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Clinical, echocardiography and cardiac catheterization study of children with congenital cyanotic heart diseases

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Abstract
Objective: To study contribution of noninvasive investigations and cardiac catheterization in making final diagnosis of patients with congenital cyanotic heart diseases.
Methods: A Prospective study carried out at a Paediatric Cardiology center at a tertiary care hospital. One hundred children with congenital cyanotic heart diseases (CCHD) from 0-10 years formed the study group. All cases with history of cyanosis or found cyanotic on clinical examination were subjected to detailed clinical examination, Hb/PCV measurement, Chest radiograph and Electrocardiogram. A provisional clinical diagnosis was made. Diagnosis was confirmed with 2D and colour Doppler echocardiography. Selected cases were taken up for cardiac catheterization where precise anatomic diagnosis and hemodynamic abnormalities could not be clearly defined by echocardiography. The contribution of each of the noninvasive and invasive modality in reaching an accurate diagnosis was calculated.
Results: Noninvasive investigations could make a diagnosis of CCHD in most of the cases but patients with DORV, TGA, and TAPVC with obstruction and complex lesions could not be differentiated. Cardiac catheterization was done in selected cases wherein aorto pulmonary collaterals, additional VSD and pulmonary artery anatomy could not be well defined on echocardiography. Cardiac catheterization gave additional information in 30.4% cases in this study.
Conclusion: Though noninvasive evaluation including an echocardiography forms the main stay diagnosis of all congenital cyanotic heart disease, Cardiac catheterization is required in complex cases where important additional information can be obtained which is important for precise diagnosis and management including the surgical point of view.

Keywords: diagnosis, congenital cyanotic heart disease, noninvasive evaluation, cardiac catheterization

1. Introduction

Congenital heart disease (CHD) refers to structural or functional heart disease that is present at birth. The incidence of CHD in various countries and different ethnic groups is almost the same. The incidence of CHD in infancy varies from 4 to 12 per 1000 live births in various communities and hospital based studies [1]. Approximately 45% of CHD cases require some form of intervention or surgery during childhood [2].

Congenital heart disease can be divided into two major groups, depending on the presence or absence of cyanosis, into congenital cyanotic heart disease (CCHD) and congenital acyanotic heart disease (ACHD) respectively [3]. Congenital cyanotic heart disease may present in immediate neonatal period, later in infancy or in childhood. Congenital cyanotic heart disease constitutes approximately 26% of all congenital heart diseases in children [4].

In most of the cases with congenital cyanotic heart diseases detailed clinical examination and easily available investigative tools like chest radiography and electrocardiography provide a possible diagnosis. However, echocardiography is a must for all the cases to confirm the detailed morphological diagnosis to plan further management [5].

Cardiac catheterization is recommended to confirm the echocardiographic findings and to define the anatomy and pressures in greater details whenever surgical intervention is contemplated [5].

The present study was undertaken to compare the clinical diagnosis, their correlation with ECG, chest radiograph, confirmation with the echocardiographic diagnosis and the findings of cardiac catheterization in selected cases where precise anatomic diagnosis and hemodynamic abnormalities could not be clearly defined by echocardiography.

2. Material and methods

A prospective study was carried out in a large tertiary care hospital.

One hundred children with congenital cyanotic heart diseases from 0-10 years, formed the material for the present study. Patients who had undergone cardiac surgery/intervention procedures before the period of present study were not included in this study. All the cases with history of cyanosis or found cyanotic on clinical examination were subjected to detailed history and thorough clinical examination. Hemoglobin (Hb) and packed cell volume (PCV) were measured; Chest radiograph and Electrocardiogram were taken in all cases. Provisional clinical diagnosis was made on the basis of clinical examination, Chest radiograph and ECG. Diagnosis was confirmed with 2D and colour Doppler echocardiography performed by a Pediatric Cardiologist.

Selected cases were taken up for cardiac catheterization before palliative or corrective surgery. This selection was done by Pediatric Cardiologist based on his need for the same in planning future management. Observations were tabulated, analyzed statistically and interpreted.

3. Observations

100 cases of Congenital Cyanotic Heart diseases were studied in a tertiary care hospital. A thorough general examination was carried out. This included anthropometric measurements, documentation of clubbing and dysmorphic features. The findings are as per table 1. Clinical cardiovascular examination of all patients were carried out and results tabulated. The clinical findings are summarized in table 2. 6% cases had dextrocardia. 86% cases had single S2. 22% of cases did not have a cardiac murmur. Hb and PCV was measured in all patients. It was observed that 89% patients had Hb between 14 and 21gm%and PCV between 45% and 65%. 3% patients had PCV of more than 65% and required partial exchange transfusion. 8% of patients had Hb less than 14gm % and PCV less than 45%. Peripheral blood smear was also examined in these patients and after confirming the presence of iron deficiency anemia they were treated with iron and folic acid. The findings are summarized in Table 3. ECG was carried out for evaluation of all patients and findings summarized as per Table 4. Majority of patients with TOF had RAD + RVH while all cases with tricuspid atresia had LAD+LVH. A chest radiograph was taken for all patients and the findings are tabulated as per Table 5. 22% patients had a normal chest radiograph. Echocardiographic findings and cardiac catheterization findings are summarized in Table 6 and 7 respectively.

Based on the above data, the findings were summarized. The Correlation between clinical, echocardiographic and cardiac catheterization diagnoses has been studied and tabulated in Table 8.

Table 1: General Examination findings of children diagnosed with congenital cyanotic heart diseases.

S. No.	Lesions	Dysmorphic Features n(%)	Clubbing n(%)	Wt<3rd centile n(%)	Ht<3rd centile n(%)
1	TOF (56)	5(8.9)	26(46.4)	33(58.9)	23(41)
2	TA (5)	0	2(40)	1(20)	1(20)
3	DORV (7)	1(14.2)	1(14.2)	2(28.5)	0
4	TGA (13)	0	5(38.4)	6(46.1)	2(15.3)
5	TAPVC (7)	0	0	0	0
6	AV Canal defect (3)	1(33.3)	1(33.3)	3(100)	0
7	Hypoplastic left heart (2)	0	0	0	0
8	Eisenmengers complex (1)	1(100)	1(100)	1(100)	1(100)
9	Complex Lesion (5)	0	0	3(60)	1(20)
10	Truncus arteriosus (1)	1(100)	0	0	0
	Total	9	36	49	28

Table 2: Cardiovascular Examination findings of children diagnosed with congenital cyanotic heart diseases.

No.	Lesions (n)	Apex RV/LV/N/ Dextro n(%)	Palp P2 n(%)	Thrill n(%)	S2 Single n(%)	Murmurs		
						ESM n(%)	PSM/ Continuous murmu	Silent n(%)
1	TOF (56)	RV-56(100)	0	12(21.4)	56(100)	46(82.14)	2(3.5)	8(14.2)
2	TA (5)	LV-5 (100)	0	0	5(100)	4(80)		1(20)
3	DORV (7)	RV 5(71.4)Dextro-2(28.5)	0	0	7(100)	6(85.7)		1(14.28)
4	TGA (13)	RV-12 (85.7)Dextro-1 (7.6)	1(7.6)	4(30.76)	11(84.6)	5(38.4)	3(23)	5(38.4)
5	TAPVC (7)	RV6 (85.7)LV-1 (14.2)	3(42.8)	0	0	2(28.5)	1(14.5)	4(57.1)
6	A V Canal defect (3)	LV-2 (66.6) Dextro-1 (33.3)	0	1(33.3)	2(66.6)	2(66.6)		1(33.3)
7	Hypo plastic left heart (2)	RV-2 (100)	0	1(50)	2(100)	1(50)		1(50)
8	Eisenmengers complex (1)	N-1 (100)	1(100)	0	0	1(100)		
9	Complex congenital cyanotic heartdisease(5)	N-3 (60)Dextro-2 (50)	0	2(40)	3(60)	3(75)	1(20)	1(25)
10	Truncus arteriosus (1)	N-1 (100)	0	1(100)	1(100)	1(100)		
	Total	N-4 RV-81 LV-9Dextro-6	5	20	86	71	7	22

- 6% cases had dextrocardia. 86% cases had single S2.
- 22% of cases did not have a cardiac murmur.

Table 3: Haemoglobin (Hb) and packed cell volume (PCV) in children diagnosed with congenital cyanotic heart diseases

Hb			PCV		
<14gm%	14-21gm%	>21gm%	<45%	45-65%	>65%
8	89	3	8	89	3

Table 4: Electrocardiographic findings in children diagnosed with congenital cyanotic heart diseases

S.No.	Lesions(n)	Mean QRS Axis				RVH n (%)	LVH n (%)	BVH n (%)
		RAD n (%)	LAD n (%)	Indeterminate n (%)	Normal n (%)			
1	TOF (56)	52(92.8)	0	0	4(7.1)	42(75)	0	0
2	TA (5)	0	5 (100)	0	0	0	5(100)	0
3	DORV (7)	4(57.1)Dextro (28.5)	0	1(14.2)	0	4(57.1)	0	0

4	TGA (13)	7(53.8)Dextro-1(7.6)	0	1(7.6)	4(30.7)	0	0	0
5	TAPVC (7)	5(71.4)	0	0	2(28.5)	5(71.4)	0	0
6	A V Canal defect (3)	Dextro-1(33.3)	0	2(66.6)	0	1(33.3)	0	0
7	Hypoplastic left heart (2)	2(100)	0	0	0	2(100)	0	0
8	Eisenmengers complex (1)	1(100)	0	0	0	1(100)	0	0
9	Complex congenital cyanotic heart disease	1(20)Dextro-2 (40)	1(20)	0	0	0	0	0
10	Truncus arteriosus (1)	0	0	0	1(100)	0	0	1(100)
	Total (100)	72Dextro-06	7	4	11	55	5	1

Table 5: Chest Radiography findings in children diagnosed with congenital cyanotic heart diseases

S. No.	Lesion (n)	Pulmonary plethora n (%)	Pulmonary Oligemia n (%)	Cardiomegaly n (%)	Normal radiograph n (%)	Classical radiograph n (%)
1	TOF (56)	0	43(76.7)	2(3.5)	11(19.6)	43(76.7)
2	TA (5)	0	2(40)	1(20)	2(40)	
3	DORV (7)	0	6(85.7)	1(14.2)	1(14.2)	6(85.7)
4	TGA (13)	5(38.47)	3(23)	6(46.1)	4(30.7)	1(7.6)
5	TAPVC (7)	6(85.7)	0	5(71.4)	1(14.2)	1(14.2)
6	A V Canal defect (3)	1(33.3)	0	1(33.3)	2(66.6)	
7	Hypoplastic left heart (2)	2(100)	0	2(100)	0	
8	Eisenmengers complex (1)	0	1(100)	0	0	
9	Complex congenital cyanotic heart disease (5)	0	0	2(40)	1(20)	
10	Truncus arteriosus (1)	1(100)	0	1(100)	0	
	Total (100)	15	55	21	22	51

Table 6: Echocardiographic findings in children diagnosed with congenital cyanotic heart diseases

S. No.	Lesions	Solitus	Atria & IAS	Ven. &IVS	Pulmonary Stenosis	PDA	Collat	Arch
1	TOF (56)	Solitus-56	ASD 2	VSD 56	Pul atr 12 PS 44	13	-	L 30 R 26
2	TA (5)	Solitus-5	ASD 5	VSD 5	PS 3	2	0	L 5
3	DORV (7)	Dextro-2 Solitus 5	PFO 1	VSD 7	PS 7 Malpos art 2	0	0	L 3 R 4
4	TGA (13)	Dextro-1 Solitus 12	ASD 9	VSD 7Intact septum 6	NRGA 7 Side by Side 6 PS 6	3	0	L10 L 3
5	TAPVC (7)	Solitus 7	ASD 7	VSD 1	Aorta from RV 1	1	0	L 7
6	A V Canal defect (3)	Solitus 2 Inver 1	ASD 3 Common Avalve 2	VSD 3	PS 3Aorta ant 1Hypo PA 2	1	0	L 3
7	Hypoplastic left heart (2)	Solitus 2	PFO 2	LV hypoplastic 2	PA dil 2 Hypo aorta 2	2	0	L 2
8	Eisenmenger complex (1)	Solitus 1	N	N	N	1	0	
9	Complex congenital cyanotic heart disease (5)	Solitus 3 Invers 2	Single atrium 2 Single AV valve 2	VSD 1 Single ven 2 DILV 1 DORV 1 Hypoplastic TV &RV 1	PS 4 Hypo PA 1	0	0	L 5
10	Truncus arteriosus (1)	Solitus 1		VSD 1	PA /annulus not seen	0	0	

Table 7: Cardiac Catheterization Findings in congenital cyanotic heart diseases.

Srl.No.	Lesions	No of VSD	Infundibulum	PA Anatomy	Status of coronary arteries	Aorto pul Collat-erals	Others
1	TOF (20)	Large, non restrictive, sub aortic VSD – 20 Additional muscular VSD – 4	Severe infundibular Hypertrophy – 18 Infundibulum normal - 2	Hypoplastic MPA- 16 LPA stenosis at origin-1 Severe stenosis at origin of Both pulm arteries- 2 Confluent pulmonary artries in all cases	Common origin coronary artery-1 Large conal branch crossing RVOT-1	MAPCAS-2 (Coiling done)	Rt aortic arch- 6 Lt aortic arch- 14
2	TGA (4)	Septum intact					Balloon septostomy Done (4)
3	TOF, Pulmonary atresia, (2)	Single VSD	RV size small Hypertrophy +	Pul atresia, Confluent PA's			PDA (2)
4	TAPVC (1)	-	-	Normal	-	-	Left sided veins to Coronary sinus

Table 8: Correlation between clinical, echocardiographic and cardiac catheterization diagnoses

S No	Lesion	Clinical diagnosis Based on examination + ECG + Chest radiograph	Additional information on echocardiography	Additional information on cardiac cath (Additional findings / Cath performed)
1	TOF (56)	TOF Physiology56	18(44.9%)	7/ 22

2	TA (5)	TA (5)	2(40%)	
3	DORV (7)	TOF (7)	7(100%)	
4	TGA (13)	TOF physiology (6)TGA (7)	13(100%)	Septostomy done-4
5	TAPVC (7)	Cyanotic heart disease (7)	7(100%)	1/ 1
6	A V Canal defect (3)	AV Canal defect(3)	3(100%)	
7	Hypoplastic left heart (2)	Cyanotic heart disease (2)	2(100%)	
8	Eisenmengers complex (1)	Eisenmengers complex (1)	1(100%)	
9	Complex congenital cyanotic heart disease (5)	Cyanotic heart disease (5)	5(100%)	
10	Truncus arteriosus (1)	TOF	1(100%)	

4. Discussion

The present study was conducted on 100 clinical cases of Congenital Cyanotic Heart Diseases (CCHD) in a tertiary care referral hospital.

Tetralogy of Fallot was found to be the commonest CCHD with a prevalence rate of 56%. In other studies its prevalence has been reported ranging from 2.3 % to 21 % out of all congenital heart diseases [6]. Cyanosis was detected clinically in 96.4% cases. In two cases cyanosis was not evident clinically but these children did have history of cyanosis. Their echocardiography showed classical features of Tetralogy of Fallot and therefore they were called as cases of "Pink Tetralogy". Cyanotic spells were present in 51.7% cases of TOF. Squatting and dyspnoea on exertion was evident in 42.8 and 39.2% cases respectively. Seizures were seen in 7.1% cases of TOF. Seizures were associated with severe cyanotic spells, and in 2 cases they led to neurological deficit also. History of recurrent chest infection was also present in 3.5% cases. However, clinical and echocardiographic findings did not provide any evidence of increased pulmonary blood flow in these cases. It appears that these patients probably had reactive airway disease, causing recurrent respiratory symptoms.

Clinical examination revealed that 58.9% cases were underweight (weight less than 3rd percentile) while 41% cases had short stature (Height < 3rd percentile). Clubbing was seen in 46.4% cases while the youngest patient having clubbing was 5 months old. Examination of cardio-vascular system revealed the presence of thrills in 21.4% cases. Second sound was single in all the patients. Ejection systolic murmur was present in 82.2% cases at left upper para sternal region, possibly due to infundibular and valvular pulmonary stenos is, while no murmur was audible in 14.2% cases. Clinically cases where no murmur was audible were suspected to be having pulmonary atresia. In ECG in the present study RAD was seen in 92.8% cases and RVH was present in 75% of cases. There were 7.1%cases of TOF where ECG could be passed as normal. Radiograph of chest had classical Cour-en-sabot pattern or boot shaped heart with upturned apex and pulmonary olegemia in 76.78 % cases. There were about 23 % cases where oligemia was not obvious and X ray chest could be passed as normal.

Analysis of the data for correlation with clinical (Examination + ECG + Chest radiograph) and echocardiography findings revealed that clinical diagnosis was correct in 55.1 % cases. ECG was normal in 7.1% cases and chest radiograph was near normal in 24 % cases. Echocardiography was contributory in demonstrating details of complete anatomy. Additional VSDs were detected in 2 cases (3.5 %), that remained undetected by clinical examination but have importance for surgical correction. Thirteen cases (23.2%) were found to be having PDA while murmur of PDA was heard only in two cases.

TOF with Pulmonary Atresia is an extreme form of TOF and was present in 9 patients in this study**.** Clinically the diagnosis was suspected in patients with presentations like TOF having no audible murmur or only an audible continuous murmur of PDA or collaterals. Echocardiography is therefore an essential investigation to diagnose these cases.

There were seven cases of double outlet right ventricle. These were clinically diagnosed as of TOF. Chest radiograph and electrocardiographic features also could not help in the exact diagnosis of these cases. Extreme right axis deviation and indeterminate axis, though described in DORV was not seen in any of these patients. The diagnosis was made by echocardiography by demonstrating aortic override of more than 50%.

Cardiac catheterization was performed in 22 cases of TOF, which confirmed the echocardiographic findings. Additional information was obtained in seven cases (31.84%) where another (muscular) VSD was detected in two cases. Large collaterals were found in two cases, which were taken up for coil embolization at the same time. Coronary artery abnormality was found in two patients (10%) which was in the form of single origin of coronary artery in one case and in another a large conal branch was found crossing RVOT. In a study Fellows *et al* [7]. found the incidence of recognized coronary anomalies as 5.0% in TOF. In one case anomalous origin of left subclavian was detected. It was arising as a separate branch from descending aorta. These findings were important from surgical point of view.

Tricuspid Atresia: There were five cases of tricuspid atresia in the present study amongst cyanotic patients making the prevalence of tricuspid atresia as 5.0%. The present observation is in agreement with the findings of. Walloopillai [8], who reported the incidence of Tricuspid Atresia as 5.9 % cases amongst total cases of CHD. Though, cyanosis was present in all these cases except one patient who only had history of cyanotic spells and squatting. Clinical examination revealed left ventricular type apex beat and single second sound. 80% cases had ejection systolic murmur while 20% did not have any murmur. ECG showed left axis deviation and left ventricular hypertrophy in all the cases. All these cases of tricuspid atresia were suspected on the basis of ECG, which was confirmed by echocardiography. Thus the presence of left axis deviation proved to be 100 % specific for tricuspid atresia in the present study. Chest x-ray showed pulmonary oligemia in 40% cases only. Echocardiography in such patients is gold standard as to demonstrate associated defects. Pulmonary stenosis was seen in three cases, PDA was present in two cases

while hypoplasia of right ventricle was demonstrated in two cases. These details are very important to plan future surgical intervention (single ventricle repair or a two ventricular repair). Thus cases of tricuspid atresia, though, suspected on basis of clinical examination and ECG, cannot be managed further without detailed echocardiographic examination. Cardiac catheterization could not be performed in any case of tricuspid atresia included in this study.

Transposition of great arteries (TGA)

There were 13 cases of TGA. Out of these six were with intact ventricular septum. They had normal birth weight and were found normal at birth. Tachypnea and cyanosis were detected after few hours of birth when the disease was suspected. In all these cases examination of cardio-vascular system was non-committal. No murmur could be audible in any of the case.

Chest radiograph showed cardiomegaly with normal lung fields. Echocardiography was done in all these cases soon after suspicion of diagnosis, which revealed TGA with intact IVS and PDA. PDA was in various stages of closing in these cases. These were duct dependent lesions. VSD was observed in seven cases of TGA in the present study. Pulmonary stenosis was found in six cases. These cases were in fact clinically suspected having TOF. Patients of TGA with pulmonary stenosis have to be managed like TOF while patients without PS require early corrective treatment so as to prevent development of pulmonary hypertension.

Total anomalous pulmonary venous connection (TAPVC)

There were seven cases of TAPVC. All of them presented with cyanosis and features of CCF. Four patients (51.7%) did not have any murmur on examination. X-ray chest showed cardiomegaly in 71.4% and plethoric lung fields in 85.5% cases. Based on clinical features, chest radiograph and ECG, a diagnosis of congenital cyanotic heart disease with increased pulmonary blood flow was made in all seven cases. Confirmed diagnosis of TAPVC was made only after echocardiography, though, it was kept as one of the important differential diagnosis. Detailed echo confirmed the diagnosis of TAPVC and also details of the type of TAPVC in all seven cases. Two cases had infracardiac obstructive TAPVC, which were sent immediately for surgery and were operated based on echocardiographic diagnosis. Four cases were of supracardiac variety and were taken up for surgery in next 1 – 3 months after their condition stabilized with medical management. One case, which presented at 1.5 years age was clinically suspected to be having a large left to right shunt. Diagnosis was made on echocardiographic examination. It was a mixed type of TAPVC. On echo examination anomalous venous drainage was suspected to be of cardiac type while on cardiac catheterization it was proved to be of mixed variety (two left pulmonary veins to coronary sinus + two right pulmonary veins to RA).

Hypoplastic left heart syndrome (HLHS)

There were two cases of hypoplastic left heart in the present series. One presented at the age of seven days with the history of cyanosis, seizures and dyspnoea. Examination revealed features of congestive cardiac failure and shock. There was a faint grade II ejection systolic murmur at left parasternal region. The diagnosis was made by echocardiography only. The other child presented at 1 ½ months of age. This patient had mild cyanosis and features of congestive cardiac failure. ECG and chest radiograph were not contributory for making the diagnosis. Diagnosis was confirmed only after echocardiographic examination.

Atrioventricular canal defect (AV Canal defect)

In this study there were three cases of AV Canal defect with pulmonary stenosis. They all presented with cyanosis and effort intolerance. On examination one had dextrocardia. Clinical diagnosis was suspected to be TOF physiology in all three of them. Chest radiograph did not contribute in diagnosis. ECG confirmed dextrocardia in one while in two cases with situs solitus, mean QRS axis was indeterminate. It gave a clue to diagnosis. However, it was only after echocardiography that a diagnosis of complete AV Canal defect could be made. Complete AV canal defect (Endocardial cushion defect) is more common in children with Down's syndrome but in this series none of the case with AV Canal defect had Downs syndrome. All these patients had associated pulmonary stenosis hence presented late in infancy and early childhood. Echocardiography further demonstrated the presence of PDA in one case. Mean QRS axis can give a clue but echocardiography is a must for confirmatory diagnosis.

Truncus Arteriosus

There was one case of Truncus arteriosus in this study. This baby was found to have cyanosis with congestive cardiac failure. Chest x-ray revealed cardiomegaly with pulmonary plethora. Diagnosis was made on echocardiography only which confirmed it to be of Type I.

Complex Congenital Cyanotic heart Disease

Five cases in the study were grouped in the category of complex congenital heart disease. Two cases had dextrocardia with normal abdominal situs. Clinical diagnosis in all these cases was Tetralogy of Fallot. There were no specific clinical, ECG or radiological features. Echocardiography showed the presence of single atrium and single ventricle (in two cases), common AV valve (in two cases), double inlet left ventricle (in one case), double outlet right ventricle (in one case), hypo plastic right heart (in one case), TAPVC (in two cases), bilateral superior vena cava (in two cases), pulmonary stenosis (in four cases) and hypo plastic pulmonary arteries and PDA (in one case). Management of these patients was solely dependent on their detailed echo based diagnosis.

Eisenmenger's Complex

A case of large PDA also presented with cyanosis at the age of 10 years. She had severe pulmonary arterial hypertension and was inoperable. Patient was included in this study because of the presence of cyanosis. Diagnosis was suspected on clinical examination. ECG and chest radiograph also pointed towards possibility of Eisenmenger's complex. Exact diagnosis was confirmed by echocardiography examination only.

5. Conclusion

Congenital cyanotic heart diseases presented at an early age. More than 60% patients presented within first year and about 98 % by 5 years of age.

Tetralogy of Fallot is the commonest congenital cyanotic heart disease and was seen in 56% of all CCHD in this study. This was followed by transposition of great arteries (13%).Apart from cyanosis, dyspnoea was the most common symptom. However, no symptom was particularly characteristic of a specific lesion. Growth retardation is commonly seen in CCHD and was present in 49 % cases. Clinical findings, ECG and Chest radiograph could make a diagnosis of CCHD in most of the cases above 1 year of age but patients with DORV, TGA, and TAPVC with obstruction and complex lesions could not be differentiated. Such cases were mostly diagnosed as of TOF on initial assessment. ECG with features of left axis deviation was a diagnostic feature in all cases of tricuspid atresia. Echocardiography forms the mainstay of diagnosis of all congenital cyanotic heart diseases. It was found to be important for confirmation of the diagnosis, defining associated lesions and for planning further management. Echocardiography provided additional information in 50 % cases of TOF that was important from the point of surgical management while in all other CCHD it was important in 100% patients. Management of CCHD was not possible without echocardiographic diagnosis in all cases. Cardiac catheterization was not required in all cases of TOF. It was done in selected cases wherein aorto pulmonary collaterals, additional VSD and pulmonary artery anatomy could not be well defined on echocardiography. Cardiac catheterization gave additional information in 30.4% cases in this study. These findings were missed on echocardiography. Cardiac catheterization was a relatively safe procedure and was required in complex cases where additional information could be obtained which was important from surgical point of view. The procedure was also used as an opportunity to embolize collaterals in cases where they posed a threat of excessive bleeding during surgery or doing a palliative procedure like balloon atrial septostomy in TGA.

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