Complete Transposition of the Great Vessels

II. An Electrocardiographic Analysis

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The electrocardiographic patterns in infants with complete transposition of the great vessels are neither so uniform nor so diagnostic as those in some other types of congenital cardiac disease. Zuckermann,1 Sodi-Pallares,2 and their respective associates have indicated that certain T-wave configurations may contribute to the diagnosis of complete transposition of the great vessels. Generally, however, the electrocardiogram is considered to be nonspecific.

Several other investigators3-5 have related a defect in the ventricular septum with (1) the degree of axis deviation, (2) the configuration or height of the R wave in lead V1, (3) the per cent R of RS in leads V4R and V1, and (4) the height of the RS voltage in the midprecordial leads. These studies, however, have directed little or no attention to the possible electrocardiographic influences of the size of the ventricular septal defect, nor have they taken into account additional forms of communication that may be present—such as a large patent ductus arteriosus.

The difficulty in evaluating electrocardiographic data in patients with complete transposition of the great vessels has stemmed from several factors. The complex assortment of malformations designated as "complete transposition of the great vessels" is bound to have resulted in confusion; thus, some conditions that include transposed great vessels are to be distinguished from the specific entity, complete transposition.

The purpose of this study was to review the electrocardiographic findings in 54 patients who had complete transposition of the great vessels and in whom confirmation of the malformation had been made at necropsy. Our particular purpose evolved into comparing results of analysis of the scalar electrocardiograms with size and type of anatomic communication between the two circulations.

Definition of Terms. As used in this report, "complete transposition of the great vessels" designates a condition in which there are two ventricles and two atrioventricular valves; the aorta arises entirely from the right ventricle, the pulmonary artery takes origin exclusively from the left ventricle, and a ventricular septum (with or without a defect) is present. Excluded from this study were all subjects having a single ventricle, as well as those showing atresia of an atrioventricular or semilunar valve, even though the great vessels were related in a transposed manner.

Materials and Methods

The basic material was a series of 54 infants with complete transposition of the great vessels in whom both scalar electrocardiographic studies and necropsy specimens were available. Each specimen of heart was examined for anatomic detail, the results of which are given in a separate report.6 The ages at time of death ranged between 1 week and 2 years: of the 54 patients, 28 were less than 6 weeks of age, 14 were between 2 and 6 months of age, 7 were between 6 and 12 months, and 5 were between 1 and 2 years.

Most of the electrocardiograms had been made with a Sanborn direct-writing multichannel unit. Readings were made of the 12 conventional leads and lead V4R. With use of the lead placement system of Schmitt,7 vectorcardiograms also were obtained in isolated cases. These are presented for

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Illustrative purposes only, to explain the various details in contour of deflection in the electrocardiographic tracings. They are not to be interpreted as an extensive study of the vectorcardiogram in complete transposition of the great vessels.

For each case the electrocardiograms were analyzed in two ways. The first consisted of studying individual components of the deflections in several leads, including a determination of the mean manifest electrical axis and direction of inscription of the QRS loop. The second consisted in viewing all leads together to determine whether one or another form of ventricular hypertrophy could be identified.

The criteria employed for signs of right and of left ventricular hypertrophy were those proposed by Vince and Keith, utilizing Ziegler's normal values. For identifying biventricular (combined right and left) hypertrophy, the sources mentioned were used but with certain modifications, yielding the following as criteria of biventricular hypertrophy:

1. Direct signs of right plus direct signs of left ventricular hypertrophy.

2. Direct signs of right ventricular hypertrophy in association with the following:
   a. Q wave 2 mm. or more in lead V6.
   b. T wave inversion in V6 after a positive T in right precordial leads.
   c. Counterelectric loops of the QRS loop in the frontal plane.
   d. Tall biphasic complexes in V1 in midprecordial leads over 50 mm. in height (Katz-Wachtel sign).

3. Direct signs of left ventricular hypertrophy associated with the following:
   a. R wave 15 mm. or more in lead V1.
   b. Tall biphasic complexes in midprecordial leads over 50 mm. in height.

There were six cases of complete transposition of the great vessels with special anatomic characteristics such as obstructive malformations of the aortic arch, left ventricular-right atrial communication, and pulmonary stenosis. These were set aside and the electrocardiographic observations on them will be described in a separate section under results. Removing these cases from the basic material left 48 cases among which comparisons could be made.

**Preliminary Study**

During the initial phases of this study, the basic 48 cases were separated into two groups: (1) those with an intact ventricular septum (26 cases), and (2) those with a ventricular septal defect, regardless of size (22 cases). At this phase of the study, no separation was made on the basis of the status of the ductus arteriosus.

According to earlier reports, defects in the ventricular septum should be suspected in patients with complete transposition who show any one or a combination of the following: (1) normal or left axis deviation, (2) an R per cent of RS in lead V1 of less than 75 per cent (in the presence of a loud murmur), (3) an R wave of less than 10 mm. in lead V4R, (4) adult progression of the QRS complexes in the precordial leads, (5) the Katz-Wachtel phenomenon in the midprecordial leads, or (6) certain T-wave configurations described by Zuckermann and Sodi-Pallares.

The aforementioned criteria for the presence of a ventricular septal defect were not confirmed by our material, as many of these electrocardiographic characteristics appeared in both anatomic groups during the pilot study. It was thus apparent that published electrocardiographic criteria for distinguishing between those cases with an intact ventricular septum and those with a ventricular septal defect were not uniformly reliable in our material.

In 47 of the 48 cases studied electrocardiographically (26 with intact ventricular septum and 22 with ventricular septal defect), some degree of right ventricular hypertrophy was observed. This existed in the form either of isolated right ventricular hypertrophy or within the framework of biventricular hypertrophy. For these reasons, the study was directed toward separation of the cases on a different basis.

Earlier studies had not taken into account the state of the ductus arteriosus. It was decided, therefore, to divide the patients into two groups according to the size, rather than the site, of the communication between the systemic and pulmonary arterial circulations.

We placed into one group (group I; "small communication"), those cases with a narrow or obliterated ductus arteriosus and either a small ventricular septal defect (subgroup I A; five cases) or an intact ventricular septum (subgroup I B; 22 cases). Into the second group (group II; "large communication") were placed those cases having either a large ventricular septal defect and closed or narrow patent ductus arteriosus (subgroup II A; 17 cases) or a widely patent ductus arteriosus and intact ventricular septum (subgroup II B; four cases). No cases were observed in which a widely patent ductus arteriosus and a large ventricular septal defect coexisted. An interatrial communication was present in each of the cases studied and took the form either of a valvular competent patent foramen ovale or a small atrial septal defect. No separation of cases was made on the basis of the type of interatrial communication.
Results

Group I—Small Communication (27 Cases)

P-R Interval and P Waves

In one of 27 cases with small communication, a Wolff-Parkinson-White pattern was observed. In this case, the P-R interval was shortened owing to the presence of a delta wave preceding the main component of the QRS complex. Among the remaining 26 patients, the P-R interval was prolonged for age and rate (0.16 second) in only one instance. The mean P vector in the frontal plane varied between +30 and +120 degrees. In most, the P vector approximated +50 degrees. The P waves were abnormal in 12 instances. Right atrial enlargement ("P pulmonale") was present in seven patients, and bialtrial enlargement was indicated in the other five.

QRS Complex (Analysis of Individual Components)

Mean Manifest Electrical Axis. The mean QRS axis in the frontal plane varied from +70 degrees to +215 degrees (fig. 1). Among the 27 patients, the average mean QRS axis was approximately +135 degrees. Right axis deviation for age was evident in 13 patients, a normal axis in 14. Vectorial analysis of the frontal plane showed the QRS loop to be directed clockwise in each of the 27 cases (fig. 1).

The QRS loop in the horizontal plane (precordial leads) was oriented anteriorly and to the right in 23 patients, anteriorly and to the left in four. Each of the latter four patients demonstrated a normal mean QRS axis in the frontal plane.

Lead V1. Various configurations of the QRS complex in lead V1 were observed. The three major configurations were: an rR's, an Rs, and a qR pattern (fig. 2). Uncommonly observed patterns were, an RS, an R, and an rsR'. The R per cent of RS in lead V1 was 75 or more in 23 of the 27 patients (85 per cent) and between 50 and 70 per cent in the remaining four.

Midprecordial Leads. The Katz-Wachtel sign (tall diphasic RS complexes at least 50 mm. in height in lead V2, V3, or V4) was observed in three instances.

Lead V6. In lead V6, essentially the following three variations in pattern were observed: rS, RS, and qRS. The most common pattern was that of a small r, and deep S wave (fig. 2). A Q wave, 2 mm. or more, was observed in four patients (15 per cent).

T Vector

The mean T vector in the frontal plane varied between +25 and +135 degrees. The majority of patients showed a mean T vector ranging between +30 and +70 degrees. The mean T vector in the horizontal plane (precordial leads) was oriented anteriorly in 15 patients (fig. 2, left) and posteriorly in six. It was perpendicular to the frontal planes in four, and it was indeterminate in two. Notched T waves in lead V6 were seen in two patients, and positive T waves, higher in the right precordial than in the left precordial leads, were observed in six patients.

Analysis for Hypertrophy Patterns

The types of ventricular hypertrophy among the 27 patients in group I were observed as follows:

Among four of five patients with a small ventricular septal defect and narrow ductus arteriosus (subgroup I A) and 18 of 22 with
Electrocardiograms and vectorcardiogram showing isolated right ventricular hypertrophy in two infants with complete transposition of the great vessels of the small communication type (intact ventricular septum and small patent ductus arteriosus). Left. The horizontal plane \((H)\) illustrates absence of the "Q loop" as shown by the first few milliseconds of the QRS sE loop directed anterior-leftward. This is reflected by absence of the Q wave in lead \(V_6\). \(F = \) frontal plane, \(S = \) left sagittal plane. Arrows denote direction of inscription. Right. Note the qR pattern in lead \(V_1\).

Table 1

Incidence of Ventricular Hypertrophy and Atrial Enlargement in Complete Transposition (48 Cases)

<table>
<thead>
<tr>
<th>P waves</th>
<th>Group I—Small communication (27 cases)</th>
<th>Group II—Large communication (21 cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Group I A: Small VSD (6)</td>
<td>Group I B: intact septum; small PDA (22*)</td>
</tr>
<tr>
<td>BVH</td>
<td>RVH</td>
<td>BVH</td>
</tr>
<tr>
<td>Normal</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>RAE</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>LAE</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>BAE</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Totals</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>

*One case with Wolff-Parkinson-White syndrome precluded evaluation of hypertrophy.

VSD, ventricular septal defect; PDA, patent ductus arteriosus; BVH, biventricular hypertrophy; RVH, right ventricular hypertrophy; RAE, LAE, and BAE, right, left, and bialtrial enlargement, respectively.
for age and rate (0.18 second) in two. The mean P vector in the frontal plane varied between +90 and +45 degrees, the average being +60 degrees. Atrial enlargement was observed in 14 of the 21 patients. Left atrial enlargement was observed in four patients, biatrial enlargement in seven, and right atrial enlargement in three (table 1).

QRS Complex (Analysis of Individual Components)

Mean Manifest Electrical Axis. In the 21 cases, the mean QRS axis in the frontal plane varied between -15 and +190 degrees, the average being approximately +120 degrees (fig. 1). Right axis deviation for age was evident in eight instances and a normal axis in 12; left axis deviation was observed in one. Among these 21 patients, the QRS loop was inscribed in a clockwise direction in 16 and counterclockwise in five (fig. 1).

The QRS loop in the horizontal plane (precordial leads) was oriented anteriorly and to the right in 13 patients, anteriorly and to the left in five, and to the left, equally anterior and posterior, in three.

Lead V₁. The QRS complex in lead V₁ showed varying configurations. The patterns were of the R, rsR', rR', and RS types. The percentage of R of RS in lead V₁ was 75 or more in 15 patients (71 per cent). In the remaining six patients the percentage of R varied between 45 and 72 (fig. 3, upper left).

Midprecordial Leads. Tall diphasic RS complexes over 50 mm. in amplitude in the midprecordial leads were observed in 10 patients (48 per cent; fig. 4, right).

Lead V₆. Essentially three patterns were observed in lead V₆. These included qRS, RS, and rS patterns. Among the 18 patients in whom a Q wave appeared in lead V₆ (85 per cent) the amplitude varied in depth from 1 to 6 mm. (figs. 3 and 4). Among 16 of these 18 patients, the Q wave was 2 mm. or more.

T Vector
The mean T vector in the frontal plane varied between zero and +160 degrees. In the vast majority of patients it ranged between +45 and +60 degrees. In the horizontal plane it was oriented anteriorly in 13 instances (fig. 3, lower left), posteriorly in five, and perpendicular to the frontal plane in three (fig. 3, upper right). Positive T waves, higher in the right precordial leads than in the left, were observed in three patients.

Analysis for Hypertrophy Patterns
In considering the type of hypertrophy patterns among the 21 patients comprising group II (large communication), the following were observed.

Among 13 of 17 patients with a large ventricular septal defect, and in each of the four patients with a large patent ductus arteriosus and intact ventricular septum, evidence of biventricular hypertrophy was observed (table 1). Thus, among 21 patients with large communication, biventricular hypertrophy was observed in 17 instances (80 per cent). In the remaining four patients with large ventricular septal defects, isolated right ventricular hypertrophy was observed.

Comparisons of Electrocardiographic Findings
In the two anatomic groups, the most reliable indication for the presence of a small communication was signs of isolated right ventricular hypertrophy, which were found in 22 of 27 (81 per cent) cases in group I (table 1); whereas, most indicative for large communication were electrocardiographic signs of biventricular hypertrophy, which appeared in 17 of 21 (80 per cent) cases in group II (table 1).

It is significant that none of the 48 patients comprising groups I and II exhibited signs of isolated left ventricular hypertrophy. Furthermore, among the patients with biventricular hypertrophy, signs of right ventricular hypertrophy predominated over signs of left ventricular hypertrophy in the vast majority of cases.

When it came to analyzing the differences among the individual components of the QRS complex, important distinctions were absent except, perhaps, for the following:

1. The direction of inscription employed by the QRS loop in the frontal plane: Among patients of groups I and II in whom the mean manifest electrical axis was normal, the QRS loop was inscribed counterclockwise only in cases with large communication (fig. 1).
Figure 3

Electrocardiograms and vectorcardiograms showing biventricular hypertrophy in three patients of the large communication type. In each, the initial cardiac vector ("Q loop") in the (H) plane is accentuated, producing relatively deep Q waves in left precordial leads. Abbreviations as in figure 2. Upper left. Large ventricular septal defect. Uncommon variety of biventricular hypertrophy. There is relatively more left ventricular predominance than is usually observed in patients with complete transposition. Upper right. Common variety of biventricular hypertrophy. Prevailing pattern is right ventricular hypertrophy [wide-open anterior-rightward clockwise QRS loop (H) plane producing extremely tall R waves in lead V1]. In contradistinction to tracings in figure 2, the "Q loop" is accentuated. Lower left and lower right. Tracings at 2 and 5 weeks respectively in patient with intact ventricular septum and large patent ductus arteriosus. In each, the prevailing pattern is right ventricular hypertrophy. Characteristically, the "Q loop" is accentuated. Note the classic figure-of-eight pattern of right ventricular hypertrophy (H plane) in lower right. N/2 = one-half standardization.

2. Q wave in lead V1: Only in patients with a small communication was a Q wave observed in lead V1. Furthermore, this sign among patients in this study is in itself significant, since other investigators4, 5, 13 rarely, if ever, observed this pattern in complete transposition.

3. Q wave in the left precordial leads: A Q
wave in lead V₆ was observed in only 15 per cent with small communication while 85 per cent with large communication exhibited this sign. This latter finding is significant, for Watson and Keith observed Q waves in lead V₆ in only 13 per cent of their patients with “transposition of the great vessels.”

**Cases with Unusual Associated Anomalies**

In addition to the foregoing 48 subjects, there were six cases with unusual associated anomalies. These will now be considered.

**Obstructive malformations of the aortic arch (two cases):** In two patients with an intact ventricular septum, a preductal coarctation was observed in one, and tubular hypoplasia of the aortic arch in another. Electrocardiographically, each showed evidence of mild degrees of left axis deviation for age (+15 degrees and -30 degrees), a counterclockwise QRS loop in the frontal plane, and coexisting left atrial enlargement. Left ventricular hypertrophy was observed in the patient with tubular hypoplasia of the aortic arch (fig. 5, upper), and biventricular hypertrophy (with predominant left ventricular hypertrophy) in the patient with coarctation of the aorta.

**Left ventricular-right atrial communication (two cases):** The typical electrocardiogram of patients with ventricular septal defects of the persistent common atrioventricular canal type was observed in two cases with a left ventricular-right atrial communication, namely, marked left axis deviation (-120 degrees and -90 degrees), a counterclockwise QRS loop in the frontal plane, and right or biventricular hypertrophy (fig. 5, lower left and lower right).

**Pulmonary valvular stenosis (two cases):** In each of the two patients with a large ventricular septal defect and pulmonary valvular stenosis, isolated right ventricular hypertrophy was observed.

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Figure 5
Upper. Electrocardiogram in a 1-month-old infant with complete transposition and coarctation of the aorta. There is mild left axis deviation for age. The precordial leads show relatively deep S waves in leads V4R and V1, indicating left ventricular hypertrophy. Lower left and lower right. Electrocardiograms in two patients with complete transposition and left ventricular-right atrial communication. Each shows features usually observed in patients with isolated ventricular septal defects of the "A-V commune" type.
Discussion

For various reasons, in patients with complete transposition of the great vessels, it is desirable to know the state of the ventricular septum.

Earlier reports have suggested that the presence of right axis deviation was a sign, among others, of an intact ventricular septum, whereas the presence of normal or of left axis deviation favored the existence of a ventricular septal defect. Our preliminary study did not establish such an association.

It seemed important, then, to work from a functional, rather than from a primary anatomic point of view, and to consider the influence of absence or presence of flow through the ductus arteriosus, as well as through the ventricular septum.

We divided our cases into two groups on this basis. Group I was composed of patients with "small communication," that is, showing either a narrow or obliterated ductus arteriosus and an intact ventricular septum or a small ventricular septal defect. Group II was formed by patients with large communication, that is, showing either a wide patent ductus arteriosus with an intact ventricular septum or a large ventricular septal defect. The essential difference between the two groups revolved about the type of hypertrophy exhibited.

In 47 of the 48 patients without unusual associated anomalies, signs of right ventricular hypertrophy were present. However, some showed only this sign while others showed signs of left ventricular hypertrophy, as well ("biventricular hypertrophy").

In group I (small communication) the electrocardiogram showed isolated right ventricular hypertrophy in 81 per cent of the cases. In group II (large communication) the electrocardiogram showed biventricular hypertrophy in 80 per cent of the cases.

These results indicate a strong (although not absolute) basis for distinguishing between cases with a small or large communication between the ventricles or great arteries.

When a large communication is present, the electrocardiogram cannot distinguish between a large ventricular septal defect and a wide patent ductus arteriosus associated with an intact ventricular septum.

To recognize signs of additional left ventricular hypertrophy in the presence of predominant right ventricular hypertrophy in complete transposition, one is dependent to a large extent upon the direction and magnitude of the initial vector forces of the QRS interval, as viewed in the horizontal plane. This feature, to our knowledge, has not been emphasized for complete transposition.

Cabrera and Gaxiola, among others, cited evidence that the initial forces of the QRS interval in the horizontal plane (termed by them a "Q loop") are signs primarily of depolarization of the ventricular septum. These initial cardiac vectors show increased magnitude when there is left or biventricular "overload." In contrast, the "Q loop" shows decreased magnitude when there is isolated right ventricular "overload." In the left precordial leads, increased magnitude of the "Q loop" is represented by significant Q waves (2 mm. or more), whereas decreased magnitude of the "Q loop" is represented simply as either an absent or small Q wave (2 mm. or less).

These views were confirmed by the presence of a Q wave, 2 mm. or more, in 16 of the 21 patients with large communication, and were further confirmed wherein we recorded both scalar electrocardiograms and vectocardiograms (fig. 2, left, and fig. 3).

In addition to the foregoing 48 patients, two showed obstructive malformations of the aortic arch, and two exhibited a left ventricular-right atrial communication. Left axis deviation was observed in each of these, whereas this electrical phenomenon was observed in only one patient with an uncomplicated large communication and in none of the patients with a small communication. Left axis deviation among patients with transposed great vessels usually suggests tricuspid atresia or common ventricle. Our observations, however, indicate that left ventricular-right atrial communication or obstructive malformations of the aortic arch should be considered if left axis deviation, associated with predominant.
signs of left ventricular hypertrophy, is observed in patients with complete transposition of the great vessels.

Summary

A correlative study of necropsy findings and electrocardiography in 54 cases of complete transposition of the great vessels was made to determine whether or not specific electrical patterns could predict the status of the ventricular septum. It became apparent that published criteria for distinguishing between patients with a ventricular septal defect and those with an intact ventricular septum were not uniformly reliable.

Rather than the location of a communication, the important factor in differentiation depended upon the size of the opening, be it between the ventricles or through a patent ductus arteriosus.

Cases were divided into two groups: In group I were those with a small communication, and in group II were those with a large communication between the greater and lesser circulations. The group with small communication showed a narrow or obliterated ductus arteriosus and an intact ventricular septum, or a small ventricular septal defect. In group II there was either a wide patent ductus arteriosus with an intact ventricular septum or a large ventricular septal defect.

In group I the electrocardiogram predominantly showed isolated right ventricular hypertrophy, whereas, in group II, biventricular hypertrophy was the usual finding.

Among the five patients in whom the mean manifest electrical axis was deviated to the left, two showed obstructive malformations of the aortic arch, and two left ventricular-right atrial communication. The fifth case was simply one of a large communication without complicating anomalies.

References

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