

The Electrocardiogram in Congenital Heart Disease

With Special Reference to Left Axis Deviation

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The incidence of the four main types of axis deviation in each diagnostic group of congenital malformations of the heart was found by an examination of 2,752 electrocardiograms from the Harriet Lane Home Cardiac Clinic. Records from patients with dextrocardia or dextrarotation of the heart, and those with complete bundle branch block were excluded. The precordial and unipolar limb leads in the 289 electrocardiograms which showed left axis deviation were examined, and the types of precordial pattern most frequently seen in the different diagnostic groups analyzed. It is concluded that the precordial patterns show considerable difference in the various malformations associated with left axis deviation, and are of diagnostic aid.

THE FREQUENT FINDING of left axis deviation in the electrocardiogram of patients with tricuspid atresia and its extraordinary rarity in other cyanotic cardiac malformations, prompted an investigation into the degree of deviation of the electrical axis of the heart in congenital heart disease.

A total of 2,874 electrocardiographic records was studied from different patients who attended the Harriet Lane Home Cardiac Clinic between 1946 and 1952. Only patients with congenital malformations of the heart are included; cases of functional murmur or acquired heart disease were omitted. The electrocardiogram described is that obtained at the first attendance. Among the 2,874 records, there were 77 instances of dextrocardia or dextrarotation of the heart and 45 instances of complete bundle branch block; eight of these showed a Wolff-Parkinson-White syndrome, three left bundle branch block and 34 right bundle branch block. All of these were excluded from the study, leaving a total of 2,752 electrocardiograms from patients with congenital heart disease.

The *clinical diagnosis* in each case was based on a complete medical history, physical examination, fluoroscopy, and x-ray films of the heart. A red blood cell count, hemoglobin and hematocrit was obtained. Many patients were subsequently re-

examined and cardiac catheterization and angiocardiology performed.

It should be emphasized that a large number of patients referred to this clinic, particularly in 1946 and 1947, were in the cyanotic group; 845 of the 2,752 records were from patients with tetralogy of Fallot. For this reason, this study cannot be used to determine the incidence of the various malformations in congenital heart disease. It is believed, however, that within each diagnostic group, the incidence of the four main types of axis deviation can be assessed and is significant.

The findings in the present series are considered in two parts. Part I is devoted to the incidence of the different types of axis deviation in this series of cases, and Part II to a detailed analysis of the electrocardiograms showing left axis deviation.

PART I.—THE INCIDENCE OF THE DIFFERENT TYPES OF AXIS DEVIATION IN VARIOUS CONGENITAL MALFORMATIONS

The Criteria Committee of the New York Heart Association¹ has adopted three simple descriptive groups for the electrical axis of QRS. These are: no deviation of the electrical axis, 30 to 90 degrees; left deviation of the electrical axis, 29 to minus 90 degrees; right deviation of the electrical axis, 91 to minus 91 degrees. This nomenclature has been adopted

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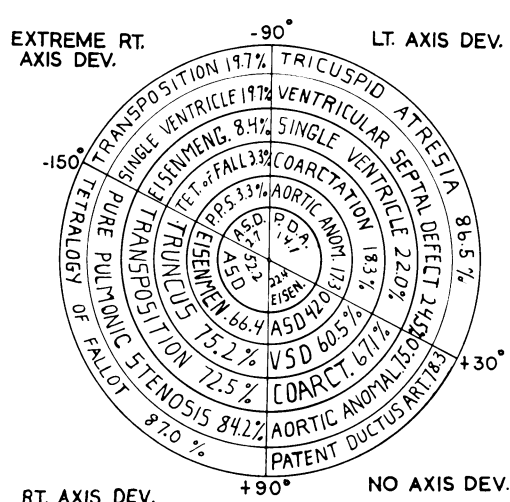


FIG. 1. The four main types of axis deviation, and the six congenital malformations most commonly associated with each type.

in this paper with a slight modification. The group of right axis deviation has been divided into: right deviation of the electrical axis, 91 to minus 149 degrees; an extreme right deviation of the electrical axis, minus 150 to minus 91 degrees.

(1) No axis deviation (plus 90 to plus 30 degrees) was present in 34.5 per cent of all cases (944 records).

(2) Left axis deviation (plus 29 to minus 90 degrees) was present in 10.5 per cent of all cases (289 records).

(3) Right axis deviation (plus 91 to minus 149 degrees) was present in 51.1 per cent of all cases (1,411 records).

(4) Extreme right axis deviation (minus 91 to minus 150 degrees) was present in 3.9 per cent of all cases (108 records).

The high incidence of right axis deviation in this series probably reflects the large number of cyanotic patients seen. The least common type of axis deviation is that lying between minus 90 and minus 150 degrees. In figure 1, the four types of axis deviation are represented as segments of a circle, and the six most common malformations in each type are shown.

Right axis deviation was present in 87 per cent of the 845 patients with tetralogy of Fallot, in 84 per cent of the 177 with pulmonary valvular stenosis, and in 75 per cent of the 85 patients with truncus arteriosus. The inci-

dence of right axis deviation in auricular septal defect, Eisenmenger's complex, and transposition of the great vessels ranged between 50 and 70 per cent. The incidence in ventricular septal defect and in coarctation of the aorta was 15 per cent; indeed, the majority of infants under 2 years of age with coarctation showed right axis deviation. In figure 2, the total number of patients with each clinical diagnosis is shown and the incidence of each type of axis deviation, in that diagnostic group, is reported.

No axis deviation was present in approximately 70 per cent of patients with patent ductus arteriosus, congenital aortic valvular lesions, or coarctation of the aorta. Isolated ventricular septal defect was diagnosed in 348 patients, 60 per cent of whom showed a normal axis. There were 224 patients with

THE DISTRIBUTION OF THE ELECTRICAL AXIS IN 2752 CASES OF CONGENITAL CARDIAC DISEASE

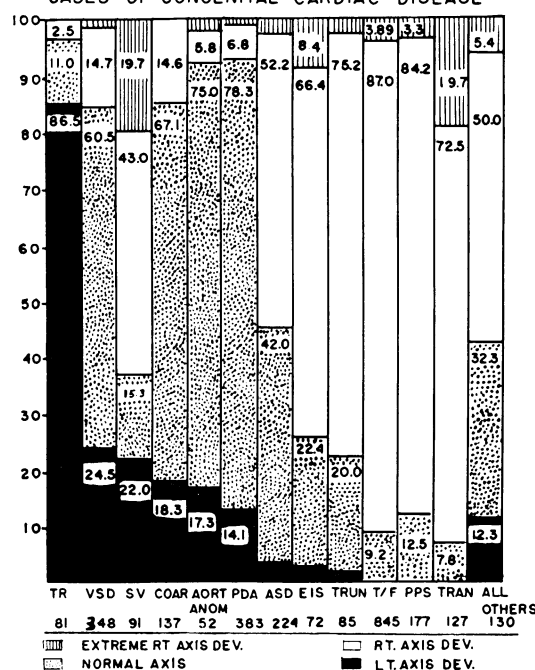


FIG. 2. The distribution of the electrical axis in 2,752 cases of congenital heart disease. (Total cases in each group is given below diagnosis.) Abbreviations: TR, tricuspid atresia. VSD, ventricular septal defect. SV, single ventricle. COAR, coarctation of the aorta. AORT ANOM, other aortic anomalies. PDA, patent ductus arteriosus. ASD, auricular septal defect. EIS, Eisenmenger's complex. TRUN, truncus arteriosus. T/F, tetralogy of Fallot. PPS, pulmonary valvular stenosis. TRAN, transposition of the great vessels.

auricular septal defects, 42 per cent with a normal axis. In Eisenmenger's complex and truncus arteriosus, the incidence of normal axis deviation was approximately 20 per cent; in tetralogy of Fallot, pulmonary valvular stenosis, transposition of the great vessels and tricuspid atresia, it was approximately 10 per cent.

Extreme right axis deviation was most often observed in transposition of the great vessels and in single ventricle (20 per cent of 127 cases and 91 cases, respectively). The incidence in Eisenmenger's complex was 8 per cent, and in tetralogy of Fallot and auricular septal defect approximately 3 per cent. It was not seen in tricuspid atresia.

Left axis deviation was present in 289 instances. A diagnosis of tricuspid atresia was made in 70 of these patients and confirmed at autopsy in 11. Left axis deviation thus occurred in 86 per cent of the 81 patients with this diagnosis. Of the 348 patients with ventricular septal defects, 84 (24 per cent) showed left axis deviation. The clinical diagnosis was confirmed by cardiac catheterization in 11 of the 84. Single ventricle was diagnosed in 91 patients of whom 20 (22 per cent) showed left axis deviation. The diagnosis was confirmed at autopsy in only 6. Coarctation of the aorta was present in 137 patients, of whom 24 (18 per cent) showed left axis deviation: in 16 of these, the diagnosis was confirmed at operation. The diagnostic group "other aortic anomalies" includes 52 patients with congenital aortic or subaortic stenosis, vascular ring, or congenital aortic insufficiency. Of these, 9 (17 per cent) showed left axis deviation. Only 54 patients (14 per cent) with a patent ductus showed left axis deviation; the diagnosis was confirmed surgically in 43.

There were 28 patients with left axis deviation not included in the six major diagnostic groups above. Of these, seven had auricular septal defects (two confirmed by cardiac catheterization).^{*} There were also two patients with Eisenmenger's complex and two with truncus arteriosus (which represented an

incidence of left axis deviation of 2.5 per cent in both these conditions).

The 17 remaining cases are included in the group of "rare malformations" shown in the last column of figure 2. There were 6 patients, out of a total of 19 patients, with situs inversus and levocardia in whom the electrocardiogram showed left axis deviation. One of the six came to autopsy and had a single ventricle and a large auricular defect. The diagnosis of atrioventricularis communis was made in 12 instances (mainly on the basis of coexisting mongolism), and in four of these left axis deviation was present. Left axis deviation was seen in one instance in each of the following diagnostic groups: Ebstein's anomaly (total 14 cases), congenital mitral insufficiency (total five cases), pulmonary hypertension with reversed flow through a patent ductus (total six cases), and underdevelopment of the right ventricle without tricuspid atresia (total three cases). Cardiomegaly of undetermined origin was associated with left axis deviation in three patients.

Among the electrocardiograms in this study, no instance was found of left axis deviation in association with tetralogy of Fallot, pulmonary valvular stenosis, or transposition of the great vessels. Since the data were collected, however, one infant with marked left axis deviation was found to have complete transposition and a small right ventricle at autopsy.

Confirmation of the clinical diagnosis in the 289 cases showing left axis deviation, was obtained in only slightly over one-third of the cases. In 20, the diagnosis was confirmed by autopsy, in 59, by surgery, and in 34 by cardiac catheterization or angiocardiology, or both.

PART II.—ANALYSIS OF ELECTROCARDIOGRAMS WITH LEFT AXIS DEVIATION

A marked degree of left axis deviation (minus 31 to minus 90 degrees) was more frequent in single ventricle and tricuspid atresia than in the other malformations. In patent ductus arteriosus, coarctation of the aorta, and other aortic anomalies, left axis deviation was frequently mild and seldom more than moderate in degree (table 1).

The 289 records which showed left axis

^{*} It has been the experience of this clinic, that in this malformation left axis deviation usually indicates a low septal defect of the ostium primum type.

TABLE 1.—*The Electrical Axis Within the Six Major Groups of Congenital Heart Disease Showing Left Axis Deviation*

Diagnosis	No. cases	Slight l. axis, $+29^{\circ}$ to 0°	Moderate, -1° to -30°	Marked l. axis, -31° to -90°
		%	%	%
Tricuspid atresia...	70	9	25	66
Single ventricle....	20	15	10	75
Vent. septal defect..	84	37	31	32
Pat. duct. art.....	54	59	26	15
Coarctation.....	24	42	46	12
Aort. anom.....	9	55	45	0
Total.....	261	37	30	33

deviation were further analysed to determine what differences, if any, were present in the precordial and unipolar limb leads in the various diagnostic groups, and whether left axis deviation was uniformly accompanied by other evidence of left ventricular preponderance.

Unipolar Limb Leads in Cases with Left Axis Deviation

The augmented limb leads of Goldberger² aV_R , aV_L and aV_F had been taken in 157 patients with left axis deviation in the electrocardiogram. These leads were studied only to determine the electrical position of the heart in relation to the antero-posterior axis of the body. The position was horizontal or semi-horizontal in 146 of the 157 cases.

Precordial Leads in Cases with Left Axis Deviation

Precordial leads were taken in 278 of the 289 patients with left axis deviation. A right and left precordial lead (V_1 and V_5 or V_6) were used for analysis and the pattern in each was classified according to the scheme set out in figure 3. The total number of cases in each clinical group with a particular type of precordial pattern is shown in figure 4. The patterns in the precordial leads give reliable information concerning the relative thickness of right and left ventricle. In this sense the term "preponderance" of one ventricle over the other has been used in the interpretation of these leads. Such preponderance may be nor-

mal and the term is not used to indicate hypertrophy.

Type I (R Pattern). The prominent R wave may be preceded by a Q or followed by an S wave, or both may be present. The R wave may be slurred but not notched and the duration of the whole complex is less than 0.10 second. When this pattern occurs in V_1 , it is taken to indicate right ventricular preponderance (fig. 3, V_1 type I). An S wave is usually present in this lead in the physiological type of right ventricular preponderance seen in infancy. Consequently in this age group Rs pattern is frequent but R or qR is rare, and when it does occur, is usually associated with pathological right ventricular hypertrophy. Only 30 instances of this V_1 type I pattern were found in the electrocardiograms showing left axis deviation, and subdivision into physiological and pathological patterns was not attempted.

An R or qR pattern in V_5 which is the pattern of normal preponderance, was found in 224 records (fig. 4).

Type II a. (rSr₁ Pattern). The r and r₁ are of equal amplitude and the duration of the whole complex less than 0.11 second. This pattern in V_1 is generally considered normal in children.^{3, 4}

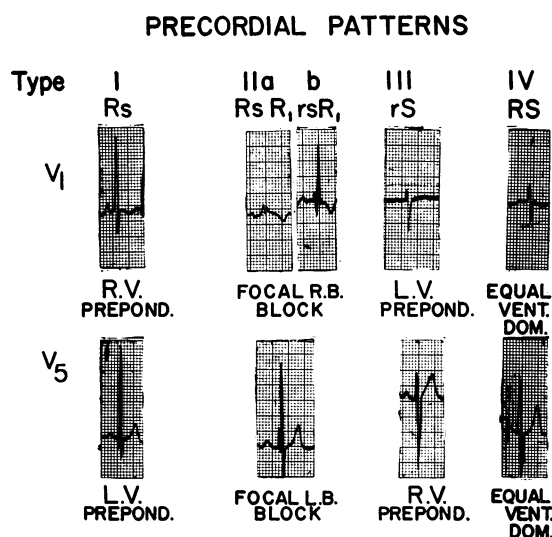


FIG. 3. Types of precordial pattern. Abbreviations: RV, right ventricle. LV, left ventricle. PREPOND., preponderance. VENT. DOM., ventricular dominance.

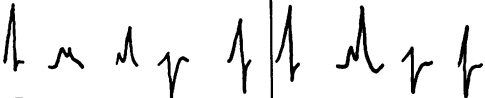
		V ₁ pattern					V ₅ or V ₆ pattern				
DIAGNOSIS	No.										
AGE	of cases	Type	I	IIA	IIB	III	IV	I	II	III	IV
TRICUSPID ATRESIA	66					59		55			
0-6 mo.	13					10	3	11		1	1
6-12 "	12					11	1	9		2	1
1-2 yr.	6					6		5		1	
2-5 "	11					10	1	9		2	
5-15 "	20					18	2	17		1	2
15 yrs. +	4					4		4			
SINGLE VENTRICLE	19					12		11			
0-6 mo.	3		1			2				1	2
6-12 "	4				3		1	2		1	1
1-2 yr.	3		1			2		3			
2-5 "	2		1			1		1			1
5-15 yr.	4					4		2		1	1
15 yrs. +	3					3		3			
VENT. SEPT.DEFECT	82					32		63			
0-6 mo.	11		5		3	3		9			2
6-12 "	7		5		1	1		4		1	2
1-2 yr.	14		2	2	4	4	2	11			3
2-5 "	20		2	4		11	3	14			6
5-15 "	24		1	4	8	11		19		1	3
15 yrs. +	6		1	3		2		6			
PAT.DUCT. ART.	52					31		50			
0-6 mo.	3		1			1	1	2			1
6-12 "	1						1	1			
1-2 yr.	5		1	2			2	5			
2-5 "	10		2			4	4	9		1	
5-15 "	22		1		1	16	4	22			
15 yrs. +	11					10	1	11			
COARCTATION	24					19		18			
0-6 mo.	0										
6-12 "	0										
1-2 yr.	1			1						1	
2-5 "	2			1	1						2
5-15 "	11			1	1	9		9			2
15 yrs. +	10					10		9			1
RARER GROUPS	35					16		27			
AORTIC LESIONS	9					9		9			
AURICULAR DEFECT	7		1	3	3			3		1	3
SITUS INV.& LEVO.	6		2		1	3		4		1	1
A.V.COMMUNIS	4		1			2	1	3			1
EISENMENGER	2		1	1				2			
TRUNCUS	2				1		1	2			
UNDER DEV. R.V.	1					1		1			
REVERSED DUCTUS	1				1					1	
EBSTEIN'S	1				1			1			
CARDIOMEG. ? CAUSE	2				1	1		2			

Fig. 4. The distribution of the different types of precordial patterns in patients with congenital heart disease with left axis deviations, showing 243 cases. There are 5 major diagnostic groups, subdivided by age; also 35 cases in the rarer diagnostic groups.

Type II b. (rSR₁ Pattern). The R₁ is of greater amplitude than the initial R. The total is less than 0.11 second. Some authors^{5, 6} consider that this type II b pattern in V₁ is usually associated with right ventricular hypertrophy.

In the present series, V₁ type II (a and b) occurred 51 times and was most frequent in children over 1 year of age with ventricular septal defects, auricular septal defects and coarctation. The pattern was not seen in tricuspid atresia.

No instances of V₅ type II pattern, indica-

tive of incomplete left bundle branch block, were found in the present series.

Type III (rS Pattern). There is a small positive R deflection followed by a large negative S wave and the total duration of the complex is less than 0.10 second. This pattern in V₁ is seen in normal adults and with increasing frequency in normal children as age advances. It reflects the normal left ventricular preponderance and occurred in 169 instances in the present series. In most malformations, the incidence was greater in the older age groups but in tricuspid atresia it was almost invariably present from birth (fig. 4).

The rS pattern in V_5 , which may mean right ventricular preponderance was seen in only 17 cases in the present series.

Type IV (RS Pattern). The R wave and S wave are of equal amplitude. In V_1 it was seen in 28 instances in the present series, including 14 of the 52 patients with patent ductus arteriosus, and all seven cases of tricuspid atresia in which a V_1 Type III pattern was not present.

An RS pattern in V_5 was present in 36 instances, that is, in the majority of records which did not show the R pattern in this lead.

Correlation of the Type of Precordial Pattern in V_1 and V_5 . Patients with congenital heart disease, who showed left axis deviation in the electrocardiogram, had a type I pattern in V_5 in 81 per cent of cases (224 of 277 records). This pattern of left ventricular preponderance was not noticeably influenced by age. The incidence of a left ventricular preponderant pattern in V_1 was lower (61 per cent); furthermore, in most malformations it was more common in the older age group.

A left ventricular preponderant pattern in both V_1 and V_5 occurred in 83 per cent of patients with tricuspid atresia, and did not vary with age. This combination was found in all cases of "other aortic anomalies" with left axis deviation and, in 75 per cent of those with coarctation, 58 per cent of those with patent ductus and 42 per cent of the 19 cases of single ventricle.

The combination of a type I pattern in V_5 and type IIa or IIb pattern (incomplete right bundle branch block) in V_1 occurred in 45 per cent of cases of ventricular septal defect, and in 20 per cent of the patients under 15 years with a patent ductus. This combination has come to be considered very typical of these two conditions; it is also frequently found in these malformations even when there is no deviation of the electrical axis.

An rS pattern, which when seen in V_1 indicates left ventricular preponderance and in V_5 indicates right ventricular preponderance, was seen in both leads only in tricuspid atresia and single ventricle. It is believed that, at least in the former condition, it indicates clockwise rotation of the heart.

A type IV pattern in both V_1 and V_5 was seen occasionally in tricuspid atresia and in ventricular septal defect.

DISCUSSION

The nomenclature adopted for the electrical axis of QRS is descriptive and does not indicate which of the subdivisions are normal or abnormal.

The presence of left axis deviation in children and even in youth may be of some significance. In a study of 1,276 electrocardiograms taken in 167 average healthy infants and children whose ages ranged from 3 weeks to 12 years, 4.73 per cent were found to have left axis deviation.⁷ The criteria for axis deviation was similar to that used in this study. In another series of 679 healthy infants and children ranging in age from 6 months to 9 years, only three were found with an electrical axis less than 0 degrees.⁸ A further study of 100 healthy children aged between a few weeks and 14 years, only two were found to have an electrical axis less than 20 degrees.⁹ Even in normal young adults left axis deviation is not a common finding; in a study of 500 young adult males no more than 22 were found to have axes less than 0 degrees.¹⁰

Little uniformity has been present in the criteria adopted for the description of axis deviation in these reports on normal individuals. In the present series of patients, 845 had tetralogy of Fallot; the right axis deviation, which occurs so frequently in this condition, would decrease the incidence of left axis deviation in the group as a whole. Also, this series of patients with congenital heart disease, although very largely composed of infants and children does contain a small number of young adults. For these reasons the true significance of left axis deviation in the electrocardiograms of children is difficult to assess. Nevertheless, the incidence of 10.5 per cent of children with left axis deviation in this series of 2,786 patients with congenital heart disease suggests that this finding is frequently associated with an abnormality and therefore whenever found warrants further investigation before the child is considered normal.

In this large group of congenital cardiac

conditions, it appears furthermore that with very few exceptions those conditions where left ventricular preponderance is expected are either in the left axis deviation group or the no axis deviation group. Those conditions with right ventricular preponderance are to be found in the right axis and extreme right axis groups.

Analysis of the precordial and limb leads in the electrocardiograms which showed left axis deviation, reveals that in tricuspid atresia and in some instances of single ventricle, there is definite associated left ventricular preponderance. These two conditions account for the great majority of patients with cyanotic congenital heart disease who show left axis deviation. More rarely, this combination of left axis deviation and cyanosis may be seen in Eisenmenger's complex, truncus arteriosus, situs inversus with levocardia, atrioventricularis communis, or underdevelopment of the right ventricle without tricuspid atresia. The precordial electrocardiograms in these conditions may show right ventricular preponderance, incomplete right bundle branch block, or equiphasic QRS complexes over the precordium indicative of equal dominance of the two ventricles.

In acyanotic patients, left axis deviation is not so markedly limited to a single malformation. It is found with varying frequency in patients with ventricular septal defects, patent ductus arteriosus, coarctation of the aorta, and other aortic anomalies. In patients with aortic anomalies, the precordial leads reveal that left axis deviation is uniformly accompanied by left ventricular preponderance. In other malformations, left axis deviation is frequently associated with a left ventricular preponderant pattern in V_5 : in ventricular septal defect and patent ductus in early childhood this pattern may be accompanied by incomplete right bundle branch block in V_1 , or even a right ventricular preponderant pattern in this lead.

SUMMARY

The electrocardiograms of 2,752 patients with congenital malformations of the heart were examined and divided into four groups:

(1) No axis deviation (34.5 per cent); (2) left axis deviation (10.5 per cent); (3) those with right axis deviation (51.1 per cent) and (4) extreme right axis deviation (3.9 per cent). Electrocardiograms from patients with dextro-rotated hearts or complete bundle branch block were excluded. Right axis deviation occurred in all the malformations studied but was most frequent in pulmonary valvular stenosis and tetralogy of Fallot.

No axis deviation was found also in association with all the malformations studied; it was seen most frequently in patent ductus, coarctation of the aorta, other aortic anomalies, and ventricular septal defects. Extreme right axis deviation was rare and occurred more frequently in transposition of the great vessels and in single ventricle than in other anomalies. It was not seen in tricuspid atresia or coarctation of the aorta. Left axis deviation in cyanotic patients was overwhelmingly associated with tricuspid atresia although it was seen in single ventricle, Eisenmenger's complex, truncus arteriosus, and a few rare anomalies. In this series, it was not seen in tetralogy of Fallot, pulmonary valvular stenosis or transposition of the great vessels. Among acyanotic patients, left axis deviation was most frequent in ventricular septal defects, coarctation of the aorta, other aortic anomalies and patent ductus arteriosus.

Unipolar limb leads were available in 157 of the 289 electrocardiograms which showed left axis deviation. In 146 of these the electrical position was horizontal or semihorizontal.

The precordial electrocardiogram was available for analysis in 278 of the 289 cases which showed left axis deviation. Among cyanotic patients, left axis deviation was accompanied by marked evidence of left ventricular preponderance in tricuspid atresia and some cases of single ventricle; more variable patterns were obtained in the other malformations. Among acyanotic patients, definite left ventricular preponderance was seen in association with aortic anomalies. In infancy and early childhood, a pattern of left ventricular preponderance in V_5 and incomplete right bundle branch block in V_1 was a frequent combination in patients with ventricular septal defect,

patent ductus arteriosus, and occasionally coarctation of the aorta.

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SUMMARIO IN INTERLINGUA

Le frequentia del quatro major typos de deviation axial in cata gruppo diagnostic de congenite malformationes del corde esseva determinate per le examine de 2.752 electrocardiogrammas obtenite al Clinica Cardiac del Harriet Lane Home a Baltimore, Md. Esseva excludite le registrationes ab patientes con dexterocardia o dexterorotation del corde e con complete bloco del branca dextere. Le derivationes precordial e unipolar extremital esseva examine in le 289 electrocardiogrammas exhibente deviation sinistrorse del axe. Le typos del configurationes precordial que esseva incontrate le plus frequentemente in le varie gruppos diagnostic esseva analysate. Nos conclude que le configurationes precordial monstra considerabile differentias in le varie malformationes associate con deviation sinis-

trorse del axe e que assi illos pote esser de adjuta diagnostic.

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