Isolated Ventricular Septal Defect of the Persistent Common Atrioventricular Canal Type

By Henry N. Neufeld, M.D., Jack L. Titus, M.D., James W. Dushane, M.D., Howard B. Burchell, M.D., and Jesse E. Edwards, M.D.

In recent years certain electrocardiographic patterns have come to be associated with the congenital cardiac malformation called "persistent common atrioventricular canal."1-4 Experience has shown, however, that the electrocardiographic features are not specific for that malformation. They may be observed also in cases in which both great vessels arise from the right ventricle without pulmonary stenosis,5 in certain cases of ventricular septal defect,6 in some cases of atrial septal defect of the ostium secundum type,7 and in certain cases of acquired heart disease. The same features are observed in cor triloculare biventriculare (common atrium), but this malformation may be considered a variant of persistent common atrioventricular canal.8

The purpose of this study was to review those cases of anatomically proved ventricular septal defect occupying the position of the ventricular component of the defect in the complete form of persistent common atrioventricular canal. An answer was sought to the question of whether this anatomic character of the ventricular septal defect could account for the electrocardiographic findings, namely, early QRS vectors being directed to the left and the QRS vectors in the frontal plane describing a counterclockwise loop and lying mostly superior to the isoelectric point.

Search of the clinical, surgical, and pathologic files of the Mayo Clinic (involving 60 necropsy specimens and more than 300 cases of ventricular septal defect observed at operation) yielded 15 cases of isolated ventricular septal defect of the anatomic AV commune type (table 1). In six cases the anatomic nature of the defect was studied only at the time of cardiectomy under the conditions of extracorporeal circulation. In the other nine cases necropsy specimens were available for study. Each specimen was examined by conventional means; in four cases the specimens were studied also by examining serial sections of the conduction system.

Pathologic-anatomic features of ventricular septal defects have been studied and described, among other investigators, by von Rokitansky,9 Becu and associates,10 Kirklin and co-workers,11,12 and Warden and associates.13 Kiely and associates14 emphasized the possibility that isolated ventricular septal defects of the common atrioventricular canal type may occur with or without involvement of the atrioventricular valves.

Pathologic Features

In the group of 15 cases described in this paper (cases 1 to 15, table 1), the pathologic features (figs. 1 and 2) were as follows. From the right ventricular aspect, the ventricular septal defect extended anteriorly to the crista supraventricularis. A part, at least, of the membranous septum was identifiable and formed the anterosuperior boundary of the defect. The defect did not extend anteriorly beyond the papillary muscle of the conus. The posterior edge of the defect extended more dorsally than does the more common type of defect, and its entire superior edge lay immediately beneath the tricuspid ring. From the left ventricular view, the defect was positioned beneath the membranous portion of the...
Figure 1

Case 2. a. Right atrium and ventricle. Ventricular septal defect (D.) is overhung by septal leaflet (S.) of tricuspid valve. Although no striking cleft is apparent in tricuspid valve, minor defect appears to be present in tissue between septal leaflet and anterior leaflet. Chordae of septal leaflet of tricuspid valve are deficient. Anterosuperior extremity of defect is formed by crista supraventricularis (C.) b. Right ventricular aspect and pulmonary trunk (P. T.). Ventricular septal defect (D.) abuts on inferior aspect of crista supraventricularis (C.). c. Left ventricular aspect. Ventricular septal defect (D.) lies in front of anterior leaflet of mitral valve (M.) and involves considerable amount of muscular part of ventricular septum where it forms wall of outflow tract of left ventricle. Beneath aortic valve is membrane representing membranous portion of ventricular septum, which forms upper boundary of defect. The mitral valve was devoid of clefts. Case 3. d. Tricuspid valve and adjacent structures. Beneath septal leaflet of tricuspid valve (S.) is ventricular septal defect (D., pointing to probe in defect). Anterosuperior extremity of defect is bounded by crista supraventricularis (C.). Tricuspid valve is devoid of clefts. e. Right ventricular aspect and pulmonary valve (P. V.). Ventricular septal defect (D.) lies between septal leaflet of tricuspid valve below and crista supraventricularis above (C.). At border of defect crista supraventricularis has an arch-shaped deformity. This corresponds to position of left ventricular aspect of defect. Right ventricular hypertrophy is pronounced. (T. O. = tricuspid orifice.) f. Left ventricular aspect. In front of anterior leaflet of mitral valve (M.) is large ventricular septal defect (D.). Defect extends for considerable distance into anterior part of muscular portion of ventricular septum, which forms wall of outflow tract of left ventricle. Aortic valve (A. V.) does not form immediate boundary for defect; instead, a fragment of membranous portion of ventricular septum is attached to aortic valve and forms upper boundary of defect.
VENTRICULAR SEPTAL DEFECT

ventricular septum. The upper edge of the defect was separated from the aortic leaflets by the membranous portion of the ventricular septum or by the musculature that corresponds to the crista supraventricularis (figs. 1 and 2). In contrast to what is found in the common type of ventricular septal defect, the aortic valve did not form a boundary of the defect in the 15 cases considered herein.

In two cases (cases 1 and 6, table 1) a cleft was found in the septal leaflet of the tricuspid valve. In two cases (cases 7 and 9) a cleft was present in the anterior leaflet of the mitral valve. In four cases (cases 11 to 14) both the septal leaflet of the tricuspid valve and the anterior leaflet of the mitral valve showed a deformity or a cleft. In three cases there was an associated atrial septal defect at the fossa ovalis, and in one case there was no cleft, but both atrioventricular valves were continuous through the defect. In no case was an atrial septal defect of the ostium primum type present.

These findings indicate that the degree of the deficiency varied. In some cases the deficiency occurred only in the junction of the lower portion of the atrioventricular endocardial cushions; the remainder of the region of the atrioventricular canal had developed normally. In other cases the deficiency apparently included the atrioventricular valves but not the atrial septum.

Embryology

The complete and partial varieties of persistent common atrioventricular canal—their embryology, pathology, and clinical picture—are now well known.15-20 It is also known that in some cases only an isolated malformation of the atrioventricular cushion may be found. This malformation may exist as an isolated ostium primum, as an isolated cleft of the mitral or tricuspid valve or of both, and as an isolated ventricular septal defect with or without cleavage of the atrioventricular valves.14

Kramer21 demonstrated that, embryologically, the endocardial cushions make a specific contribution to the closure of the ventricular septum. The atrioventricular cushions at the base join the muscular part of the ventricular septum to obliterate the interventricular communication in this region. Final closure of the embryonic interventricular foramen results from fusion of elements of the endocardial cushions of the atrioventricular canal, the conus septum, and the endocardium of the muscular septum.15, 21 The ventricular septal defect of the type described herein appears to result from failure of fusion of these three elements. Failure of fusion not only allows a defect to remain, but it also deprives the developing muscular septum of its anchors. As a result of the mechanical tensions of the growing heart, the unanchored muscular septum is pulled away from the atrioventricular region, causing enlargement of the defect.

If we are correct in reasoning that the ventricular septal defect under consideration is primarily a result of deficiency of atrioventricular cushion, it seems logical to consider the defect a variant of persistent common atrioventricular canal. The anatomic position of the defect, and, as will be shown, the similarity of the unusual excitatory pattern in the electrocardiogram as compared with electrocardiographic phenomena found in common atrioventricular canal, as well as the similarity in the conduction system and associated lesions in the atrioventricular valves, support our assumption.

The Conduction System

Mönckeberg22, 23 found that in patent foramen ovale and ventricular septal defect, the location and distribution of the atrioventricular node, bundle of His, and bundle branches were normal. He and Keith24 also found that the bundle always passed below the defect. Morison,25 in a report of a case of a malformed heart with atrial septal defect, which, from the illustrations, appeared to be of the ostium primum type, found the atrioventricular node displaced posteriorly and below the coronary sinus. The fibers of the bundle split into an abortive left branch, which failed to reach the left ventricle, and a large right branch, which divided to supply...
Figure 2

Case 9. a. Right side of heart. Ventricular septal defect (D.) is overhung by septal leaflet of tricuspid valve (S.), which has been partly retracted upward to expose defect. Chordae from under aspect of atrioventricular valvular tissue are inserted in lower edge of defect. Anterosuperior extremity of defect is formed by crista supraventricularis (C.). b. Left ventricular aspect. Ventricular septal defect (D.) cuts into muscular portion of outflow tract of ventricular septum. Beneath aortic valve (A. V.) is membranous portion of ventricular septum, which is continuous with anterior leaflet of mitral valve (M.). Chordae from anterior aspect of atrioventricular valvular tissue insert into edges of ventricular septal defect, as shown in a. c. Close-up view of ventricular septal defect (D.) from left ventricular aspect, showing extension of chordae across ventricular septal defect and at same time extensive deficiency of anterior portion of muscular part of ventricular septum in this region. (A. V. = aortic valve.) Case 14. d. Right side of heart. Beneath septal leaflet of tricuspid valve (S.), which is cleft, is ventricular septal defect (D.). Anterosuperior boundary of defect is formed by crista supraventricularis (C.). e. Left ventricular aspect. Anterior leaflet of mitral valve (M.) is cleft. Chordae from edges of cleft insert into anterior wall of ventricular septal defect (D.). Aortic valve (A. V.) lies above defect and is separated from it by elements of membranous portion of ventricular septum. f. Left atrium and left ventricle. Ventricular septal defect (D.) is overhung by cleft of anterior leaflet of mitral valve (M.). Chordae from edges of crest of mitral valve insert into anterior wall of ventricular septal defect on its right ventricular aspect.
both the right and left sides. Wallgren and Winblad,\textsuperscript{26} studying a case of persistent ostium primum associated with complete atrioventricular block, were unable to find continuity between the atrioventricular bundle and the node. Lev\textsuperscript{27} found in four cases of the complete variety of persistent common atrioventricular canal that the conduction system presented a fairly common pattern. The sinoatrial node was normal. The atrioventricular node was displaced posteriorly, originating mainly from the posterior wall of the atrium. In the absence of a well-formed atrial and ventricular septum, the atrioventricular bundle was displaced posteriorly, skirting the lower margin of the defect. In some instances the right bundle branch gave off several strands rather than continuing as a single trunk.

In two of our cases (cases 1 and 5) the conduction system was studied histologically by making serial sections of blocks taken from the region of the atrioventricular junction; the posterior limit of the blocks was approximately the ostium of the coronary sinus, and the anterior limit was the crista supraventricularis. The blocks included approximately 1 cm. of interatrial septum superiorly and 2 cm. of interventricular septum inferiorly. By study of serial sections of such blocks, the locations of the atrioventricular node, common atrioventricular bundle (bundle of His), and right bundle branch could be determined. This method is similar to that used in other histologic studies of the conduction system (fig. 3).

The atrioventricular node appeared to be in its usual location. The bundle arose from the anterior extension of the node and penetrated the central fibrous body in the usual fashion. This course placed the most anterior extension of the bundle just above the posterior-superior angle of the ventricular septal defect. At this point the conduction fibers turned downward posteriorly behind the defect and followed the posterior edge of the ventricular septal defect in the form of an arc (fig. 3). These abnormalities are explicable in the light of what is known of the embryogenesis of the conduction system. In the mammal the atrioventricular node and bundle originate from the posterior part of the musculature of the atrial canal that lies

\textbf{Figure 3}

\textit{a. Interior of right side of heart; conduction system. Atrioventricular node (N.) is in usual location. Main bundle (M. B.) arises from ventral extent of node and runs to branching point (B.); here right bundle (R. B.) begins and continues in an arc dorsally to defect. b. Photomicrograph of conduction tissue. M. V. = mitral valve; T. V. = tricuspid valve; B. = bundle; B. B. = branching bundle; V. S. = ventricular septum; V. S. D. = ventricular septal defect.}
of persistent common atrioventricular canal considered herein are similar to those found in the fully developed stage of the malformation as reported by Lev.27

Electrocardiographic Findings (Table 1)

Rhythm. In one case (case 3) there was an ectopic atrial rhythm. In the other 14 cases normal sinus rhythm was recorded, with multiple premature ventricular beats occurring in two of these.

P-R Interval. In all 15 cases the P-R interval could be measured. In seven cases it was considered to be normal, and in eight cases the value was at the upper limit of normal or considered to be prolonged in relation to heart rate and age28 (table 1).

P Wave. Alteration of either the P wave or the QRS complex, or both, suggestive of enlargement of the right atrium, was found in one case (case 1). In two cases (cases 7 and 9) configuration of the P wave was indicative of enlargement of the left atrium. In another case (case 2) the alteration was suggestive of enlargement of both atria, and in the remaining five cases that demonstrated anatomic evidence of malformed atrioventricular valves, the changes in the P wave were not diagnostic.

The electrocardiographic signs of enlargement of the left atrium or right atrium or both were compatible with the anatomic findings of mitral insufficiency or tricuspid insufficiency, or both, as noted at necropsy or at operation. In two cases (cases 7 and 12) wedge pressures were obtained; they showed increased pressure (mean pressure of 20 and 26), with the V waves being prominent. In case 12, normal P waves were seen on electrocardiograms.

QRS Complex. The mean electrical axis of the QRS complex in the frontal plane was estimated in all cases. The mean electrical axis lay between -30° and -160° (table 1 and fig. 4). The duration of QRS was 0.07 second or less in five cases, between 0.07 and 0.08 second in five cases, and greater than 0.08 second in five cases. Vector analysis of the QRS complex in the frontal plane (table 1 and figs. 5 to 7) revealed in all cases that the
<table>
<thead>
<tr>
<th>Age and sex</th>
<th>Heart rate</th>
<th>PR interval (II), second</th>
<th>QRS (II), second</th>
<th>QRS axis</th>
<th>Frontal vector loop</th>
<th>QRS pattern</th>
<th>Lead V₁*</th>
<th>Lead V₅*</th>
<th>Lead I</th>
<th>Hemodynamic data</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 19 yr. F</td>
<td>100</td>
<td>0.16 0.08</td>
<td>-110° C$</td>
<td>$</td>
<td>rR's</td>
<td>r = 2</td>
<td>2.0</td>
<td>-</td>
<td>0.5</td>
<td>8.0</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 25.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 126/0 39% (O₂)</td>
</tr>
<tr>
<td>2 7 yr. F</td>
<td>95</td>
<td>0.28 0.07</td>
<td>-160° C</td>
<td>Figure-8</td>
<td>rsR'</td>
<td>r = 2</td>
<td>-</td>
<td>2.0</td>
<td>7.0</td>
<td>23.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 23.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 95/5</td>
</tr>
<tr>
<td>3 24 yr. M</td>
<td>67</td>
<td>0.24 0.10</td>
<td>-60° C</td>
<td></td>
<td>rsR'</td>
<td>r = 2</td>
<td>-</td>
<td>7.0</td>
<td>23.0</td>
<td>10.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 23.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 113/6 72% (air)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>39% (air)</td>
</tr>
<tr>
<td>4 24 yr. M</td>
<td>74</td>
<td>0.16 0.09</td>
<td>-50° C</td>
<td></td>
<td>rR'</td>
<td>r = 1.0</td>
<td>+</td>
<td>2.0</td>
<td>25.0</td>
<td>13.0 +</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 18.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 109/3 64% (air)</td>
</tr>
<tr>
<td>5 7 yr. M</td>
<td>96</td>
<td>0.18 0.07</td>
<td>-100° C</td>
<td></td>
<td>RS</td>
<td>20.0</td>
<td>13.0</td>
<td>+</td>
<td>4.0</td>
<td>22.0 +</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 80/0</td>
</tr>
<tr>
<td>6 28 yr. F</td>
<td>82</td>
<td>0.16 0.10</td>
<td>-60° C</td>
<td></td>
<td>rsSr'</td>
<td>r = 2</td>
<td>-</td>
<td>11.0</td>
<td>10.0</td>
<td>3.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 2.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 25/0 52% (air)</td>
</tr>
<tr>
<td>7 1-2/3 yr.</td>
<td>130</td>
<td>0.14 0.07</td>
<td>-160° C</td>
<td></td>
<td>Rs</td>
<td>25.0</td>
<td>5.0</td>
<td>-</td>
<td>6.0</td>
<td>32.0 1.0 -</td>
</tr>
<tr>
<td>F</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 60/0 W.P.1</td>
</tr>
<tr>
<td>8 5-1/2 yr.</td>
<td>112</td>
<td>0.20 0.09</td>
<td>-70° C</td>
<td></td>
<td>Rs</td>
<td>21.0</td>
<td>2.0</td>
<td>-</td>
<td>23.0</td>
<td>8.0 -</td>
</tr>
<tr>
<td>F</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 74/0</td>
</tr>
<tr>
<td>9 11 mo. M</td>
<td>138</td>
<td>0.16 0.07</td>
<td>-50° C</td>
<td></td>
<td>RS</td>
<td>14.0</td>
<td>10.0</td>
<td>-</td>
<td>3.5</td>
<td>23.0 18.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 87/3 33% (air)</td>
</tr>
<tr>
<td>10 5 yr. F</td>
<td>100</td>
<td>0.17 0.08</td>
<td>-90° C</td>
<td></td>
<td>RS</td>
<td>11.5</td>
<td>12.0</td>
<td>+</td>
<td>2.0</td>
<td>14.0 7.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 86/3</td>
</tr>
<tr>
<td>11 5-1/2 yr.</td>
<td>106</td>
<td>0.21 0.08</td>
<td>-30° C</td>
<td></td>
<td>Rs</td>
<td>8.5</td>
<td>14.5</td>
<td>-</td>
<td>2.0</td>
<td>13.0 1.0 -</td>
</tr>
<tr>
<td>M</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>-</td>
</tr>
<tr>
<td>12 11 yr. F</td>
<td>95</td>
<td>0.16 0.08</td>
<td>-60° C</td>
<td></td>
<td>rsR's</td>
<td>r = 2</td>
<td>s = 10</td>
<td>1.5</td>
<td>25.0</td>
<td>9.0 +</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 56/6 30% (air)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>s' = 3</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>20% (air)</td>
</tr>
<tr>
<td>13 2 yr. F</td>
<td>108</td>
<td>0.14 0.08</td>
<td>-70° C</td>
<td></td>
<td>rsR's</td>
<td>r = 1.5</td>
<td>3.0</td>
<td>-</td>
<td>0.5</td>
<td>11.0 5.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 26.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>- 120/14</td>
</tr>
<tr>
<td>14 7 yr. F</td>
<td>90</td>
<td>0.16 0.09</td>
<td>-160° C</td>
<td></td>
<td>Figure-8</td>
<td>r = 1.5</td>
<td>-</td>
<td>0.5</td>
<td>15.0</td>
<td>21.0 +</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>+ 100/5 70% (O₂)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>21% (O₂)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>35% (air)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>54% (air)</td>
</tr>
<tr>
<td>15 2 mo. M</td>
<td>123</td>
<td>0.10 0.06</td>
<td>-100° C</td>
<td></td>
<td>rsR's</td>
<td>r = 1.0</td>
<td>s = 1.0</td>
<td>0.5</td>
<td>20.0</td>
<td>20.0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>R' = 17.0</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>-</td>
</tr>
</tbody>
</table>

*Values for Q, R, and S are in millimeters.
†Expressed as percent of total pulmonary flow.
‡Expressed as percent of systemic flow.

§C = Counterclockwise.

| | | | W.P. = Wedge pressure. |
instantaneous vector advanced in a counterclockwise direction and was mainly superior to the isoelectric point. In two cases (cases 2 and 14, fig. 5) the vectorcardiogram showed a narrow figure-of-eight configuration of the QRS loop placed horizontally across the isoelectric point, with the initial loop rotating in a counterclockwise direction.

Chest leads were obtained in 14 of the 15 cases. Various configurations of the QRS complex in lead V₁ were found. In eight cases the configuration of QRS complexes in lead V₁ (rsR', rSr') was suggestive of “diastolic (or volume) overloading” of the right ventricle. In the remaining cases the configuration of the QRS complex was RS. The ratio of R to S in lead V₆ was indicative of overloading of the right ventricle in 10 cases.

In lead V₆, two different types of QRS configurations were observed: RS in two cases and QRS in 12. In only three cases was the ratio of R to S in lead V₆ equal to or less than 1 (cases 1, 14, and 15). One of these three was the case of a 2-month-old child. In the remaining cases the ratio of R to S in lead V₆ was greater than 1.

T Wave. The T wave was found to be negative in lead I in eight cases (table 1, figs. 6 and 7). In two cases it was biphasic; the meaning of this finding is not clear to us. These changes in lead I were found to be rare in an electrocardiographic-anatomic study of 50 cases of the usual type of ventricular septal defect, and they are not emphasized in the literature.

Special Anatomic Features

The ventricular septal defect described herein and considered to be of the persistent common atrioventricular canal type has an anatomic peculiarity: the upper edge of the defect is not bordered by muscle but rather by the tricuspid ring. There are other defects of the ventricular septum that, because of their location, resemble the one described; however,

![Electrocardiogram in case 8, in which anatomic diagnosis was proved at operation. (See table 1.)](image)

![Electrocardiogram from case 11, in which anatomic diagnosis was proved at operation. (See table 1.)](image)
VENTRICULAR SEPTAL DEFECT

these defects are completely surrounded by muscle and so have a band of muscle separating them from the tricuspid ring.

Findings in Two Related Cases

Special study of two cases was of interest from the point of view of the conduction tissue and the electrocardiograms, although they did not fall into the category of the 15 cases presented in this paper. In one case the pathologic examination revealed two defects (fig. 8). One was located posteriorly, close to the tricuspid ring, and the other was found more anteriorly; a band of muscle surrounded the posterior defect and separated it from the anterior one. Study of the conduction system revealed that the bundle of His and its right branch lay between the two defects, in the muscle separating them (fig. 8). The electrocardiogram in this case showed the usual pattern found in ventricular septal defect. The mean manifest electrical axis of the QRS complex was $+100^\circ$, with a clockwise loop of the QRS in the frontal plane on the scalar electrocardiogram.

In the second case a large ventricular septal defect was located posteriorly. A band of musculature was found between the ventricular septal defect and the tricuspid ring (fig. 9). Although in this case the defect was unusually positioned and suggested the atrioventricular canal type, careful observation proved it to be of the muscular type of defect. Study of the conduction system showed that the right bundle lay ventral to the defect, in its usual position. The electrocardiogram showed a mean electrical axis of the QRS complex of $+100^\circ$ and a clockwise loop in the frontal plane. There were signs of right ventricular overloading and, in some cases, of left ventricular overloading as well.

Discussion

The electrocardiographic findings in our 15 cases have been thought by some authors to be virtually diagnostic of persistent common atrioventricular canal. This study indicates that such electrocardiograms are also characteristic of isolated ventricular septal defect of the types described in this paper. They have also been noted in cases having both great vessels arising from the right ventricle without pulmonary stenosis, occasionally in other types of ventricular septal defect, and in some cases of atrial septal defect of the ostium secundum type; opportunity has not yet been afforded for careful histologic study of the conduction system in the latter conditions. This defect is different anatomically and hemodynamically from the so-called common persistent atrioventricular canal. It differs surgically also, the surgical risk in these cases apparently being higher than in the usual type of ventricular septal defect.

The electrocardiographic findings were striking. In all instances the mean electrical axis of the QRS complex lay above the isoelectric point, with a vector loop directed in a counterclockwise direction; the chest leads indicated right ventricular “diastolic (volume) overloading” or right ventricular hypertrophy. These electrocardiographic changes
cardiographic findings do not correlate either with the size of the left ventricle or with the degree of mitral regurgitation estimated during operation.2

Toscano-Barboza and co-workers1 assumed that the position of the "defect at the top of the ventricular septum" causes the alteration in excitation. In 1959 Ellis and associates6 described five cases of common atrium in which the electrocardiographic findings were similar to those found in persistent common atrioventricular canal. Neufeld and associates5 found similar electrocardiographic changes in patients with origin of both great vessels from the right ventricle without pulmonary stenosis. All these findings, together with the electrocardiographic patterns of alteration in excitation in cases of ventricular septal defect of the atrioventricular type, suggest the presence of a congenital anomaly or distortion of the conduction system.

Burchell and associates12 opinion, based on recordings of electrocardiographic potentials from the cavities and surfaces of the ventricles, is that the different orientation of the advancing fronts of depolarization is the result of congenital absence of a portion of the left bundle-branch system.

A factor to be considered is the constant finding of a change in the anatomic location of the conduction system in cases of persistent common atrioventricular canal, as described by Lev27 and as observed in our cases (fig. 3). The bundle retains its primitive relationship to the posterior endocardial cushion and is mislocated posteriorly, skirting the posterior rim of the defect. The elongation and looping of the bundle around the defect (fig. 3) are congenital anomalies, and the developmental basis of these anomalies of the conduction system is probably the same. Such anomalies may be the cause of the alteration in excitation that gives the characteristic pattern on the electrocardiogram.

**Summary**

Among 60 necropsy specimens of isolated ventricular septal defects and more than 300 cases of ventricular septal defect observed at

*Circulation, Volume XXIII, May 1961*
operation, 15 cases demonstrating unusual anatomic positions of the defect were found. The ventricular defect differed from the ordinary ventricular septal defect in that it usually occupied the position of the ventricular component of the defect in persistent common atrioventricular canal. For this reason it was named ventricular septal defect of the persistent common atrioventricular canal type.

Deformities of one or both atrioventricular valves were common (nine of 15 cases). No atrial septal defects of the ostium primum type were present. Anatomic studies of the conduction tissue revealed that this tissue skirted the posterior and inferior aspects of the ventricular septal defect and that, as in persistent common atrioventricular canal, the course taken by the conduction tissue was unusually long, as a result of the peculiar posteroinferior position of the lower edge of the defect.

The electrocardiographic features were striking. In all cases the mean electrical axis of the QRS lay above the isoelectric point; the vector loop obtained in the frontal plane from the scalar electrocardiogram was directed counterclockwise, and its main mass was above the zero line. In addition, in all cases there were signs of right ventricular overload, and in some cases of left ventricular overload as well.

Electrocardiographic findings of this pattern have been thought by some authors to be diagnostic of persistent common atrioventricular canal, but we observed that they also occurred in each of the cases of isolated ventricular septal defect of the variety described herein. We recognize that in the usual variety of ventricular septal defect this electrocardiographic pattern occurs, but it does so uncommonly. We have not studied its exact incidence. The anatomy, hemodynamics, and surgical considerations are different in cases with this defect from those with persistent common atrioventricular canal. The surgical risk in these cases has been higher than that in the usual type of ventricular septal defect.

In the discussion of the electrophysiologic theories that seek to explain the unusual electrocardiographic patterns in this group of cases, a new theory is offered, based on studies of the conduction system. In our opinion, the different orientation of the advancing fronts of depolarization is the result of congenital displacement of the bundle of His in its relation to the ventricular septal defect.

References


Isolated Ventricular Septal Defect of the Persistent Common Atrioventricular Canal Type
HENRY N. NEUFELD, JACK L. TITUS, JAMES W. DUSHANE, HOWARD B. BURCHELL and JESSE E. EDWARDS

Circulation. 1961;23:685-696
doi: 10.1161/01.CIR.23.5.685
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1961 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/23/5/685

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/