The Electrocardiogram in Tricuspid Atresia and Pulmonary Atresia with Intact Ventricular Septum

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MOST PATIENTS with cyanotic congenital heart disease show right axis deviation and right ventricular hypertrophy of more or lesser degree in the electrocardiogram. Left axis deviation by contrast has long been recognized as the cornerstone of the diagnosis of tricuspid atresia.¹ Although occasionally the A QRS is between 0° and +90° or may deviate slightly to the right in this condition, anatomic left ventricular hypertrophy is always noted at postmortem examination and the chest leads invariably indicate an adult R/S progression.

Within recent years another type of cyanotic congenital heart disease was described in which the A QRS is not deviated to the right and right ventricular hypertrophy is the exception rather than the rule.² ³ In patients with pulmonary atresia with intact ventricular septum, the frontal plane axis is between 0° and +90° in the vast majority. The chest leads in these patients show left ventricular hypertrophy if the right ventricular cavity is truly reduced; whereas, in the rare individual with adequate right ventricular volume, the chest leads will indicate right ventricular hypertrophy.

Although the electrocardiograms usually allow relatively easy differentiation of the two conditions, certain overlaps may occur. The present study was undertaken to analyze carefully the electrocardiographic features of tricuspid atresia and pulmonary atresia with intact ventricular septum. It is hoped that by careful attention to detail and correlation with anatomic findings the clinical diagnosis of the two entities may be placed on a firmer basis. It is also hoped that electrophysiological explanations of the observed differences may be offered.

Group Studied

All patients seen at the Children's Hospital Medical Center from the period January 1953 to June 1965 with the diagnosis of tricuspid atresia or pulmonary atresia with intact ventricular septum, proven by postmortem study or angiocardiography, were included in this investigation. In 37 the diagnosis was tricuspid atresia and in 17 of these the diagnosis was confirmed at postmortem examination. Of 20 patients with pulmonary atresia, 17 were examined at autopsy. The ages of the patients with tricuspid atresia, at the time of the electrocardiographic analysis, ranged from 10 days to 20 years with a median of 13 months (table 1). The ages of the patients with pulmonary atresia ranged from 2 days to 7 months with a median of 6 days (table 2). Twenty-five of the 37 patients with tricuspid atresia and nine of the 20 with pulmonary atresia were male.

Methods

Standard 12-lead electrocardiograms were obtained in all patients. Vectorcardiograms, using the Frank lead system, were obtained in 20 patients with tricuspid atresia and eight with pulmonary atresia. The maximal spatial voltage of the QRS and P loops, as well as the 0.01-second vector, was calculated by a technique previously described.⁴ For purposes of description left axis deviation was considered to be present when the QRS axis fell between 0° and

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Case 4, tricuspid atresia, type 1B. (A) Electrocardiogram showing the P “tricuspidale.” Note the first peak. Left axis deviation and an adult type of QRS progression in precordial leads. (Recorded at half standardization). (B) Vectorcardiogram. Note the early posterior orientation of the QRS loop, the counterclockwise inscription of the frontal plane projection, and the large, triangular-shaped P loop in the sagittal plane.

-90°; “normal” axis +1° to +90°, and right axis deviation from +91° to −91°. Electrocardiographic diagnosis of ventricular and atrial hypertrophy was defined according to criteria outlined by Nadas. Normal quantitative vectorcardiographic data for comparison was obtained from 15 infants aged 4 to 12 months.

The patients with tricuspid atresia were classified according to the anatomic types described by Keith and associates.2

Type 1. Without transposition of the great vessels
   A. With pulmonary atresia
   B. With pulmonary hypoplasia and a small ventricular septal defect
   C. No pulmonary hypoplasia and a large ventricular septal defect

Type 2. With transposition of the great vessels
   A. With pulmonary atresia
   B. With pulmonary or subpulmonary stenosis
   C. With a large pulmonary artery

The patients with pulmonary atresia were classified according to the types of Greenwold and associates.6

Tricuspid Atresia (Table 1)
Electrocardiographic Data

The P waves were peaked in lead I or II in all cases and exceeded 2.5 mm in height in 86%. Notching of this wave was observed in 30 patients (81%) with the first peak being taller in each instance (figs. 1 to 3*).

The distribution of the mean QRS axis in

*For purposes of publication figures 1, 2, 3, and 6 have been retouched.
Table 1

Anatomic, Electrocardiographic, and Hemodynamic Data in Patients with Tricuspid Atresia

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<th>Anatomic type</th>
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<th>P Wave (mv)</th>
<th>P Loop (mv)</th>
<th>Duration (sec)</th>
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Abbreviations: RAP (s/m) = right atrial pressure, systolic/mean; LMSV = left maximal spatial voltage.

*Intraventricular conduction defect.

the frontal and horizontal projections for each individual case is shown in figure 4 (A). Left axis deviation in the frontal plane was evident in 30 patients (81%), while normal or right axis deviation was present in seven cases, six of which were type 2C and the seventh was type 1A. The standard precordial leads presented an adult type of QRS progression in 32 (86%) out of the 37 patients. The five patients in whom this type of progression was not present showed an rS pattern in leads V1 through V6. The criteria for left ventricular hypertrophy proposed by Nadas was fulfilled in 85% of the cases, and a strain pattern in lead V6 was present in 51%.

Vectorcardiographic Features

The P loop was available for quantitative analysis in 18 of the 20 patients in whom vectorcardiograms were recorded. This loop...
showed an increased spatial magnitude in every case with a range of 0.23 to 0.54 mv and a mean value of 0.39 mv (normal mean, 0.19 ± 0.06 mv). A sizable area both anteriorly and posteriorly, diagnostic of biatrial hypertrophy, was observed in 14 patients and 10 of these displayed irregularities in contour (figs. 1 to 3).

The QRS loop presented characteristic features: the spatial 0.01-second vector was of increased magnitude in 15 cases of the 20 with a range of 0.14 to 0.37 mv and a mean of 0.23 mv (normal mean, 0.16 ± 0.07). The left maximal spatial voltage (LMSV) ranged from 0.3 to 4.1 mv with a mean of 2.4 mv. It was above the mean normal value (1.60 ± 0.50 mv) in all but three patients, two of them in group 2C who also presented reduced leftward voltage. The initial portion of the horizontal QRS loop (first 0.02) in 12 patients consisted of a narrow loop pointing anteriorly at first and turning posteriorly later. The body of the loop was posteriorly oriented in all 20 patients; in half of them it was inscribed counterclockwise.
Anatomic, Electrocardiographic, and Hemodynamic Data in Patients with Pulmonary Atresia

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<th>Anatomic</th>
<th>No.</th>
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<th>P Wave (mv)</th>
<th>P Loop (mv)</th>
<th>Duration (sec)</th>
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*Intraventricular conduction defect.

Table 2

The frontal QRS loop displayed a counterclockwise rotation in 16 patients.

Pulmonary Atresia (Table 2)

Electrocardiographic Data

Peaked P waves were observed in 14 of the 20 patients, and only in seven of these did the height of this exceed 2.5 mm. Notching, observed commonly in tricuspid atresia, was present in only two cases.

The distribution of the mean QRS axis in the frontal and horizontal plane is shown in figure 4 (B). Normal frontal plane axis was present in 12 patients, all of whom were type I (fig. 5). Right axis deviation was evident in eight patients including the three infants with type II deformity (fig. 6). There were no instances of left axis deviation. The standard precordial leads presented an adult type of QRS progression in 11 out of the 20 patients; whereas, right ventricular hypertrophy pattern was observed in five (three in type I, and two in type II). In four patients rS patterns were observed from V₁ through V₆. On the basis of the precordial voltage the diagnosis of left ventricular hypertrophy could be made in 10 patients, and right ventricular hypertrophy in five. Strain pattern in V₆ was present in seven infants.

Vectorcardiographic Features

The P loop showed leftward and anterior orientation in all eight patients for whom vectorcardiograms were available; the spatial magnitude was in the higher range of normality in seven and above normal suggestive of right atrial hypertrophy in one (range, 0.18 to 0.30 mv; mean, 0.22). No patient showed bialtrial hypertrophy.

In all eight patients the frontal QRS loop rotated clockwise. The horizontal loop was inscribed counterclockwise in six patients, all of them with type I lesions, and in each of these instances was posteriorly oriented indicating left ventricular hypertrophy. On the other hand, the two patients with anterior orientation and clockwise rotation, suggesting right ventricular hypertrophy, were of type...
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II. Contrary to the findings in patients with tricuspid atresia, there were no abnormalities of the initial forces in this group of infants. The mean left maximal spatial voltage was above normal (2.4 mv) similar to that in tricuspid atresia but with a narrower range (1.0-3.0 mv).

There was no significant relationship either in patients with tricuspid atresia or those with pulmonary atresia between the size of the atrial septal defect, right atrial pressure, or arterial \( O_2 \) saturation, and the height of the P wave or voltage of the P loop.

**Discussion**

Patients with tricuspid atresia and reduced pulmonary blood flow (types 1A, 1B, 2A and 2B) and those with increased pulmonary flow (type 1C) without transposition of the great vessels comprised 73% of the present series. In all but one patient (no. 2, table 1), the QRS axis was superiorly oriented and an adult progression of the QRS complex was seen in all but three (nos. 4, 6, 25, table 1) patients. Right ventricular hypertrophy was not present in any. True hypoplasia of the right ventricle was found in all patients, anatomically, in whom a postmortem examination was available. Cut section invariably revealed a small, thin-walled, right ventricle consisting essentially only of a pulmonary artery outflow tract. This absence of significant right ventricular muscle mass may well explain by itself the electrical left ventricular preponderance in the precordial chest leads. It might be expected that in the presence of hypertrophied left ventricle, as in tricuspid atresia, these effects would be further enhanced. The left axis deviation in these infants, however, cannot be adequately explained on the basis of left ventricular hypertrophy, and it still remains to be clarified. The frequent presence of left axis deviation could be related to conduction abnormalities in the left ventricle. Uhley and Rivkin produced left axis deviation in dogs, without prolongation of the QRS duration, by localized interruption of peripheral branches of the left ventricular conduction system. Furthermore, in tricuspid atresia, types 1C and 2B, the ventricular septal defect has
been described as being in a high membranous position similar to that found in cases of endocardial cushion type of ventricular septal defects, in which left axis deviation is a common finding.

Two features of the vectorcardiographic pattern in tricuspid atresia deserve comment. The characteristic first 0.02-second inscription of the QRS loop, with increased anterior and rightward forces followed by sharp posterior direction is present in 12 patients with tricuspid atresia. The counterclockwise inscribed body of the horizontal plane oriented leftward and posteriorly, noted in 10 infants, resembles the pattern of anterior myocardial infarction.

A possible explanation for both these phenomena may be found in the experiments of Horan and co-workers. These authors concluded that the right septal mass as well as the normal right ventricular wall contribute to the normal development of the counterclockwise rotation of the horizontal QRS loop. Presumably, in patients with tricuspid atresia, the lack of significant right septal dipoles leads to unopposed, stronger than normal, left septal forces, and as a consequence, the initial 0.01-second vector presents its characteristic pattern of increased rightward voltage followed by an earlier posterior displacement of the QRS loop due to the absence of right ventricle.

Figure 2
Case 19, tricuspid atresia, type 1C. (A) Electrocardiogram. Note the P "tricuspidale" in V1 and V6. Left axis deviation, and rS pattern in V1, followed by QS in V2 and qRS in V3, suggesting a clockwise rotation of the horizontal QRS loop. (B) Vectorcardiogram showing increased initial forces, clockwise rotation of the initial limbs in the horizontal plane, and counterclockwise inscription of the frontal plane. (C) Enlarged P loop showing sizable anterior and posterior areas and irregularities on its contour.
Figure 3

Case 30, tricuspid atresia, type 2C. (A) Electrocardiogram showing right axis deviation, and RS pattern in V_{1} through V_{6}. Note the wide P waves "tricuspidale" type. (B) Vectorcardiogram demonstrating a right ventricular hypertrophy pattern.

Figure 4

Distribution of the mean QRS axis in the frontal and horizontal projections for each individual case. (A) Tricuspid atresia (TA) with and without transposition of the great vessels (TGV). (B) Pulmonary atresia (PA) with small right ventricular cavity (type I), and with normal or enlarged right ventricular cavity (type II).
This behavior of the initial forces has been described by Cabrera and Gaxiola\(^7\) as suggestive of hypoplasia of the right ventricle in the presence of mid and terminal QRS forces which are diagnostic of left ventricular hypertrophy.

Seven patients in these series did not display the usual pattern of left axis deviation. All but one of these had tricuspid atresia (type 2C) characterized by high pulmonary flow and transposition of the great vessels (fig. 4A). The high degree of variability displayed by this group might well be expected in that the physiology of this defect is more in keeping with that of transposition of the great vessels with single ventricle than with that of tricuspid atresia. Indeed, the great variability in the frontal plane axis (\(-30^\circ\) to \(+150^\circ\)) in these patients is in agreement with the previously reported vectorcardiographic findings in single ventricle.\(^1\) The remaining case of tricuspid atresia without left axis deviation occurred in type 1A, the rare lesion in which both pulmonary and tricuspid atresia are present.

The syndrome of pulmonary atresia with intact ventricular septum is often referred to as "hypoplastic right heart syndrome."\(^2\) Anatomically true muscle hypoplasia, however, is never observed in this lesion. On the contrary, there is markedly thickened right ventricle with a small cavity in type I, and a normal or enlarged right ventricular cavity with thick wall in type II. The presence of a significant right ventricular muscle mass, as well as the integrity of the ventricular septum seems to account for the most obvious differences be-

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**Figure 5**

*Case 1, pulmonary atresia, type I. (A) Electrocardiogram. Note the normal frontal QRS axis (60°), the "adult" progression of precordial leads, and the P "pulmonale." (B) Vectorcardiograms showing normal morphology and voltage.*

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between the electrocardiograms of pulmonary atresia and tricuspid atresia. In the former, a normal or rightward QRS frontal axis was found, as opposed to the previously described left axis deviation which occurs in tricuspid atresia with true hypoplasia of the right ventricular wall.

The fact that in pulmonary atresia type I right precordial patterns of hypertrophy are absent, despite the markedly hypertrophied right ventricle (fig. 5), whereas in type II the right precordial voltages are significantly increased (fig. 6) suggests that the size of the right ventricular cavity may be a major contributing factor to the presence of increased right ventricular potentials. The theoretical studies of Brody, and the experimental data published by Nelson and associates, Bayley and Berry, Horan and associates, and Angelakos and Gokhan contribute to the understanding of the electrophysiological mechanism involved. According to these authors, the highly conductive intracavitary blood mass (approximately 10 times that of the surrounding tissue) exerts a powerful distorting effect on the heart muscle-lead relationship. Blood within the ventricle produces effective augmentation of radially directed dipoles, which in turn increases precordial voltages and reduces tangentially directed dipoles or waves of excitation. In contrast, when the volume of the intracavitary blood mass is decreased, or when the blood in the right ventricular cavity is replaced by gas, mineral oil, or a medium with higher resistivity, the magnitude of the dipoles oriented radially to the heart surface is decreased, and that of the tangentially oriented ones is exaggerated, independent of right ventricular wall thickness. In other words, blood volume with its lower resistivity is a major factor in the production of precordial potentials. Furthermore, according to Bayley and Berry, not only the blood mass resistivity must be considered, but also the

![Figure 6](http://circ.ahajournals.org/)

**Figure 6**

*Case 20, pulmonary atresia, type II. (A) Electrocardiogram shows right axis deviation and Rs pattern in V1 and V2. (B) Vectorcardiogram showing the characteristic features of right ventricular hypertrophy.*

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radii of the heart cavity. The analysis of equa-
tions 3 and 4 given by these authors\textsuperscript{14} shows that increments of the heart cavity would in-
crease the body surface potential, and reduc-
tion of this cavity would decrease those poten-
tials. These experimental studies are remark-
ably borne out by the electrocardiographic
studies in the present cases of pulmonary
atresia as well as those in previous reports.\textsuperscript{3}

The electrocardiographic findings in patient
3 (table 2) contribute further insight to the
understanding of the electrophysiology of
these lesions and constitute another link be-
tween an observed clinical situation and
previous experimental observations. The
post-mortem examination of this infant revealed
typical pulmonary atresia of type I with a
greatly hypertrophied right ventricular wall
with an extremely small right ventricular
cavity (fig. 7). The initial electrocardiogram
taken on this patient (fig. 8A) would not lead
to suspect the presence of right ventricular
hypertrophy. The electrocardiogram taken 2
days later (fig. 8B) shows absolute “right ven-
tricular preponderance” certainly due to a
profound conduction disturbance (right bun-
dle-branch block). These observations seem to
confirm the theoretical predictions of Brody\textsuperscript{12}
and Nelson and associates\textsuperscript{13} and the experi-
mental findings of Horan and associates\textsuperscript{9}
on the effect of intracavitary blood upon normal
and abnormal depolarization. It can be seen
that the presence of “block,” changing the pat-

\begin{figure}
\centering
\includegraphics[width=\textwidth]{figure7.png}
\caption{Lateral wall of right ventricle cut to show (1) shallow chamber under tricuspid valve, (2) hypertrophic right atrium, and (3) markedly thick right ventricular wall. Same case as figure 8.}
\end{figure}

\begin{figure}
\centering
\includegraphics[width=\textwidth]{figure8.png}
\caption{Case 8, pulmonary atresia, type I. (A) Electrocardiogram showing QRS axis in 95° and P “pulmonale.” (B) Two days later the patient developed right bundle-branch block. Note the change in QRS axis, and the appearance of right ventricular hypertrophy pattern in precordial leads.}
\end{figure}
tern of depolarization from radial toward a more tangentially oriented sequence of excitation,9 favors the diagnosis of "hypertrophy," whereas during normal activation these forces were not recorded.

Of 17 patients with type I pulmonary atresia, three displayed the precordial pattern of right ventricular hypertrophy in exception to the general rule. The presence of mild right bundle-branch block may well account for the increased right precordial voltage in these cases. The fact that the QRS complex was not prolonged in these patients does not necessarily militate against this theory in that in patients with or without right ventricular hypertrophy and mild right bundle-branch block, increased right ventricular precordial potentials can be produced without significant prolongation of the QRS duration.16

The findings outlined call our attention to the extremely complicated heart muscle-lead relationship, when attempts are made to correlate body surface potentials with anatomic situations. The simplified relationship between hemodynamic parameters such as intraventricular pressure17 and body surface potentials should be cautiously considered and restricted to a very selected group of patients in whom the many other variables are relatively constant.

Behavior of atrial depolarization in both tricuspid atresia and pulmonary atresia deserves special comment. Our observations in patients with tricuspid atresia confirm Zuckermann and associates,18 description of a double-peaked P wave with the first peak taller than the second due to bialtrial hypertrophy with preponderance of the right atrium. This characteristic pattern could be referred to as P "tricuspidale" and represents the mirror image of the P "mitrale." The spatial P loop presents irregularities on its contour, increased voltage, and a sizable area both anteriorly and posteriorly (figs. 1 to 3). These findings are suggestive of abnormal conduction and bialtrial hypertrophy. The hemodynamic status of the atria in pulmonary atresia is similar to that observed in tricuspid atresia. This could lead to the expectation that the P wave would be have in an identical manner in the two conditions. But as a matter of fact in only two instances was a P "tricuspidale" pattern noted in the two oldest patients with pulmonary atresia. It does not seem unreasonable to assume that due to the lethal nature of this defect most infants do not live long enough to develop sufficient left atrial hypertrophy to display the characteristic P wave.

Whereas a qualitative analysis of the atrial depolarization may give information leading to differentiation of tricuspid atresia and pulmonary atresia, quantitative analysis is far from ideal. According to Taussig,1 there is an indirect relationship between the height of the P wave and the size of the interatrial communication in tricuspid atresia. However, Keith and associates2 failed to demonstrate this correlation. In order to supersede the limitations of the standard electrocardiogram as far as quantitative data are concerned,4 an attempt was made to correlate the maximal spatial vectorial voltage of the P loop with the size of the atrial septal defect in eight cases of tricuspid atresia in whom postmortem examinations were available. Even with this approach, no significant correlation was demonstrated. This lack of correlation could well be expected in view of the numerous variables which would affect the surface potentials arising from the atrium. As discussed, relative to the QRS complex, the effects of blood mass would also influence atrial potentials and in view of the known tangential spread of excitation in the atrium,14 P wave voltages in atrial dilatation would be expected to be inconstant in a quantitative sense. In hypertrophy, however, the excitation tends to be more radial, and therefore, one could observe a more predictable effect on body surface potential. Since in most instances a combination of dilatation and hypertrophy is present, it is not surprising that quantitative analysis of P wave measurement does not prove to be fruitful. In addition, evaluation of the size of the atrial septal defect may be distorted at postmortem examination and may be an unreliable reflection of the antemortem situation.
Summary

Electrocardiograms were studied in 37 patients with tricuspid atresia and in 20 with pulmonary atresia and intact ventricular septum. The results were correlated with angiocardiographic findings and postmortem examinations. Differential features were as follows:

1. Biatrial hypertrophy with characteristic P “tricuspidale” was noted in 81% of the electrocardiograms in patients with tricuspid atresia, whereas this pattern was seen in only two infants with pulmonary atresia and intact ventricular septum.

2. In tricuspid atresia, left axis deviation was usually directed posteriorly; however, abnormal or right axis deviation was present in seven cases, six of whom were type 2C and the seventh, type 1A. In every case the horizontal QRS axis was oriented posteriorly. By contrast, in patients with pulmonary atresia there were no instances of left axis deviation in the frontal plane. Normal frontal plane axis was present in 12 patients, all of whom were type I; right axis deviation was evident in eight patients including the three infants with type II deformity. The horizontal QRS axis was usually directed posteriorly; however, anterior orientation occurred in several patients.

3. A typical feature of the vectorcardiogram in patients with tricuspid atresia was the increased anterior magnitude of the initial forces followed by a sharp posterior shift. The initial forces in patients with pulmonary atresia were not characteristic.

4. The electrocardiographic features in patients with tricuspid atresia and normal position of the great vessels showed decreased right ventricular potentials consistent with true hypoplasia of the right ventricle, but in patients with pulmonary atresia similar lack of right ventricular potentials could be correlated with decreased right ventricular cavity, whereas the ventricular wall was actually hypertrophied.

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The evolutionary course that has put the profession of medicine where it is may not be the one that would have been pursued by conscious wisdom, but it has had the result of putting medicine in the very small class of professions that, in this tame world, can still be called jobs for men. When I speak of jobs for men, I use a traditional phrase that nowadays needs a little definition. By it I mean professions in which it is possible for people—men or women—to pursue the dying ideal that an occupation for adults should allow of intellectual freedom, should give character as much chance as cleverness, and should be subject to the tonic of difficulty and the spice of danger.—The Collected Papers of Wilfred Trotter. London, Oxford University Press, 1941, p. 4.
The Electrocardiogram in Tricuspid Atresia and Pulmonary Atresia with Intact Ventricular Septum

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