The Electrocardiogram in Congenital Heart Disease

A Preliminary Report

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Adequate unipolar electrocardiograms have been recorded on 101 patients with congenital heart disease and the findings analyzed. It has been found that the chief value of such tracings rests in the determination of ventricular preponderance, the evidence being obtained from study of the QRS complexes in unipolar limb and multiple precordial leads, with relatively little help from the RS-T segments and T waves. Auricular hypertrophy, encountered chiefly in association with pulmonic stenosis and tricuspid valve disease, could be best detected by analysis of P waves seen in the right precordial leads rather than in the limb leads. Intraventricular block was observed both with auricular and with ventricular septal defects, and was also found in Ebstein's disease and with coartation of the aorta. Auriculoventricular block and arrhythmias were rare.

Accurate diagnosis of congenital cardiac lesions no longer must await evidence supplied at the autopsy table. Today, in the majority of such cases, specific and precise data as to the nature of single or multiple cardiovascular anomalies can be obtained as the result of a careful assessment of clinical and laboratory findings. It is well known that the history and physical examination by themselves are often inadequate to provide a satisfactory diagnosis; and information secured through roentgenologic study, including angiocardiography, and cardiac catheterization, including blood gas analysis, are necessary in many instances. The electrocardiogram is an additional and valuable laboratory tool. It does not by itself provide a final answer, except in instances of uncomplicated dextrocardia of the situs inversus type, but it does indicate whether gross preponderance and hypertrophy of the right or left ventricles exist, whether there is evidence of auricular hypertrophy, and whether abnormalities of rhythm or conduction are present.

The analysis of electrocardiographic tracings in the congenital cardiac group is not altogether easy. In order to obtain maximum help in determining the presence of an abnormal right or left ventricular preponderance, at least three and preferably more precordial leads should be available; in this regard, the unipolar limb leads are of value as a guide to the electrical position of the heart. Furthermore, while it is not difficult to detect abnormally high voltage of the QRS complexes in adults, it is a different matter in infants and young children, where the thinness of the chest wall and proportions of the heart and thoracic cage are such that relatively high voltage complexes may normally be found. Reliable standards for the normal P and QRS amplitudes have not been fully established for this age group. It is also well known that a vertical or a mild right axis deviation is a more common finding in normal infants and children than in adults, and in the past an undue and unjustified emphasis has been placed on axis deviation as such. It is our belief that the significance of axis deviation should be considered in the light of other findings in the limb and chest leads.

The following data has been obtained by analysis of the electrocardiograms of 101 patients with congenital cardiovascular lesions ranging in age from 1 day to 52 years (53 of the group were over the age of 12 years).
diagnoses (table 1) have been confirmed in 96 patients by cardiac catheterization, surgical exploration, autopsy findings, or by a combination of these methods. In 5 instances (1 case of dextrocardia with situs inversus and 4 cases of aortic or subaortic stenosis) this type of proof as to the nature of the lesion is not at hand; however, precise clinical observations are available.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. of Cases</th>
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<tbody>
<tr>
<td>Patent Ductus Arteriosus</td>
<td>24</td>
</tr>
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<td>Tetralogy of Fallot</td>
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<tr>
<td>Ventricular Septal Defect</td>
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<td>Coarctation of the Aorta</td>
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<td>Auricular &amp; Ventricular Septal Defects</td>
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<tr>
<td>Dextrocardia without Situs Inversus</td>
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<tr>
<td>Pulmonic Stenosis &amp; Aortic Regurgitation</td>
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<td>Transposition Great Vessels</td>
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<td>Ebstein's Disease</td>
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<tr>
<td>Coronary Arteriovenous Fistula</td>
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<tr>
<td>Patent Ductus Arteriosus with Mitral Valvulitis</td>
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<tr>
<td>Tricuspid Atresia</td>
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<tr>
<td>Tricuspid Stenosis, Pulmonic Stenosis, Patent Foramen Ovale, Endocardial Sclerosis Right Ventricle</td>
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</tr>
<tr>
<td>Total</td>
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Standard limb leads, augmented unipolar limb leads, and at least three unipolar chest leads were recorded in each case.

**Dextrocardia**

The diagnosis of uncomplicated dextrocardia of the situs inversus type is readily made by examination of the electrocardiogram. The findings are absolutely characteristic and, if chest leads are available, cannot be confused with any other entity. There is seen a total inversion of lead I. Leads II and III appear to have been interchanged as do leads aVR and aVL, and the precordial leads taken in the usual fashion show an absence of the normal QRS progression, with small complexes as one approaches the left axilla. However, precordial leads recorded over the right side of the thorax are quite normal in appearance.

Dextrocardia without situs inversus and without other anomalies is occasionally encountered and in such cases the standard and unipolar limb leads are essentially within normal limits. The precordial leads are also normal, except that they may show a shift of the QRS transitional zone to the right.

**Auricular Septal Defect Including Lutembacher's Syndrome**

McCulloch in 1916 published the electrocardiogram of a 23 month old infant whose heart showed at autopsy a large auricular septal defect. This tracing was characterized by right axis deviation with peaked P waves in leads II and III. In 1934, Rosler discussed the problem of auricular septal defect at length, citing 62 cases, 5 of which were studied with apparently adequate electrocardiograms. Four of these revealed a moderate right axis deviation and 1 showed a marked right axis deviation. Bedroom, Papp, and Parkinson in their 1941 article include 10 cases proved at autopsy. In their uncomplicated septal defect group, there were no arrhythmias and most of the tracings showed a right axis deviation. Their cases of auricular septal defect associated with mitral stenosis (Lutembacher's syndrome) were characterized electrocardiographically by the presence of auricular fibrillation or flutter with right axis deviation, and one is stated to have shown a right bundle branch block. Brown has reported that a slight right axis deviation is common and that the P waves may be large, and Taussig, remarking that right axis deviation is "the rule" and that the P waves are high, adds that the P-R interval is frequently prolonged and that notching and widening of the QRS complexes may occur. A right axis deviation was observed in the electrocardiograms of 3 of 4 patients with this diagnosis studied by Brannon, Weens and Warren, and the tracing of the fourth showed a tendency to right axis deviation.

We have studied 5 patients with auricular septal defects, all diagnosed by cardiac catheterization, and believe that on clinical grounds 3 are associated with mitral stenosis. The age
range of this group was 8 to 30 years. Four of the 5 patients were males. No arrhythmias were seen. The most significant findings in the electrocardiograms were the presence of right bundle branch block in 2 cases (the intraventricular conduction times measured 0.12 second), right axis deviation (+120°) in a third accompanied by a right bundle branch block pattern in the precordial leads but with normal QRS duration, and similarly right axis deviation (+125°) in a fourth case with precordial leads consistent with right ventricular hypertrophy. The electrocardiogram in the fifth patient revealed a vertical axis but did not suggest enlargement of the right ventricle and no block was present. The P waves were normal in 1 tracing and in 4 others were peaked and slightly high (0.15 to 0.2 millivolt) in leads V1, and/or V2 (and in 1 case V3 and V4).* In no instance were the P waves strikingly abnormal and no A-V block was seen. The QRS voltages were not remarkable.

Figure 1 illustrates an electrocardiogram on a 16 year old male with an uncomplicated auricular septal defect, showing a characteristic right bundle branch block.

![Electrocardiogram](image)

**Fig. 1.** Electrocardiogram of a 16 year old male with uncomplicated auricular septal defect, showing a characteristic right bundle branch block.

Ventricular Septal Defect

In no aspect of the field of congenital heart disease may such wide clinical, x-ray, and electrocardiographic variations be found as in lesions of the ventricular septum. Despite our textbook concepts, the "simple" ventricular septal defect (Roger's disease) often blends into Eisenmenger's complex, and the latter, in turn, in its advanced form, may be difficult to distinguish from cor triloculare biaatriatum unless the heart is examined for disease. For this reason, we are grouping all lesions of the ventricular septum under one main head.

* We have reviewed a series of electrocardiograms recorded on normal children and adults and have observed that the P waves do not exceed 0.15 millivolt in amplitude in unipolar precordial leads.

Our present series includes 4 noncyanotic patients, all studied by cardiac catheterization, who have been shown to have ventricular septal defects probably best labeled as Roger's
disease. The age range was from 6 to 11 years and three were females. Except for left axis deviation (−20° and −47°) in 2 of the tracings, the electrocardiograms showed no characteristic findings and in particular no intraventricular block was seen.

We have also seen 4 patients (3 studied by cardiac catheterization and 1 by angiocardiography and later by operation) with ventricular septal defects which might be classified as Eisenmenger's complex. Three of these patients were cyanotic with low arterial oxygen saturation, and the fourth, although not cyanotic, had an arterial oxygen saturation of 86 per cent. All showed clubbing and prominent pulmonary vessels on fluoroscopy. The youngest was 6 years of age and the oldest was 43. All of these had electrocardiograms characterized by a right axis deviation. Three tracings showed the picture of right ventricular hypertrophy in the precordial leads (fig. 2), one being associated with the right bundle branch block pattern without actual QRS prolongation, and another revealing definite intraventricular block (QRS duration of 0.12 second) of an indeterminate type. A mixed QRS progression of uncertain significance was found in the precordial leads of the fourth patient. Variable A-V block with at times a nodal rhythm was noted in one electrocardiogram but the P waves were otherwise abnormal in only one record in which high peaked P waves were seen in leads II, III, and aVr with deeply inverted P waves in V5 and upright peaked P waves in V6.

Also to be included in this group are 3 patients, 2 diagnosed as cor triloculare biaatriatum (one aged 1 day studied at autopsy, and one aged 25 years diagnosed by cardiac catheterization), and one aged 8 years, diagnosed also by cardiac catheterization as having a huge ventricular septal defect. Prominent P waves, usually peaked, were observed in two of the three tracings in the limb and precordial leads. A complete right bundle branch block was found in one, a probable incomplete left bundle branch block in another, and a high degree of right axis deviation (+150°) with precordial leads indicative of right ventricular preponderance was present in the infant's electrocardiogram. The voltage of the QRS complexes was increased in two of the tracings.

![Figure 2. Electrocardiogram of a 43 year old female with Eisenmenger's complex, showing right axis deviation, a right ventricular hypertrophy pattern in the precordial leads, and slight prolongation of the intraventricular conduction time.](http://circ.ahajournals.org/figshare/circulation_by_guest.2017-11-11_99_99_99)
Combined Auricular and Ventricular Septal Defects

Our 3 examples of combined auricular and ventricular septal defects were 3 month and 14 month old male infants, both studied at autopsy, and a 6 year old girl who was diagnosed by cardiac catheterization. The electrocardiograms taken on these patients were not distinctive, except that two showed slightly prominent P waves, and all three had a vertical axis. The precordial leads indicated an essentially balanced bilateral ventricular hypertrophy in 2 cases, which was confirmed at autopsy, and in the third case bilateral ventricular hypertrophy was evident but with preponderance of the left ventricle. There was no auriculoventricular or intraventricular block present and there were no arrhythmias.

Ebstein's Disease

Few cases of Ebstein's disease (congenital downward displacement and deformity of the tricuspid valve) have been reported. The electrocardiogram of the patient described by Yater and Shapiro was characterized by intraventricular block, probably left bundle branch block, with paroxysms of supraventricular tachycardia. The 3 cases reported by Engle, Payne, Bruins, and Taussig showed high peaked P waves, A-V block, and right bundle branch block in 2 patients (and probably also in the third).

Our patient with Ebstein's disease, a 16 year old boy whose heart was examined pathologically, also had right bundle branch block in his electrocardiogram (fig. 3), the intraventricular conduction time measuring 0.12 second. Very high peaked P waves were found, being especially prominent in the precordial leads, and it was seen at autopsy that these were related to a greatly enlarged right auricle. No septal defect was present.

Tricuspid Atresia

The clinical diagnosis of tricuspid atresia depends on an unusual extent upon the presence of characteristic electrocardiographic findings. Blackford and Hoppe in 1931, and Taussig in 1936, commented upon the presence of a left axis deviation as a most valuable distinctive feature, serving to differentiate this group of cyanotic patients with a small right ventricle, from the much larger cyanotic group associated with right ventricular hypertrophy.

We have seen one such patient, a young woman aged 24 years, in whom the clinical impression of tricuspid atresia was confirmed by cardiac catheterization. Her electrocardiogram (fig. 4) showed a left axis deviation of high degree, broad, high, notched P waves, and slight prolongation of the intraventricular conduction time (QRS, 0.11 second). The voltage of the qR complex in lead aV1 was slightly increased, and the precordial leads were characteristic of left ventricular preponderance.

Isolated Pulmonic Stenosis

In 1945, Currens, Kinney, and White reported the findings in a series of 11 cases of pulmonic stenosis with an intact ventricular septum. Electrocardiograms were available on 4 of these, three of which showed a marked right axis deviation (1 of these cases had a good-sized patent foramen ovale) whereas the fourth, recorded on a 43 year old patient with only moderate stenosis, showed a normal axis. On reviewing the tracings, it is seen that the P waves tend to be high and peaked, and that in 2 instances, a definite T2 and T3 inversion...
is present with some RS-T segment depression and sagging. Greene and co-workers have described 4 patients with pure pulmonic stenosis and observed that a right axis deviation was present in only 1 of the 4; it is of interest that this one patient, although the youngest in the group, had the highest right ventricular systolic pressure. Blackford and Parker have reported a case of a 23 year old male with uncomplicated pulmonic stenosis whose electrocardiogram, limited to the three standard limb leads, showed an apparent complete right bundle branch block with a high peaked P2 and P3.

right axis deviation was observed in the 3 others. The precordial leads were characteristic of right ventricular hypertrophy in five electrocardiograms and suggestive in a sixth, but only 1 of these showed RS-T and T wave abnormalities of the typical right ventricular "strain" type.

This evidence of a right ventricular preponderance could be correlated with the right ventricular pressures observed at the time of cardiac catheterization. In the 2 instances in which the resting right ventricular systolic pressure was less than 50 mm. Hg, the QRS progression in the precordial leads was normal for the age of the patient. The 5 cases with definite signs of right ventricular hypertrophy in their electrocardiograms (one is illustrated in figure 5) were associated with resting right ventricular systolic pressures of 100 mm. Hg or more; the patient whose electrocardiogram was merely suggestive of right ventricular hypertrophy showed a resting right ventricular systolic pressure of 100 mm. of mercury.

PULMONIC STENOSIS AND AURICULAR SEPTAL DEFECT

The clinical and laboratory features of patients with pulmonic stenosis and auricular septal defect have been well summarized by Selzer and his associates, who comment on the great frequency of prominent tall P waves

Fig. 4. Electrocardiogram of a 24 year old female with tricuspid atresia. The P waves are broad and notched, a left axis deviation is present, the intraventricular conduction time is slightly prolonged, and the unipolar leads are characteristic of left ventricular preponderance.

The 8 patients with isolated pulmonic stenosis whom we have studied form an especially interesting group. Their ages ranged from 3 to 43 years and the diagnosis was established in each case by cardiac catheterization. No arrhythmias were observed and no auriculoventricular or intraventricular block was seen with the exception of a possible incomplete right bundle branch block in 1 case. The P waves were normal in three tracings and in the remaining five were peaked with slight to moderate increases in amplitude (0.17 to 0.3 millivolt), notably in leads V1, V2 and V3, and less often in the limb leads. The axis was entirely normal (+50°) in one case, a vertical to mild right axis deviation (+90° to +110°) was present in 4 cases, and a high degree of
and right axis deviation. Engle, Taussig, and Bruins have also recently reviewed this syndrome and have described similar findings. Diagnosis was established by autopsy. Their ages ranged from 3 weeks to 28 years. Analysis of the electrocardiograms showed a sinus rhythm.

![Electrocardiogram of a 7 year old female with isolated pulmonic stenosis](image1)

**Fig. 5.** Electrocardiogram of a 7 year old female with isolated pulmonic stenosis, showing right axis deviation with precordial leads typical of a high degree of right ventricular hypertrophy, very high R waves being present in leads V1 through V4.

![Electrocardiogram of a 9 year old boy with pulmonic stenosis and auricular septal defect](image2)

**Fig. 6.** Electrocardiogram of a 9 year old boy with pulmonic stenosis and auricular septal defect. The P waves are tall, and the precordial leads, as in figure 5, are typical of right ventricular hypertrophy.

including the presence of right bundle branch block or evidences of right ventricular hypertrophy.

In our own series of 7 cases, 5 were studied by cardiac catheterization and in 2 the diagnosis in all, without auriculoventricular or intraventricular block. The P waves were peaked and increased in amplitude in five instances, being most abnormal in leads V1 through V4. Right axis deviation (greater than +115°) was
seen in all cases, and the voltage was increased in the precordial leads in three of the seven tracings. In addition, the precordial leads were characteristic of right ventricular hypertrophy in four records and suggested it in one other. Figure 6 shows an electrocardiogram of a 9 year old boy with this condition, illustrating the abnormal tall P waves, the high degree of right axis deviation, and the evidences of right ventricular hypertrophy.

The Tetralogy of Fallot

Maude Abbott’s Atlas of Congenital Cardiac Disease\(^2\) includes the electrocardiograms of 3 patients with the tetralogy of Fallot. All three of these tracings, which were limited to the standard bipolar limb leads, showed a well marked right axis deviation, and high P waves in lead II. In one, a partial variable auriculoventricular block was described. More recently, Taussig\(^3\) has stated that in this condition, the electrocardiogram always shows a “marked right axis deviation and usually the P waves are abnormally high and pointed,” and Dry\(^4\) has commented that right axis deviation of moderate to marked degree is “nearly always present,” that high and pointed P waves are common, and that conduction disturbances may also occur.

We have studied the electrocardiograms of 15 patients, aged 9 months to 37 years, all of whom were diagnosed by cardiac catheterization, operation, autopsy or combinations of these methods as having the tetralogy of Fallot.\(^*\) The rhythm was normal in 13 of these, one showed a wandering auricular pacemaker, and one revealed auricular fibrillation (coexisting thyrotoxicosis was suspected clinically). Prominent and typically peaked P waves were seen in 13 tracings, being found chiefly in leads V\(_3\)R, V\(_1\), V\(_2\) or V\(_3\), and less often in leads II, III, V\(_4\), V\(_5\) and in lead aV\(_R\) (where they were sharply inverted). In one electrocardiogram, recorded on an 18 month old patient, the axis was +90°; in another fourteen right axis deviation over +100° was present and in nine, the axis was +120° or more. The voltage appeared high in all but three tracings. Evidences of right ventricular hypertrophy were also suggested by the presence of a high R or R’ in lead aV\(_R\) in nine electrocardiograms, and in twelve, the precordial leads showed findings typical of right ventricular hypertrophy with a reversal of the usual R/S ratio (one of the twelve also demon-

\(^*\) In 1 of these patients, an associated patent ductus was suspected at operation, although not demonstrated by cardiac catheterization, and in another, an associated auricular septal defect was suggested but not proved by cardiac catheterization.

![Electrocardiogram of a 37 year old female with the tetralogy of Fallot. The P waves are peaked in leads V\(_2\), V\(_3\), and V\(_4\), a right axis deviation is present, and the precordial leads show right ventricular preponderance.](image-url)
strated a slightly prolonged QRS interval with the right bundle branch block type of QRS, i.e. rsR' in lead V1). The precordial leads in one tracing showed RS complexes of similar form in all positions recorded, and in two electrocardiograms a mixed progression was found. RS-T and T wave changes consistent with right ventricular "strain" were seen in only five electrocardiograms.

Figure 7 illustrates the electrocardiogram of a 37 year old woman with this condition, showing peaked P waves in leads V3, V4 and V5, a right axis deviation, and the signs of right ventricular hypertrophy described above. The intraventricular conduction time is slightly prolonged and lead V1 shows the right bundle branch block pattern.*

PULMONIC STENOSIS AND AORTIC REGURGITATION

The combination of pulmonic stenosis and free aortic regurgitation must be rare indeed. By chance, we have had an opportunity to observe and study with data obtained by cardiac catheterization 2 such cases. Both patients were males, aged 16 and 17 respectively, both were known to have heart murmurs recognized in early infancy (one at 3 weeks of age and the other at 4 weeks), and both showed wide pulse pressures without evidence of peripheral arteriovenous fistulas, large hearts, and loud basal systolic and diastolic murmurs. Neither patient gave a history of rheumatic fever or chorea, and it is possible that they represent congenital endocardial sclerosis with valvular deformities.

Their electrocardiograms have certain similarities in that no arrhythmias were observed, the axes were within normal limits, and high voltage QRS complexes were present throughout the precordial leads, which, although indicating left ventricular preponderance, suggested bilateral ventricular hypertrophy by virtue of high voltage, diphasic QRS complexes in leads V1 and V2. One tracing in addition reveals broad (0.12 second) P waves and intraventricular block (QRS duration of 0.12 second) without findings characteristic of either right or left bundle branch block.

TRANSPOSITION OF THE GREAT VESSELS

The electrocardiographic findings in patients with transposition of the great vessels are said to be usually those of a right ventricular preponderance with large P waves.22, 23 One case with complete auriculoventricular block has been described.

We have analyzed the tracings of 2 male patients, aged 10 days and 4 months, both studied at autopsy. A sinus rhythm was present with normal P-R and QRS intervals, right axis deviation (+115° and +160°), and peaked P waves which in 1 case were high. Tall R waves were seen in lead aVR and the precordial leads showed high voltage diphasic QRS complexes and were consistent with a right ventricular preponderance. Figure 8 illustrates the electrocardiogram of the 10 day old baby with this condition.

AORTIC AND SUBAORTIC STENOSIS

The literature does not contain many reproductions of electrocardiograms from patients with congenital aortic or subaortic stenosis. Schnitker24 reported 1 case with standard limb leads suggesting the presence of left ventricular hypertrophy, and Taussig,4 and Brown4 state that left axis deviation may be found.

We have reviewed the electrocardiograms of 4 patients with presumed aortic or subaortic stenosis. These patients, all males and aged from 2 to 28 years, were diagnosed clinically by the following criteria: (1) presence of a loud cardiac murmur known since infancy or childhood (4 months to 7½ years of age); (2) absence of a history of rheumatic fever or chorea; (3) presence of a grade III or IV systolic murmur, loudest in the second right interspace adjacent to the sternal border, and accompanied by a thrill; (4) absence of evidence of other valvular involvement, and (5) lack of cyanosis or clubbing.

* Since writing the above, we have also seen and studied by cardiac catheterization a 5 year old girl with the tetralogy of Fallot plus an auricular septal defect ("pentalogy"). Her electrocardiogram also showed right axis deviation, high pointed P waves, and evidence of a high degree of right ventricular hypertrophy.
The electrocardiograms of our 4 patients are relatively normal. There are no arrhythmias, auriculoventricular or intraventricular block, unusual P waves, or abnormal axis deviation. The precordial leads are consistent with left ventricular preponderance and the voltage is at the upper limit of normal or slightly increased in all four tracings, but in only one electrocardiogram (that of the oldest patient) are the T waves in leads I, aVL, V5, and V6 characteristic of the classic left ventricular hypertrophy or “strain” pattern. From the electrocardiographic point of view, this lesion seems to be well tolerated.

Coarctation of the Aorta

Hamilton and Abbott25 in their comprehensive survey of 200 cases of coarctation of the aorta (1928) commented that in this condition “the electrocardiogram . . . almost invariably shows a left-sided preponderance.” In 1942, Rhodes and Durbin26 summarized the electrocardiographic findings in an additional 116 cases recorded in the literature and observed that 32 of the tracings were normal, 57 showed a left axis deviation, 8 a right axis deviation and 17 revealed prolongation of the intraventricular conduction time; various other findings were listed for 14 of the cases. Of the 8 patients whose tracings were characterized by a right axis deviation, 4 had intraventricular block and three of these were associated with a patent ductus arteriosus, and of the 4 cases without block, 3 lacked pathologic proof of the diagnosis and 1 of these probably also had an associated patent ductus arteriosus. On the basis of their data, it would appear that a right axis deviation is very rare in uncomplicated examples of coarctation of the aorta but that it may occur in association with a patent ductus, presumably by greatly increasing the pressure in the pulmonary artery leading in

Fig. 8. Electrocardiogram of a 10 day old baby with transposition of the great vessels. The P waves are somewhat peaked, a marked right axis deviation is present, and leads V3, V4, and V6 are consistent with the presence of right ventricular hypertrophy.
grams in a series of 64 patients studied at the Mayo Clinic. Recently, Sokolow and Edgar have referred to the electrocardiographic findings in 24 cases of coarctation of the aorta, observing five normal records, fourteen with evidence of left ventricular hypertrophy, three (with other congenital defects) with right ventricular hypertrophy, and two with right bundle branch block in association with left ventricular hypertrophy.

We have reviewed the electrocardiograms of 8 male and 3 female patients with coarctation of the aorta. Their ages ranged from 8 to 24 years and the diagnosis was proved by aortic catheterization in one and by operation in the remaining 10 cases. The rhythm was normal in all but one tracing, which showed inverted P waves with a normal P-R interval in leads II, III, and aVr in a vertical heart, evidence of an ectopic auricular pacemaker. Five of the electrocardiograms showed no signs whatever of left ventricular hypertrophy; three others suggested left ventricular hypertrophy (deep S waves in right precordial leads in two and inverted T waves in leads I, V₅ and V₆ in another; see fig. 9). On the whole, no very impressive evidence of left ventricular enlargement was found. One electrocardiogram recorded on a 15 year old male with a blood pressure of 240/120 revealed a typical incomplete right bundle branch block. Two others showed intraventricular conduction times at the upper limit of normal associated with an rSr' type of QRS complex in leads V₁ and V₂ with persistent S waves in leads V₃ and V₆, also suggesting an incomplete right bundle branch block. There was no precise correlation of electrocardiographic abnormality with age or with blood pressure readings, although the patients with the highest pressures tended to have abnormal tracings.

**Fig. 9.** Electrocardiogram of a 17 year old male with coarctation of the aorta. The tracing is consistent with left ventricular hypertrophy in a vertical heart.

**Patent Ductus Arteriosus**

It is generally agreed that the electrocardiogram is normal in the vast majority of patients with the diagnosis of uncomplicated patent ductus arteriosus. As is clear from the report of Keys and Shapiro on patency of the ductus arteriosus in adults, hypertrophy of both ventricles eventually occurs and therefore a relative balance is usually maintained with neither an undue right nor an undue left ventricular preponderance pattern appearing (although high voltage QRS complexes may be found). It is of the greatest importance, however, to stress the fact that a mild right
axis deviation is by no means a rarity in this condition. Bullock, Jones and Dooley, Steinberg, Grishman, and Sussman, Shapiro, and many others have reported proved cases whose electrocardiograms showed some degree of right axis deviation. It is in this type of situation that the mistake must not be made of assuming that a right axis deviation is synonymous with right ventricular hypertrophy, and that hence the patient cannot be considered as having an isolated patent ductus arteriosus; adequate study of unipolar limb and chest leads should be made and it will usually be found—as is clear in the data reported below—that there is merely a vertical heart pattern without evidence of a right ventricular preponderance. Exceptionally, a true right ventricular hypertrophy may occur in patients in the older age group in whom a widely patent ductus is found associated with pulmonary arteriolar sclerosis (as reported by both Levine and Ulrich). As to other uncommon findings, Levine and Geremia have described some prolongation of the QRS interval and inversion of the T waves in leads I and II, each occurring twice among 34 cases, and Dry, Harrington, and Edwards have reported a case associated with auricular fibrillation similar to one we have seen.

We have analyzed the electrocardiograms of 24 patients, aged 4 to 36 years, in all of whom the diagnosis of patent ductus arteriosus was confirmed by operation or cardiac catheterization or by both of these methods. Except for frequent ventricular premature beats in one tracing, the rhythm was normal in each case and neither auriculoventricular nor intraventricular block were seen. The P waves were not remarkable in twenty-one records, and were slightly prominent in three others. One electrocardiogram showed a tendency to left axis deviation (−10°), two showed a vertical axis, and in one there was a tendency to right axis deviation (+95°). The axis was entirely normal in the remainder. It is important to stress the fact that a vertical axis or slight right axis deviation may occur with an uncomplicated ductus arteriosus and, indeed, two tracings had been wrongly interpreted as showing a predominant right ventricle because of this finding. The QRS progression in the precordial leads indicated predominance of the left ventricle in all cases and actual left ventricular hypertrophy was suggested in 6 cases either by the presence of deep S waves in leads V₁, V₂ or V₃ or by high R waves in V₅ or V₆. In addition, three tracings revealed high-voltage diphasic RS complexes over the right, or right and middle precordial leads, with high R waves in leads V₄, V₅ or V₆ sug-

\[ \begin{align*}
\text{FIG. 10. Electrocardiogram of a 14 year old female with an uncomplicated patent ductus arteriosus, showing the presence of a slight right axis deviation. A normal left ventricular preponderance is seen in the precordial leads.}
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suggesting that combined ventricular hypertrophy existed. The RS-T segments and T waves showed no characteristic changes.

Figure 10 illustrates a slight right axis deviation occurring in the electrocardiogram of a 14 year old patient with an uncomplicated patent ductus arteriosus.

Although it is not included in this series, we wish to mention the electrocardiogram of a 42 year old woman known to have had heart disease since infancy in whom the diagnosis of patent ductus arteriosus had been made at the Massachusetts General Hospital on the basis of characteristic clinical findings with a typical x-ray picture. The diagnosis was confirmed by independent studies at another hospital. The patient declined surgery and permission for autopsy was refused at the time of her death from congestive heart failure, so that final proof as to the anomaly is lacking although there is little doubt as to the nature of the lesion. Her electrocardiograms showed auricular fibrillation associated with a considerable degree of left axis deviation (-30°) with a characteristic left ventricular hypertrophy pattern in the limb and chest leads including a very high R in lead CFg.

We have not correlated the electrocardiographic findings with the ductus size as estimated at operation. It has been our experience that what appears to be grossly a ductus of large dimensions shunting a great volume of blood may be found to be pathologically associated with a variable and often small aortic or pulmonary arterial opening.

**Miscellaneous Cases**

We have had an opportunity to observe 1 unusual cyanotic female infant from birth until her death at the age of 6 months. At autopsy, a congenital endocardial sclerosis of the right ventricle was found associated with pulmonic stenosis, slight tricuspid stenosis, and a patent foramen ovale. The right ventricle was thick walled but of small capacity; the left ventricle and right auricle were both hypertrophied. Electrocardiograms on this patient were confusing, initially suggesting left ventricular hypertrophy in a vertical heart, and later indicating a ventricular balance with RS complexes of similar contour in leads V1 through V6. The axis was vertical and the P waves were peaked and high.

Another female patient, aged 15 years, was found at autopsy to have a combination of a patent ductus arteriosus plus saccing of the mitral valve and adjacent endocardium (rheumatic?, endocardial sclerotic basis?). Both ventricles were hypertrophied, the left being thicker than the right. Her electrocardiogram showed a sinus rhythm, a vertical axis, normal P-R and QRS intervals, and high voltage R waves in leads aVF and V6 with a deep S in V2 consistent with left ventricular hypertrophy. The T wave in lead V6 was slightly diphasic.

Finally, we have followed for some years a 9 year old boy, previously reported,40 who at operation was found to have an arteriovenous fistula between the right coronary artery and the coronary sinus. The electrocardiogram on this patient was within normal limits.

**Discussion**

Certain general conclusions can be drawn from such a group of electrocardiograms. It is apparent that arrhythmias are not common in patients with congenital heart disease; indeed, they are seen with frequency only in Lutembacher's syndrome in which others have reported a considerable incidence of auricular fibrillation and auricular flutter. The study of P waves has been rewarding, and it has been observed that analysis of the precordial leads, notably V6R, V1, and V2, often yielded more information as to unusual size and contour of these waves than did the limb leads which have received so much attention in the past. Abnormal P waves were most often associated with pulmonic stenosis, in which they were characteristically high and peaked. High peaked P waves also have been found in Ebstein's disease, and we have noted broad and high P waves in tricuspid atresia.

It is important to appreciate the value of multiple precordial leads in a determination of relative ventricular preponderance, as compared with axis deviation. This is of course a familiar observation, but it will bear particular repetition in the field of congenital heart disease in which so much emphasis has been
placed on right axis deviation as contrasted with left axis deviation. As pointed out above, axis deviation should be interpreted in the light of findings in unipolar limb and multiple precordial leads, and the presence of a vertical or right axis is not adequate evidence of right ventricular hypertrophy. Where clear signs of right ventricular preponderance are seen electrocardiographically, cardiac catheterization has usually revealed an elevation of the right ventricular systolic pressure above 100 mm. of mercury. It has also been noted that the incomplete right bundle branch block pattern may occur in association with right ventricular hypertrophy, leads aV₆ and V₄ quite commonly showing a late R′ with a late S wave in leads V₅ and V₆. RS-T segment and T wave changes of the “strain” type were infrequent, even in the presence of definite unilateral ventricular hypertrophy.

Our series includes only one electrocardiogram with A-V block, recorded on a patient with Eisenmenger’s complex. Intraventricular block, usually right, was observed with auricular septal defects, ventricular septal defects, Ebstein’s disease, and with coarctation of the aorta.

SUMMARY

1. The electrocardiographic findings encountered in a group of 101 patients with congenital heart disease have been reviewed.
2. Arrhythmias were rare.
3. Abnormal P waves were best seen in leads V₃R through V₅ and were most often associated with pulmonic stenosis, complicated or uncomplicated, where they were peaked. They were also found in association with tricuspid valve disease.
4. The most valuable information as to relative ventricular preponderance was obtained from the QRS complexes in multiple chest leads; axis deviation was of lesser importance and at times misleading.
5. Analysis of the RS-T segments and T waves was usually unrewarding; the “strain” pattern was not common, even in the presence of a high degree of unilateral ventricular hypertrophy.
6. Intraventricular block was seen with auricular septal defects, ventricular septal defects, Ebstein’s disease, and with coarctation of the aorta.

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REFERENCES

14 Taussig, H. B.: Clinical and pathological findings in congenital malformations of the heart due to


24 SCHNITZER, M. A.: The electrocardiogram in congenital cardiac disease: a study of one hundred and nine cases, one hundred and six with autopsy. Cambridge, Mass., Harvard Univ. Press, 1940.

25 HAMILTON, W. F., AND ABBOTT, M. E.: Coarctation of aorta of adult type; complete obliteration of descending arch at insertion of ductus in boy of fourteen; bicuspid aortic valve; impending rupture of aorta; cerebral death; statistical study and historical retrospect of 200 recorded cases, with autopsy, of stenosis or obliteration of descending arch in subjects above age of two years. Am. Heart J. 3: 381, 1928.


