THE INCIDENCE AND SIGNIFICANCE OF SUPERIOR QRS AXIS IN CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE

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SYNOPSIS

The incidence of superior QRS axis in 200 consecutive infants and children with cyanotic congenital heart disease who underwent cardiac catheterization is 26%. Patients with asplenia syndrome, transposition of great arteries, double outlet right ventricle and dextrocardia complex are most likely to have superior QRS axis. The frequency of superior QRS axis in patients with tetralogy of Fallot is 13%, a finding that has not been previously high-lighted. Transposition of great arteries accounts for more than a quarter, and tetralogy of Fallot about one-fifth, of all the cases of cyanotic congenital heart disease with superior QRS axis. Tricuspid atresia is extremely rare, accounting for only 2% in our series. Further classification of these cases according to the mean frontal QRS axis and the roentgenologic patterns provides further diagnostic informations. Definitive diagnosis of cyanotic congenital heart disease requires, however, cardiac catheterization and angiography.

INTRODUCTION

The electrocardiogram (ECG) provides important diagnostic information pertaining to cyanotic congenital heart disease (CHD) (1).Cyanotic CHD with a superior QRS axis (between -1° and -180°) in the ECG is not uncommon (2) and forms a diagnostically challenging group. The term "superior axis" (Liebman and Nadas 1971) (3) is preferable to the old terminology of "let axis deviation" (4), since it accurately describes the mean QRS vector as being above the horizontal line, represented by lead I, in the frontal plane. Goldman (5) also believes that a "marked left axis deviation is in actual fact an abnormally superior vector". Moreover, an axis between -90° and -180° may actually be extreme right axis deviation rather that left axis deviation.

Since the superior QRS axis is readily recognised by a dominantly negative deflection (S wave) in lead aVF, a practical diagnostic approach to this group of children with cyanotic CHD would be useful to neonatologists and general paediatricians, who usually first see these children before they are referred to the paediatric cardiologist.

The purpose of the present study is to assess the incidence and to consider the diagnostic implications of superior QRS axis in children with cyanotic CHD, with a view to formulate practical diagnostic approach, so that neonatologists and paediatricians can have a good idea of the likely anatomical diagnosis before invasive investigation by cardiac catheterization and angiography.

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MATERIALS AND METHODS

Two hundred consecutive children with cyanotic CHD who were subjected to cardiac catheterization from January 1976 to end of September 1982 were studied. The full medical history, complete physical examination, chest roentgenogram, 12 leads ECG, full blood count including haemoglobin and haematocrit, peripheral blood smear to look for Howell-Jolly bodies and arterial blood gas study of each patient were carried out before cardiac catheterization which was performed by either one of us. Since 1981 two dimensional echocardiography was also performed before cardiac catheterization. The criteria for diagnosis of cyanosis in CHD in general and the actual practical diagnostic approach in these 200 cases in particular have been published elsewhere (6). In general, all patients with cyanotic CHD were catheterized with the exception of a very few who perished before catheterization. These were excluded from analysis as the underlying cardiac diagnosis was not accurately documented.

The 12 leads ECG were carefully analysed with special reference to determination of mean frontal plane QRS axis and hypertrophy pattern. Those children with superior QRS axis will be further analysed, especially in relation to roentgenologic pattern. The superior QRS axis was defined by the presence of a dominantly negative deflection (S wave) in lead aVF. The mean frontal plane QRS axis was calculated, using a Casio fx-310 scientific calculator from the vectors in leads I and aVF, using the computational formulae previously published (7) (Table 1).

RESULTS

Table 2 shows the age, sex and ethnic distributions of 200 consecutive children with cyanotic CHD who underwent cardiac catheterization from January 1976 to September 1982. Their age ranged from 4 days to 14 years at the time of cardiac catheterization. Note the 30% of the cases were catheterized in the first year of life, with the remaining number of cases fairly evenly distributed in the toddler, preschool and schooling age groups. The male to female ratio is 1.4 to 1. The ethnic distribution roughly conforms to that of the hospitalised children with slight over-representation of the Chinese children.

Table 3 shows the major diagnostic categories of the 200 children with cyanotic CHD. Tetralogy of Fallot constitutes more than one-third and together with pulmonary atresia, they comprise more than one-half of the 200 cases. Other relatively common conditions include transposition of great arteries (12%), pulmonary stenosis with right to left shunting at the atrial and/or ventricular level (8%), dextrocardia complex (6%), asplenia syndrome (4%), double outlet right ventricle (3%) and total anomalous pulmonary venous drainage (3%).

Table 4 shows the incidence of superior QRS axis in major diagnostic categories of cyanotic CHD in descending frequency. The incidence of superior QRS axis in each diagnostic category is also compared with that in tetralogy of Fallot by Chi-square test or Fisher's exact test.

Table 5 shows the frequency of individual category of cyanotic CHD with superior QRS axis. Note that although the incidence of superior QRS axis in children with tetralogy of Fallot is the lowest (12.8%, Table 4), it is the second most common amongst all cyanotic children with superior QRS axis.

Table 6 shows the classification of cyanotic CHD with superior QRS axis according to the mean frontal QRS axis. Less than 10% of the cases have the mean frontal plane QRS axis situated between -1° to -44°. The only case of tricuspid atresia with superior QRS axis has a mean frontal plane axis of -19°. Note that all the cases of tetralogy of Fallot with superior QRS axis have the mean frontal plane axis situated between -136° to -180°. All the 3 cases of double outlet right ventricle with superior QRS axis have the mean frontal plane QRS axis situated between -45° to -135°. Transposition of great arteries, on the other hand, are equally distributed in these two quadrants.

Table 7 shows the clinical and roentgenologic classification of all cases of caynotic CHD with superior QRS axis. Asplenia syndrome, dextroxardia complex and laevocardia with situs inversus can be easily delineated by clinical and roentgenologic examination and peripheral blood smear showing the presence of Howell-Jolly bodies. Further subclassification according to pulmonary vascular mark-

TABLE 1
COMPUTATION OF MEAN FRONTAL PLANE QRS AXIS FROM VECTORS OF LEAD I AND LEAF AVF

If the QRS vector(s) in lead(s) —

(a) I and aVF are both positive : $\theta = \tan^{-1}\left(\frac{aVF}{I}\right)$

(b) I and aVF are both negative : $\theta = -180 + \tan^{-1}\left(\frac{aVF}{I}\right)$

(c) I only is negative : $\theta = 180 - \tan^{-1}\left(\frac{aVF}{I}\right)$

(d) aVF only is negative : $\theta = -\tan^{-1}\left(\frac{aVF}{I}\right)$

where θ is the mean frontal plane QRS axis measured in degrees and I and aVF represent the QRS vectors in leads I and aVF respectively.

Ref: Tay JSH, Yip WCL: Polar equations for the QRS factors in the frontal plane. J Singapore Paediatr Soc 1981; 23:139-141.

TABLE 2
AGE, SEX AND ETHNIC DISTRIBUTIONS OF ALL CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE WHO UNDERWENT CARDIAC CATHETERIZATION FROM JANUARY 1976 TO SEPTEMBER 1982

AGE (AT CATHETERIZATION)		SEX		ETHNIC GROUP	
< 1 wk	2 (1.0)	Male	115 (57.5)	Chinese	166 (83.0)
1 wk — 1 mo	15 (7.5)	Female	85 (42.4)	Malay	23 (11.5)
1 mo — 1 yr	55 (27.5)			Indian	8 (4.0)
1 yr — 3 yr	39 (19.5)			Others	3 (1.5)
3 yr — 6 yr	41 (20.5)				
6 yr — 12 yr	39 (19.5)				
> 12 yr	9 (4.5)				
TOTAL	200 (100.0)		200 (100.0)		200 (100.0)

Figures in parenthesis indicate percentages.

TABLE 3
ANATOMICAL DIAGNOSIS OF 200 CONSECUTIVE CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE AT CARDIAC CATHETERIZATION

	NO	%
Tetralogy of Fallot	78	39.0
Pulmonary atresia	28	14.0
Transposition of great arteries*	24	12.0
Pulmonary stenosis with VSD and/or ASD	16	8.0
Dextrocardia complex	12	6.0
Asplenia syndrome	8	4.0
Double outlet right ventricle	6	3.0
Total anomalous pulmonary venous drainage	6	3.0
Eisenmenger's syndrome	5	2.5
Ebstein's anomaly	4	2.0
Laevocardia with situs inversus	3	1.5
Endocardial cushion defect	3	1.5
Coarctation syndrome	2	1.0
Persistent truncus arteriosus	2	1.0
Tricuspid atresia	2	1.0
Double outlet left ventricle	1	0.5
TOTAL	200	100.0

*Five out of 24 had single ventricle.
ASD = Atrial septal defect
VSD = Ventricular septal defect

TABLE 4
INCIDENCE OF SUPERIOR QRS AXIS IN MAJOR DIAGNOSTIC CATEGORIES OF CYANOTIC CONGENITAL HEART DISEASE

DIAGNOSIS	No.	%	*Significance of difference
Asplenia syndrome	7/8	87.5	$p < 0.0001$
Transposition of great arteries†	14/24	58.3	$p < 0.0001$
Double outlet right ventricle	3/6	50.0	$p < 0.05$
Dextrocardia complex	5/12	41.7	$p < 0.05$
Others#	6/28	21.4	NS
Pulmonary stenosis with VSD and/or ASD	3/16	18.8	NS
Pulmonary atresia"	4/28	14.3	NS
Tetralogy of Fallot	10/78	12.8	—
Total	52/200	26.0	

* Incidence of superior QRS axis in each diagnostic category compared with that in tetralogy of Fallot by Chi-square test or Fisher's exact test

† Two out of 14 had associated single ventricle. The other 12 had associated VSD.

Include 2 cases of endocardial cushion defect, 2 cases of laevo-cardia with situs inversus and 1 case each of tricuspid atresia and Ebstein's anomaly

" Two out of 4 had asssociated VSD, 1 had endocardial cushion defect and 1 had ASD

ASD = Atrial Septal Defect NS = Not Significant

VSD = Ventricular Septal Defect

TABLE 5
FREQUENCY OF INDIVIDUAL CATEGORY OF CYANOTIC CONGENITAL HEART DISEASE WITH SUPERIOR QRS AXIS

	NO.	%
Transposition of great arteries	14	26.9
Tetralogy of Fallot	10	19.2
Asplenia syndrome	7	13.5
Dextrocardia complex	5	9.6
Pulmonary atresia	4	7.7
Double outlet right ventricle	3	5.8
Pulmonary stenosis + VSD and/or ASD	3	5.8
Others*	6	11.5
TOTAL	52	100.0

*Include 2 cases of endocardial cushion defect, 2 cases of laevocardia with situs inversus and 1 case each of tricuspid atresia and Ebstein's anomaly

ASD = Atrial Septal Defect

VSD = Ventricular Septal Defect

TABLE 6
CLASSIFICATION OF CYANOTIC CONGENITAL HEART DISEASE WITH SUPERIOR QRS AXIS ACCORDING TO THE MEAN FRONTAL AXIS

-136° to -180°		-45° to -135°		-1° to -44°	
TOF	10	TGA	7(1)	TGA	1(1)
TGA	6	ASP	4	TA	1
PA	3	DC	4	PS + ASD	1
ASP	3	DORV	3	PS + ASD + VSD	1
DC	1	ECD	2		
PS + ASD	1	PA	1		
LC + SIV	1	LC + SIV	1		
		EA	1		
TOTAL	25	TOTAL	23	TOTAL	4

ASD = Atrial Septal Defect, ASP = Splenia Syndrome, DC = Dextrocardia, DORV = Double Outlet Right Ventricle, EA = Ebstein's Anomaly, ECD = Endocardial Cushion Defect, LC = Laevocardia, PA = Pulmonary Atresia, PS = Pulmonary Stenosis, SIV = Situ Inversus, TA = Tricuspid Atresia, TGA = Transposition of Great Arteries, TOF = Tetralogy of Fallot

Figure in parenthesis indicates number with associated single ventricle.

TABLE 7
CLINICAL AND ROENTGENOLOGIC CLASSIFICATION OF CYANOTIC CONGENITAL HEART DISEASE WITH SUPERIOR QRS AXIS

CLINICALLY DELINEABLE SUBGROUPS		ROENTGENOLOGIC SUBGROUPS		
ASPLENIA SYNDROME	7	WITH PULMONARY PLETHORA		16
DEXTROCARDIA COMPLEX	5	TRANSPOSITION OF GREAT ARTERIES	12	
LAEVOCARDIA WITH SITUS INVERSUS	2	ENDOCARDIAL CUSHION DEFECT	2	
		DOUBLE OUTLET RIGHT VENTRICLE	2	
		WITH PULMONARY OLIGAEMIA		22
		TETRALOGY OF FALLOT	10	
		PA	4	
		PS + VSD AND/OR ASD	3	
		TRANSPOSITION OF GREAT ARTERIES + PS OR PA	2	
		EBSTEIN'S ANOMALY	1	
		TRICUSPID ATRESIA + PS	1	
		DOUBLE OUTLET RIGHT VENTRICLE + PS	1	
TOTAL	14	TOTAL		38

ASD = ATRIAL SEPTAL DEFECT; PA = PULMONARY ATRESIA; PS = PULMONARY STENOSIS; VSD = VENTRICULAR SEPTAL DEFECT

ings helps to diagnostically separate further the remaining cases of cyanotic CHD.

Table 8 shows the comparison of the present series with that of Shinebourne et al's cardiac patients with superior QRS axis. Note that the incidence of superior QRS axis is more than twice higher but tricuspid atresia is extremely rare in our series. The frequency of transposition of great arteries with superior QRS axis in the two series is identical, but tetralogy of Fallot is not present in Shinebourne's series.

QRS axis less than 0°, the presence of superior QRS axis in an infant or a child should always alert the presence of significant CHD.

The incidence of superior QRS axis in children with CHD in different series is difficult to compare, as different workers used different criteria in their classification of the mean QRS axis. Shinebourne et al (2), using the same criteria as ours, found that 10% of their patients with cyanotic as well as acyanotic CHD in the first 3 months of life had superior QRS axis, an

TABLE 8
COMPARISON OF TWO SERIES OF CHILDREN WITH SUPERIOR QRS AXIS

	SHINEBOURNE ET AL*	YIP ET AL
Incidence of Superior QRS Axis	47/473 (10%)	52/200 (26%)
Age of Patients	First 3 months	4 days to 14 years
Type of CHD	Cyanotic and acyanotic	Cyanotic
Two most common cyanotic CHD with superior QRS axis	Tricuspid atresia 13/26 (50%)	Transposition of great arteries 14/52 (27%)
	Transposition of great arteries 7/26 (27%)	Tetralogy of Fallot 10/52 (19%)
Incidence of tricuspid atresia	13/26 (50%)	1/52 (2%)

* Shinebourne EA, Haworth SG, Anderson RH, Ulgur A: Differential diagnosis of congenital heart disease in the first 3 months of life. Significance of a superior (left) ARS axis. Arch Dis Child 1974; 49:729-733.

CHD = Congenital Heart Disease

DISCUSSION

The classification of electrical QRS axis has not been standardised. The Criteria Committee of the New York Heart Association (8) first adopted three simple descriptive groups, viz. no deviation (+30° to +90°), left axis deviation (+29° to -90°) and right axis deviation (+91° to -91°). This classification has been adopted by Brink and Neill to study the electrocardiograms of children with CHD with special reference to left axis deviation (4). Liebman and Nadas (3), however, maintained that standard definitions divided the mean frontal QRS vector in three groups: normal axis (0° to +90°), left axis deviation (0° to -90°) and right axis deviation (+90° to +180°). This classification leaves the quadrant between -90° to -180° unspecified, which is really indeterminate as the axis could either be left or extreme right. As previously observed by Goldman (5), "a marked left axis deviation" is in fact an abnormally superior vector, the term superior axis is substituted for left axis deviation by Liebman and Nadas. (3). Shinebourne et al (2) have thus defined superior axis as one with the mean frontal plane QRS axis between -1° and -180° in their study of differential diagnosis of congenital heart disease in the first 3 months of life.

In the present study, we have adopted the same definition of superior QRS axis as by Shinebourne et al. Since Maroney and Rantz (9) found that, in their series of 679 healthy infants and children, ranging in age from 6 months to 9 years, only three had mean

incidence significantly lower that the 26% of our series. Tricuspid atresia, a CHD well recognised to be associated with left axis deviation (4, 10, 11) which, however, is not present in all the cases (12, 13), accounts for 13 out of 26 cyanotic patients with superior QRS axis (50%) in Shinebourne et al's series. It is however, distinctly rare in our series, accounting for only 2% of all our cyanotic patients with superior QRS axis. This is largely due to the small number of cases of tricuspid atresia in our series.

Transposition of great arteries, all associated with either a ventricular septal defect or single ventricle, and tetralogy of Fallot account for nearly half of all our cases with superior QRS axis. It is interesting to note that not a single case of tetralogy of Fallot was found in Shinebourne et al's series (2), as they are usually studied after the first three months of life. On the other hand, Brink and Neill (4) found that 3.89% of their patients with tetralogy of Fallot had a mean QRS axis situated between -91° to -150°, i.e. superior axis by our definition. Like in all our cases, this superior QRS axis is attributed to extreme right rather than left axis deviation.

In terms of probability, asplenia syndrome, transposition of great arteries and double outlet right ventricle have more than 1 in 2 chance of having a superior QRS axis in our series. Brink and Neill (4) noted that 19.7% of their cases with transposition of great arteries had a mean frontal plane QRS axis situated between -91° to -150°. Other workers, however, observed that an abnormally superior vector

occurs only very occasionally and only when a ventricular septal defect is present (14, 15, 16). All of our cases of transposition of great arteries are associated with either a ventricular septal defect or sinlge ventricle. Double outlet right ventricle was not mentioned by Brink and Neill in their 289 cases with left axis deviation (4), while in Shinebourne et al's series, only 1 case of double outlet right ventricle with superior QRS was noted. Other workers, however, had noted that abnormal superior vector did occur in patients with double outlet right ventricle (17, 18) and the cause was found to be related to conduction defect as a result of the abnormally posteriorly and inferiorly placed left conduction bundle (19).It is now generally accepted that the cause of the so called "left axis deviation" is not due to left ventricular hypertrophy (3).On the other hand, conduction defect has been shown to be the most probable cause in endocardial cushion defect (20, 21, 22), tricuspid atresia (23), double outlet right ventricle (19), transposition of great arteries (2), and even isolated ventricular septal defect (24, 25) and tetralogy of Fallot (25).Superior QRS axis in asplenia syndrome has never been documented previously. Although its clinical usefulness is limited in this condition as the anatomical diagnosis can be easily suspected on clinical, roentgenologic and haematological ground, further studies on the conduction system in these cases may shed more light on the pasthogenesis of superior QRS axis in children with congenital heart disease.

Further classification of cyanotic children with superior QRS axis according to the mean frontal axis and roentgenologic patterns will provide more diagnostic informations. For example, all the cases of tetralogy of Fallot have the mean QRS axis situated between -136° to -180° and the most likely diagnosis in a cyanotic child with the mean QRS axis situated between -45° to -135° who has an oligaemic lung fields will be transposition of great arteries with severe pulmonary stenosis or pulmonary atresia. However, it should be emphasized that while identification of superior axis greatly facilitates differential diagnosis of cyanotic CHD, exact diagnosis is still dependent on cardiac catheterization and angiography.

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