovascular complications during pregnancy are common, such as supraventricular tachycardia (SVT), pulmonary embolism and heart failure. In 1 retrospective study, 74 women have 157 pregnancies, and cardiovascular events occur in 8.1% of these pregnancies.[8] Fetal risks in mothers with TOF include spontaneous abortion, intrauterine growth restriction, prematurity, perinatal mortality, and CHD in the fetus.[9] Our patient had an early surgical correction, and the pregnancy was found in April 2017. After pregnancy, she had regular prenatal visits and gave birth to a healthy baby. Fortunately, our patient and her baby had no complications.

3.3. Severe AV block and TPM

AV block represents a delay or disturbance in the transmission of an impulse from the atria to the ventricles. In general, there are 3 degrees of AV nodal blocks: 1st degree, 2nd degree, and 3rd degree. Our patient had complete AV block with no symptoms. Therefore, she refused to have a permanent pacemaker before hospitalization. She walked 50 meters without chest tightness and breathing difficulties. However, she had chest tightness occasionally when she climbed 3 staircases. According to New York Heart Association,[10] her heart function was classified as level I. She had 3rd degree AV block so the cardiologist advised a TPM before CS in order to reduce the risk of anesthesia and surgery. Although there is currently no established indication for preoperative prophylactic pacemaker implantation,[11] TPMs can be very useful. TPM implantation is an important and potentially life-saving intervention in the treatment of symptomatic bradycardia and for overdrive pacing of some life-threatening tachydysrhythmias. TPMs can also improve the hemodynamic parameters. Implantation of TPMs is usually safe. Our pregnant SIT patient with complete AV block underwent

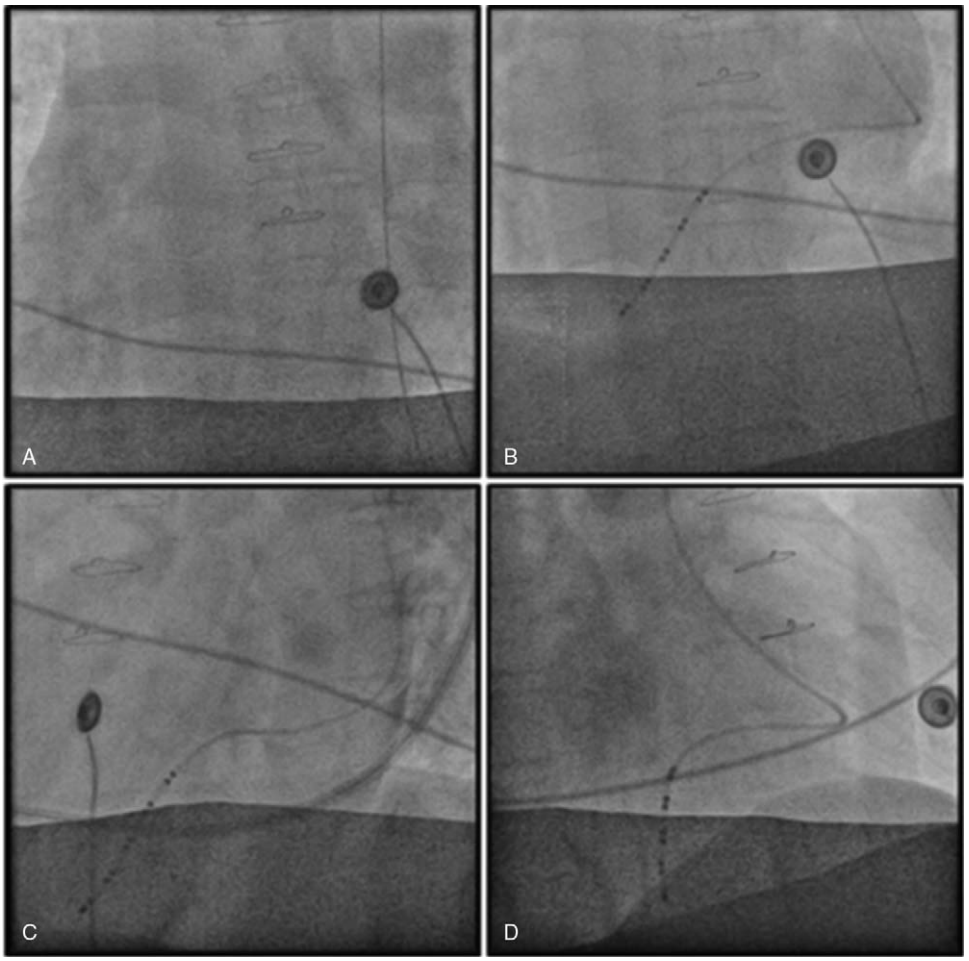

Figure 3. A temporary pacemaker implanted with DSA guidance. (A) anterior and posterior (before surgery); (B) anterior and posterior (after surgery); (C) left anterior oblique (after surgery); (D) right anterior oblique (after surgery). DSA = digital subtraction angiography.

TPM implantation via her left subclavian vein with DSA guidance. The advantages of this approach included minimal incision size and blood loss, so it could be performed under local anesthesia.[12] In fact, appropriate care for women with CHD requires knowledge of cardiac physiology during pregnancy, the common lesions of CHD, and coordinated care from cardiologists and maternal-fetal medicine specialists.[13]

3.4. Cesarean section

Pregnancy increases venous return, thus increasing preload. Pregnancy with CHD is a high-risk condition, which may increase maternal and fetal complications. These patients require more frequent antenatal examination and restriction of physical activity.[9] SIT with severe AV block may cause cardiac arrest, especially during pregnancy. Therefore, further research on cardiac arrest risk stratification in dextrocardia in general and during pregnancy in particular is highly necessary.[14] CS was selected as the delivery method due to the minor increase in minute volume (30%) as compared with natural vaginal birth (50%), even though CS has a lot of possible complications too.[7] The patient required detailed pre-delivery evaluation with closely monitored blood pressure, heart rate and blood loss during the operation. The patient also needed detailed evaluation, which was under joint supervision of an obstetrician, a cardiologist and an anesthetist.[9] The CS was successful, and her heart rate was stable with the TPM. Moreover, the 24-hour blood loss was 400 mL after birth. Postpartum maternal complications may include SVT, pulmonary embolism, and heart failure.[12] We monitored our patient carefully postpartum, and she had no complications in 72 hours.

Collectively, the incidence of CHD in people with SIT is not high. Corrective surgery is the prerequisite for delivery. Severe AV block is very dangerous. Therefore, these patients require more frequent antenatal examination and restriction of physical activity. A TPM may be implanted before delivery in order to regulate heart rate and reduce the risk of anesthesia. Blood circulation changes significantly during perinatal periods. Therefore, it is important to monitor the operation closely by cardiologists, anaesthesiologists, and obstetricians in order to prevent the occurrence of uterine bleeding and heart failure after delivery.

Author contributions

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Software: Xiaoqing Wang.
Supervision: Jinggang Zhang.
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Writing – review & editing: Wenfeng Ye.

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