Differential diagnosis of congenital heart disease in the first 3 months of life

Significance of a superior (left) QRS axis

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Shinebourne, E. A., Haworth, S. G., Anderson, R. H., and Ulgur, A. (1974). *Archives of Disease in Childhood*, 49, 729. **Differential diagnosis of congenital heart disease in the first 3 months of life: significance of a superior (left) QRS axis.** The ECGs of 473 infants under the age of 3 months who were referred to a paediatric cardiological unit were analysed; 47 (10%) of the ECGs showed a superior axis (dominantly negative deflection S wave, in lead aVF). Of these, the majority of noncyanosed patients with plethora on chest x-ray proved to have either an atrio-ventricular canal defect or a large ventricular septal defect. When cyanosis and pulmonary plethora on x-ray were present, tricuspid atresia with increased pulmonary flow (types Ic or IIc) or d-transposition with ventricular septal defect accounted for most cases. With cyanosis and pulmonary oligoemia on x-ray, tricuspid atresia (types Ia and b) or pulmonary atresia with ventricular septal defect accounted for all cases. Finally, 2 patients with superior axis presenting in a shocked condition were found to exhibit the hypoplastic left heart syndrome.

Recognition of superior axis in the ECG provides a useful diagnostic aid in congenital heart disease in early infancy.

Congenital heart disease in the first 3 months of life (excluding asymptomatic infants with heart murmurs), usually presents with cyanosis or heart

failure or a combination of both. A small group of patients with hypoplastic left heart syndrome present with shock due to a low cardiac output. Thus limited types of physiological disturbance are produced despite the multitude of anatomical abnormalities seen in congenital heart disease.

The ECG provides additional diagnostic information pertinent to congenital heart disease, particularly after calculation of the mean frontal QRS axis. During the first months of life this axis alters from $+150^\circ$ to $+135^\circ$ at birth (Ziegler, 1966) to between $+30^\circ$ and $+90^\circ$ at 3 months (Criteria Committee of the New York Heart Association, 1964). A superior axis (between -1° and -180°) is always abnormal and is recognized by a dominantly negative deflection (S wave) in lead aVF. The term 'superior axis' (Liebman and Nadas, 1971) is preferable to 'left axis deviation' since it accurately describes the mean QRS vector as being above the horizontal line, represented by lead I, in the frontal plane. The present investigation considers the

Abbreviations

AVCD: atrioventricular canal defect
DORV: double outlet right ventricle
ECG: electrocardiogram
LAH: left anterior hemiblock
LVH: left ventricular hypertrophy
PDA: persistent ductus arteriosus
PVM: pulmonary vascular markings
RVH: right ventricular hypertrophy
TAPVD: total anomalous pulmonary venous drainage
TGA: transposition of great arteries
VSD: ventricular septal defect

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diagnostic implications of a superior axis in the first 3 months of life when assessed in relation to clinical presentation and plain chest x-ray.

Patients and methods

The ECGs of all children under the age of 3 months admitted to the Brompton Hospital between January 1967 and December 1972 were examined. The case histories, chest x-rays, cardiac catheterization data, and angiocardiograms of those with a superior axis were then evaluated and the anatomical diagnosis determined. The patients were then placed in the following clinical groups.

Group 1. Acyanotic with increased PVM on the plain chest x-ray.

Group 2. Cyanosed with increased PVM on chest x-ray.

Group 3. Cyanosed with decreased PVM on chest x-ray.

Group 4. Shocked with poor peripheral pulses.

Results

Distribution of diagnoses. 47 out of 473 patients (10%) had a superior axis and all had congenital heart disease. The principle anatomical diagnoses are shown in the Table.

TABLE

Principal clinical groupings and anatomical diagnoses in patients under 3 months of age with superior axis

Group I	AVCD	13
	VSD	6
	Coarctation + PDA	2
Group II	Tricuspid atresia type Ic and IIc*	7
	d-TGA + VSD	7
	DORV	1
	TAPVD	1
Group III	Tricuspid atresia type Ia* and Ib	6
	Pulmonary atresia + VSD	2
Group IV	Hypoplastic left heart syndrome	2
Total		47

*Classification of tricuspid atresia (Keith, Rowe, and Vlad, 1958): type I, normally related great vessels; type II, transposed great vessels; (a) with pulmonary atresia, (b) with pulmonary stenosis (hypoplasia), (c) with large VSD and no pulmonary stenosis.

Group 1. Acyanotic with increased PVM. Of the 21 noncyanosed patients with plethora on chest x-ray, 13 had an AVCD, 6 VSD, and 2 had coarctation plus PDA. Those with coarctation were distinguished clinically by the presence of reduced femoral pulses but neither on clinical nor radiological examination, nor by other ECG features

could patients with AVCD be distinguished from those with VSD. Approximately 50% of patients in each group had the pattern of LAH. In this condition there is a Q wave in lead I and a deep S in lead III (Fig. 1). The disturbance in ventricular depolarization, which will be discussed subsequently, reflects an abnormality in part of the left branch of the bundle of His. Similarly, first-degree block, right, left, or biventricular hypertrophy were inconstant features of each group. Final differentiation between AVCD and VSD was made only by left ventricular angiography.

Group 2. Cyanosis with increased PVM. 7 of the 16 patients in this group had tricuspid atresia (types Ic or IIc) (Table), 7 had d-transposition of the great arteries (d-TGA) associated with VSD, 1 had TAPVD, and 1 DORV. Dominant LVH was found in all patients with tricuspid atresia, as was the Q1 S3 pattern of LAH (Fig. 1). The remainder of the cases in this group all had right or biventricular hypertrophy. RVH was observed in the cases of DORV and TAPVD and in 2 cases of d-TGA + VSD. The other 5 patients with d-TGA + VSD showed biventricular hypertrophy.

Group 3. Cyanosis with decreased PVM. Of the 8 patients in this group, 6 had tricuspid atresia with normally related great vessels (type I) and 2 had pulmonary atresia with VSD. Those with tricuspid atresia had either intact ventricular septum and pulmonary atresia (type Ia), or a small VSD and pulmonary hypoplasia (type Ib). In both, the axis was between -10° and -85° , there was left ventricular dominance, and the Q1 S3 pattern of LAH was present. Of the 2 patients with pulmonary atresia and VSD, 1 had an axis of -70° with LAH and the other had an axis of -120° in the absence of LAH.

Group 4. Shocked with poor peripheral pulses. The 2 patients in the group both had aortic atresia with hypoplastic left heart syndrome.

Discussion

In young infants with heart disease the presence of a superior axis (i.e. dominant S in aVF) is of diagnostic importance. When taken in conjunction with the major clinical and radiological findings, more specific anatomical diagnosis is facilitated before cardiac catheterization and angiography.

In this series most acyanotic infants with heart failure who show pulmonary plethora and cardiomegaly on the chest x-ray have an AVCD or large VSD. Of the 13 with AVCD, 4 had Down's

FIG. 1.—ECG from patient with tricuspid atresia, type Ic, with a large VSD and normally related great vessels. The mean frontal QRS axis is -30° . The 3 mm P wave in lead II indicates right atrial hypertrophy and 70 mm R wave in V_6 indicates left ventricular hypertrophy. ST segment changes reflect digoxin therapy. The combination of a Q wave in lead I and a deep S wave in lead III indicates left anterior hemiblock.

syndrome. The association of this lesion with trisomy 21 is well documented (Rowe and Uchida, 1961), as is the presence of a superior axis (Brink and Neill, 1955; Blount, Balchum, and Gensini, 1956; Milnor and Bertrand, 1957). In our series none of the 6 patients with large VSD had Down's syndrome. Final distinction between AVCD and large VSD could only be made by left ventricular angiography.

In the conditions characterized by cyanosis and plethora on chest x-ray, clinical detection of arterial desaturation may be difficult. As indicated by the pulmonary plethora, total pulmonary blood flow is increased and effective pulmonary flow (the systemic venous return that is oxygenated by passage through the lungs) is high. Under these circumstances arterial desaturation will be slight and may only be proven by measurement of arterial blood gases.

In these patients LVH characterizes both tricuspid atresia with a large VSD and normally related great vessels (type Ic) and tricuspid atresia with TGA and no obstruction to pulmonary blood flow (type IIc). In the latter, the pulmonary artery arises from the left ventricle and in the absence of valvar or subvalvar pulmonary stenosis pulmonary flow exceeds systemic. In DORV and TAPVD, RVH rather than LVH will be found as was also the case in 2 patients with d-TGA+VSD. The remaining 5 patients with superior axis and d-TGA+VSD had biventricular hypertrophy. Despite these and other clinical or radiological features, definitive diagnosis in the group with cyanosis and plethora is made at cardiac catheterization and angiography.

In our series, if the chest x-ray showed reduced PVM in a cyanosed infant with a superior axis,

tricuspid atresia (types Ia or b) was the commonest diagnosis. Tricuspid atresia with normally related great vessels is well established as having a superior axis (Neill and Brink, 1955; Somlyo and Halloran, 1962; Puri and Neill, 1966), though tricuspid atresia with transposed great vessels may have a normal QRS axis (Gamboa, Gersony, and Nadas, 1966).

In addition to hypoplastic left heart syndrome, the clinical features of a shocked infant could be produced by sepsis, haemorrhage, hypothermia, or other noncardiac catastrophies. Other possible intracardiac diagnoses are paroxysmal supra-ventricular tachycardia, aortic stenosis, or severely obstructed (infradiaphragmatic) TAPVD. The final differential diagnosis between severe aortic stenosis and hypoplastic left heart syndrome is only made at cardiac catheterization and angiography.

Pathogenesis of superior axis. According to Wigle and Baron (1966) most investigators agree that activation of the mid-left surface of the ventricular septum dominates the initial phase of septal depolarization and produces a normal axis in the ECG. In a study of epicardial excitation of patients with ostium primum defect, Roos and Durrer (1964) showed that the posterobasal portion of the septum was excited earliest, and that subsequent activation of the anterior and lateral portions of the left ventricle produced the associated superior axis. The anatomical studies of Feldt, DuShane, and Titus (1970) of AVCD (ostium primum can be considered to be an incomplete AVCD) provided a morphological basis for this abnormal excitation sequence. They showed that the entire atrio-ventricular conduction system was displaced

FIG. 2.—Diagrammatic representation of the conducting tissue (hatched) in the normal heart and a specimen with AVCD. The heart has been bisected in its sagittal plane, and the right-hand portion is viewed from the left side. Thus the diagram shows the left side of the atrial and ventricular septa, together with the anterior septum (AIVS) between infundibulum (Inf) and the left ventricle which has been transected. (PA—pulmonary artery; Ao—aorta; LA—left atrium; MV—mitral valve.) In the normal the left bundle descends beneath the aortic valve, and splits some way down the septum into three branches which are usually interconnected. The anterior branches pass towards the anterior papillary muscle (APM) and the posterior towards the posterior muscle (PPM).

In the canal defect, the large septal defect (D) is in three parts, between atrium and ventricle, above and below the atrioventricular valve (AVV), and a segment beneath the aorta. The defect displaces the conducting elements posteriorly so that they ramify as a continuous sheet, mainly over the posterior septum. The position of the conducting tissue is based on personal studies (R. H. Anderson, unpublished observations), together with the works of Feldt et al., 1966, 1970, and Demoulin and Kulbertus (1972).

posteriorly by the defect, and that the left bundle branch was distributed principally to the postero-basal portion of the septum (Fig. 2). These workers had earlier shown similar patterns of origin of the left bundle branch in hearts with isolated VSDs and Fallot's tetralogy exhibiting a superior axis. In these hearts they again concluded that the distribution of the left bundle branch favoured initial activation of the posterior septum (Feldt, DuShane, and Titus, 1966). Few detailed studies of the conduction tissue have been made in the other anomalies presently studied which possessed superior axes. However, Lev (1968) indicated that in tricuspid atresia both left bundle branch and septum were involved in fibrosis, while preliminary studies by one of us suggest that in some cases of d-TGA the left bundle may be deviated posteriorly (R. H. Anderson, unpublished observations). Both of these situations could favour preferential activation of the posterior portion of the ventricular septum.

Further studies of conducting tissue in these conditions are indicated. The exact pathogenesis of the LAH pattern (Q1 S3) seen in some of our cases with superior axis is also not clear. The fact that it was not present in all cases indicates that LAH probably has a different genesis from superior axis, though the two may be related. In 'normal' hearts the presence of LAH has been explained on the basis of trauma to the vulnerable anterior division of the left bundle branch (Rosenbaum, 1969). However, when writing recently, Wigle and Baron (1966) intimated that they were unaware of any study which had shown whether the vital mid-left septal surface was activated by the reported anterior, posterior, or intermediate divisions of the left bundle. The study of the isolated human heart by Durrer *et al.* (1970) showed that there were in fact three early areas of activation in the left ventricle, one being mid-septal, and this finding correlates with the histopathological study of Demoulin and Kulbertus (1972), emphasizing the importance of the septal division of the left bundle branch. In the histopathological studies of superior axis already referred to (Feldt *et al.*, 1966, 1970), the left bundle is shown to be a continuous sheet of fibres, but a paucity of fibres arising from its anterior portion has been noted. It may well be that the presence of LAH pattern is dependent upon the degree of paucity of these fibres, in turn deciding the degree of activation of the anterior septum relative to the predominant activation of the posterior septum. Additional histopathological studies will be required to elucidate this problem.

In conclusion, we state that while identification of

superior axis facilitates differential diagnosis of congenital heart disease, exact diagnosis is dependent upon cardiac catheterization.

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REFERENCES

- Blount, S. G., Balchum, O. J., and Gensini, G. (1956). The persistent ostium primum atrial septal defect. *Circulation*, **13**, 499.
- Brink, A. J., and Neill, C. A. (1955). The electrocardiogram in congenital heart disease; with special reference to left axis deviation. *Circulation*, **12**, 604.
- Criteria Committee of the New York Heart Association (1964). *Diseases of the Heart and Blood Vessels: Nomenclature and Criteria for Diagnosis*, 6th ed. Little, Brown, Boston.
- Demoulin, J. C., and Kulbertus, H. E. (1972). Histopathological examination of concept of left hemiblock. *British Heart Journal*, **34**, 807.
- Durrer, D., van Dam, R. T., Freud, G. E., Janse, M. J., Meijler, F. L., and Arzbaecher, R. C. (1970). Total excitation of the isolated human heart. *Circulation*, **41**, 899.
- Feldt, R. H., DuShane, J. W., and Titus, J. L. (1966). The anatomy of the atrioventricular conduction system in ventricular septal defect and tetralogy of Fallot; correlations with the electrocardiogram and vectorcardiogram. *Circulation*, **34**, 774.
- Feldt, R. H., DuShane, J. W., and Titus, J. L. (1970). The atrioventricular conduction system in persistent common atrioventricular canal defect. Correlations with electrocardiogram. *Circulation*, **42**, 437.
- Gamboa, R., Gersony, W. M., and Nadas, A. S. (1966). The electrocardiogram in tricuspid atresia and pulmonary atresia with intact ventricular septum. *Circulation*, **34**, 24.
- Keith, J. D., Rowe, R. D., and Vlad, P. (1958). Tricuspid atresia. In *Heart Disease in Infancy and Childhood*, p. 437. Macmillan, New York.
- Lev, M. (1968). Conduction system in congenital heart disease. *American Journal of Cardiology*, **21**, 619.
- Liebmman, J., and Nadas, A. S. (1971). An abnormally superior vector. (Formerly called marked left axis deviation.) *American Journal of Cardiology*, **27**, 577.
- Milnor, W. R., and Bertrand, C. A. (1957). The electrocardiogram in atrial septal defect. A study of 24 cases with observations on the RSR'-V₁ pattern. *American Journal of Medicine*, **22**, 223.
- Neill, C. A., and Brink, A. J. (1955). Left axis deviation in tricuspid atresia and single ventricle; the electrocardiogram in 36 autopsied cases. *Circulation*, **12**, 612.
- Puri, P. S., and Neill, C. A. (1966). Vectorcardiographic study in 10 cases of tricuspid atresia. In *Electrocardiography in Infants and Children*, p. 268. Ed. by D. E. Cassels and R. F. Ziegler. Grune and Stratton, New York; Heinemann, London.
- Roos, J. P., and Durrer, D. (1964). Epicardial excitation in ventral atrial septal defect. *British Heart Journal*, **26**, 136.
- Rosenbaum, M. B. (1969). Types of left bundle branch block and their clinical significance. *Journal of Electrocardiology*, **2**, 197.
- Rowe, R. D., and Uchida, I. A. (1961). Cardiac malformations in mongolism. A prospective study of 184 mongoloid children. *American Journal of Medicine*, **31**, 726.
- Somlyo, A. P., and Halloran, K. H. (1962). Tricuspid atresia: an electrocardiographic study. *American Heart Journal*, **63**, 171.
- Wigle, E. D., and Baron, R. H. (1966). The electrocardiogram in muscular subaortic stenosis. *Circulation*, **34**, 585.
- Ziegler, R. F. (1966). Introduction and clinical application. In *Electrocardiography in Infants and Children*, p. 1. Ed. by D. E. Cassels and R. F. Ziegler. Grune and Stratton, New York; Heinemann, London.

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