Journal of Cardiovascular Diseases & Diagnosis
ISSN: 2329-9517

Mishin et al., J Cardiovasc Dis Diagn 2019, 7:2
DOI: 10.4172/2329-9517.1000359

Case Report Open Access

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

A complete anatomical correction of the full form of the unroofed coronary sinus syndrome type II was performed, with the transfer of coronary sinus (CS) to the right atrium. The anatomical features for a successful procedure were the proximity of the CS mouth to the atrial septal defect, the absence of intersection between the tunnel and the mouths of the pulmonary veins and the clear visualization of the CS mouth with diagnostic 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). It is characterized by partial or complete absence of the roof of the coronary sinus (CS) and communication between the CS and the left atrium [1]. 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]. When complete (URCS type I and II), due to anatomical features, the standard surgical procedure is to close the defect with a transfer of the CS to the RA. We present a case of a complete anatomical correction of a full type II URCS with a transfer of the CS to the RA.

Case Report

In October 2016, an 8-year-old boy was admitted to our department with dyspnea, consistent with New York Heart Association class II symptoms. The transthoracic echocardiography reveled ostium secundum type ASD with a left to right shunt, dilation of the coronary sinus (1.3 × 1.3 cm). 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 [5], 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, enlarged coronary sinus without walls and communication with the ostiums of the pulmonary veins were detected, confirming a completely 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 absence of an intersection between the tunnel and the mouths of the pulmonary veins, which were located far from the reconstruction AV node zone. 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 left atrium. We used one patch of autologous pericardium and formed a tunnel from the ostium of the CS into the cavity of the RA using Prolene 6-0 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 formed was simultaneously closed with the ASD using one patch. Control cardioplegia – 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 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, oxygen saturation 100% [6,7].

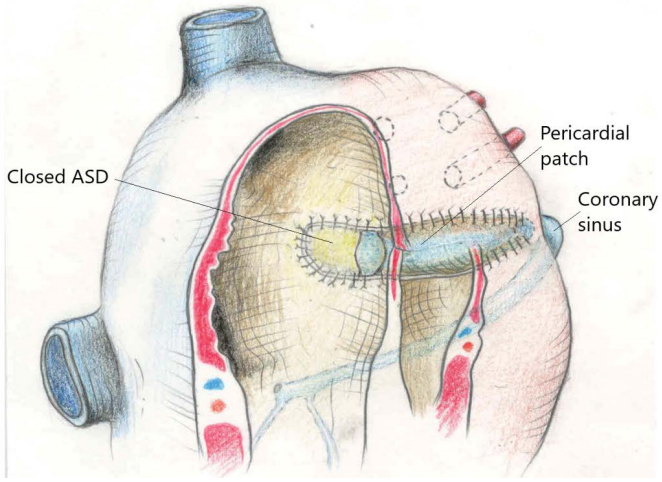

Figure 1: Scheme of operation.

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Received February 06, 2019; **Accepted** February 26, 2019; **Published** March 05, 2019

Citation: Mishin A, Kanzhigalin D, Sarsembayeva A (2019) A Complete Anatomical Correction of an Atrial Septal Defect Coronary Sinus with an Incidental Finding of the Unroofed Coronary Sinus Syndrome Type II. J Cardiovasc Dis Diagn 7: 359. doi: 10.4172/2329-9517.1000359

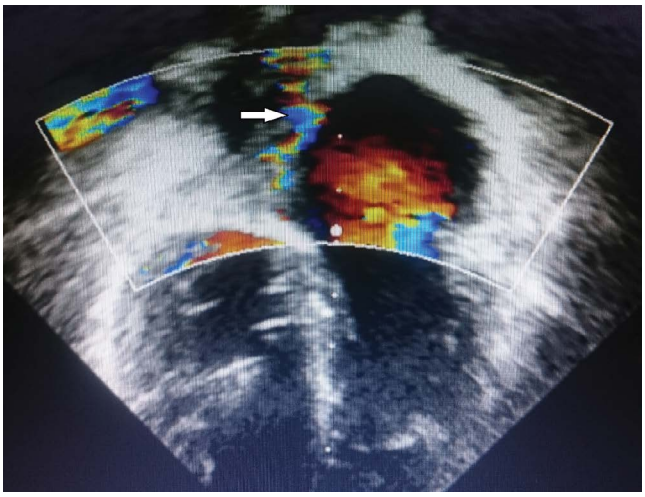

Figure 2: Echocardiographic picture showing the free drainage of the coronary sinus into the right atrium (arrow).

Discussion and Conclusion

In summary, 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 ASD could be surgically reconstructed with the simultaneous creation of a tunnel with 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 absence of intersection between the tunnel and the mouths of the pulmonary veins and the clear visualization of the CS mouth with diagnostic cardioplegia. There was no risk of damage to the AV node, which was located far from the reconstructed zone, and the seams placed far from the mouth of the CS 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.

Acknowledgments

We thank Danielle Libersan, PhD for her help in preparing the manuscript.

Conflicts of Interest

None declared.

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