The Genesis and Importance of the Electrocardiogram in Coarctation of the Aorta

By ROBERT F. ZIEGLER, M.D.

The electrocardiogram in uncomplicated coarctation of the aorta in infants reflects the pattern of the fetal circulation and serves as a useful prognostic guide. When the fetal ductus inserts proximally to the region of coarctation and closes at birth, right ventricular enlargement is present at birth but is retrogressive. Furthermore, in this situation collateral circulation has begun to develop before birth so that the left ventricle can compensate on assumption of the postnatal circulation, and the prognosis is good. When the insertion of the fetal ductus is distal to the coarctation, progressive, and frequently fatal, left ventricular enlargement and failure develop shortly after birth. Modifications of these electrocardiographic patterns in the production of right bundle branch block are also discussed and illustrated.

LITTLE has been written by way of defining the precise value of the electrocardiogram in the various forms of congenital heart disease. A few papers have described the form of the auricular and ventricular deflections in autopsied cases, but this type of information has added little to the solution of clinical problems in living patients. Advances that are continuing to be made in the surgical correction of congenital cardiovascular defects obviously must be accompanied, or better, preceded, by similar advances in accuracy of differential diagnosis, prognosis, and the selection of cases for operation. To this end a number of clinical and laboratory procedures are available. Against a background of clinical, physiologic, roentgenographic, surgical, and pathologic data, it is our purpose to evaluate critically the clinical importance of the electrocardiogram in coarctation of the aorta.

MATERIAL AND DIAGNOSIS

Material for this study includes observations from a total of 57 patients with coarctation of the aorta, 54 of whom had complete electrocardiograms recorded. The age range of these patients was from 10 days to 36 years. In 38 cases the coarctation was uncomplicated; in eight there was only an associated patent ductus arteriosus (two proximal, four distal to the coarctation, and two in an indeterminate position); in nine there were other associated intracardiac anomalies; two occurred in association with a double aortic arch and two with a right retroesophageal subclavian artery attached distal to the region of coarctation. Physiologic studies were done in 15 cases, retrograde aortography in 2, and surgery in 34 with only one operative mortality.* In the entire group there were 15 deaths and 11 autopsy examinations.

It must be generally agreed that the electrocardiogram is not in any way essential for establishing the diagnosis of coarctation of the aorta. The clinical diagnosis depends on a demonstration of the mechanical effects of a localized obstruction to aortic blood flow, to which the electrocardiogram can only give confirmatory information, namely, evidence of unilateral ventricular enlargement. An important question will therefore concern itself with the accuracy with which the electrocardiogram can provide this information.

It is interesting to note that in 57 patients with coarctation of the aorta the clinical diagnosis was made correctly in 46 and missed in 11. The diagnosis of coarctation was made incorrectly in one case not included in this report (a 2 week old infant in whom the autopsy diagnosis of an endocardial tumor of the left ventricle was established). All 11 patients in whom the clinical diagnosis was missed were infants, and all had complicated combinations of defects in which the primary diagnostic evidence of aortic obstruction was diminished or absent. These included: (1) double aortic arch with coarctation of the anterior segment; (2) patent ductus arteriosus with aortic insertion distal to the region of coarctation; (3) retroesophageal right subclavian artery originating distal to the coarctation and carrying blood into the descending aorta; and (4) transposition of the great vessels with other associated intracardiac defects. In most of these instances the diagnosis of coarctation probably could have been made by roentgenographic visualization (retrograde

* All operations were done by Dr. Conrad R. Lam.

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aortography). Furthermore, diagnostic roentgenography has the additional advantage of making evident the exact location and extent of the coarcted segment, a fact which might occasionally be of importance in adults but is seldom necessary in small children.

**Electrocardiographic Findings**

One of the ways in which the electrocardiogram should be most helpful in establishing or confirming the diagnosis of any particular form of congenital heart disease is the indication it gives of unilateral ventricular enlargement consistent with the defect in question. Furthermore, it might reasonably be expected that in uncomplicated coarctation of the aorta the electrocardiogram should display some evidence of left ventricular enlargement. But when one observes the high incidence of right as well as left ventricular hypertrophy in such patients (table 1), the problem of reconciling the diversity of electrocardiographic patterns within what appears to be a single clinical entity justifiably arises. An explanation for these observations in terms of other correlative data should help to establish the significance and usefulness of the electrocardiogram in this condition.

It is to be noted that all of the patients with uncomplicated coarctation of the aorta whose electrocardiograms were characteristic of right ventricular hypertrophy were infants less than 6 months of age. Several observations in this group of patients are worthy of particular mention: (1) Two infants, age 10 days and 2 weeks, respectively, when first seen were in severe right-sided congestive heart failure which rapidly and permanently disappeared following the administration of digitalis, diuretics, and oxygen (fig. 1). (2) Two babies, age 2 weeks and 5 months, respectively, had electrocardiographic evidence of right ventricular hypertrophy but a normal instead of an elevated right ventricular mean pressure. (3) All of the electrocardiograms in this group which were originally characteristic of right ventricular hypertrophy progressed during the first year to patterns of right bundle branch block with or without associated evidence of left ventricular hypertrophy. (4) All of the infants survived and later had successful surgical correction of the aortic coarctation. (5) At the time of operation in these cases the aortic insertion of the ligament of the ductus was located at or just proximal to the site of the coarctation.

In contrast to these infants with uncomplicated coarctation of the aorta and electrocardiographic evidence of right ventricular hypertrophy are a comparable number in the same age group with electrocardiograms characteristic of left ventricular hypertrophy or left bundle branch block (fig. 2). The most notable observation in this group is the high mortality rate of 60 per cent during early infancy. Unfortunately, autopsy information is lacking with regard to the aortic attach-

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**Table 1.**—Electrocardiographic Findings in Coarctation of the Aorta

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>RVH</th>
<th>RVH→Rt. BBB</th>
<th>Rt. BBB</th>
<th>LVH</th>
<th>Lt. BBB</th>
<th>Indeterminate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uncomplicated coarctation</td>
<td>38</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Birth-18 months</td>
<td>12</td>
<td>4</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>3 years-adult</td>
<td>26</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>11</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Coarctation with patent ductus</td>
<td>8</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Proximal to coarctation</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1</td>
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<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Distal to coarctation</td>
<td>4</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Indeterminate position</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Complicating defects</td>
<td>11</td>
<td>0</td>
<td>4</td>
<td>1</td>
<td>4</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>57</td>
<td></td>
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</tbody>
</table>

RVH—Right ventricular hypertrophy; Rt. BBB—Right bundle branch block; LVH—Left ventricular hypertrophy; Lt. BBB—Left bundle branch block.
Fig. 1. A series of precordial leads in a baby with uncomplicated coarctation of the aorta, in whom the fetal ductus attached proximal to the region of coarctation and closed at birth. When first seen at the age of 10 days there was evidence of right ventricular enlargement and right-sided congestive heart failure which cleared rapidly and permanently under medical management. Surgical resection of the coarctation was done successfully at the age of 3½ years. Note the electrocardiographic progression from right ventricular hypertrophy to right bundle branch block.

Fig. 2. The precordial-lead electrocardiogram in a baby with uncomplicated coarctation of the aorta. Although the electrocardiogram displays evidence of left ventricular hypertrophy and at 2 years left bundle branch block, there is also evidence at 3 weeks of associated right bundle branch block. At no time was there evidence of cardiac failure, and surgery was done successfully at the age of 2 years. It is impossible to state the exact functional position of the fetal ductus in this case. At operation the ligament of the ductus attached almost exactly into the region of coarctation.
ELECTROCARDIOGRAM IN COARCTATION OF THE AORTA

ment of the ligamentum arteriosum, which might be assumed to have been distal to the region of coarctation.

DISCUSSION

The very minimum that can be learned from the foregoing observations is that the electrocardiogram in uncomplicated coarctation of the aorta has considerable prognostic significance. Evidently, whatever mechanism produces predominant left ventricular enlargement in infants with coarctation also predisposes them to an early fatal termination, usually cardiac (left ventricular) failure. Infants with coarctation and predominant right ventricular enlargement are apparently not so predisposed, cardiac strain and failure in these cases being retrogressive rather than progressive as in the former group. An explanation which has previously been suggested involves the position of the fetal ductus with relation to the region of coarctation and the resulting pattern of the circulation before and just after birth.¹

Assuming that coarctation of the aorta is present during fetal life, the circulatory dynamics and resulting work-load of right and left ventricles would depend on the location of the aortic insertion of the ductus arteriosus. If this were distal to the region of coarctation, the circulatory dynamics would not be basically different from that of normal infants in utero, and the size of each ventricle at birth would be normal. Further, since the descending aorta would be supplied with blood from the pulmonary artery via the ductus arteriosus, there would be no stimulus for the development in utero of any collateral circulation around the region of coarctation. At birth, particularly if the ductus closes normally, a severe strain is placed on the left ventricle, which cannot be well borne because of the lack of adequate collateral circulation, with the result that progressive left ventricular enlargement and failure are likely to occur and will frequently result in early death.

If the fetal ductus inserts proximally to the region of coarctation, or in such a manner that the coarcted segment obstructs the ductus as well as the aorta, pressure within the pulmonary vascular circuit would be increased, and the resulting right ventricular enlargement would be present at birth. If the lesion were sufficiently severe, right-sided cardiac failure might even be present at this time. At birth, particularly if the ductus closes, the abnormal

![Fig. 3. The precordial-lead electrocardiogram in a child with coarctation of the aorta with a patent ductus arteriosus inserting proximal to the region of coarctation. At 5 months the electrocardiogram shows right ventricular hypertrophy probably in combination with right bundle branch block; at 2 years right bundle branch block predominates, possible with the addition of left ventricular hypertrophy (ventricular activation time of 0.04 second in lead V₄). Cardiac catheterization at 5 months indicated an elevated right ventricular pressure. Surgical correction of both patent ductus arteriosus and the coarctation was done successfully at the age of 2 years.](image-url)
work-load imposed on the right ventricle in utero is abruptly transferred to the left ventricle which usually does not dilate and fail since adequate collateral circulation is supposed to have developed antenatally under these circumstances. Cardiac failure and right ventricular enlargement in this type of case should be and usually is retrogressive and thus bears a favorable prognosis without any need for immediate surgical interference.

If the original thesis concerning the relation between the incidence of neonatal right ventricular hypertrophy in coarctation of the aorta and the proximal position of the fetal ductus arteriosus is correct, then one would expect electrocardiograms characteristic of right ventricular hypertrophy or right bundle branch block in infants with coarctation and persistent patency of a proximal ductus. This is indeed the case as observed in two such infants of this series (fig. 3). One essential difference between these comparable groups of cases with electrocardiographic evidence of right ventricular hypertrophy is the normal pulmonary artery and right ventricular pressures in those in which the ductus closed normally at birth and higher than normal pulmonary artery and right ventricular pressures in those in whom the ductus remained patent. These pressures in the latter circumstance are, in fact, higher than in infants of a comparable age with only an uncomplicated patent ductus arteriosus, presumably reflecting the increased pulmonary blood flow resulting from the high aortic-pulmonary pressure gradient. It is in this situation, that is, coarctation of the aorta with a proximal patent ductus, that pulmonary hypertension and progressive pulmonary vascular sclerosis have been reported.  

Because of the pulmonary arterial and right ventricular hypertension when the proximal ductus remains patent, neonatal right ventricular enlargement and possible congestive changes may be expected to regress less rapidly than when the proximal ductus closes normally at birth. In this situation surgical intervention may be necessary much earlier than in uncomplicated coarctation of the aorta (with a closed proximal ductus) and, in some cases at least, earlier than in uncomplicated patent ductus arteriosus.

When the ductus remains patent, the aortic

![Fig. 4. Precordial-lead electrocardiograms of two infants (A, 2½ months; and B, 5 months) with coarctation of the aorta and a patent ductus arteriosus inserting distal to the region of coarctation. Both had anatomic left ventricular hypertrophy at autopsy with some enlargement of the right ventricle as well. Both babies had right ventricular hypertension as indicated by cardiac catheterization. In the first patient (A) the pulmonary artery pressure was lower than right ventricular pressure, indicating a functional pulmonary stenosis. At autopsy this was found to be due to encroachment on the outflow tract of the right ventricle by a greatly hypertrophied interventricular septum, (apparently a true Bernheim syndrome).]
insertion of which lies distal to the region of coarctation, certain reasonably consistent observations may be made. These include the following: (1) Predominant left ventricular hypertrophy was uniformly present (on necropsy evidence in two cases and on clinical evidence in one), usually in combination with some degree of right ventricular enlargement as well. (2) The electrocardiogram rarely displayed evidence of left ventricular hypertrophy, but in all three cases was characteristic of right bundle branch block instead (figs. 4 and 5). (3) Pulmonary artery and right ventricular hypertension were regular features. The mortality rate in this group can hardly be compared with the rate in those in whom the ductus, supposedly attached distal to the coarctation, closed. Of the three infants with a distal open ductus, one died in heart failure at 2½ months, one died during operation as the ductus was being closed, and one survived successful surgery (division of the patent ductus and resection of the coarctation) at the age of 5 months (fig. 5).

From the observations made thus far, it would appear that coarctation of the aorta, regardless of where the ductus inserts and whether or not it remains patent, is regularly associated after the neonatal period with anatomic left ventricular enlargement, providing of course that there is no other intracardiac defect that would disturb the usual relation between right and left ventricular output. The actual appearance of the electrocardiogram, however, seems to depend on other factors. Apparently the only clinical circumstance which favors the development of the classic electrocardiographic pattern of left ventricular hypertrophy is coarctation with a distal closed ductus, or anatomic left ventricular enlargement together with normal right ventricular size and pressure. Currently accepted electrocardiographic evidence of right ventricular hypertrophy was observed in infants with coarctation of the aorta only in two circumstances: (1) transiently during the immediately neonatal period when the fetal ductus attached proximal to the coarctation

![Fig. 5. The precordial leads in a baby with coarctation of the aorta and a distal patent ductus arteriosus. Right ventricular pressure was elevated, and the degree of cardiac enlargement plus the presence of congestive heart failure made operation seem imperative. Division of the ductus followed by resection of the coarctation was done successfully at the age of 5 months. Note the change from predominant right to left ventricular hypertrophy. One would probably be correct in assuming the presence of both right and left ventricular enlargement at 5 months although this could hardly be detected by the electrocardiogram at this time. A, preoperative (5 months); B, postoperative (6 months).]
but closed normally at birth, and (2) infrequently during later infancy when the ductus attached distal to the coarctation and remained patent.

The most common electrocardiographic pattern in infants with coarctation of the aorta is that of right bundle branch block which appears under two fundamentally different

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**Fig. 6.** Precordial lead electrocardiograms in three older children with uncomplicated coarctation of the aorta. All show evidence of left ventricular hypertrophy. For further discussion, see text.

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**Fig. 7.** Precordial lead electrocardiograms in three patients with uncomplicated coarctation of the aorta. All display evidence of right bundle branch block. For further discussion, see text.
circumstances. The first is the stage of retrogression of the right ventricular hypertrophy which characterizes the electrocardiograms of infants with coarctation and a proximal closed ductus. The second is in association with the combination of left ventricular enlargement and right ventricular hypertrophy or hypertension. The latter combination occurs with

Fig. 8. Precordial leads in a child with an uncomplicated interauricular septal defect as diagnosed by cardiac catheterization. Note again the early pattern of right ventricular hypertrophy changing to one of uncomplicated right bundle branch block. For further discussion, see text.

Fig. 9. Precordial leads $V_4R$ and $V_1$ through $V_6$ in a child with a normal heart. Note the evidence of apparent right ventricular hypertrophy at the age of 2 weeks and the progression to a pattern of uncomplicated right bundle branch block. For further discussion, see text.
coarctation of the aorta and a patent ductus either proximal or distal to the region of aortic obstruction.

The explanation for the electrocardiographic pattern of left ventricular hypertrophy in older children and adults with uncomplicated coarctation of the aorta seems obvious. However, in older children and adults nearly half of the electrocardiograms are characteristic of right bundle branch block instead of left ventricular hypertrophy (table 1, figs. 6 and 7). This pattern, that is, right bundle branch block, might reasonably be assumed to represent a stage in the progression from neonatal right ventricular hypertrophy, and has been observed in at least three different clinical situations: (1) in uncomplicated coarctation of the aorta with the fetal ductus attached proximal to the coarctation and closed at birth (fig. 1); (2) in uncomplicated interauricular septal defect (fig. 8); and (3) in the normal electrocardiographic progression in infants without any evidence of cardiovascular disease (fig. 9). Without precise information regarding the electrophysiological mechanism involved, these observations provide a description of one consistent electrocardiographic sequence in the development of the pattern of right bundle branch block. An important associated problem, which is the subject of a separate study, is to determine criteria for the detection of right or left ventricular hypertrophy in combination with defects of intraventricular conduction. Mention should also be made that in older children and adults the different electrocardiographic patterns do not have the same prognostic value as stated for the infant group.

Summary and Conclusions

Correlated data have been assembled in 57 patients with coarctation of the aorta, ranging in age from 10 days to 36 years. On the basis of this material the following observations have been made.

1. Electrocardiographic evidence of right ventricular hypertrophy or right bundle branch block was observed frequently in infants with uncomplicated coarctation of the aorta. The following statements regarding this group of patients seemed justified, and evidence was cited: (a) Right ventricular enlargement at birth results from the fetal circulatory pattern with the ductus inserting proximal to the region of coarctation. (b) Neonatal right ventricular enlargement and occasionally right-sided congestive heart failure are retrogressive and do not necessitate immediate surgical interference if the ductus closes normally at birth. (c) The prognosis in these infants is excellent.

2. Electrocardiographic evidence of left ventricular hypertrophy (and occasionally left bundle branch block) was observed less frequently in infants with uncomplicated coarctation of the aorta than in older children and adults with this defect. The following observations appeared to characterize this group of infants: (a) Left ventricular enlargement and failure are progressive and usually fatal. (b) This sequence of events is believed to represent the pattern of the fetal circulation with the ductus in utero inserting distal to the region of coarctation and closing after birth.

3. The genesis of the electrocardiographic pattern of right bundle branch block in older children and adults with uncomplicated coarctation of the aorta is believed to result, in part at least, from the sequential progression of right ventricular hypertrophy to right bundle branch block as observed when the fetal ductus inserts functionally proximal to the region of coarctation and closes at birth.

4. The same electrocardiographic sequence, from right ventricular hypertrophy to right bundle branch block, has also been observed in normal infants and children and in babies with uncomplicated interauricular septal defect.

5. Right bundle branch block is apparently a characteristic electrocardiographic pattern when there is combined right and left ventricular enlargement or when there is anatomic left ventricular hypertrophy and functional right ventricular hypertension. This pattern was observed in coarctation of the aorta with persistent patency of the ductus arteriosus regardless of the exact site of its insertion.

Sumario Español

El electrocardiograma en casos de coartación de la aorta en infantes sin complicaciones refleja el patrón de circulación fetal y sirve...
como un guía de pronóstico de valor. Cuando el ducto fetal se inserta próximamente a la región de la coartación y se cierra al nacer, el ventrículo derecho se encuentra agrandado a la vez, pero este engrandecimiento es retrogrado. Además, en esta situación la circulación colateral se ha empezado a desarrollar antes de nacer de manera que el ventrículo izquierdo puede compensar al comenzar la circulación post-natal, y el pronóstico es favorable. Cuando el ducto fetal se inserta distal a la coartación, engrandecimiento y decompensación progresiva del ventrículo izquierdo a veces fatal se desarrollan prontamente después del nacimiento. Modificaciones de estos patrones electrocardiográficos en la producción de bloqueo de la rama derecha del haz de His también se discuten y se ilustran.

REFERENCES


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