A PRACTICAL DIAGNOSTIC APPROACH TO CYANOTIC CONGENITAL HEART DISEASE

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SYNOPSIS

Cyanotic congenital heart disease is a diagnostic challenge to many general paediatricians. This paper discusses how a working diagnosis of the underlying anatomical defects in a patient with cyanotic congenital heart diseases can be arrived at by careful clinical evaluation, supplemented by simply laboratory investigations. The major clinical, radiological and electrocardiographic findings together with the definitive anatomical diagnoses of two hundred consecutive children with cyanotic congenital heart diesease who underwent cardiac catheterization from 1976 to 1982 were analysed. After exclusion of 6 clinically delineable subgroups (34 cases), viz. dextrocardia complex, laevocardia with situs inversus, asplenia syndrome, coarctation syndrome, Ebstein's anomaly and Eisenmenger's syndrome, the remaining 166 cases were classified according to a combined electrocardiographic (by frontal plane QRS axis and ventricular hypertrophy pattern) and roentgenographic (by pulmonary vascularity) scheme. The diagnosis of more than 90% of these 200 cases can be accounted for by this practical diagnostic approach.

INTRODUCTION

Congential heart disease (CHD) with its incidence around 6 to 8 per 1000 live births(1) is the single most important major congenital abnormality. It is responsible for much mortality and morbidity in infants and children today. From the analysis of the mortality record of the University Department of Paediatrics, Singapore General Hospital, it is gratifying to note a steady decline in the case fatality rate, defined as the total number of dealth per 100 hospitalised children aged below 12 years in the past 15 years (Table 1) (2). This is partly due to better general health status of our children in recent years as reflected by the declining infant mortality rate (3) and partly due to improved standard of preventive as well as therapeutic and intensive paediatric care. However, when the major causes of death in hospitalised children in the department are examined more closely (Table 2) (2), it is revealing that CHD has becomes the top killer in the last five years. Among these children who succumbed, a great majority died from the more severe, cyanotic form of the heart defect.

Because of the overwhelming importance of this problem, the Department has set up a cardiac clinic registry and a cardiac catheterization registry for proper documentation and to facilitate on going research. From the year 1974 to end of September 1982, there were a total of 1158 cases under the cardiac registry, and of these about half were subjected to cardiac catheterization (Table 3).

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TABLE 1

TOTAL MORTALITY IN HOSPITALISED CHILDREN (0-12 YEARS) FROM 1966 TO 1980 IN THE UNIVERSITY DEPARTMENT OF PAEDIATRICS SINGAPORE GENERAL HOSPITAL

YEAR	TOTAL ADMISSION	TOTAL DEATH	CASE FATALITY RATE (%)
1966	6873	273	3.972
1967	6586	244	3.705
1968	6308	216	3.424
1969	6524	179	2.744
1970	6356	169	2.659
1971	6380	158	2.477
1972	6200	144	2.323
1973	6637	178	2.682
1974	4876	144	2.953
1975	3820	86	2.251
1976	4787	85	1.776
1977	4318	73	1.691
1978	4375	59	1.349
1979	5484	70	1.277
1980	5124	44	0.858
TOTAL	84648	2122	2.507

REF: YIP WCL, TAY JSH, TAN NC: CONGENITAL HEART DISEASE IN SINGAPORE — PRESENT PROBLEMS AND FUTURE PERSPECTIVES. SINGAPORE MED J 1982; 22: 133-139.

TABLE 2

MAJOR CAUSES OF DEATH IN HOSPITALISED CHILDREN (0-12 YEARS) FROM 1966 TO 1980 IN THE UNIVERSITY DEPARTMENT OF PAEDIATRICS SINGAPORE GENERAL HOSPTIAL

YEAR	MAJOR CAUSES OF DEATH					TOTAL DEATH
	NO. 1	NO. 2	NO. 3	NO. 4	NO. 5	
1966-1970	BRONCHO PNEUMONIA	CHD*	GASTRO-ENTERITIS	MALIGNANCY	KERNICTERUS	1081
	288 (27%)	164 (15%)	90 (8%)	62 (6%)	45 (4%)	(100%)
1971-1975	BRONCHO PNEUMONIA	CHD*	GASTRO-ENTERITIS	MALIGNANCY	SEPTICAEMIA	710
	127 (18%)	109 (15%)	63 (9%)	43 (6%)	41 (6%)	(100%)
1976-1980	CHD*	MALIGNANCY	BRONCHO PNEUMONIA	SEPTICAEMIA	GASTRO-ENTERITIS	331
	72 (22%)	56 (17%)	53 (16%)	16 (5%)	7 (2%)	(100%)

* CHD = CONGENITAL HEART DISEASE

REF: YIP WCL, TAY JSH, TAN NC: CONGENITAL HEART DISEASE IN SINGAPORE — PRESENT PROBLEMS AND FUTURE PERSPECTIVES. SINGAPORE MED J 1982; 22: 133-139.

Since 1976, full cardiac catheterization records were available for analysis. A survey of the type of heart disease at catheterization (Table 4) reveals that 97% of the total 434 cases from January 1976 to end of September 1982 were due to CHD. There were about equal numbers of cyanotic as well as acyanotic form of CHD. In recent years, significant rheumatic heart disease has become so rare in Singapore children that we only catheterize about one patient per year.

There are many problems facing a child, and his family, who suffers form the severe cyanotic form of CHD, e.g. diagnosis, hyperviscosity syndrome, relative anaemia, haemorrhagic diasthesis, hyperuricaemia (4), timing of surgery, pre- and post-operative prognosis (5,6) and psychological trauma. Cyanotic CHD is often considered difficult and tanglesome, as far as diagnosis of the anatomical defect is concerned, by general paediatricians. The purpose of this study is to focus on clinical diagnosis of cyanotic CHD and to disucss how a working diagnosis of the underlying anatomical defects can be arrived at by clinical evaluation, supplemented by simple laboratory investigations.

TABLE 3

DEPARTMENT CARDIAC CLINIC REGISTRY AND CARDIAC CATHETERIZATION REGISTRY

YEAR	TOTAL NO. REGISTERED	TOTAL NO. CATHETERIZED
1974	210	30
1975	110	41
*1976	68	40
1977	102	63
1978	96	80
1979	131	69
1980	109	40
1981	160	79
†1982	172	63
TOTAL	1158	505

* FULL CARDIAC CATHETERIZATION RECORD AVAILABLE SINCE JANUARY 1976

† REGISTRY UP TO END OF SEPTEMBER 1982

TABLE 4

TYPES OF HEART DISEASE AT CARDIAC CATHETERIZATION

YEAR	CHD†		RHD=	OTHERS	TOTAL
	CYANOTIC	ACYANOTIC			
1976	19	20	0	1[a]	40
1977	31	32	0	0	63
1978	38	40	2	0	80
1979	39	28	2	0	69
1980	17	21	1	1[b]	40
1981	36	39	3	1[c]	79
*1982	20	41	1	1[d]	63
TOTAL	200	221	9	4	434

* REGISTRY UP TO END OF SEPTEMBER 1982
† CHD = CONGENITAL HEART DISEASE
= RHD = RHEUMATIC HEART DISEASE
a CHRONIC CONSTRICTIVE PERICARDITIS
b CONGESTIVE CARDIOMYOPATHY
c MYOTONIC DYSTROPHY WITH PULMONARY HYPERTENSION
d HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY

TABLE 5

AGE, SEX AND ETHNIC DISTRIBUTION OF ALL CHILDREN WITH CYANOTIC 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 (42.5)	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.

MATERIALS AND RESULTS

The cardiac catheterization reports of the 200 children with cyanotic CHD were reviewed and their major clinical, radiological and electrocardiographic findings together with the difinite anatomical diagnoses were tabulated.

Table 5 shows the age, sex and ethnic distribution of the 200 cyanotic CHD who underwent cardiac catherterization from January 1976 to September 1982. Thirty % of the cases were catheterized in the first year of life with the remaining number of cases fairly evenly spread out in the toddler, preschool and schooling age groups. The male to female ratio is 1.4:1. The ethnic distribution roughly conforms to that of the hospitalised childred with slight overrepresentation of the Chinese children.

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

DIAGNOSIS OF CYANOSIS IN A CHILD WITH CHD

Recognition of cyanotic CHD is easy when there are obvious central cyanosis and clubbing with concomittant cardiac signs (Table 7). However, diagnostic difficulty does arise not infrequently. Both false negative and false positive states (Table 8) have been encountered. Mild cyanosis is frequently missed especially in darkly pigmented races. On the other hand, peripheral cyanosis due to cold with a soft innocent murmur may lead the unwary to diagnose cyanotic CHD. Anaemia, commonly due to iron deficiency, is well known in masking cyanosis as there is inadequate reduced haemoglobin for clinical recognition. On the other hand, central cyanosis may be due to hypoxaemia as a result of severe cardiac failure in patients with large left to right shunting or due to lung infection rather than due to right to left shunting as in the various forms of cyanotic CHD. Here vigorous treatment of cardiac failure or lung infection and hyperoxia test wil be useful. Other helpful criteria for cyanosis in a child with CHD are haemoglobin concentration higher than normal for age and low arterial oxygen saturation.

CYANOTIC CHD: THE DIAGNOSTIC LADDER

It must be stressed from the very beginning that the diagnostic work up of cyanotic CHD must be approached in a systematic manner (Table 9). Like any diagnostic problem in medicine, the analysis of cyanotic CHD must commence from careful clinical evaluation. Further diagnostic investigations by more sophisticated techniques, either invasive or non invasive, should be planned carefully, depending on the availability of such facilities and expertise, and only after simple laboratory investigations such as chest roentgenogram and electrocardiogram have been carried out.

In the clinical evaluation (Table 10), a full medical history is of utmost importance. Special emphasis must be given to the onset of cyanosis, the presence of hypoxic spells, the degree of exercise intolerance, the history of squatting, frequent chest infections and cardiac failure. A thorough physical examination often yields sufficient informations to enable delineation of certain subgroups of cyanotic CHD. For example, the presence of brachiofemoral pulse delay and of a large systolic pressure gradient between the brachial and popliteal blood pressures easily singles out coarctation syndrome from the rest of the cyanotic CHD. Again determination of cardio-viscaral situs will enable one to identify the dextrocardia complex and the syndrome of laevocardia with situs inversus.

The simple laboratory investigations which are of great diagnostic importance are listed in Table 11. In the reading of the chest roentgenogram, particular attention should be paid to the pulmonary vascularity, cardio-visceral situs and cardiothoracic ratio. Certain radioloical peculiarities are of diagnostic significance, although their absence does not negate the respective diagnosis if the other clinical, electrocardiographic and radiological features are consistent. Classical examples are the boot-shaped heart of tetralogy of Fallot (Fig. 1), the egg-shaped heart of transposition of great arteries (Fig. 2) and the snow-man appearance of total anomalous pulmonary venous drainage (Fig. 3). In the analysis of electrocardiogram as part of the diagnostic work up of cyanotic CHD, the mean frontal plane QRS axis and the ventricular hypertrophy patterns are the most important. Certain other electrocardiographic peculiarities are also helpful. For example, giant P waves (Fig. 4) in the right praecordial leads alert the possibility of Ebstein's anomaly. Similarly P tricuspidale (Fig. 5), if present, suggests the possibility of tricuspid atresia as the cause of the cyanotic CHD. Finally the presence of Howell Jolly bodies in the peripheral blood smear singles out asplenia syndrome from the rest of cyanotic CHD.

EXCLUSION OF CLINICALLY DELINEABLE DIAGNOSITC SUBGROUPS OF CYANOTIC CONGENITAL HEART DISEASE

There are 6 cathegories of cyanotic CHD among the 200 cases which can be easily delineated from the rest by the simple practical approach outlined above (Table 12). Dextrocardia complex (Fig. 6) and the syndrome of laevocardia with situs inversus (Fig. 7) can be identified from a careful examination of cardio-visceral situs. The cardiovascular malformations in these two groups of patients are listed in Table 13 and Table 14 respectively. The asplenia syndrome has one pathognomonic haematological feature and that is the presence of Howell-Jolly bodies in the peripheral blood smear. Their underlying cardiovascular malformations (listed in Table 15) are very complex. Coarctation syndrome may or may not be cyanotic. However, this should be promptly identified if the examination of femoral pulses and brachial and popliteal blood pressure measurements are part of the routine in paediatric cardiac examination. Both of our patients had double intracardiac shunts and pulmonary hypertension and one patient had in addition patent ductus arteriosus. The Eisenmenger's syndrome ususally reveals itself by a relatively long history with late onset cyanosis which is initially mild, by booming pulmonary second heart sound, prominent pulmonary conus and peripheral vascular prunning in the chest roentgenogram (Fig. 8) and by electrocardiographic evidence of right ventricular hypertrophy or biventricular hypertrophy. Finally Ebstein's anomaly can be easily identified by the presence of large and relatively silent heart, huge right atrium and oligaemic lung fields in the chest roentgenogram (Fig.9) and the characteristic giant P waves in the electorcardiogram.

DIFFERENTIAL DIAGNOSIS OF CYANOTIC CONGENITAL HEART DISEASE BY ELECTROCARDIOGRAM AND CHEST ROENTGENOGRAM

Having excluded the 6 clinically delineable subgroups, the remaining cases of the cyanotic CHD can be further differentiated by careful analysis of the electrocardiogram and roentgenogram. A practical diagnostic scheme of

TABLE 6

MAJOR DIAGNOSTIC CATEGORIES OF CYANOTIC CONGENITAL HEAT DISEASE

	NO.	%
TETRALOGY OF FALLOT	78	39.0
PULMONARY ATRESIA	28	14.0
TRANSPOSITION OF GREAT ARTERIES	24	12.0
PULMONARY STENOSIS WITH VSD &/OR ASD	16	8.0
DEXTROCARDIA COMPLEX	12	6.0
ASPLENIA SYNDROME	8	4.0
TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE	6	3.0
DOUBLE OUTLET RIGHT VENTRICLE	6	3.0
EISENMENGER SYNDROME	5	2.5
EBSTEIN ANOMLY	4	2.0
LAEVOCARDIA WITH SITUS INVERSUS	3	1.5
ENDOCARDIAL CUSHION DEFECT	3	1.5
CAORCTATION SYNDROME	2	1.0
PERSISTENT TRUNCUS ARTERIOSUS	2	1.0
TRICUSPID ATRESIA	2	1.0
DOUBLE OUTLET LEFT VENTRICLE	1	0.5
	200	100.0

TABLE 7

CRITERIA FOR CYANOSIS IN A CHILD WITH CONGENITAL HEART DISEASE

1. Obvious central cyanosis and clubbing clinically
2. Haemoglobin concentration higher than normal for age
3. Low arterial oxygen saturation (< 93%)

TABLE 8

CYANOSIS: DIAGNOSTIC DIFFICULTY

A. FALSE NEGATIVE
   1. Mild Cyanosis
   2. Skin Pigmentation
   3. Anaemia

B. FALSE POSITIVE
   1. Peripheral cyanosis
   2. Cardiac failure
   3. Lung infection

TABLE 9

CYANOTIC CONGENITAL HEART DISEASE: THE DIAGNOSTIC LADDER

4. INVASIVE CARDIAC EVALUATION
   (a) Haemodynamic quantitation
   (b) Angiocardiographic studies
3. NON INVASIVE CARDIAC EVALUATION
   (a) Echocardiography: M-Mode, 2-D, Doppler
   (b) Radionuclear studies
2. SIMPLE LABORATORY INVESTIGATIONS
   (a) Chest roentgenogram
   (b) Electrocardiogram
   (c) Haemoglobin concentration
   (d) Peripheral blood film
1. CLINICAL EVALUATION
   (a) History
   (b) Physical examination

TABLE 10

A PRACTICL APPROACH TO CYANOTIC CONGENITAL HEART DISEASE: CLINICAL EVALUATION

1. FULL MEDICAL HISTORY:
   especially onset of cyanosis, hypoxic spell, squatting, cardiac failure
2. THOROUGH PHYSICAL EXAMINATION:
   including palpation for femoral pulse, blood pressure measurements, determination of cardio-visceral situs

TABLE 11

A PRACITCAL APPROACH TO CYANOTIC CONGENITAL HEART DISEASE: SIMPLE LABORATORY INVESTIGATIONS

1. CHEST ROENTGENOGRAM (INCLUDING UPPER ABDOMEN):
   (a) pulmonary vascularity
   (b) cardio-visceral situs
   (c) cardiothoracic ratio
   (d) other radiological peculiarities
2. ELECTROCARDIOGRAM:
   (a) Mean frontal plane QRS axis
   (b) Ventricular hypertrophy patterns
   (c) Other electrocardiographic peculiarities
3. PERIPHERAL BLOOD FILM:
   (a) Howell-Jolly bodies

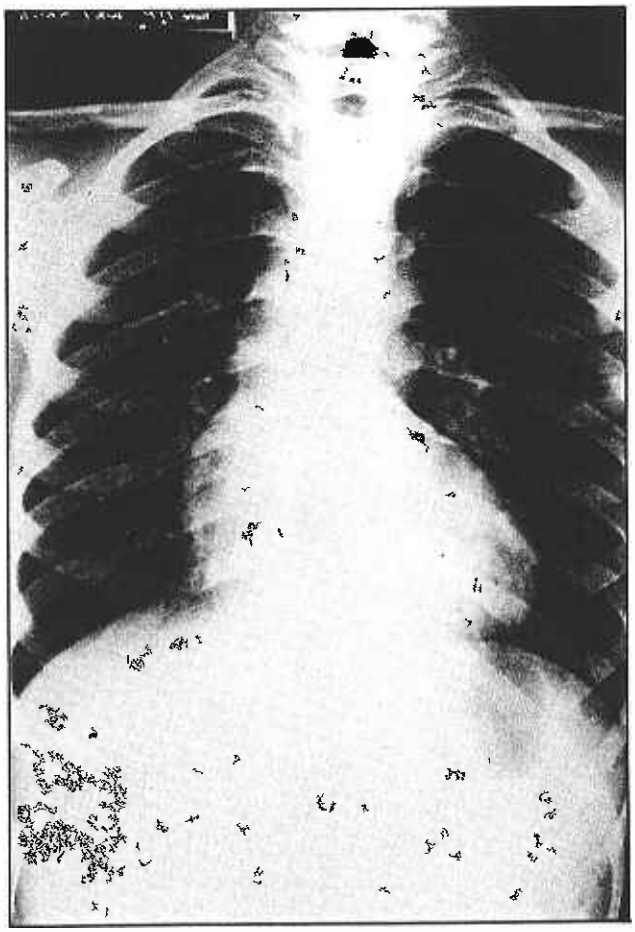


Fig. 1 :Chest roentgenogram showing typical boot-shaped heart of tetralogy of Fallot with oligaemic lung fields and concave pulmonary segment.

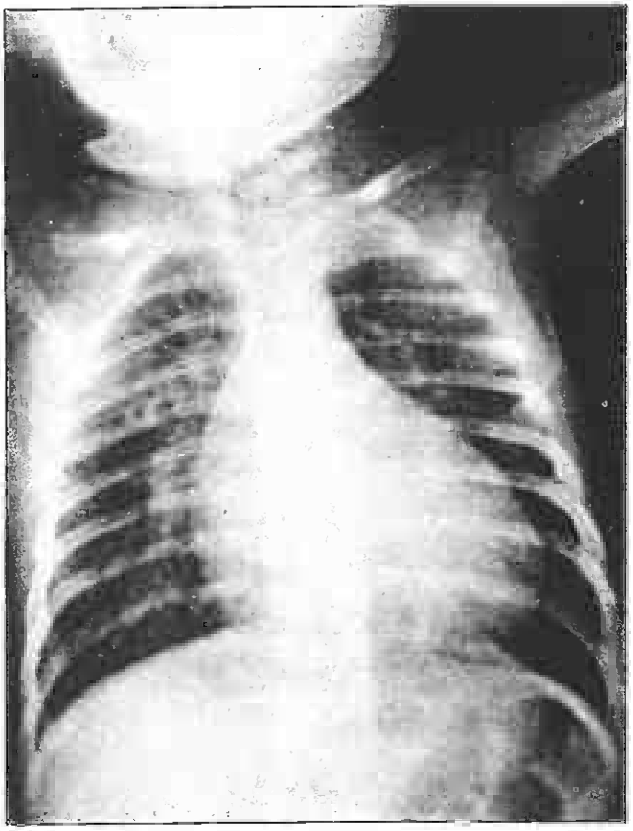


Fig. 2 : Chest roentgenogram showing egg-shaped heart of transposition of great arteries with narrow pedicle and gross pulmonary plethora.

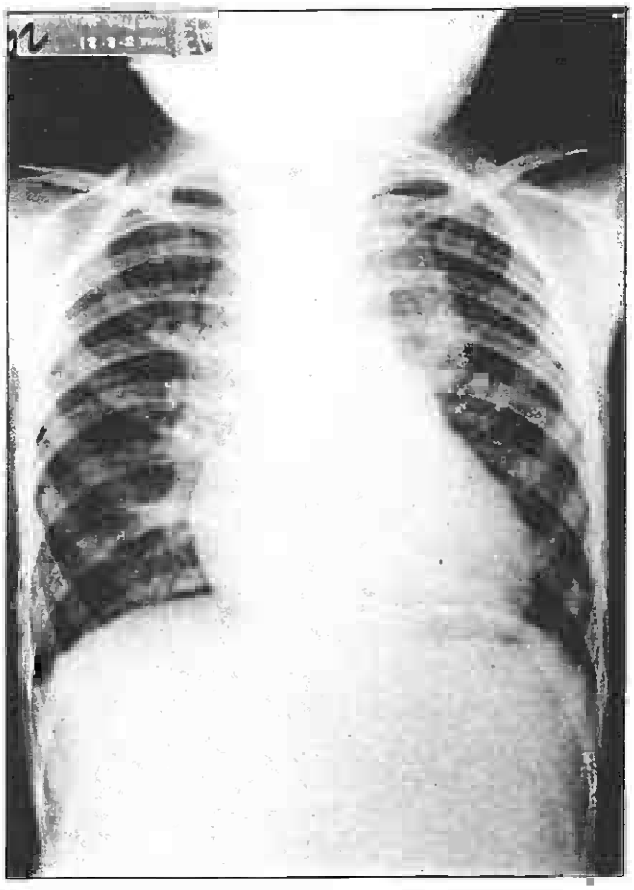


Fig.3 : Chest roentgrenogram showing classical snow-man appearance of total anomalous pulmonary venous drainage.

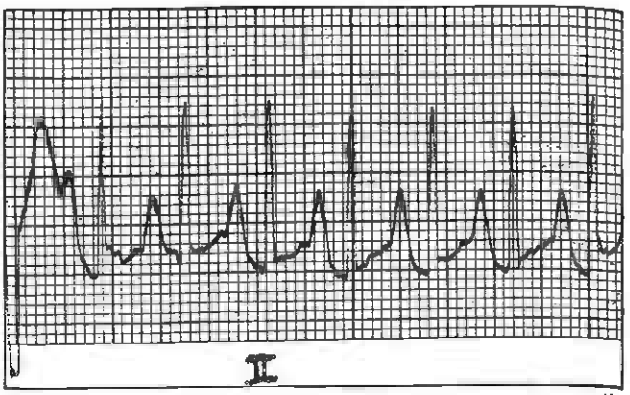


Fig. 4 : Electrocardiogram showing gian P waves of Ebstein's anomaly.

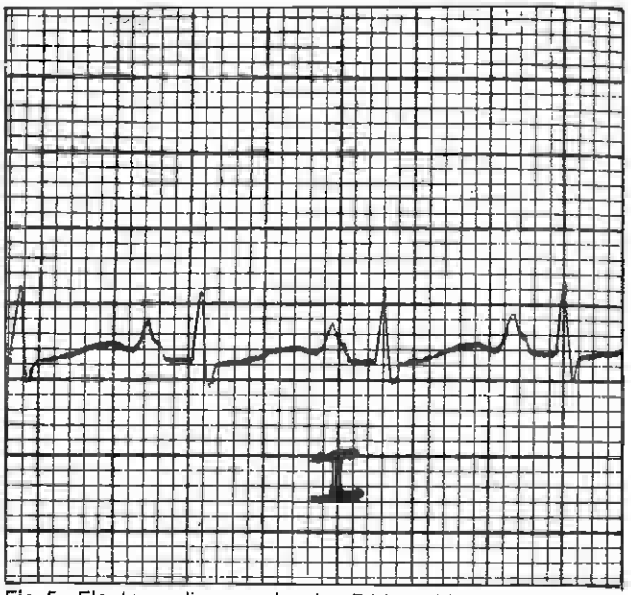


Fig. 5 : Electrocardiogram showing P tricuspidale with taller first peak in tricuspid atresia.

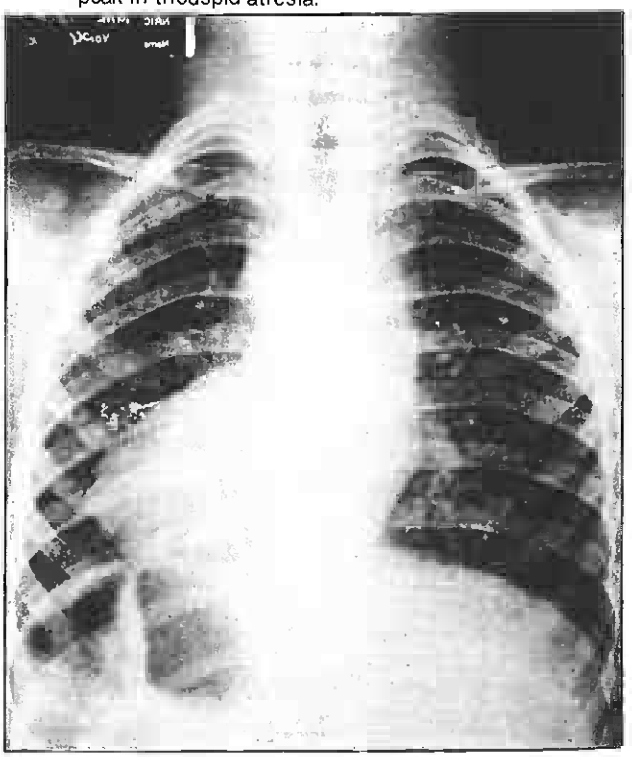


Fig. 6 : Chest roentgenogram showing heart in the right hemithorax with liver shadow situated in the left upper abdomen (dextrocardia with situs inversus).

TABLE 12

EXCLUSION OF CLINICALLY DELINEABLE DIAGNOSTIC GROUPS OF CYANOTIC CONGENTIAL HEART DISEASE

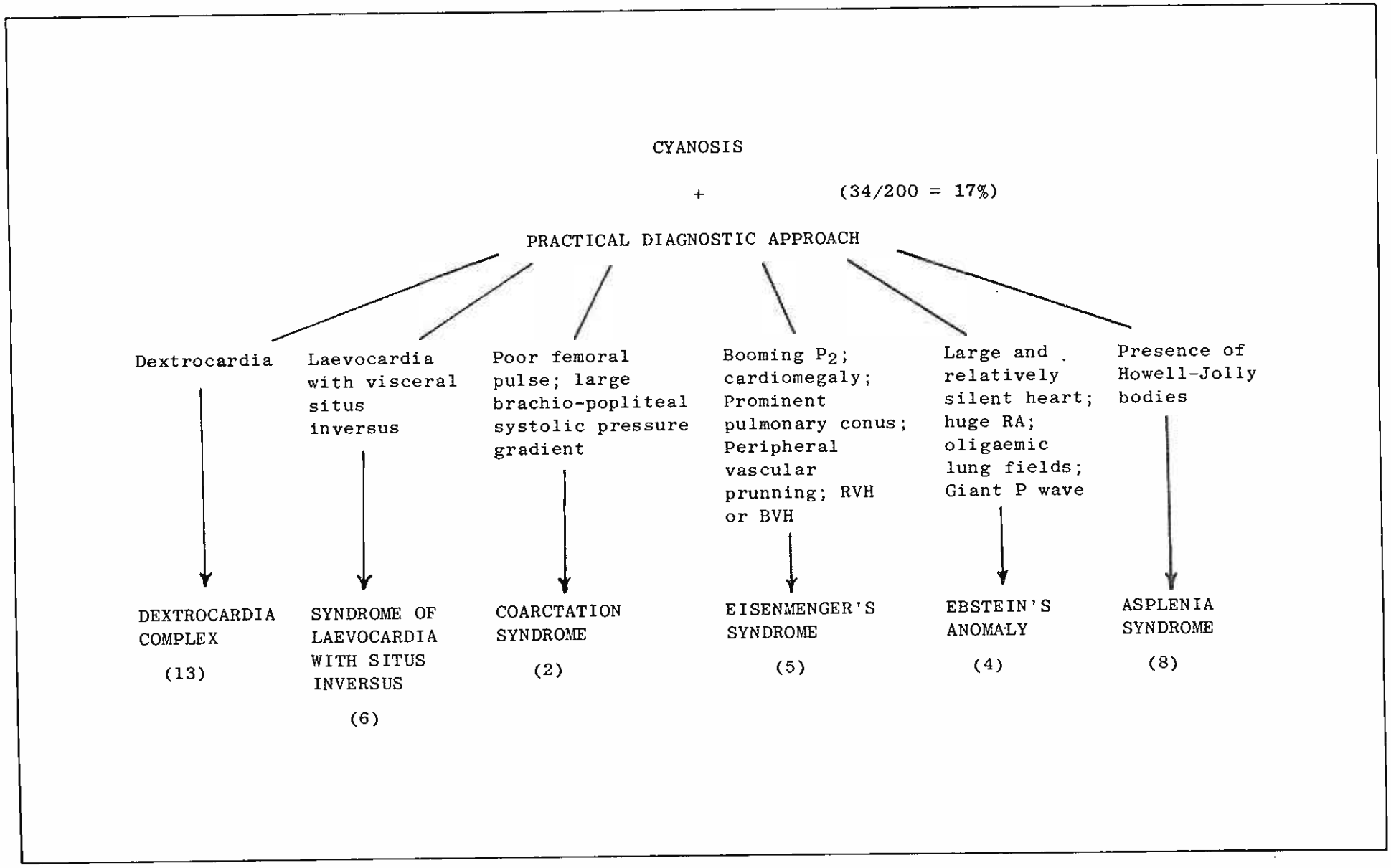


TABLE 13

CARDIOVASCULAR MALFORMATIONS IN DEXTROCARDIA COMPLEX (13 CASES)

VISCERAL SITUS:		13
INVERSUS	8	
SOLITUS	4	
AMBIGUOUS	1	
TRANSPOSITION OF GREAT ARTERIES		12
ATRIAL SEPTAL DEFECT		10
PULMONARY STENOSIS		8
VENTRICULAR SEPTAL DEFECT		7
PULMONARY ATRESIA		4
SINGLE VENTRICLE		4
AORTOPULMONARY COLLATERALS		2
ABSENT INFERIOR VENA CAVA		2
TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE		1
DOUBLE OUTLET RIGHT VENTRICLE		1
PATENT DUCTUS ARTERIOSUS		1

TABLE 14

CARDIOVASCULAR MALFORMATIONS IN SYNDROME OF LEAVOCARDIA WITH SITUS INVERSUS (6 CASES)

TRANSPOSITION OF GREAT ARTERIES	6
ATRIAL SEPTAL DEFECT	6
PULMONARY ATRESIA	5
VENTRICULAR SEPTAL DEFECT	3
PATENT DUCTUS ARTERIOSUS	3
SINGLE VENTRICLE	2
ABSENT INFERIOR VENA CAVA	1

TABLE 15

CARDIOVASCULAR MALFORMATIONS IN ASPLENIA SYNDROME (8 CASES)

CARDIOVISCERAL SITUS:		8
LAEVOCARDIA WITH SITUS SOLITUS	4	
LAEVOCARDIA WITH SITUS INVERSUS	3	
DEXTROCARDIA WITH SITUS SOLITUS	1	
PULMONARY ATRESIA		7
TRANSPOSITION OF GREAT ARTERIES		5
BILATERAL SUPERIOR VENA CAVA		5
VENTRICULAR SEPTAL DEFECT		4
PATENT DUCTUS ARTERIOSUS		4
TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE		4
ATRIAL SEPTAL DEFECT		3
SINGLE ATRIUM		3
AORTO-PULMONARY COLLATERALS		3
SINGLE VENTRICLE		2
BILATERAL INFERIOR VENA CAVA		2

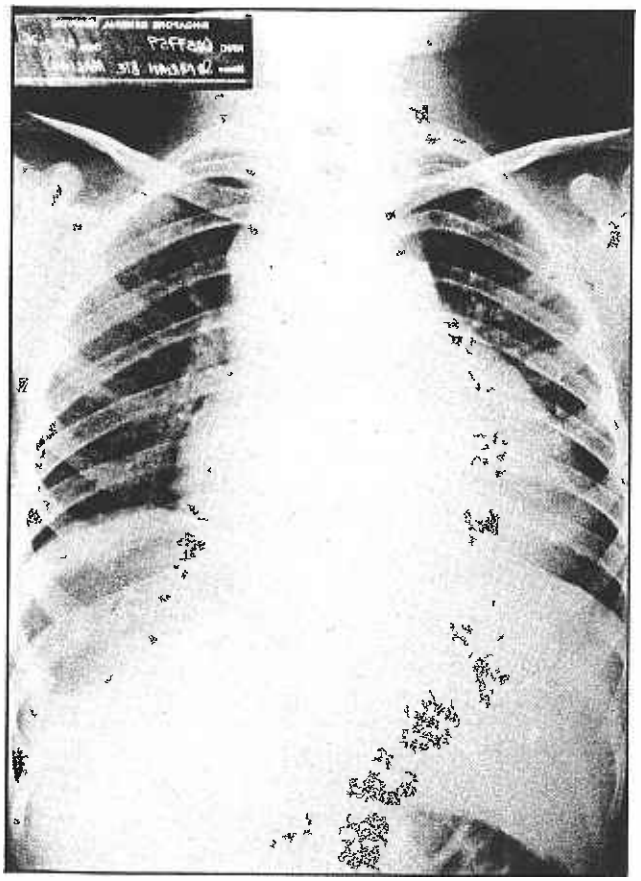


Fig. 7 : Chest roentgenogram showing heart in the usual left hemithorax but the liver shadow also situated in the left upper abdomen (laevocardia with situs inversus).

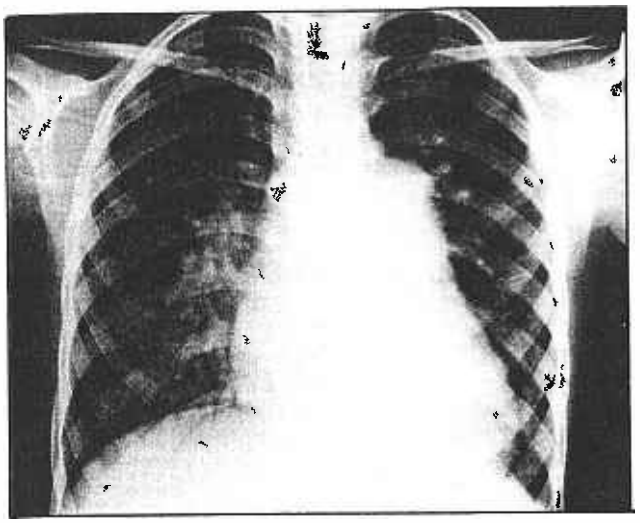


Fig. 8.Chest roentgenogram showing cardiomegaly, prominent pulmonary conus with proximal pulmonary vascular plethora and peripheral vascular prunning (Eisenmenger's syndrome).

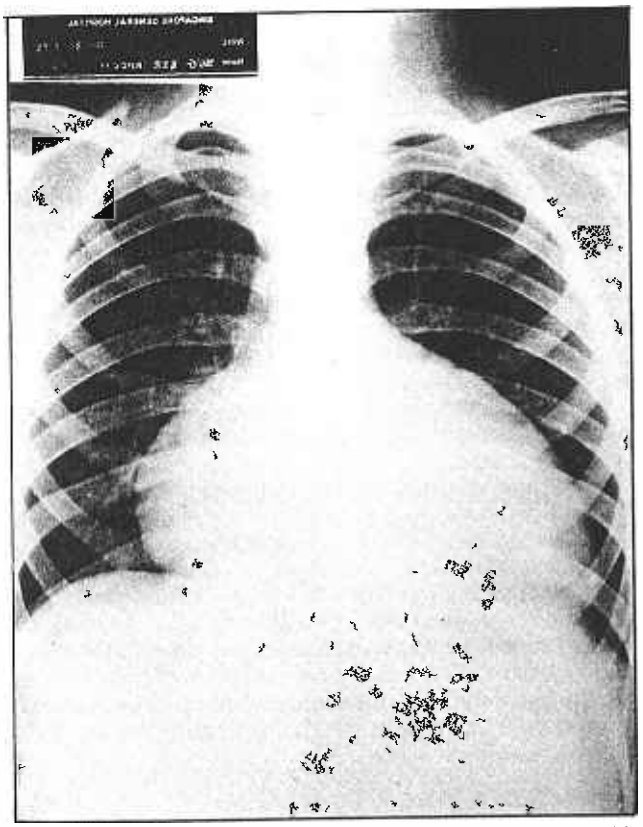


Fig. 9 : Chest roentgenogram showing gross cardiomegaly with giant right atrium and oligaemic lung fields (Ebstein's anomaly).

cyanotic heart disease, based on the electrocardiographic and roentgenographic features (listed in Table 16), has been proposed by Elliott and Schiebler (7).

(1) Computation of the frontal plane QRS axis

The analysis begins with the determaination of the frontal plane QRS axis. Although there are many methods to accomplish this, the easiest way is to compute from the QRS vectors in lead I and aVL. If a hand-held calculator with simple scientific functions is available, the frontal plane QRS axis can be easily calculated to the nearest degree (Table 17) (8).

TABLE 16

DIFFERENTIAL DIAGNOSIS OF CYANOTIC CONGENITAL HEART DISEASE BY ELECTROCARDIOGRAM AND CHEST ROENTGENOGRAM

1. ELECTROCARDIOGRAM
   (a) RAD, LAD or NAD
   (b) RVH, LVH, BVH or NVH
2. CHEST ROENTGENOGRAM
   (a) Plethora or Oligaemia

RAD = Right Axis Deviation
LAD = Left Axis Deviation
NAD = Normal Axis Deviation
RVH = Right Ventricular Hypertrophy
LVH = Left Ventricular Hypertrophy
BVH = Biventricular Hypertrophy
NVH = No Ventricular Hypertrophy

TABLE 17

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

If the QRS vectors 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.

(2) Determination of normal axis, or right axis or left axis deviation

The normal mean frontal plane QRS axis changes with age (Table 18) (9). In the first 24 hours of life, the average mean axis is at plus 137 degrees but the normal range varies from 75 to plus 190 degrees. From then on it gradually moves from the relatively right axis to the normal axis. For the paediatric cases after infancy, the normal axis is situated within minus 15 degrees to plus 120 degrees. Hence in children, the changing standard of normality must be borne in mind when one is deciding whether the QRS axis is normal, or deviated to the right or to the left. When the axis is situated in the range from minus 90 degrees to minus 180 degrees or from plus 180 degrees to plus 270 degrees, i.e. in the fourth quadrant (Table 19), it is not easy to differentiate extreme right axis from extreme left axis deviation. One simple way, when one has only the standard 12 leads electrocardiogram, is to look for the presence or absence of q wave in lead aVL. The presence of q wave in lead aVL indicates counter-clockwise QRS loop in the frontal plane which signifies extreme left axis deviation.

TABLE 18

NORMAL MEAN FRONTAL PLANE QRS AXIS* ACCORDING TO AGE (MODIFIED FROM ZIEGLER)

	Average	Minimum	Maximum
0 — 24 hours	137	75	190
1 day — 1 month	116	-5	190
1 month — 6 month	72	35	135
6 months — 1 year	64	30	135
1 year — 5 years	63	0	110
5 years — 12 years	66	-15	120
12 years — 16 years	66	-15	110

* Measured in degrees

REF: NADAS AS, FYLER DC. PEDIATRIC CARDIOLOGY, 3RD EDITION. PHILADELPHIA: W.B. SAUNDERS CO, 1972.

TABLE 19

MEAN FRONTAL PLANE QRS AXIS IN THE 4TH QUADRANT: RIGHT AXIS DEVIATION OR LEFT AXIS DEVIATION?

	EXTREME RIGHT AXIS DEVIATION	EXTREME LEFT AXIS DEVIATION
q wave in aVL	absent	present
QRS loop in frontal plane	clockwise	counter-clockwise

TABLE 20

SELECTED ELECTROCARDIOGRAPHIC VOLTAGE CRITERIA FOR VENTRICULAR HYPERTROPHY

A. RIGHT VENTRICULAR HYPERTROPHY
   1. R in V_1, V_2 or aVR greater than normal for age
   2. S in I or V_6 greater than normal for age

B. LEFT VENTRICULAR HYPERTROPHY
   1. R in I, II, aVL, aVF or V_6 greater than normal for age
   2. S in V_1 or V_2 greater than normal for age

C. COMBINED VENTRICULAR HYPERTROPHY
   1. Positive voltage criteria for right and left ventricular hypertrophy

REF: GUNTHEROTH WG: INITIAL EVALUATION OF THE CHILD FOR HEART DISEASE. PEDIATR CLIN NORTH AM 1978; 25(4): 657-675.

(3) Determination of ventricular hypertrophy patterns

Generally ventricular hypertrophy patterns can be inferred by two sets of interrelated criteria, namely, the so-called orientation criteria and voltage criteria(10). However these two sets of criteria are not necessary concordant in children with congenital heart disease e.g. left axis deviation does not always indicate left ventricular hypertrophy. Indeed in patients with endocardial cushion defect, left axis deviation is accompanied by right ventricular hypertrophy. For this reason, only selected voltage criteria listed in Table 20 are used in the analysis in this paper. Again because of the changing right ventricular preponderance in infancy to the left ventricular preponderance in later childhood, it will be important to follow the upper limits of R and S voltages of the relevant leads for the diagnosis of right, left or combined ventricular hypertrophy (Table 21 and Table 22) (10).

DIFFERENTIAL DIAGNOSIS OF CYANOTIC CHD BY ELECTROCARDIOGRAM AND ROENTGENOGRAM

After exclusion of the clinically delineable subgroups, the remaining of the 200 cases of cyanotic CHD are analysed according to the modified Elliott-Schiebler scheme (7). In the subgroup with right ventricular hypertrophy with right axis deviation or normal axis, the 3 common lesions are transposition of great arteries, total anomalous pulmonary venous drainage and double outlet right ventricle, if the chest roentgenogram also shows pulmonary plethora (Table 23). If the lung fields are oligaemic, then the two commonest lesions are tetralogy of Fallot and pulmonary atresia. The rest of the lesions are made up of those with severe pulmonary stenosis and either simple intracardiac right to left shunting at the atrial and/or ventricular level, or more complicated lesions like transposition of great arteries and double outlet right or left ventricle. This subgroup accounts for 61% of the 200 cases.

In the subgroup with normal axis and either no ventricular hypertrophy, biventricular hypertrophy or left ventricular hypertrophy, there are only 5 lessions, namely transposition of great arteries, pulmonary atresia, double outlet right ventricle, persistent truncus arteriosus and tricuspid atresia (Table 24). This subgroup accounts for 9% of the 200 cases.

Finally, in the subgroup with left axis deviation which is an uncommon occurrence, the differential diagnoses are rather limited. In the presence of left ventricular hypertrophy, tricuspid atresia and transposition of great arteries are the two most likely diagnoses. If the left axis deviation is associated with right ventricular hpertrophy, the 3 dif-

TABLE 21

RELEVANT UPPER LIMITS OR R AND S VOLTAGES* FOR DIAGNOSIS OF RIGHT VENTRICULAR HYPERTROPHY (ADAPTOR FROM GUNTHEROTH WG)

VOLTAGE	LEAD	0-1/12 YEAR	1/12-6/12 YEAR	6/12-1 YEAR	1-3 YEARS	3-8 YEARS	8-12 YEARS	12-16 YEARS
R	V1	25	20	20	18	18	16	16
	V2	30	30	28	25	28	22	19
	aVR	7	6	6	6	5	4	4
S	I	10	9	9	8	8	8	8
	V6	12	6	4	4	4	4	5

* MEASURED IN MILLIMETERS, WHEN 1 MW = 10 MM PAPER

REF: GUNTHEROTH WG: INITIAL EVALUATION OF THE CHILD FOR HEART DISEASE. PEDIATR CLIN NORTH AM 1978; 25(4) : 657-675.

TABLE 22

RELEVANT UPPER LIMITS OR R AND S VOLTAGES* FOR DIAGNOSIS OF LEFT VENTRICULAR HYPERTROPHY (ADAPTOR FROM GUNTHEROTH WG)

VOLTAGE	LEAD	0-1/12 YEAR	1/12-6/12 YEAR	6/12-1 YEAR	1-3 YEARS	3-8 YEARS	8-12 YEARS	12-16 YEARS
R	I	8	12	16	16	15	15	13
	I I	14	24	27	23	22	24	24
	aVL	7	8	10	10	10	10	12
	aVF	14	20	16	20	19	20	21
	V5	30	30	30	36	36	36	33
	V6	21	20	20	24	24	24	22
S	V1	20	18	16	27	30	26	24
	V2	35	30	30	34	38	38	48

* MEASURED IN MILLMETERS, WHEN 1 MV = 10 MM PAPER

REF: GUNTHEROTH WG: INITIAL EVALUATION OF THE CHILD FOR HEART DISEASE. PEDIATR CLIN NORTH AM 1978; 25(4): 657-675

TABLE 23

CORRELATION OF ELECTROCARDIOGRAPHIC AND ROENTGENOGRAPHIC FINDINGS IN CYANOTIC CONGENITAL HEART DISEASE (ADAPTED FROM ELLIOTT & SCHIEBLER)

CYANOSIS

+

RIGHT VENTRICULAR HYPERTROPHY (122/200 = 61%)

+

NORMAL AXIS OR RIGHT AXIS DEVIATION

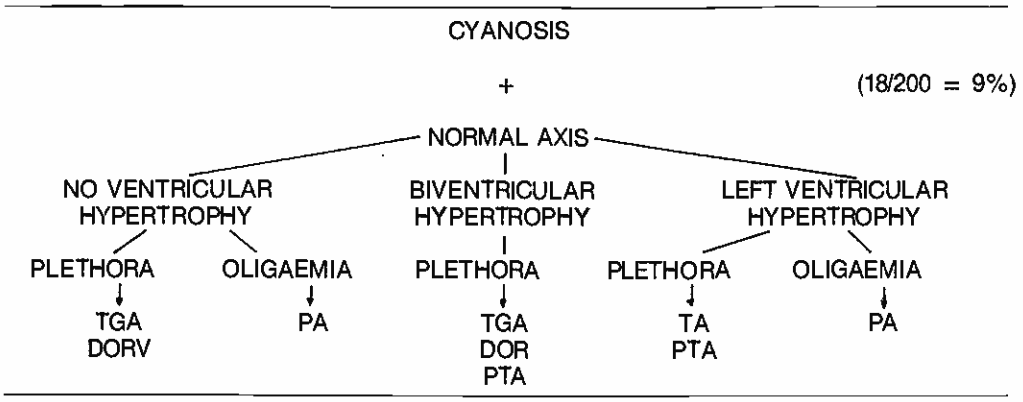
PLETHORA	OLIGAEMIA
↓	↓
TGA	TOF
TPAVD	PA
DORV	PS + ASD &/OR VSD
	PS + TGA/DORV/DOLV

TGA = Transposition of Great Arteries
TAPVD = Total Anomalous Pulmonary Venous Drainage
DORV = Double Outlet Right Ventricle
DOLV = Double Outlet Left Ventricle
TOF = Tetralogy of Fallot
PA = Pulmonary Atresia
PS = Pulmonary Stenosis
ASD = Atrial Septal Defect
VSD = Ventricular Septual Defect

REF: ELLIOTT LP, SCHIEBLER GL. A ROENTGENOLOGIC — ELECTROCARDIOGRAPHIC APPROACH TO CYANOTIC FORMS OF HEART DISEASE. PEDIATR CLIN OF NORTH AM 1971; 18(4) : 1133-1161.

TABLE 24

CORRELATION OF ELECTROCARDIOGRAPHIC AND ROENTGENOGRAPHIC FINDINGS IN CYANOTIC CONGENITAL HEART DISEASE (ADAPTED FROM ELLIOTT & SCHIEBLER)

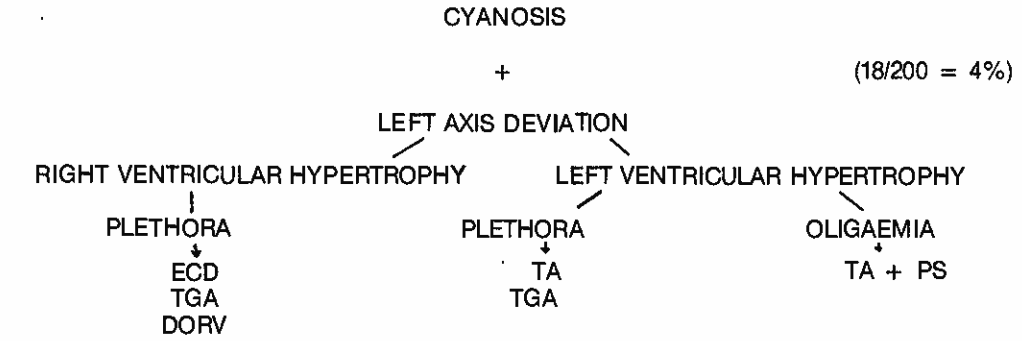


TGA = TRANSPOSITION OF GREAT ARTERIES
DORV = DOUBLE OUTLET RIGHT VENTRICLE
PTA = PERSISTENT TRUNCUS ARTERIOSUS
PA = PULMONARY ATRESIA
TA = TRICUSPID ATRESIA

REF: ELLIOTT LP, SCHIEBLER GL. A ROENTGENOLOGIC — ELECTROCARDIOGRAPHIC APPROACH TO CYANOTIC FORMS OF HEART DISEASE. PEDIATR CLIN OF NORTH AM 1971; 18(4) : 1133-1161.

TABLE 25

CORRELATION OF ELECTROCARDIOGRAPHIC AND ROENTGENOGRAPHIC FINDINGS IN CYANOTIC CONGENITAL HEART DISEASE (ADAPTED FROM ELLIOTT & SCHIEBLER)



ECD = Endocardial Cushion Defect
TGA = Transposition of Great Arteries
DORV = Double Outlet Right Ventricle
TA = Tricuspid Atresia
PS = Pulmonary Stenosis

REF: ELLIOTT LP, SCHIEBLER GL. A ROENTGENOLOGIC — ELECTROCARDIOGRAPHIC APPROACH TO CYANOTIC FORMS OF HEART DISEASE. PEDIATR CLIN OF NORTH AM 1971; 18(4) : 1133-1161.

ferential diagnoses to be considered are endocardial cushion defect, transposition of great arteries and double outlet right ventricle (Table 25). This subgroup only accounts for 4% of the 200 cases.

USEFULNESS AND LIMITATIONS OF THE ELECTROCARDIOGRAPHIC AND ROENTGENOGRAPHIC APPROACH

How useful is this electrocardiographic and roentgenographic approach as a guide to the differential diagnosis of cyanotic CHD? THis can be assessed by the accountability of the 7 common cyanotic heart lesions by this scheme (Table 26). THese 7 lesions constitute 97% of the 166 cases (after exclusion of the 34 clinically delineable cases). Of the 3 commonest lesions, namely tetralogy of Fallot, pulmonary atresia and transposition of great arteries, the accountability by this scheme is 100%, 86% and 79% respectively. Hence, the overall accountability of all the 200 children with cyanotic CHD by this proposed practical diagnositc approach is more than 90% (Table 27).

There are 18 cases (9%) which do not fit into the modified Elliott-Schielbler scheme. These are listed in Table 28. The reasons for the misfit are tabulated in Table 19. There are two patients with pulmonary atresia who had large patent ductus arteriosus which accounts for the unexpected plethora. The other reason for the misfit for

TABLE 26

ACCOUNTABILITY OF INDIVIDUAL CYANOTIC CONGENITAL HEART DISEASE BY THE MODIFIED ELLIOTT-SCHIEBLER SCHEME

	NO.	%
TETRALOGY OF FALLOT	78/78	100
TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE	6/6	100
PULMONARY ATRESIA	24/28	86
TRANSPOSITION OF GREAT ARTERIES	19/24	79
DOUBLE OUTLET RIGHT VENTRICLE	4/6	67
ENDOCARDIAL CUSHION DEFECT	2/3	67
PULMONARY STENOSIS + ATRIAL SEPTAL DEFECT &/OR VENTRICULAR SEPTAL DEFECT	10/16	63

TABLE 27

ACCOUNTABILITY OF 200 CHILDREN WITH CYANOTIC CONGENITAL HEART DISEASE BY DIAGNOSTIC APPROACH

	NO.	%
CLINICALLY DELINEABLE GROUP	34/34	100
CORRELATION BY ECG & CXR	148/166	89
TOTAL	182/200	91

TABLE 28

CYANOTIC CONGENITAL HEART DISEASE: THE MISFIT

NO.	
TRANSPOSITION OF GREAT ARTERIES	5
PULMONARY STENOSIS + ATRIAL SEPTAL DEFECT AND/OR VENTRICULAR SEPTAL DEFECT	5
PULMONARY ATRESIA	4
DOUBLE OUTLET RIGHT VENTRICLE	2
PERSISTENT TRUNCUS ARTERIOSUS	1
TRICUSPID ATRESIA	1
TOTAL	18 (9%)

TABLE 29

CYANOTIC CONGENITAL HEART DISEASE: THE REASONS FOR MISFIT

	NO.
Unexpected plethora*	2
Unexpected relationship between axis deviation and ventricular hypertrophy pattern	16
TOTAL	18

*Two patients with pulmonary atresia who had large patent ductus arteriosus

the remaining 16 cases is due to unexpected relationship between axis deviation and ventricular hypertrophy pattern.

CONCLUSION

Cyanotic CHD is often regarded as such complex anatomical lesions by general paediatricians that pre-catheterization diagnosis is considered difficult or impossible. Although it is true that even for paediatric cardiologists occasional surprise still occurs when the definitive diagnosis revealed at cardiac catheterization differs from the clinical diagnosis, it is demonstrated by this study, however, that in 9 out of 10 occasions a fair idea of the probable anatomical diagnosis can be derived from clinical and simple laboratory investigations.

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