Cor Triatriatum

Review of the Surgical Aspects with a Follow-up Report on the First Patient Successfully Treated with Surgery

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SUMMARY
A follow-up report on the first patient with cor triatriatum to undergo successful surgical treatment is presented. A second procedure was necessary 9½ years later because of stenosis of the initial surgically created orifice in the anomalous diaphragm. A review and analysis have been made of 17 cases from the literature reporting successful operations for this lesion. Total excision of the accessory septum utilizing cardiopulmonary bypass is presently the appropriate surgical treatment of this entity. Preoperative and postoperative hemodynamic data are discussed. Recognition and correction of this defect are emphasized in order to avoid permanent pulmonary vascular changes attended by a relatively fixed cardiac output.

Additional Indexing Words:
Cardiac output Pulmonary vascular disease Congenital heart disease

Cor TRIATRIATUM is a rare lesion. Information regarding its course, diagnosis, and potential correction is relatively sparse. Reported herein is the case of an adult which demonstrates several facets of interest in the surgical treatment of the condition. A review of the literature on operative correction also accompanies this report.

Report of Case
A 24-year-old man was admitted to the University of Minnesota Hospitals in August 1954 with the complaint of progressive exertional dyspnea of 2 years’ duration. A heart murmur had been discovered at the age of 17 years during a bout of “pneumonia.” At the time of admission he had severe dyspnea on climbing one to two flights of stairs but he had no other complaints.

The pertinent physical findings in this normally developed male included a right ventricular thrust in the absence of cardiomegaly, normal sinus rhythm with infrequent extrasystoles, an accentuated second sound in the second left interspace with the relative intensities of the two components not described, and a blowing diastolic murmur along both sternal borders. No other abnormalities were noted.

The thoracic roentgenogram showed the heart to be normal in size with a suggestion of mild left atrial enlargement. The pulmonary artery segment was prominent. Many small, discrete densities were scattered throughout both lung fields suggesting hemosiderosis. The electrocardiogram revealed occasional premature atrial contractions, right axis deviation, P pulmonale, and right ventricular hypertrophy with strain.

The initial right heart catheterization was performed at rest on August 12, 1954 (table 1),
Table 1

*Heart Catheterization Data*

<table>
<thead>
<tr>
<th></th>
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<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Resting hemodynamics</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pressures (mm Hg)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery wedge</td>
<td>m = 26</td>
<td>m = 8</td>
<td>m = 2-12</td>
<td>(1-4)‡</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>36/15, m = 23</td>
<td>92/44, m = 56</td>
<td>22/14, m = 18</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>35/3</td>
<td>82/0-3</td>
<td>17-30/0-7</td>
<td>(2, 3)</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>97/4-6</td>
<td>115-165</td>
<td>ED = 5-12</td>
<td>(4)</td>
</tr>
<tr>
<td>Oxygen consumption (cc/min/m²)</td>
<td>134</td>
<td>123</td>
<td>166</td>
<td>(3, 5)</td>
</tr>
<tr>
<td>Cardiac index (L/min/m²)</td>
<td>2.8</td>
<td>2.1</td>
<td>2.5</td>
<td>(6)</td>
</tr>
<tr>
<td>Total pulmonary res. (dynes sec cm⁻³)</td>
<td>368</td>
<td>705</td>
<td>120-350</td>
<td>(5, 7, 8)</td>
</tr>
<tr>
<td>Pulmonary arteriolar res. (dynes sec cm⁻³)</td>
<td>205</td>
<td>160</td>
<td>50-160</td>
<td>(3, 5, 7)</td>
</tr>
</tbody>
</table>

| **Exercising hemodynamics**          |               |               |              |          |
| Pressures (mm Hg)                    |               |               |              |          |
| Pulmonary artery wedge               | m = 9         |               |              |          |
| Pulmonary artery                     | 36/22, m = 27 |              |              |          |
| Right ventricle                      | 42/2-2.5      |              |              |          |
| Oxygen consumption (cc/min/m²)       | 504           |              |              |          |
| Cardiac index (L/min/m²)             | 3.4           |              |              |          |
| Total pulmonary res. (dynes sec cm⁻³) | 318           |              |              |          |
| Pulmonary arteriolar res. (dynes sec cm⁻³) | 212           |              |              |          |

*Data from three separate procedures with normal values from the literature for comparison. Findings on the systemic side of the circulation were normal on each occasion. Surgery was performed April 1955 and November 1964.
†Zero base line for supine pressure measurements taken as one half of chest thickness measured from table top at the second costocondral junction.¹
‡Numbers in parentheses are the references from which the normal values were taken or calculated.
Abbreviations: m = mean; ED = end diastolic.
and revealed a normal cardiac index with mild pulmonary hypertension and a mildly elevated total pulmonary resistance. An increase in oxygen saturation from the superior vena cava (64%) to the right atrium (74%) suggested the diagnosis of atrial septal defect.*

Surgical correction was recommended but declined by the patient. In April 1955, atrial fibrillation and congestive heart failure developed during a bout of “pneumonia.” These conditions were controlled by medical therapy, and the patient was prepared for surgery. In May 1955, using hypothermia and inflow occlusion, the atria were explored and the third chamber was discovered, as previously described.9,10 An incision was made in the diaphragm separating this chamber from the left atrium, a new communication 2.5 cm in diameter resulted.9 Recovery from the operation was uneventful.

The patient became asymptomatic and led a physically vigorous life until early in 1964 when he again noted the insidious onset of progressively severe dyspnea on exertion. Diuretic therapy yielded only temporary improvement in his symptoms and by October 1964 his exercise tolerance was reduced to ascending one flight of stairs. Additionally, he was intermittently troubled with two-pillow orthopnea.

Physical examination revealed occasional moist inspiratory rales at both lung bases. A mild right ventricular heave was felt, and the second sound was normally split with a normal respiratory variation, the pulmonic component being much louder than the aortic component. A faint blowing systolic murmur was present along the left sternal border.

The thoracic roentgenogram now showed definite right ventricular hypertrophy. The electrocardiogram was similar to the previous tracings but with more prominently notched P waves.

Repeat right and left heart catheterizations were carried out on October 20, 1964 (table 1). The cardiac index was low, but there was no right or left ventricular failure. There was marked elevation in the pulmonary artery wedge pressure to a mean level of 26 mm Hg and a 16 mm Hg end-diastolic gradient existed between the pulmonary artery wedge (that is, pulmonary venous pressure) and the left ventricular diastolic pressure. There was also moderate elevation of the pulmonary artery pressure, the total pulmonary resistance, and the pulmonary arteriolar resistance. The pulmonary artery pressure failed to fall after a slow infusion of 65 mg of tolazoline hydrochloride into the pulmonary artery. This suggested a fixed pulmonary vascular resistance.11 A pulmonary arteriogram was done for visualization of the venous phase, but filling of the left side was inadequate to define detail. The findings were deemed compatible with restenosis of the opening in the anomalous diaphragm in the left atrial chamber causing recurrent obstruction to the pulmonary outflow at the left atrial level.

Re-operation was performed in November 1964 for which cardiopulmonary bypass and moderate hypothermia were utilized. Exploration of the left atrium revealed a thick calcified diaphragm that divided the left atrium into two chambers except for two small communicating holes, 4 mm and 5 mm in diameter (fig. 1). The diaphragm was completely excised back to the atrial wall, converting the left atrium into one large single chamber.

Figure 1

Specimen removed at surgery in 1964. Excised diaphragm divided the left atrium and caused cor triatriatum. The two small holes provided the only communication between the pulmonary veins and mitral valve. The superior hole was the residual of the original incision in 1955.

*In retrospect the increase in oxygen saturation from the superior vena cava to the right atrium was probably the result of mixture with more saturated blood from the inferior vena cava.
Dramatic relief of symptoms followed surgery, and the patient's exercise tolerance increased markedly. However, he suffered frequent episodes of paroxysmal atrial tachycardia in the immediate postoperative interval.

At follow-up examination in December 1965, the right ventricular heave had disappeared, but the intensity of the pulmonic component of the second sound was undiminished.

Right heart catheterization was repeated on December 9, 1965 (table 1). During catheterization, he developed paroxysmal atrial tachycardia with a 2:1 block and a ventricular rate of 140 beats/minute while the catheters were being placed. The arrhythmia did not respond immediately to treatment, but the procedure, including a study of supine bicycle ergometer exercise, was completed. The resting cardiac index was at the lower limit of normal although oxygen consumption was at the upper limit of normal. The pulmonary artery wedge and right-sided pressures, as well as the total pulmonary resistance, had returned to normal. The pulmonary arteriolar resistance, however, had dropped less significantly and remained at the upper limit of normal. The persistent elevation of the pulmonary arteriolar resistance confirmed the previous conclusions of some fixed pulmonary hypertension drawn from the tolazoline hydrochloride infusion study. The exercise study was quite vigorous, raising the oxygen consumption over three times to 1,008 cc/min, but the cardiac output did not rise proportionally. That is, the cardiac index rose to only 3.4 L/min/m² compared to an expected value of at least 4.5 L/min/m² (based on a normal increase in cardiac index of 0.6 L/min/m² for each 100 cc/min/m² increase of oxygen consumption).5, 6, 12, 13 The inability to increase the cardiac output with exercise was attributed to the residual increased pulmonary arteriolar resistance since both this variable and the total pulmonary resistance increased with exercise.5, 7 The pulmonary artery wedge pressure remained normal during exercise indicating that left ventricular failure was not a factor.

Obviously, the presence of the arrhythmia complicates interpretation of the data.

Discussion

Cor triatriatum is a congenital cardiac anomaly in which there is a division of the left atrium by a diaphragm into (1) an upper accessory chamber into which the pulmonary veins drain, and (2) a lower chamber which communicates with the mitral valve.14 The two chambers communicate through an opening, and when it is small, pulmonary venous obstruction results. It is generally agreed that this lesion results primarily from failure of the pulmonary venous stems to incorporate into the definitive left atrium.10, 14, 15

The clinical manifestations of this lesion depend upon the degree of narrowing of the communication. Survival is directly related to the size of the orifice in the diaphragm and approximately 50% of the patients with this lesion die in infancy.16 Death almost always results from pulmonary edema.

Fontana and Edwards16 had only one case in their review of 357 congenital defects studied pathologically (0.3%) and Jegier and associates17 encountered two cases in 474 autopsies in cases of congenital heart disease (0.4%) over a 25-year period. Although rare, an awareness of this entity is important because of its potential correctability.

Although various authors early discussed the possibility of surgical treatment of cor triatriatum,18, 19 it was not until 1956 that the first reports of successful operative treatment appeared.9, 20 A review of the literature reveals 17 cases of successful surgery to date,9, 10, 17, 20-34 This count excludes the six "type B" cases of Lam and associates28, 35 which might better be designated anomalous pulmonary venous connection to the coronary sinus,36 since there are significant anatomic, pathological, and physiological differences from what has been defined as cor triatriatum.14 These 17 cases are summarized in table 2. Almost all of the successful surgical cases were those of older children and adults; this indicates that the obstructive component was relatively mild. Six had associated defects with three of these being atrial septal defects. Niwawama37 reported 33 cases in which 12 associated defects occurred, the most common being atrial septal defect and patent ductus arteriosus. In addition, calcium deposits occurred around the orifice of the diaphragm in four of his cases.

The correct preoperative diagnosis was made or suspected in only eight of the cases in table 2. Indeed, the diagnosis can be missed at the time of cardiotomy with definition
Table 2

Summary of Reported Cases of Successful Surgical Treatment of Cor Triatriatum

<table>
<thead>
<tr>
<th>Case</th>
<th>Reference no.</th>
<th>Age (yr.), sex</th>
<th>Associated defects</th>
<th>Preoperative diagnosis</th>
<th>Operative technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9, 10</td>
<td>29 M</td>
<td>Questionable atrial septal defect</td>
<td>Hypothermia, inflow occlusion, new opening cut under direct vision</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>20, 21</td>
<td>21 F</td>
<td>Questionable mitral stenosis and insufficiency, primary pulmonary hypertension, pulmonary venous obstruction, left atrial myxoma</td>
<td>Finger pushed through anomalous septum</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>22</td>
<td>17 M</td>
<td></td>
<td></td>
<td>Septum split from one end to the other with mitral commissurotomy knife</td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>28 F</td>
<td>Mitral disease</td>
<td></td>
<td>Septum divided from side to side with mitral commissurotomy knife</td>
</tr>
<tr>
<td>5</td>
<td>24</td>
<td>33 F</td>
<td>Mitral stenosis</td>
<td></td>
<td>Hole clipped in membrane with transventricular scissors then opened on both sides with finger</td>
</tr>
<tr>
<td>6</td>
<td>10, 25</td>
<td>3 F</td>
<td>Cor triatriatum</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>26, 27</td>
<td>9 M</td>
<td>Atrial septal defect</td>
<td>Cardiopulmonary bypass, septum excised, atrial septal defect closed</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>28</td>
<td>6 M</td>
<td>Cor triatriatum</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>29</td>
<td>7 M</td>
<td>Atrial septal defect</td>
<td>Cardiopulmonary bypass, septum excised, atrial septal defect closed</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>30</td>
<td>33 F</td>
<td>Mitral stenosis</td>
<td>Cardiopulmonary bypass using mitral commissurotomy knife</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>17, 31</td>
<td>1½ M</td>
<td>Cor triatriatum or mitral stenosis</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>32</td>
<td>1 F</td>
<td>Cor triatriatum</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>32</td>
<td>19 M</td>
<td>Cor triatriatum</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>10</td>
<td>12 F</td>
<td>Situs inversus</td>
<td>Accessory chamber originally joining venous atrium was communicated with arterial atrium by creating atrial septal defect and walling off new channel from venous atrium</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>33</td>
<td>12 F</td>
<td>Persistent left superior vena cava</td>
<td>Cardiopulmonary bypass, septum excised, persistent left superior vena cava ligated</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>34</td>
<td>22 F</td>
<td>Atrial septal defect</td>
<td>Cardiopulmonary bypass, septum excised, atrial septal defect closed</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>34</td>
<td>4 F</td>
<td>Hemianomalous connection of left pulmonary veins</td>
<td>Cardiopulmonary bypass, septum excised</td>
<td></td>
</tr>
</tbody>
</table>
later at the time of another surgical procedure\textsuperscript{19, 22, 38} or at autopsy. The finding of normal left atrial pressure in the presence of elevated pulmonary artery wedge pressure suggests the diagnosis if the left atrium can be entered at catheterization.\textsuperscript{34} Pulmonary arteriography with delayed exposure has been emphasized recently as an aid to the making of the correct diagnosis.\textsuperscript{32–34} However, the left side of the heart may be inadequately filled in some instances, as in the case reported herein, with failure to yield a definitive diagnosis.\textsuperscript{21, 25, 38} A later report\textsuperscript{39} tells of making the diagnosis through visualization of the left atrium by means of contrast medium regurgitated through the mitral valve at the time of transient arrhythmia induced by the injection into the left ventricle, but this seems to represent a fortuitous occurrence rather than a reproducible diagnostic procedure.

Several different surgical procedures have been performed for this entity (table 2). Five of the patients, primarily those of the early reports, had closed procedures similar to those done for mitral stenosis. The diaphragm was split with the finger or mitral commissurotomy knife. Ten patients have had complete excision of the diaphragm utilizing cardiopulmonary bypass. In one patient with situs inversus a more complex procedure was involved in the correction and consisted of creating a new channel between the accessory chamber, which had joined the venous atrium and the arterial atrium.\textsuperscript{10}

It seems quite possible that patients operated on by a closed technique (patients 2 to 5 and 10; table 2) have or will develop a recurrence of their lesion as exemplified by the patient reported on herein (patient one; table 2). Cardiopulmonary bypass with complete excision of the membrane should be the procedure of choice for patients with cor triatriatum.

An anatomic corrective procedure does not imply an immediate return to a normal hemodynamic state. Several authors have reported the results of cardiac catheterization studies performed in patients with this lesion at intervals 4 to 12 months after surgery.\textsuperscript{10, 17, 20, 23, 25, 32} Their findings in general have been a normal pulmonary artery wedge pressure, normal or slightly elevated pulmonary artery pressure, and slight elevation in the pulmonary vascular resistance with a normal cardiac output at rest. The catheterization findings in our patient were similar. However, with exercise definite functional impairment was manifest. The inability to increase cardiac output appropriately with exercise was probably due to the residual pulmonary arteriolar changes.\textsuperscript{5, 7} Despite this finding the patient reports excellent symptomatic relief. Miller and associates\textsuperscript{32} reported similar results in their patient catheterized at both 11 days and 9 months postoperatively. It will be of interest to recatheterize this patient again in the remote postoperative interval to assess further the reversibility of these arteriolar changes.

References

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