Mind the Gap: Open repair of iatrogenic Cor Triatriatum Dexter After Prior Atrial Septal Defect Repair

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Mind the Gap: Open repair of Iatrogenic Cor Triatriatum Dexter After Prior Atrial Septal Defect Repair

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GLOSSARY OF ABBREVIATIONS
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<thead>
<tr>
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<th>Abbreviation</th>
<th>Full Form</th>
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</thead>
<tbody>
<tr>
<td>24</td>
<td>ASD</td>
<td>Atrial Septal Defect</td>
</tr>
<tr>
<td>26</td>
<td>CT</td>
<td>Computed Tomography</td>
</tr>
<tr>
<td>27</td>
<td>CTD</td>
<td>Cor Triatriatum Dexter</td>
</tr>
<tr>
<td>28</td>
<td>EV</td>
<td>Eustachian Valve</td>
</tr>
<tr>
<td>29</td>
<td>IVC</td>
<td>Inferior Vena Cava</td>
</tr>
<tr>
<td>30</td>
<td>POD</td>
<td>Post-Operative Day</td>
</tr>
<tr>
<td>31</td>
<td>RA</td>
<td>Right Atrium</td>
</tr>
<tr>
<td>32</td>
<td>RHC</td>
<td>Right Heart Catheterization</td>
</tr>
<tr>
<td>33</td>
<td>SV</td>
<td>Sinus Venosus</td>
</tr>
<tr>
<td>34</td>
<td>MR</td>
<td>Magnetic Resonance</td>
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<td>35</td>
<td>TEE</td>
<td>Transesophageal Echo</td>
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<td>36</td>
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Central Picture Legend: A/B. MRI; dilated IVC, thickened EV and IVC obstruction.

C.ASD  D.Bovine pericardial patch.

Central Message: Right-heart failure symptoms and history of cardiac intervention may suggest an ‘iatrogenic’ cor triatriatum dexter. Although rare, clinical awareness is important to avoid extra-cardiac complications.
1. Introduction

Cor triatriatum dexter (CTD) is a congenital cardiac anomaly whereby a membranous structure divides the right atrium (RA) into two chambers. Normal embryological development involves regression of the right valve of the sinus venosus (SV); however, failure of regression can lead to CTD with varying clinical manifestations. We present a case of what we term ‘iatrogenic’ CTD, where the RA was divided into two chambers from a prior atrial septal defect (ASD) repair with persistent, thickened EV leading to liver cirrhosis.

2. Case Presentation

A 55-year-old male with a congenital ASD presented to the office, reporting an asymptomatic cardiac murmur from childhood and was diagnosed with an ASD at age 15. Right heart catheterization (RHC) showed an elevated Qp/Qs (pulmonary/systemic blood flow) suggestive of left-to-right shunting. The patient underwent surgical ASD closure at age 17 at another institution. The operative report is unavailable. Patient provided written consent for this publication; IRB approval not required.

The patient was asymptomatic throughout much of adulthood. At age 45, his elevated liver enzymes prompted liver imaging and biopsy, which showed liver fibrosis due to venous congestion. He exhibited increasing fatigue with exertion, abdominal bloating, and lower extremity edema. Cardiac magnetic resonance (MR) showed normal biventricular function and a possible obstructive membrane at the IVC/RA junction. A local computed tomography (CT) angiography demonstrated a patent IVC without a membrane.
At our institution, repeat MR and CT confirmed liver disease with mechanical obstructive etiology. RHC with intracardiac echo showed a thick band of tissue at the IVC/RA junction, with evidence of flow acceleration at the CTD and an SV versus residual ASD (Figure 1 and 2). The IVC pressure was 17-18 mmHg (normal 5-10 mmHg), with normal pressures above the level of obstruction. The Qp/Qs was 1.05. On cardiac angiogram, the catheter could advance from the RA to the pulmonary vein, confirming a residual ASD.

While ballooning could have enlarged the IVC to RA opening, the benefit and durability of repair were unknown given the EV thickness, degree of liver dysfunction, and need to also address a residual ASD. Moreover, the patient desired definitive correction as he came from overseas. Our multidisciplinary consensus was that open heart surgery with excision of the remnant EV and closure of the ASD would be optimal.

In the operating room, transesophageal echo (TEE) confirmed the expected anatomy. The IVC was dilated; a very large azygous vein drained into the SVC. No cannulation sites were identified, suggesting the initial operation was completed under inflow occlusion. The RA was opened away from the prior transverse incision. A thickened EV, which was sewn to the inferior edge of a residual ASD with near occlusion of the IVC orifice, was excised to expose the entire IVC orifice. The ASD was enlarged, then a bovine pericardial patch sewn in. The patient was uneventfully weaned from bypass. TEE demonstrated no residual turbulence through the IVC; bubble study was negative.
Postoperative course was uneventful. The patient is doing well one year later with normalization of liver enzymes and significant improvement in liver stiffness, measured by MRI elastography showing average liver stiffness of 5.5 kPa and CAP score of 180.

3. Discussion

CTD is an extremely rare heart conditions owing to failure of tissue to regress during embryological development. In this patient, the RA being divided into two chambers by a membrane from prior ASD repair - an iatrogenic rather than developmental failure – is even rarer. We present a patient who underwent an ASD repair as a teenager and developed iatrogenic CTD that caused right-sided heart failure and liver cirrhosis, requiring surgical repair in his 50s.

Few papers highlight the varied clinical presentation of iatrogenic CTD. A 2011 case featured a 29-year-old female with asymptomatic CTD found on follow-up TEE after transcatheter ASD closure.2 Another case report of a 63-year-old man demonstrated CTD with exertional fatigue, lower extremity edema, and orthostatic hypotension.3 The patient had severe tricuspid stenosis and RA dilation, due to CTD-like anatomy caused by pacemaker lead fibrosis.

Our case report also illustrates the risks of inflow occlusion used in earlier era cardiac surgery.4 Surgeons in the 1970s would have considered our patient’s simple ASD closure a complete repair. However, systemic and cerebral malperfusion during inflow occlusion can cause cardiac and neurological complications, thereby heightening the risk of error caused by the initial ASD closure.5
Symptoms of right-sided heart failure, including hepatic cirrhosis, and history of cardiac intervention may suggest a diagnosis of iatrogenic CTD. Although rare, clinicians should know the causes and clinical manifestations of this anomaly to determine the most optimal treatment.
REFERENCES


FIGURE LEGENDS

Figure 1: A. Intracardiac echocardiogram (ICE) demonstrates a thick band of tissue at the inferior vena cava/right atrial (IVC/RA) junction, suggesting a prominent eustachian valve (EV) leading to cor triatriatum dexter (CTD). B. CT demonstrating the eustachian valve thickness and obstruction.

Figure 2: Enlarged atrial septal defect before (A) and after (B) bovine pericardial patch.