**COR TRIATRIUM**

**BY**

N. R. BARRETT AND JOHN B. HICKIE

London

Sydney

Cor triatrium is a congenital malformation of the heart in which a transverse septum divides the left atrium into two chambers, separating the opening of the pulmonary veins from the mitral orifice. It is a rare cardiac anomaly. In an analysis of 1,000 necropsies of congenital heart disease Abbott (1946) found it seven times, although in 10 specimens anomalous cordae were present in the left atrium.

The condition was mentioned by Andral (1829), and described originally by Church (1868). The latter reported a woman of 38 years who died with pulmonary congestion and congestive cardiac failure, and necropsy revealed a membranous septum stretched across the left atrium. Since then 23 cases have been described. Barnes and Finlay (1952) state that this septum “appears to be so eminently suitable for surgical treatment that every effort should be made to recognize the abnormality during life.”

The following patient is interesting in that mitral valve disease was suggested by clinical and physiological studies, but the true diagnosis was made after cardiotomy, and subsequently a successful operation was performed. The pressures in the various cardiac chambers were recorded before and after division of the left atrial septum.

**CASE REPORT**

A man aged 17 years had mild exertional dyspnoea and bronchitis for many years, with increasing breathlessness and productive cough for two years. One week before his first admission he had an haemoptysis.

On examination he had a pigeon chest and a dorsal kyphosis. The pulse was regular, the blood pressure 110/60 mm. Hg, and the jugular venous pressure normal. There was no cardiac enlargement, but a right ventricular impulse was palpable. There was an accentuated split second sound at the base, a normal mitral first sound, no opening snap, and a grade one mitral diastolic murmur. In the electrocardiogram there was right ventricular hypertrophy and broad notched P waves. Radiography showed enlarged pulmonary arteries, right ventricular enlargement, and no valve calcification. A diagnosis of mitral stenosis was made.

Six months later he had a further haemoptysis and was readmitted. The signs were unaltered except that the mitral diastolic murmur was heard with great difficulty and its presence doubted by most observers. Cardiac catheterization (Table I) showed pulmonary hypertension, an elevated “pulmonary capillary wedge pressure” with a rise on exercise, and no evidence of an intracardiac shunt. The form of the “pulmonary capillary wedge tracing” was consistent with tight mitral stenosis. As the diagnosis was doubtful, an angiocardiogram was attempted, but rapid auricular fibrillation developed early in the procedure and it had to be abandoned. Normal rhythm was restored with “digoxin” and quinidine. Oximetric studies following Valsalva’s manoeuvre, before and after exercise, showed no evidence of an atrial septal defect.

**OPERATIVE FINDINGS**

The first thoracotomy was done without an accurate pre-operative diagnosis. The history and the physical signs were not typical of rheumatic heart disease, and it was upon probability that mitral stenosis was suggested as the most likely cause of the high pressure which was known to exist in the pulmonary vascular bed. This diagnosis was made in the absence of the typical murmurs, without evidence of left atrial enlargement, and without an opening snap.

The possibility that the obstruction in the atrium was caused by a myxoma, arising from the septum, was entertained, but was not considered to be probable.

We may wonder, in retrospect, if left atrial puncture, done either at bronchoscopy or by direct needling, would have clarified the issue. If the catheter or needle had entered the upper chamber a high pressure (different from that of the pulmonary artery) would have been recorded; if it could then have been jugged through the orifice in the septum, which seems doubtful considering its position, an accurate diagnosis

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**Table I**

**RESULTS OF INITIAL CARDIAC CATHETERIZATION**

<table>
<thead>
<tr>
<th>Position</th>
<th>Pressures</th>
<th>Oxygen (vol.)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
<td>Diastolic</td>
</tr>
<tr>
<td>S.V.C.</td>
<td></td>
<td>5</td>
</tr>
<tr>
<td>R.A.</td>
<td>92</td>
<td>0</td>
</tr>
<tr>
<td>P.A.</td>
<td>92</td>
<td>62</td>
</tr>
<tr>
<td>P.C.</td>
<td>52</td>
<td>32</td>
</tr>
<tr>
<td>P.C.E.X.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>B.A.</td>
<td>110</td>
<td>60</td>
</tr>
</tbody>
</table>

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COR TRIATRIUM

might have been made; in these circumstances there
would have been a zone of high pressure, then one of
normal pressure before the typical ventricular tracings
were obtained. If, on the other hand, the needle had
entered the lower chamber, normal atrial pressures
would have been recorded and a diagnosis of primary
pulmonary hypertension would have been suggested.

The left chest was opened as for a mitral valvotomy.
The lung was congested and did not deflate easily.
The heart appeared to be normal in size; but the pul-
monary artery was dilated and so were the left pul-
monary veins. The pericardium was normal. The left
atrium was enlarged and its upper surface was
dark in colour and firm to palpation; the auricular
appendage felt, and looked, normal; a thrill was pal-
ipple at the top of the left atrium in the groove
between it and the pulmonary artery. No anomalous
vessels were seen.

The auricular appendage was opened in the usual
way. It contained no clot and the mitral valve felt
normal in anatomy and movement. Exploration of
the atrium itself showed it to be unusual, but it was
not obvious what the abnormality was. The lower
part of the chamber presented nothing suspicious, but
the upper two-thirds was tense. It felt as though this
area were occupied by a large, soft, doughy "tumour;
but this diagnosis was improbable because the ex-
ploring finger could not be passed to the top of the
chamber. The lower surface of the "mass" was
smooth and bulged downwards: it could be indented
and compressed, but immediately regained its shape.
On further palpation it became obvious that the lower
surface was, in fact, a septum passing from a point
just below the entry of the left pulmonary veins, across
the chamber to the inner side of the atrium above the
mitral ring. At this time the exploring finger un-
expectedly entered a small defect in the abnormal
septum: the aperture was about 1 cm. by ½ cm. in
extent and the margins were firm and contained prickly
calcification. In retrospect it is surprising that this
foramen was not discovered sooner, but it lay at some
distance from the surface of the atrium. Not having
the possibility of cor triatrium in mind, I (N. R. B.)
assumed that the lesion was one in which the inter-
atrial septum had grown in an unusual site and that
a defect had persisted in it. Because of this opinion
nothing further was done and the operation was con-
cluded. Convalescence was uneventful.

On talking the matter over with a colleague at the
Brompton Hospital (Dr. Gibson) the true diagnosis
was suggested, and it was obvious that the findings
at the first operation exactly fitted the bill. It should
be emphasized that the pre-operative investigations
had not shown evidence of an intratrial shunt, a fact
which made the diagnosis of atrial septal defect ex-
tremely unlikely. In retrospect the writer would sub-
mit that a surgeon familiar with this deformity would
have no difficulty in coming to a correct diagnosis at
operation.

A second thoracotomy was done in March, 1956.
The left atrium was explored and opened without
difficulty. The diagnosis of cor triatrium was con-
irmed by palpation and by taking the pressures in
the upper and lower chambers of the left atrium.
The foramen in the septum was too tough to split
or enlarge by finger fracture, but it was easily cut
with a mitral knife. Once the edges of the hole had
been incised the split in the septum was completed
from one wall of the atrium to the other. There was
no difficulty about the operation, although the possi-
bility of dislodging a spicule of calcification into the
systemic blood stream, and so of causing cerebral
embolism, was ever present.

When the septum had been split there was no differ-
ence in pressure between the top and the bottom of
the left atrium and the thrill had disappeared.

Convalescence was uneventful, and 10 months later
the boy was normal in every way; he plays football
with interest and efficiency, and says he has no
symptoms.

ANATOMY

In this condition a transverse fibrous or fibro-
muscular septum divides the left atrium into two
chambers, an upper or postero-superior one which
receives the pulmonary veins and a lower or
antero-inferior one which contains the orifice of
the atrial appendix and the mitral opening
(Palmer, 1930). The auricular appendix always
opens into the lower chamber. The upper
chamber is usually the larger and is often funnel-
or wedge-shaped. The membrane extends from
the interatrial septum in the region of the foramen
ovale to the antero-lateral wall of the atrium just
below the left inferior pulmonary vein. It usually
arises above the foramen ovale (Parsons, 1950), but
may arise below it (Edwards, DuShane, Alcott,
and Burchell, 1951) or at it, the medial edge of
the septum dividing into two to enclose the
foramen (William and Abrikossoff, 1911). The
two chambers communicate through one or
multiple small perforations in the dia-
aphragm usually near its centre or peripherally
between a free margin and the atrial wall (Palmer,
1930). The size of the opening may vary from a
pin hole (Parsons, 1950) to such a large opening
that there is no appreciable obstruction to the

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**TABLE II**

RESULTS OF CARDIAC CATHETERIZATION BEFORE AND
AFTER OPERATION

<table>
<thead>
<tr>
<th>Position</th>
<th>Pressure Before Operation</th>
<th>Pressure After Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
<td>Diastolic</td>
</tr>
<tr>
<td>L.V.</td>
<td>87</td>
<td>0</td>
</tr>
<tr>
<td>Aorta</td>
<td>86</td>
<td>58</td>
</tr>
<tr>
<td>P.A.</td>
<td>65</td>
<td>43</td>
</tr>
<tr>
<td>R.V.</td>
<td>65</td>
<td>0</td>
</tr>
<tr>
<td>L.A. 1</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>L.A. 2</td>
<td>—</td>
<td>—</td>
</tr>
</tbody>
</table>

L.A. 1 = antero-inferior chamber of left atrium.
L.A. 2 = postero-superior chamber of left atrium.
circulation (Potter and Ranson, 1904; Loeffler, 1949). In two cases the septum was complete (Stoebner, 1908; Tannenberg, 1930). The border of the perforation is often thickened and may contain small calcareous deposits (Pedersen and Therkelsen, 1954), or there may be calcification in the wall of the upper chamber (Hartmann, 1955), and areas of endocardial fibro-elastosis in the upper chamber, pulmonary veins, and mitral valve (Doxiadis and Emery, 1953). In all cases but one (Parsons, 1950) the mitral valve was normal. In another case (Patten and Taggart, 1929) the extra chamber was situated anteriorly communicating with both atria.

On the exterior of the atrium a depressed groove may make the line of the septum (Church, 1868) and the pulmonary veins appear dilated and have thick, pale walls (Church, 1868; Doxiadis and Emery, 1953; Pedersen and Therkelsen, 1954).

 Associated defects are not frequent, the most common being patent foramen ovale and atrial septal defect. In 10 of those cases in which it is mentioned the foramen ovale was closed, and in five it was open or there was an atrial septal defect. Abnormal drainage of the pulmonary veins was reported in three cases (Stoebner, 1908; Patten and Taggart, 1929; Becu, Tauxe, DuShane, and Edwards, 1955) and Fallot’s tetralogy in two (Preisz, 1890).

The effect on the circulation and the ultimate prognosis depends on the degree to which the intra-auricular septum prevents blood flow to the mitral valve, the size and patency of the foramen ovale, and the relationship of the septum to the pulmonary veins and the foramen ovale. In most cases the obstruction to blood flow from the lungs causes pulmonary congestion, pulmonary hypertension, right ventricular hypertrophy, and ultimately systemic congestion.

Those cases in which abnormal cords are present in the left atrium are related to the true cor triatrium (Rolleston, 1896; Hepburn, 1896; Martin, 1899; McNamara, Baker, and Costich, 1947). Most often there is a thin, thread-like cord, but there may be a broad band (Fowler, 1882; Griffith, 1903; Loeffler, 1949) or a reticular structure (de Vecchi, 1901; Helwig, 1933). The cord is always attached by one end, at or in the neighbourhood of the valve or limbus of the fossa ovalis, while the other end may be attached anywhere in the atrium, at the mitral valve, or even through the left ventricle into the aorta (Goforth, 1926). Such cords or bands have no effect on the circulation and do not produce symptoms or diagnostic problems.

**Embryology**

Cor triatrium is a congenital anomaly. Fowler (1882) suggested that it was due to overgrowth of the valve of the foramen ovale, while Martin (1899) thought that it was the result of incomplete disappearance of the wall between the common sinus of the pulmonary veins and the left atrium. Palmer (1930) considered the abnormal septum to be an inversion or direct extension of the wall of the pulmonary vein, so that the postero-superior chamber was the dilated end of the pulmonary vein and the smaller antero-inferior chamber the normal left atrium. Loeffler (1949) came to similar conclusions and called it the “heart with a pulmonary sinus.” Parsons (1950), in an extensive review of the literature, concluded that there was developmental arrest in the second month of foetal life causing a defect at the junction between the pulmonary vein and the left atrium. Edwards and others (1951) have suggested the alternative title of “congenital stenosis of the common pulmonary vein,” but cor triatrium remains the commonly used term.

**Clinical Symptoms and Signs**

Clinically there are two variants. First, those patients in whom there is little or no connexion between the accessory chamber and the left atrium: in them the foramen ovale is usually patent (Doxiadis and Emery, 1953) or there is anomalous pulmonary venous drainage. All die in infancy, usually under 4 months of age, of pulmonary congestion and heart failure. In the second group the septum produces an organic obstruction to pulmonary vein blood flow. The foramen ovale is closed and there are no other anomalies. The most prominent feature of this group is respiratory embarrassment secondary to pulmonary congestion (Parsons, 1950) with secondary pulmonary hypertension and right ventricular failure. Recurrent haemoptyses may occur (Church, 1868; McLester, Bush, and DuBois, 1940; Hartmann, 1955). On auscultation, a systolic murmur has been reported in eight patients, a diastolic murmur in two, and no murmur in four. In this case, a questionable diastolic murmur was present. In no case has prominence of the mitral first sound or an opening snap been reported. Radiographic findings in 11 patients have revealed either a generalized globular cardiac enlargement or, as in this patient, right ventricular, left auricular, and pulmonary arterial enlargement with pulmonary congestion (Parsons, 1950).
Hartmann, 1955). Cardiac catheterization (Petersen and Therkelsen, 1954) is suggestive of mitral disease with pulmonary hypertension, an elevated pulmonary capillary wedge pressure, and a rise on exercise. In our patient, the form of the tracing was consistent with mitral stenosis. Angiocardiography has not been reported; it was attempted in this patient, but auricular fibrillation caused it to be abandoned.

**DISCUSSION**

Cor triatrium is a congenital lesion which is particularly amenable to surgical treatment. The majority of the patients reported have succumbed in early childhood, at which time the diagnosis is difficult. The remainder, as in this patient, presented with symptoms of pulmonary congestion, pulmonary hypertension, and ultimately of right heart failure. The clinical picture is indistinguishable from mitral valve disease, but the mitral murmurs may be atypical or absent. The mitral first sound is normal, and there is no opening snap. Radiography, electrocardiography, and cardiac catheterization are of little or no assistance; all are consistent with mitral valve disease.

The operative findings are the most important, and depend upon an awareness of the condition, the external features of the atrium, and digital exploration of that chamber. The condition should be considered in obscure or suspicious cases of pulmonary hypertension associated with a raised pulmonary capillary wedge pressure. The diagnosis is assisted by pressure measurements in the pulmonary veins and left atrium, and they also serve as an indication of the effectiveness of the surgical procedure.

**SUMMARY**

The successful operative treatment of a case of cor triatrium in a youth of 17 years is reported. This is a rare congenital anomaly in which a transverse septum divides the left atrium into two chambers. The anatomy, embryoology, symptoms, signs, electrocardiographic, radiographic, and cardiac catheterization findings in this condition are reviewed.

**REFERENCES**


