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Corrected Transposition of Great Arteries with Cor Triatriatum and Atrial Septal Defect-Case Report

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ABSTRACT

A 37-year-old male patient with corrected transposition of great arteries (ccTGA) with cor triatriatum sinister (CTS), left superior vena cava, and atrial septal defects is reported in our case. None of these impacted the patient's growth or development, nor daily work until age 33. Later, the patient developed symptoms of obvious impaired heart function, which improved after medical treatment. However, the symptoms reappeared and gradually worsened two years later, and we decided to treat it with surgery. In this case, we selected tricuspid mechanical valve replacement, cor triatriatum correction, and atrial septal defect repair. During the follow-up of five years, the patient had no obvious symptoms, ECG did not change significantly from five years ago, and the cardiac color Doppler ultrasound showed RVEF 0.51.

INTRODUCTION

The cor triatriatum sinister is a rare congenital malformation with an incidence of 0.1% to 0.4% in congenital heart disease, often in combination with other malformations, including atrial septal defect and left superior vena cava. The previous data suggest that the left superior vena cava with the cor triatriatum sinister is more connected to the true left atrium, and the left superior vena cava is connected to the right atrium in this case [Rasemann 2007]. Corrected transposition of great arteries is a relatively rare congenital heart disease with an incidence of approximately 0.05% in all congenital heart defects [Wallis 2011]. It is rare to see corrected transposition of great arteries with cor triatriatum sinister, left superior vena cava, and atrial septal defects.

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CASE DESCRIPTION

In November 2016, a 37-year-old male patient weighing 62 kg was admitted to the Cardiac Surgery Department. The patient presented with a complaint of palpitations after activity for more than three years and aggravation for more than one year. Cardiac ultrasound revealed that the great arteries were transposed correctly at birth, without apparent symptoms, and no treatment was given. He complained of palpitations, fatigue, and sweating three years ago and was diagnosed with corrected transposition of the great arteries and anatomical tricuspid regurgitation. Following medical treatment, his symptoms began to improve. However, it took nearly a year for the symptoms to worsen; the palpitations and sweating also appeared while at rest. He had a history of pleurisy for more than 30 years and primary immune thrombocytopenia for two years. On physical examination, the heart rate was 78 times/min. The heart rate was neat, with extensive conduction, and III/6 systolic murmurs were heard in the sternum's fourth intercostal space of the left margin. The electrocardiogram (ECG) demonstrated intra-atrial block and change in ST-T. The chest radiograph revealed increased bilateral striations, an enlarged hilum, and a spherical heart. The results of the magnetic resonance examination are shown in Figure 1. (Figure 1) Echocardiography (UCG) showed the following: congenital heart disease, dextrocardia, corrected transposition of the great arteries, cor triatriatum sinister, severe pulmonary hypertension, left-sided aortic arch, double superior vena cava, mild aortic stenosis, anatomy of mitral regurgitation (mild), and anatomy of tricuspid regurgitation (severe). (Figure 2)

Treatment: Under general anesthesia, the patient underwent tricuspid mechanical valve replacement, cor triatriatum correction, and atrial septal defect repair at the median sternal incision.

A large heart with dextroversion cardis SLL and moderately elevated pulmonary artery pressure was discovered during extracardic examination. Routine heparinization was used to establish cardiopulmonary bypass. The heart was stopped by directly infusing cold-blooded cardioplegia into the aortic root. No thrombus was observed when the left atrial appendages were opened and examined, and the two openings were visible. The opening of the additional atrium is near the head, whereas the atrial septal defect is located close to the tricuspid valve. The interatrial septum was cut open, and the superior, inferior, and left upper vena cava openings in the right atrium were examined.

Atrial septal defects of 1 x 1 cm and 0.5 x 0.3 cm were found between the paraventricular and right atrium and sutured continuously. (Figure 3) There was a small opening between the additional atrium and the true left atrium, but no septate tissue was found on the lower left or right sides. The left atrium was open upward to the top of the left atrium, and the diseased tricuspid valve was removed to preserve the rest of the underlying chordae tendineae. The aortic valve was detected through the tricuspid valve orifice without any abnormalities. The tricuspid valve was replaced with a 27M Medtronics mechanical valve using the intermittent mattress suture 13 stitches, and the opening and closing of the valve leaf were tested. Slices of the bovine pericardium were used to suture the top of the left atrium. The tricuspid valve was removed and sent to the pathology department for analysis. According to the report, the valve tissue had hyperplasia with vitreous degeneration and fibrous necrosis.

Follow up and outcomes: The patient was discharged from the hospital 7 days after surgery. In the follow up of 5 years after surgery, the patient recovered well, with no obvious symptoms, no obvious limitation of physical activity, and no occasional chest tightness. The postoperative review of cardiac color ultrasound at the 5th year suggested RVEF 0.51, and the patient's electrocardiogram graphics did not change significantly compared with results 5 years ago.

DISCUSSION

Cor triatriatum is a rare congenital cardiac malformation characterized by the presence of a fibrous tissue space in the atrium that divides it into two chambers: the additional atrium and true atrium. The incidence of congenital heart disease is 0.1%~0.4% and is more prevalent among males (1.5:1). It primarily affects the left atrium, while the right atrium is not commonly affected [Rasemann 2007]. Infants and children are more likely to be affected than adults [Işık 2019]. The additional atrium lies directly above the fibrous tissue septum and receives blood flow from the pulmonary veins. The true chamber receives blood from the additional atrium and the atrial septal defect, and this blood flows into the left ventricle via the mitral valve. Typically, cor triatriatum has no apparent symptoms, and in severe cases, the symptoms resemble those of mitral stenosis. Most of the time, the severity of symptoms is primarily determined by the degree of blood flow obstruction related to the size of the opening on the fibrous tissue septum and the size of the atrial septal defect.

The corrected transposition of the great arteries (ccTGA) is a relatively rare congenital heart defect that accounts for approximately 0.05% of all congenital heart defects [Wallis 2011]. According to Van Praagh's classification, ccTGA can be classified into four types. This patient is of SLL type in which the ventricle is on the left, the atrium is correctly positioned, and the aorta is located in the left front of the pulmonary artery. The dissected tricuspid valve is prone to reflux and right ventricular dysfunction because the anatomic right ventricle is subjected to systemic circulatory pressure [Mongeon 2011].

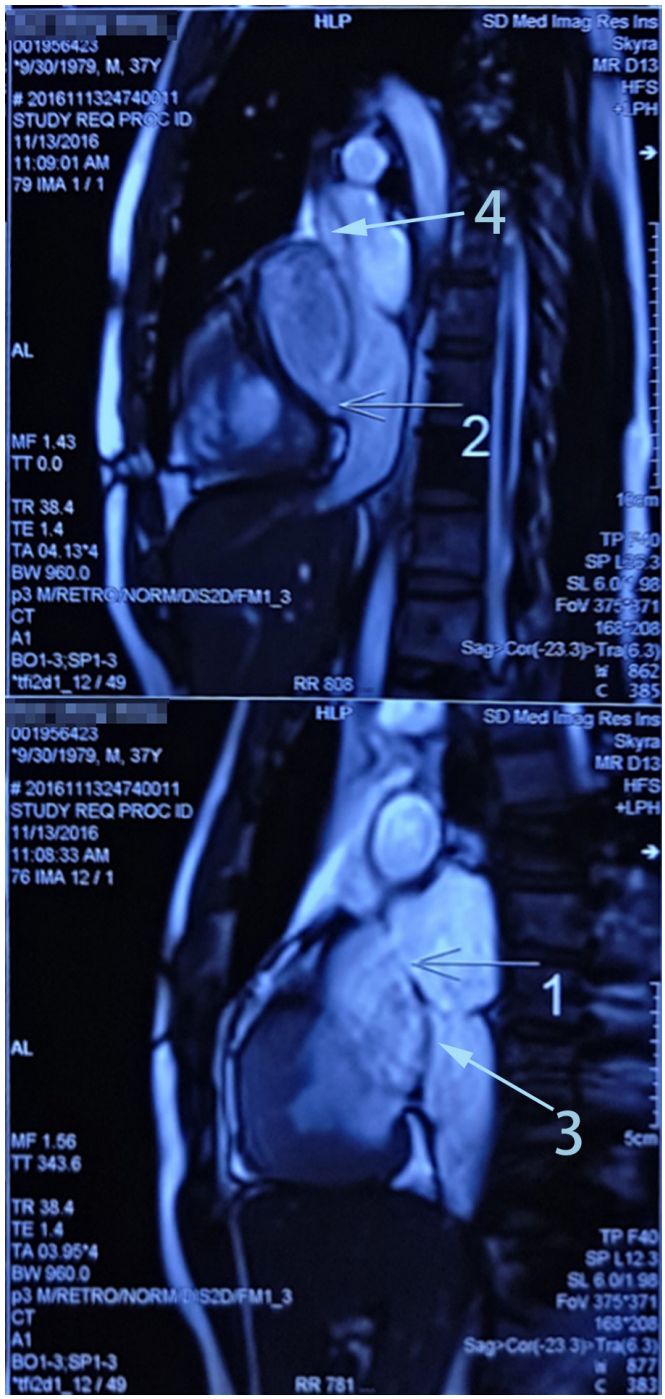

Figure 1. 1) left atrial diaphragm; 2 and 3) atrial septal defects; 4) left upper vena cava

In this case, a patient with a combination of cor triatriatum sinister, left superior vena cava, atrial septal defect, and corrected transposition of great arteries, and who survived asymptomatically for more than 30 years, is very rare. We actively improved the examination before surgery to further identify other comorbid deformities in the patient. The presence of

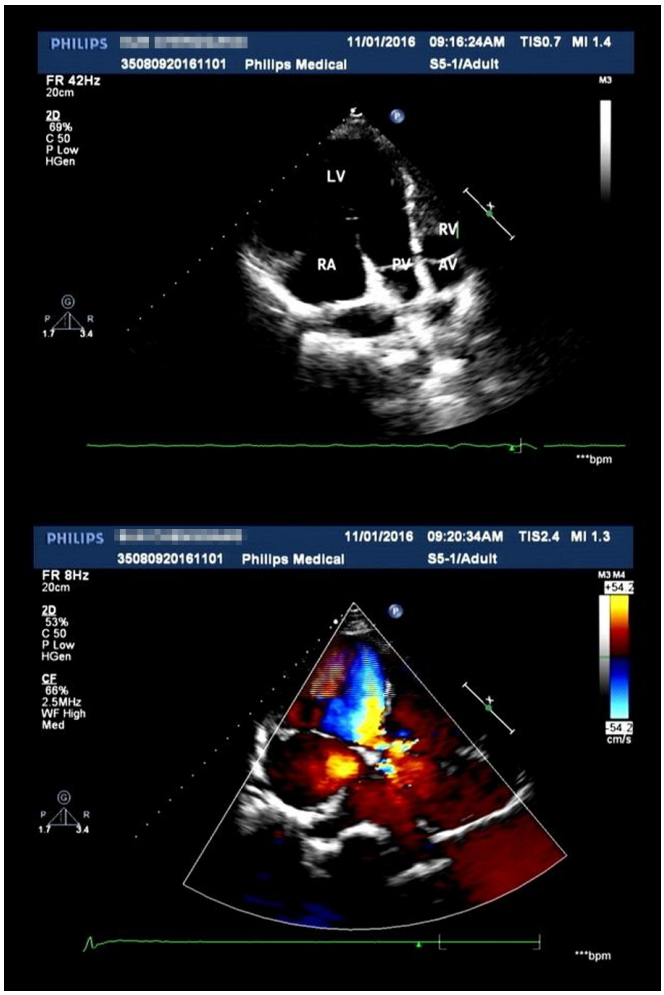

Figure 2. Ultrasound results before surgery

heart failure in patients is due to the fact that the anatomical right ventricle in the corrected transposition of great arteries assumes the function of systemic circulation, and in the long run, the ventricle expands, resulting in tricuspid regurgitation, and the tricuspid regurgitation further increases the burden on the right ventricle, forming a malignant cycle [Kutty 2018].

In previous relevant literature, there are two major categories of traditional surgery and anatomical correction surgery for the treatment of corrected transposition of great arteries, and the traditional surgical method is mainly for the correction of concomitant intracardiac malformations, including the repair of ventricular septal defects, correction of pulmonary artery outflow tract stenosis, and correction of tricuspid valve insufficiency [Filippov 2016; Cui 2021]. The purpose is to reduce the capacity and resistance of the right ventricle and delay the enlarged reconstruction of the right ventricle. The effect of anatomy correction surgery in the near and medium term is obvious, and the long-term effect is currently inconsistent, and the difficulty and trauma of the operation are very large, and the indications for surgery also have certain limitations [Hornung 2010].

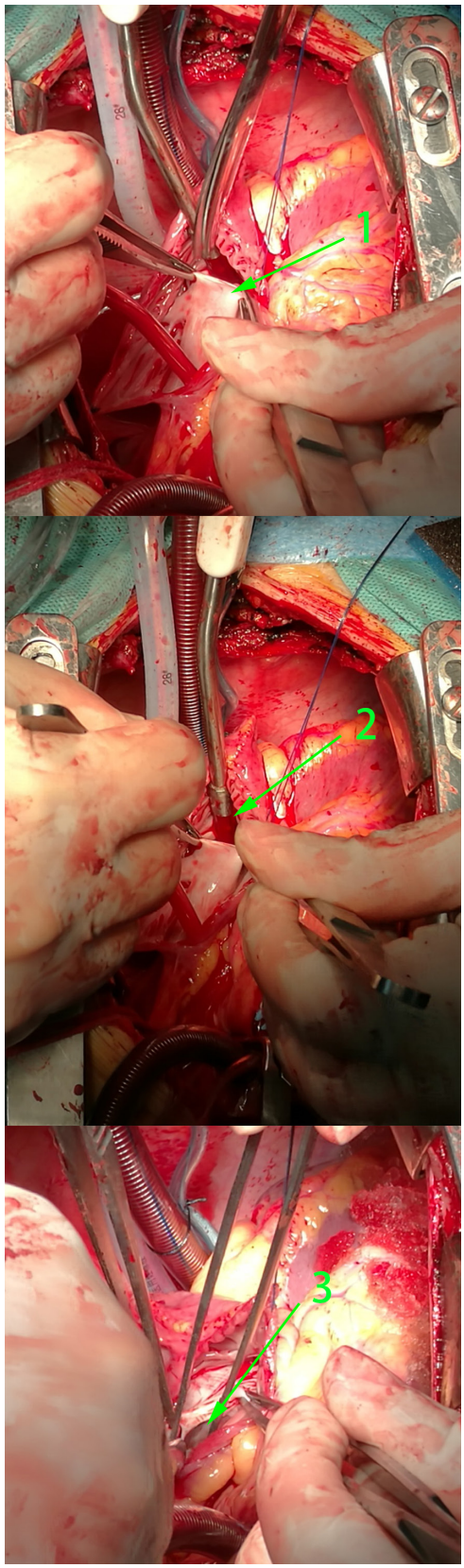

Figure 3. 1) Diaphragm of the left atrium; 2 and 3) atrial septal defects

In this case, the patient was 37 years old, the course of the disease was long, there was no significant enlargement of the morphological right ventricle, the morphological right ventricle function was 0.48 after symptomatic treatment, and the ventricular septum was complete, so we chose tricuspid mechanical valve replacement [Cui 2021; Hornung 2010]. For other cardiac malformations, whether to deal with them together, we must actively judge before surgery, think of possible problems, and make a decision based on the specific circumstances of the operation. Cor triatriatum sinister and atrial septal defects have a significant effect on hemodynamics and need to be managed together during surgery. In this patient, the atrial septal defect was located in the additional atrium, which is the most common location relationship of the cor triatriatum sinister combined with the atrial septal defect. Blood flow from the pulmonary veins first returns to the additional atrium, where a portion of the blood flows through the atrial septal defect into the right atrium. Most of the blood flows through the left atrium and into the right ventricle through the tricuspid valve.

At the same time, due to the presence of the cor triatriatum sinister diaphragm, it may increase the blood entering the right atrium through the atrial septal defect during the course of the disease, reducing the burden on the right ventricle, which may play a role in the asymptomatic survival of patients [Işık 2019]. The cor triatriatum sinister and atrial septal defects can be preliminarily judged in the cardiac ultrasound, and it should be carefully explored during surgery. We should pay special attention to the number of atrial septal defects and not miss small defects.

Small defects in cardiac color ultrasound often are difficult to distinguish, if they are closer to the location of large defects.

At the beginning of the procedure, the cardiopulmonary bypass was established. Different from ordinary practice, the patient had two superior vena cava veins, and in most cases the left superior vena cava runs along the posterior part of the left atrium, and venous blood enters the right atrium through the coronary sinus opening. During the operation, we found that the left and right upper vena cava veins were relatively thick and adopted intubation for the right superior vena cava and the left superior vena cava was blocked, so as to better the field of vision in the leakage surgery. After blocking the left superior vena cava, attention should be paid to the patient's face and whether there are signs of obstruction of venous return [Rigatelli 2007].

According to the preoperative cardiac ultrasound and the thickness observed during the operation that the blood is returned to the right atrium from the left superior vena cava, the amount of blood return may be comparable to that of the right side. Therefore, it will flow back through the left and right traffic branches to the right upper vena cava after blocking, but it may cause the reflux to be blocked if the vein of communication is too thin, affecting the patient's brain blood return, and causing damage to the nervous system. Because the right superior vena cava is thick enough and the left superior vena cava returns to the right atrium, the difficulty in pacemaker implantation by interventional surgery does not increase, which is sufficient to cope with the gradual increase in AVB tissue of major complications in the future.

CONCLUSION

In this case, there are many congenital malformations combined, and we must conduct comprehensive preoperative examinations for these kinds of patients, so as to figure out the existing malformations and the problems caused by various deformities before surgery. Our goal is to help the patient to smoothly pass the perioperative period and have a good prognosis, rather than to only correct the malformation. As far as this patient is concerned, we did not correct all malformations, but his cardiac function was not significantly affected during 5-year follow up.

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