Archives of Clinical Trials and Case Reports

Potnis AR, et al., 2023- Arch Clin Trial & Case Rep
Review Article

Extracardiac Pericardial Conduit Repair: A Novel Approach to Complex Scimitar Syndrome with Dextrocardia and intact Inter-Atrial Septum

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Abstract

Scimitar syndrome (SS) has been repaired using different surgical procedures like intracardiac baffle and redirection (directly or by using synthetic grafts) of the scimitar vein to the right or left atrium. However, several accompanying anatomic abnormalities make these interventions difficult. The most common variations complicating reimplantation of the scimitar vein (SV) include dextrocardia, hypoplasia of the right pulmonary artery and lung sequestration. Here, we present a novel method for extracardiac repair of SS with dextrocardia and intact atrial septum in which the right superior and inferior pulmonary veins formed a common venous chamber at the diaphragmatic surface and opening at the right atrium (RA) and inferior vena cava (IVC) junction. Although the use of pericardial tube grafts is seen in literature before, its use for rerouting a complex venous complex into left atrium is first reported in this case.

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Received Date: 11-01-2023.

Accepted Date: 11-07-2023.

Published Date: 11-20-2023.

Keywords: Scimitar syndrome; congenital heart disease; Pericardial conduit; Dextrocardia with intact atrial septum; Complex cardiac anomaly

Introduction

Potnis AR | Volume 2; Issue 1 (2023) | Mapsci-ACTCR-2(1)-024 | Review Article
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DOI: https://doi.org/10.37191/Mapsci-ACTCR-2(1)-24

Scimitar syndrome is defined as anomalous right pulmonary veins draining in the inferior vena cava or the right atrium. It derives its name from a curved Turkish sword (scimitar) which resembles the gently curved vertical vein in this anomaly. The scimitar vein is usually single and passes anterior to the hilum and runs below diaphragm to pierce IVC just superior, posterior and lateral to hepatic vein orifice. SS has an incidence of 20 per million births and a female preponderance of 2:1. It is reported to be present in 3% to 6% cases with sinus venosus atrial septal defects.1 Multiple approaches to scimitar syndrome (SS) have been developed in order to create an unobstructed low-flow pathway for the scimitar vein (SV) to drain in the left atrium. The procedures available are intracardiac baffling or reimplantation of SV (directly or via synthetic grafts) into the right atrium or left atrium (LA). In rare cases, affected lung lobectomy might also be required. However, these procedures cannot be used in case of unusual anatomic variants of SS. In this case study, a novel approach for the extracardiac repair of SS with dextrocardia and intact atrial septum having a common right venous chamber at the diaphragmatic surface and further opening at the RA- IVC junction is presented.

Case Description

An 8-year-old boy presented with New York Heart Association (NYHA) functional class II with a history of tachypnea and recurrent respiratory infections. The chest X-ray of the patient showed dextrocardia with plethoric lung fields. An echocardiogram confirmed dextrocardia and revealed anomalous drainage of right pulmonary veins into a common venous chamber at the diaphragmatic surface further draining into the inferior vena cava (IVC)–right atrium (RA) junction. There were no other intracardiac anomalies and the interatrial septum was found intact. Cardiac computed tomography (CCT) revealed complex scimitar anatomy. The right inferior pulmonary vein dilated to form a venous chamber. The right superior pulmonary vein drained into this chamber to form a common chamber at the diaphragmatic level and this complex was draining into RA- IVC junction.

Surgical intervention

Median sternotomy and trans pleural approach were taken. An intraparenchymal course of both veins was identified and the formation of a common venous chamber was appreciated. SV was located posterior to the hilum and dextrocardia was present, which pushed the left atrium further away from the common chamber. IVC was mobilized from diaphragmatic attachments.

Cardiopulmonary bypass (CPB) was instituted with aorto-bicaval cannulation with moderate hypothermia and aortic cross-clamping with cold root cardioplegia (Modified Del Nido Cardioplegia) was given. RA was opened and examined. Interatrial septum (IAS) was stabbed to the vent left heart. No opening of the common venous chamber was seen in the RA; however coronary sinus opening was seen to be dilated (no persistent left superior vena cava observed). Thus, the decision of extracardiac pericardial conduit (EPC) repair was taken. Common venous chamber disconnected from

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connection with coronary sinus and open end overrun with Prolene 6-0 (13mm) continuous suturing.

Preparation and anastomosis of EPC

Pericardial patch was harvested. Adequate size Hegar dilator was identified (using pulmonary vein z scores and Halgen Poiseulle equation) so as to match the opening of the disconnected end of the common chamber (14mm graft). Harvested prepared untreated pericardium was wrapped around Hegar dilator to make a tube using Prolene 8-0 9mm suture in a single layer continuous manner (Figure 1).

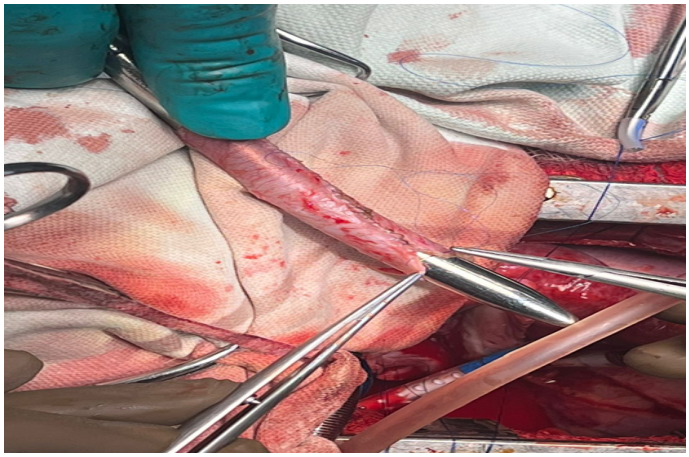

Figure 1: Construction of extracardiac pericardial conduit (black arrow)

The proximal end was anastomosed to open end with Prolene 6-0 13mm passed behind the IVC (Figure 2) and the distal end anastomosed to posterolateral wall of LA after rotating apex of the heart cephalad. IAS and RA closed, and the patient was weaned off cardiopulmonary bypass (CPB). Transesophageal echo confirmed unobstructed laminar flow across the graft tube.

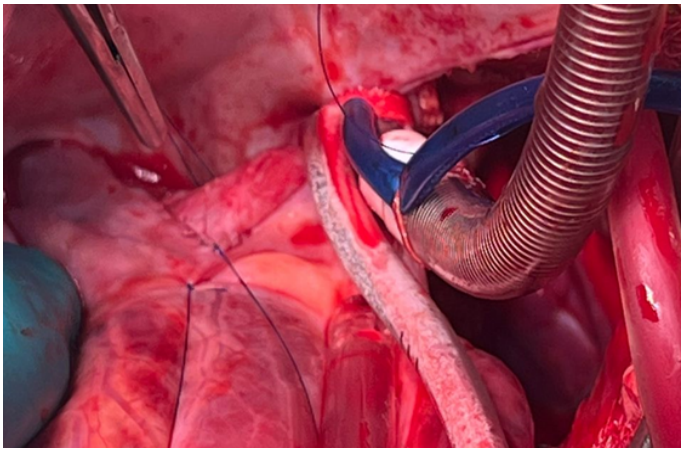

Figure 2: Pericardial conduit brought posterior to IVC (black arrow) anastomosed to LA

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The patient had an uneventful postoperative course. He started on an angiotensin-converting enzyme inhibitor (Ramipril), oral Digoxin, and antiplatelet drug (Acetyl Salicylic Acid) for 3 months in the postoperative period.

Postoperative 2D Echo performed on day 7 revealed good flow across the tube graft and no signs of leak. Postoperative central corneal thickness (CCT) after 6 months revealed a patent opacified pericardial tube graft (Figure 3).

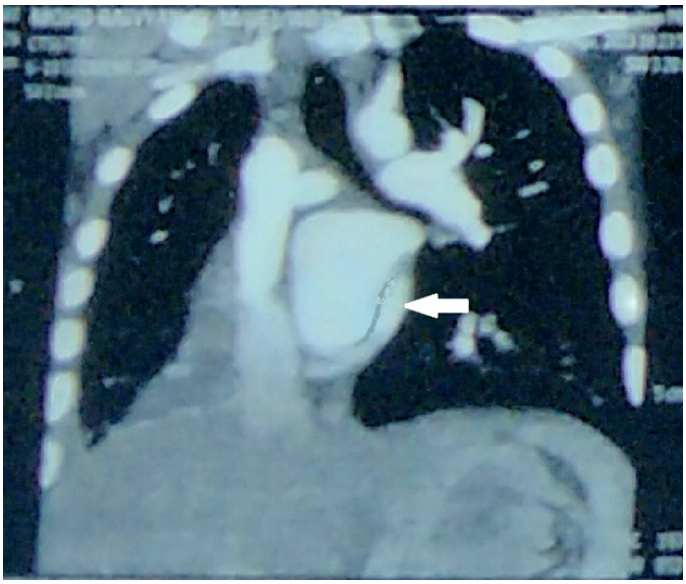

Figure 3: Patent well opacified pericardial conduit (white arrow) in cardiac CT done at 6 months.

Discussion

SS is a rare congenital anomaly with an incidence of two per 1,00,000 live births.1 While multiple methods are described for the repair of SS, intracardiac rerouting remains the most commonly used method owing to its advantage of expansion of venous orifice with growth. Nevertheless, the need for deep hypothermia and difficult LA access for anatomic variations like dextrocardia or abnormal situs and baffle thrombosis cannot be overlooked.[1] The other method most commonly used is reimplantation of the SV in LA or RA (either directly or with the use of ringed polytetrafluoroethylene (PTFE) conduits). In spite of being a complete anatomic repair with satisfactory intermediate results, the disadvantages of this technique include the requirement of extensive mobilization of the SV (complicated in dextroposition), change in angulation at hilum from acute to right angle after reimplantation leading to kinking on inflation of right lung and doubtful growing potential.[2-4] The choice of RA or LA depends on the length and mobility of SV and its relation to the lung hilum.[5] Risk of stenosis is similar in both the aforementioned methods.[6] The authors believe that the problems of deep hypothermia for rerouting, the debatable growing potential of conduits, inaccessibility in dextrocardia or anatomic variations like SV opening in the coronary sinus and high cost of synthetic grafts can be effectively tackled using an EPC graft.

Limitations

Delayed complication of anastomotic stenosis and collapse in EPC at LA end during systole needs to

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be looked for at follow up as mentioned by Lam et al.[7]

Conclusion

Authors would like to conclude by saying autologous pericardial tube grafts can prove to be an effective alternative to costlier options of synthetic grafts and show good midterm patency as evidenced in the report. Long term patency rate remains a topic of further study. This is the first study elaborating use of extracardiac pericardial tube graft for rerouting pulmonary venous complex to left atrium in scimitar syndrome as per review of literature.

References

1. Ak K, Cevik BS, Arsan S, Akalin F, Cobanoglu A. Repair of Scimitar Syndrome Using an Extracardiac Conduit in Pediatric Patients: Report of Two Patients. Ann Pediatr Cardiol. 2019;12(1):49-52. PubMed | CrossRef
2. Brown JW, Ruzmetov M, Minnich DJ, Vijay P, Edwards CA, Uhlig PN, et al. Surgical Management of Scimitar Syndrome: An Alternative Approach. J Thorac Cardiovasc Surg. 2003;125(2):238-45. PubMed | CrossRef
3. Cobanoglu A, Hanlon JT, Combs DT, Starr A. Combined Intracardiac and Extracardiac Repair of Scimitar Syndrome with Anomalous Pulmonary Veins to Both Cavae. J Thorac Cardiovasc Surg. 1984;88(1):141-6. PubMed
4. Guerra NC, Pernot M, Nesseris G, Al-Yamani M, Roques X, Thambo JB, et al. Scimitar Syndrome Repair in Adults: Intermediate-Term Results Using an Extracardiac Conduit. Ann Thorac Surg. 2016;102(6):2070-2076. PubMed | CrossRef
5. Pelletier GJ, Spray TL. Repair of Scimitar syndrome. Operative Techniques in Thoracic and Cardiovascular Surgery. 2001;6(1):32-49. CrossRef
6. Vida VL, Padalino MA, Boccuzzo G, Tarja E, Berggren H, Carrel T, et al. Scimitar Syndrome: a European Congenital Heart Surgeons Association (ECHSA) Multicentric Study. Circulation. 2010;122(12):1159-66. PubMed | CrossRef
7. Lam TT, Reemtsen BL, Starnes VA, Wells WJ. A Novel Approach to the Surgical Correction of Scimitar Syndrome. J Thorac Cardiovasc Surg. 2007;133(2):573-4. PubMed | CrossRef

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