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Case Report

A complete anatomical correction of an atrial septal defect coronary sinus with an incidental finding of the unroofed coronary sinus syndrome type II

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Abstract

The atrial septal defect coronary sinus with unroofed coronary sinus syndrome type II is a rare clinical finding. In this case report, we presented a complete anatomical correction of the full form of the unroofed coronary sinus syndrome type II with the transfer of the coronary sinus to the right atrium. The anatomical features for a successful procedure were the proximity of the coronary sinus mouth to the secundum type atrial septal defect, tunnel plastic did not block the flow of blood through all pulmonary veins into the left atrium, and the clear visualization of the coronary sinus mouth with diagnostic antegrade cardioplegia.

Keywords: Unroofed coronary sinus; Physiological correction; Oxygen desaturation; Tunnel

INTRODUCTION

Unroofed coronary sinus syndrome (URCS) is the rarest type of atrial septal defect (ASD) (less than 1% of all ASDs) [1,2]. It is characterized by a communication between the coronary sinus (CS) and the left atrium (LA) as a result of the partial or complete absence of the roof of the CS [3]. A complete anatomical correction with a transfer of the CS to the right atrium (RA) is usually performed in the case of partial URCS [2,4,5]. The standard surgical procedure is to close the ASD with an autologous patch of pericardium thereby leaving the CS in the LA. We present the case of a complete anatomical correction of a full type II URCS with a patch of autologous pericardium forming a tunnel from the mouth of the CS into the RA. A thorough review of the current literature did not yield any evidence of such a procedure to repair a type II URCS with CS drainage into the RA.

CASE REPORT

In October 2016, an 8-year-old boy was admitted to our department with dyspnea and an 94% oxygen saturation, consistent with New York Heart Association class II symptoms. The transthoracic echocardiography (ECHO) reveled an ostium secundum type ASD with a left to right shunt and dilation of the coronary sinus (1.3 × 1.3 cm). On the ECHO, the CS was between the middle and lower third of the interatrial septum. Dilatation of the CS indicated the possibility of an persistent left superior vena cava (LSVC) entering it. Cardiac catheterization was performed to rule out partial abnormal drainage. A right heart ventriculography showed an atrial septal defect with no superior vena cava and partial abnormal drainage. A computed tomography scan, which allows the visualization of the posterior structures of the heart, was not performed because there was no suspicion of URCS. The operative plan was aimed to surgically reconstruct the ostium secundum type ASD. However, during the intra-operative inspection, an ostium secundum type ASD and an enlarged CS without walls and communication with the ostiums of all pulmonary veins were detected, confirming a complete type II unroofed CS without a persistent left superior vena cava. The anatomical features for a complete anatomical correction were the proximity of the CS mouth to the ASD, the tunnel did not obstruct all 4 pulmonary veins inflow, the zone of the seam line during the formation of the tunnel was not in close proximity to the AV node, which prevented damage. Additional diagnostic cardioplegia revealed the true CS ostium (4 mm in diameter) located close to the left ostium of the pulmonary veins of the lateral wall of the LA. Using Prolene 6-0 suture, we used a patch of autologous pericardium to form a tunnel from the ostium of the CS into the cavity of the RA while taking care to separate it from the ostium of the pulmonary veins (Figure 1). The stitches were 5 mm from the edge of the ostium of the CS. Most of the circumference of the neo-coronary sinus was composed of the patch material and the tunnel was simultaneously closed with the ASD using another patch of autologous pericardium. Control cardioplegia confirmed the outflow from the ostium of the CS was not hampered. The aortic cross clamp time was 50 min and CPB time was 80 min. Three hours after the operation, the patient was extubated and, 20 hours later, transferred to the ward. The transthoracic echocardiography showed an intact atrial septum and the CS draining freely into the RA. The follow-up transthoracic echocardiography (11 months after the surgery) demonstrated a sealed ASD with the CS draining freely into the RA (Figure 2). The chambers of the heart were not dilated and the

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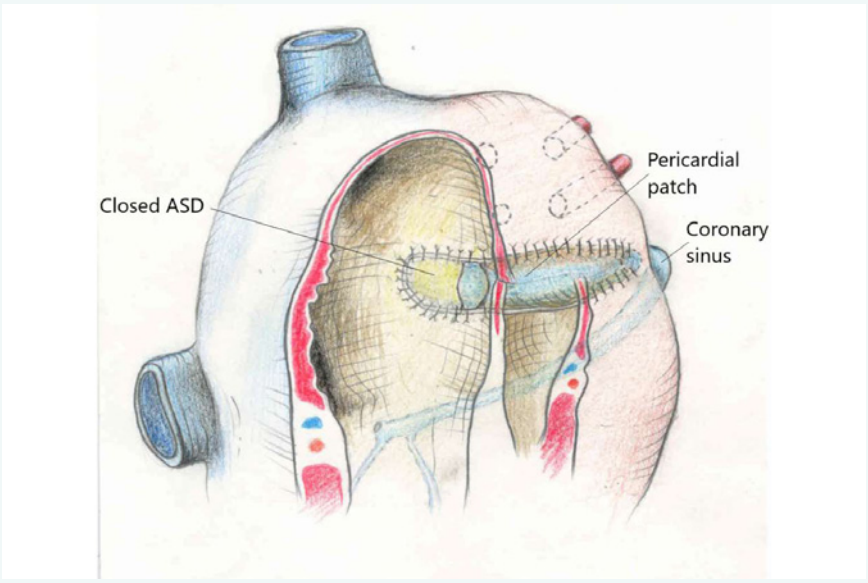

Figure 1 Scheme of operation.

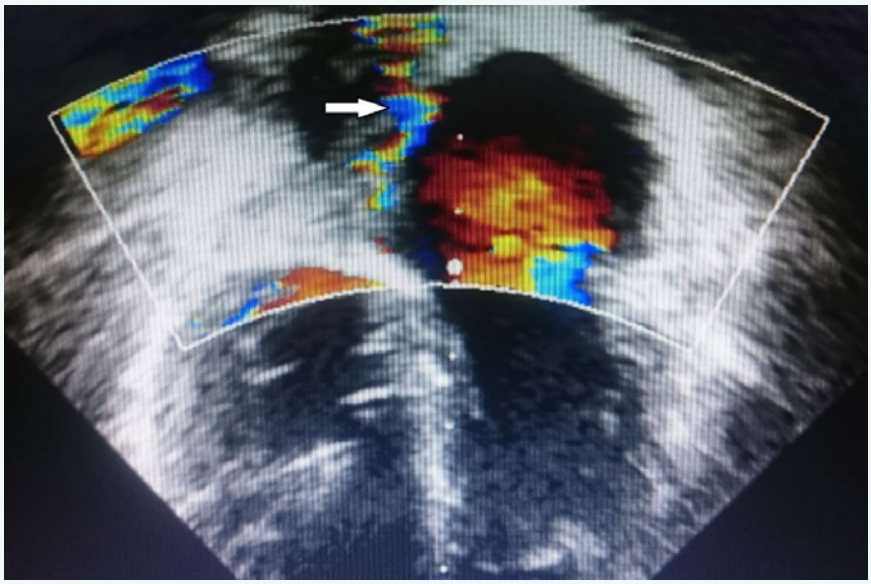

Figure 2 Echocardiographic picture showing the free drainage of the coronary sinus into the right atrium (arrow).

contractile function satisfactory with a left ventricular ejection fraction of 65%. No myocardial hypertrophy was observed. There was minimal mitral, tricuspid (1 degree) and pulmonary regurgitation (the systolic pulmonary artery pressure was 33 mmHg). One year after the operation, the patient feels well and is physically active, with an oxygen saturation of 100%.

DISCUSSION

There are several standard URCS correction methods [1,2] when the ASD is closed and the CS remains in the LA. This is the first case of radical correction of the URCS type II in the setting of an incidental finding. In this case, we performed a complete anatomical correction of the full type II URCS with preservation of the outflow of venous blood to the right heart. With intraoperative discovery, it was decided that the ASD could be surgically reconstructed with the simultaneous creation of a tunnel and the transfer of the CS to the RA. The anatomical features for allowing a successful procedure were the close location of the CS mouth to the ASD, the tunnel did not obstruct all pulmonary veins inflow, and the clear visualization of the CS mouth with diagnostic cardioplegia. The risk of AV node damage was minimal as it was located far from the reconstructed zone, and the seams were also placed far from the mouth of the CS which allowed us to avoid the potential risks of its narrowing.

These anatomical features allowed us to perform a complete anatomical correction without technical difficulties, similar to the correction of partial anomalous pulmonary vein drainage. Safe and effective complete anatomical correction of URCS with prevention of hypoxemia (5% oxygen desaturation) was also performed with other types of URCS [6-8].

In summary, the correction of an atrial septal defect coronary sinus with an incidental finding of the URCS type II with the transfer of CS to the RA was safe and feasible. Further clinical follow-up will be conducted to evaluate long-term outcomes.

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The authors have no conflicts of interest to declare.

COMPLIANCE WITH ETHICAL STANDARTS

Conflict of Interest The authors report no conflicts of interest in this work.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

This case report does not contain any studies with animals performed by any of the authors. Research involving human participants.

Informed consent Informed consent was obtained from the patient's mother.

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