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Original Research Article

Evaluation of congenital heart disease clinically and by echocardiography in children of age group 0-12 years

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ABSTRACT

Background: The present study was conducted to evaluate clinical and echo-cardio graphic findings of congenital heart disease in children of age-group 0-12 years attending present hospital.
Methods: A study was done at Katuri Medical College and hospital in 65 patients aged 0-12 years; both sexes; for a period of 2 years. study design is exploratory, with proforma been designed to study congenital heart diseases. Alexander Nada's criteria is used.
Results: Out of 4145 cases; 65 cases aged 0-12 years have congenital heart disease over a period of 2 years; (acyanotic 55 cases and cyanotic 10 cases). 23 cases are between 1-5 years and 20 cases are >5 years; 1/3rd cases diagnosed before 1 year of age. male 40 cases and female 25 cases. Urban cases-27 and rural cases are 38.48 cases have consanguinity and family history in 12 cases. extra cardiac manifestations in 6 cases. Normal sized heart with normal vascularity of lung was seen in 14 / 65 cases. 51/65 cases showed various radiological features depending on type of lesion based on echo findings. Among the ACHDs VSD was the commonest CHD seen in children. Among the cyanotic heart diseases TOF was the commonest CHD observed, comprising of 6 cases.
Conclusions: Chest X-ray was abnormal in 80% of cases with increased CT ratio in 58.3% cases. ECG was abnormal in 73.3% of cases and majority of them had left axis deviation.95%of CHDs, including ACHDs and CCHDs which were diagnosed clinically include ASD, VSD, PDA, AS, PS, CoA, TOF, TGA. Most common clinically diagnosed CHD was VSD. Clinico echo correlation was accurate in 75% cases. Clinico echo correlation was highest with isolated lesions like VSD, ASD and in cases with typical findings like CoA, PDA, AS, TOF and least with multiple lesions or atypical findings.

Keywords: Congenital heart disease, Echo in children, Echo vs clinical

INTRODUCTION

Congenital heart disease (CHD) is defined as an abnormality in 'cardiocirculatory' structure or function that is present since birth, even though it may be discovered later.[1] CHDs remains the leading cause of death in children with malformation.[2] Incidence of CHDs being 8 per 1000 live births and is the most common severe congenital abnormality. With the currently available treatment modalities over 75% of infants born with critical heart disease can survive beyond the first year of life and many can lead a near normal life thereafter.[3] The clinical profile may be studied under two broad headings cyanotic and acyanotic heart disease. Clinical presentations vary through these groups. Further, few lesions are particularly common in particular age and sex groups. Physical diagnosis consists of synthesis of information from FIVE sources: physical appearance,

arterial pulse, JVP, precordial examination and ausculatation.[4]

With newer diagnostic modalities including echocardiography, it is now considered as near confirmatory in diagnosing most of the congenital heart lesions.[1] X ray chest and ECG are complimentary to echocardiography. It needs to be ascertained as to what would be the diagnostic reliability of clinical diagnosis versus Echocardiography diagnosis.

The aim and objectives of the study are to study the correlation between physical diagnosis [clinical examinations/ chest X-ray/ Electrocardiogram (ECG)] and echocardiography diagnosis; to study the clinical profile including modes of presentations of congenital heart disease in children; to study the correlates of different congenital heart diseases and to study the common presenting symptoms and signs in children with congenital heart diseases.

METHODS

This study was conducted in the Department of Paediatrics at the Katuri Medical College and General Hospital, Guntur. 65 patients from the defined age groups in both sexes were studied during the study period of 2 years. The study design is exploratory in nature, with a proforma has been designed to study patients suspected with features of congenital heart diseases. All patients suspected to have CHDs were studied thoroughly by taking detailed history, thorough physical examination and all the cases were investigated by taking X-ray, ECG and ECHO. All the cases suspected to have CHDs were evaluated by applying by Alexander Nada's criteria. One major/ 2 minor criteria are essential for diagnosis of heart diseases.

Major criteria

- Systolic murmur grade III or more especially with thrill
- Diastolic murmur
- Cyanosis
- CCF.

Minor criteria

- Systolic murmur less than grade III
- Abnormal S2
- Abnormal ECG
- Abnormal X-ray
- Abnormal BP.

The following features will further help to identify CHD:

- Murmurs of obstructive and regurgitant lesions should be audible immediately at birth whereas murmurs of left to right shunt tend to appear later.
- Murmur presenting within the first year of life is a strong pointer towards a congenital lesion.
- Murmurs of congenital lesions tend to be parasternal rather than apical.

Inclusion criteria

- Features suggestive of CHD in the age group of 0 month to 12 years.

Exclusion criteria

- Age greater than 12 years of age
- Features not suggestive of CHD
- Innocent murmurs (functional).

As per objectives of the study, three major groups of data were collected:

- Clinico echo correlation data
- Modes of presentations of congenital heart disease in children and
- Clinical data

History and physical examination revealed a provisional clinical impression and subsequently patient was subjected to routine test include ECG and roentgenographic studies. After initial evaluation, the final diagnosis was assigned and designated termed clinical diagnosis.

Socio-demographic data were recorded and included age/ sex/ area of residence/ religion/ parental literacy and age. Occupation of head of the family/ per-capita income/ social class. Considering the referral pattern of the institute, the demography varied widely hence no single socio-economic scale was considered perfect. For instance, Kuppuswami (modified) scale adequately measured the socio-economic status in the community.

Correlation data was obtained after assigning a final clinical diagnosis to each patient and then only subjecting them to echocardiographic studies.

Echocardiographic evaluation was done in all the cases. All the CHDs were evaluated by 2D transthoracic echocardiography and colour Doppler to assess the anatomical, physiological nature of the CHDs. The clinico echo correlation was obtained and results analysed.

Statistical analysis

Include calculation of rates, ratios, chi-square tests of significance, student's 't' test, and analysis of variants test (ANOVA) for significance. All the statistical operations were performed through SPSS for Windows, Version 21.0 (Statistical Presentation System Software), SPSS Inc.

RESULTS

4145 cases were admitted during the study period of 2 years in the Department of Paediatrics, Katuri Medical College and Hospital, Guntur in the age group of 0-12 years.

During the above period 65 cases of CHDs were selected, after satisfying the inclusion criteria and exclusion criteria.

Table 1: Hospital incidence of CHDs.

Total no. of admissions (during study period)	4,145
No. of cases of CHD	65
Percentage of CHDs	1.56

From Table 1, among the 4145 cases admitted, 65 cases were found to have CHDs, comprising1.56%.

Table 2: Distribution of CHDs by major types.

Type	No. of cases	Percentage
ACHD	55	84.6
CCHD	10	15.4
Total	65	100

Chi-square value-62.31; p<0.001-HS

From Table 2 analysis of 65 cases of CHDs, it was observed that ACHDs comprising of 55 cases and CCHDs comprising of 10 cases.

Among the CHDs the incidence of ACHDs were more than the CCHDs comprising of 84.6% and 15.4% respectively.

Table 3: Distribution of CHDs by age groups.

		Frequency	Percent
Age groups	Below 1 year	22	33.84
	1-5 years	23	35.38
	>5 years	20	30.7
	Total	65	100.0

Chi-square=0.40; p=0.819 (NS)

From Table 3 analysis of presentation of CHDs, by considering the age 22 cases were diagnosed below 1 year of age, 23 cases were diagnosed between 1-5 years of age and the remaining 20 cases were diagnosed after 5 years of age. Nearly 1/3rd of cases were diagnosed before 1 year of age because of early onset of symptoms after decrease in the pulmonary pressures and establishing the left to right shunt.

From Table 4 analysis, it was observed that total 22 cases were symptomatic before 1 year of age mainly VSD and PDA.

The symptoms in both the cases must be due to early establishment of left to right shunts. In ASD all the cases were symptomatic only after 1 year of age because of lack of acceleration of blood in the pulmonary artery.

In the other heart diseases both ACHD and CCHD were symptomatic in various age groups, which cannot be considered because the incidence is only less in the present study.

Sex incidence of CHDs showed that the incidence is more in male children comprising of 61.54% as against the incidence in the female was 38.46% (25).

The sex ratio from male to female is 1.6:1 (Table 5).

The mean age of presentation in male children is 3.92 years, as against the mean age of presentation in female is 4.04 years.

With standard deviations 4.04 and 4.15 respectively. The total average in both sexes was 3.99 with standard deviation 4.07 (Table 6a).

It was observed that the mean age of presentation of ACHD was 3.58, with SD of 3.76, as against CCHD the mean age was 6.29, with SD 5.15.

For ACHDs the mean age for presentation was less than the CCHDs because of early establishment of left to right shunt.

The mean age for all the heart diseases was 3.99 with SD of 4.07 (Table 6b).

AREA wise analysis of CHDs 38 cases were from rural areas comprising of 58.46% and 27 cases from urban areas comprising 41.54%.

The difference in increased incidence of CHDs in rural areas must be studied in detail whether the consanguinity or other factors might be playing a role (Table 7).

It was observed 6 cases with CHDs were born to mothers of less than 20 years 34 cases were born to mothers between 21-30 years and 25 children with CHDs were born to mothers above 30 years (Table 8).

Children with CHDs 48 children were born to non-consanguineous parents comprising of 73.8% whereas 17 children were born to mothers with consanguinity comprising of 26.2%.

Among the 17 children born to consanguineous parents, 15 cases had the history of born to mothers with first- and second-degree consanguinity (Table 9).

Table 4: Distribution of CHDs (individual) by age groups.

ECHO		Age			
		<1 year	**1-5 year**	**>5 year**	**Total**
VSD	Frequency	8	10	4	22
	% within defect	36.36	45.45	18.18	100.0
	% within age	36.4	45.45	19.0	33.8
ASD	Frequency	2	3	3	8
	% within defect	25	37.5	37.5	100.0
	% within age	9.09	15.0	16.7	10.0
PDA	Frequency	4	3	2	9
	% within defect	44.4	33.3	22.2	100.0
	% within age	18.2	15.0	11.1	15.0
COA	Frequency	1	1		2
	% within defect	50.0	50.0		100.0
	% within age	4.5	5.0		3.3
TOF	Frequency	1	2	3	6
	% within defect	16.7	33.3	50.0	100.0
	% within age	4.5	10.0	16.7	10.0
PS	Frequency		1	2	3
	% within defect		33.3	66.7	100.0
	% within age		5.0	11.1	5.0
AS	Frequency			2	2
	% within defect			100.0	100.0
	% within age			11.1	3.3
VSD + ASD	Frequency	2			2
	% within defect	100.0			100.0
	% within age	9.1			3.3
VSD + PS	Frequency	1			1
	% within defect	100.0			100.0
	% within age	4.5			1.7
Common atrium	Frequency		1	1	2
	% within defect		50.0	50.0	100.0
	% within age		5.0	5.6	3.3
PAPVC + ASD	Frequency	1			1
	% within defect	100.0			100.0
	% within age	4.5			1.7
AS + COA + AR	Frequency			1	1
	% within defect			100.0	100.0
	% within age			5.6	1.7
VSD + AR	Frequency		1		1
	% within defect		100.0		100.0
	% within age		5.0		1.7
Dextrocardia + inverted GV	Frequency	1			1
	% within defect	100.0			100.0
	% within age	4.5			1.7
TGV+VSD+PDA	Frequency	1	1		2
	% within defect	50.0	50.0		100.0
	% within age	4.5	5.0		3.3
TAPVC + ASD	Frequency	1			1
	% within defect	100.0			100.0
	% within age	4.5			1.7
Total	Frequency	22	22	21	65
	% within defect	36.7	33.3	30.0	100.0
	% within age	100.0	100.0	100.0	100.0

Table 10 showed that 53 children with CHDs were not having any family history of heart diseases comprising of 81.53%. Only 12 children with CHDs, were having positive family history of CHDs comprising 18.46%.

Table 5: Distribution of CHDs by sex ratio.

		Frequency	Percent
Sex	Male	40	61.54
	Female	25	38.46
	Total	65	100.0

Chi-square =6.92; p<0.01(HS)

Table 6a: Distribution of CHDs by mean age and by age and sex- mean ages of CHDs by gender.

	N	Minimum	Maximum	Mean age	SD
Male	40	0	12 year	3.92	4.04
Female	25	0	12 year	4.04	4.15
Total	60	0	12 year	3.99	4.07

Table 6b: Distribution of CHDs by mean age and by age and sex- mean ages of major groups in CHDs.

ACHD-CCHD	No. of cases	Mean age	SD
ACHD	55	3.58	3.76
CCHD	10	6.29	5.15
Total	65	3.99	4.07

Table 7: Distribution of CHDs by area.

		Frequency	Percent
Area	Urban	27	41.54
	Rural	38	58.46
	Total	65	100.0

Table 8: Distribution of the CHDs by maternal age.

		Frequency	Percent
Age in years	Less than 20	6	9.23
	21-30	34	52-3
	Above 30	25	36-9
	Total	65	100.0

Chi-square-28.29; p<0.01: HS

Table 9: Distribution of the CHDs by consanguineous marriage.

		Frequency	Percent
Type of marriage	Non-consanguineous	48	73.8
	1 grade	7	10.7
	2 grade	8	12.3
	3 grade	2	3.07
	Total	65	100.0

Chi-square-29.57; p<0.01 (HS)

Table 10: Distribution of CHDs by family history.

		Frequency	Percent
Family history of CHDs	Absent	53	81.53
	Present	12	18.46
	Total	65	100.0

Chi-square-51.72; p <0.01 (HS)

Table 11: Distribution of the CHD by extra cardiac malformations.

		Frequency	Percent
ECA	Absent	59	90.76
	Present	6	9.23
	Total	65	100.0

Chi-square-86.43; p<0.01 (HS)

Morphological and anatomical abnormalities were noted only in 6 out of 65 cases were having morphological abnormalities, and majority of 59 children were not having any morphological abnormality (Table 11).

Table 12: Distribution of CHDs by modes of presentation.

Complaints	No. of cases	Percent involvement of (within all CHD)
Chest retraction	37	56.9
Cough and breathlessness	23	35.4
Breathlessness*	25	38.46
Chest pain	5	7.7
Cyanosis*	9	13.8
Cyanotic spells*	1	1.53
Feeding difficulty	18	27.7
Edema	5	7.7
Fever	21	32.3
FTT	22	33.9
Pulsation in the chest	16	24.6

Analysis of symptomatology of various CHDs, of both ACHDs and CCHDs, the common symptoms were breathlessness, cough and breathlessness, feeding failure, and failure to thrive. The major signs were chest retractions, pulsations of the chest, and fever. The less common signs were cyanosis, cyanotic spell, edema and chest pain (Table 12).

Table 13: Distribution of CHDs by past symptomology.

Type	RRTI	CCF
ACHD (n=55)	21	7
CCHD (n=10)	0	0
Total (n=65)	21	7
Total percentage	32.3	10.7

It was observed that RRTIs (>10 below 1 year and >6 above 1 year) and CCF were noted in 21 (32.3%) cases and 7 (10.7%) cases respectively out of 55 acyanotic heart diseases cases, but no case of CCHD was associated with either RRTIs or CCF (Table 13).

Table 14 showed the various radiological features of CHDs both ACHDs and CCHDs. Normal sized heart with normal vascularity of lung was seen in 14 cases out

of 65. The rest of the 51 cases showed various radiological features depending on type of lesion.

Table 14: Distribution of CHDs by X-ray findings.

	No. of cases	Percent
Normal	14	21.5
Cardiomegaly	51	78.5
Cardiomegaly + Plethora (ASD, VSD, PDA, TGATAPVC)	31	47.69
Cardiomegaly + Oligemia	1	1.53
Cardiomegaly + predominant right ventricle (TAPVC, ASD)	7	10.76
Cardiomegaly + predominant left ventricle (VSD, PDA)	25	38.46
Normal cardia size + Oligemia	7	10.76
Dextrocardia	1	1.53
Left Lung agenesis	1	1.53

As seen in Table 15, 65 cases were evaluated by echocardiography, which is supposed to be the confirmatory investigation for heart diseases authors observed common acyanotic heart diseases, the incidence of various heart diseases were arranged in Table 15. Among the 65 cases VSD PDA and ASD were the common ACHDs comprising of 21(31.7%), 9 (15%) and, 8 (10%) cases respectively in order of incidence.

Table 15: Distribution of CHDs by echocardiography.

	No. of cases	Percent
VSD	21	31.7
ASD	8	10.0
PDA	9	15.0
COA	2	3.3
TOF	6	10.0
PS	3	5.0
AS	2	3.3
SV	1	1.7
VSD + ASD	2	3.3
VSD + PS	1	1.7
Common atrium	2	3.3
PAPVC + ASD	1	1.7
AS + COA + AR	1	1.7
VSD + AR	2	1.7
Dextrocardia + inverted GV	1	1.7
TGV + VSD + PDA	2	3.3
TAPVC	1	1.7
Total	65	100.0

Among the ACHDs VSD was the commonest CHD seen in children Among the cyanotic heart diseases TOF was the commonest CHD observed in the study comprising of 10% (6 cases). The other heart diseases were less common, so the commonest CHDs were observed both CCHD and ACHD were VSD, ASD, PDA and TOF comprising more than 75% of the cases.

DISCUSSION

The present study was undertaken to know the incidence of congenital heart diseases attending the Department of Paediatrics Katuri medical college and hospital, Guntur over a period of 2 years. All the cases were studied in detail by taking proper history clinical examination and routine investigations like X ray chest and ECG. All the cases were subjected for ECHO evaluation to identify the site of lesion, size of lesion, chamber enlargement, amount of blood flow to the lungs and associated cardiac anomalies.

During the study period of 2 years, four thousand one hundred and forty-five (4145) children (<12 years) attended the department of paediatrics out of which, 65 cases were found to have congenital heart diseases comprising of 1.56%. It may not reflect the true incidence of CHDs in children because it is a hospital-based study. To know the exact incidence of CHDs in the society a large based study over a period of time is required by using the uniform criteria.

The patients were studied in three broad areas include:

- Modes of presentations of congenital heart disease in children.
- Clinical data as defined by the objectives of the study and
- Clinico echo correlation data.

Socio-demographic data was generated studying 11 variables across all types of CHDs in the study, include age, sex, hospital incidence, area of residence, religion, maternal education, paternal education, maternal age, paternal age, social class and family size.

The incidence of CHDs in the patients attending the pediatric department was 1.56%, which is comparable with studies of others like Gupte S et al (1.1%), Bai S et al (1.7%), Gupta et al (2.3%).[5-7]

Relative frequencies of various CHDs in the present study in order of frequency were VSD (33.8%), PDA (15%), ASD (10%), TOF (10%), CoA (3.3%). The results of this study is comparable with study of others, especially with a large community based study of 8,15,669 subjects (Samanek et al).[8]

The incidence of PDA in present study was 15% which was in correlation with the studies of other authors like Wallopillaiet al and Shresta et al where the incidence was 14% and 11% respectively.[9,10]

Latha V et al observed in CCHDs, cyanosis in 100% of cases, clubbing in 100% cases, dyspnea (breathlessness) in 96% cases, squatting episodes in 44% cyanotic spells in 33.33% cases and respiratory tract infection in 11.11% cases.[11]

Statistically presence of cyanosis and cyanotic spell as a mode of presentation in significant in cases of CCHDs. Tank et al studied 147 subjects and deduced the symptomology of CHDs and found breathlessness to be present in 74.83% of all CHDs.[12]

However, interestingly 58% of all CHDs were found to be asymptomatic in a study conducted by Kasturi et al in Bombay perhaps due to increased cases of neonatal period detected by routine examination due to non-establishment of left to right shunts in ACHDs patients like ASD, VSD, PDA.[13]

Analysis of 65 cases in the present study it was observed that only 19 cases presented with CCF (31%), 13 cases with respiratory tract infection (20%). The presenting features of recurrent respiratory tract infections and CCF were noted in 31.67% which is statistically significant($p<0.009$), whereas Pai et al found CCF in 16% of cases which was low when compared with the present study.[14] Of the analysis of 65 cases extracardiac anomalies (ECA) were observed in 13.3% of cases include Anotia (VSD), CTEV (VSD), Polydactyly (common atrium), Downs syndrome (ASD and VSD), Cataract, suggestive of rubella syndrome (PDA) among others. Kasturi et al and Pai et al found ECA in 20% in 24% of cases respectively which were higher than the incidence of extra cardiac manifestations in present study.

All the 65 cases of CHDs were evaluated by ECG found abnormality in 73% of cases. The most common abnormalities noted were the presence of LAD/ LVH followed by RAD/ RVH. ECG was normal in 26.7% cases of CHDs. This distribution was found to be statistically significant. The common eight CHDs which comprise 80-85% of CHD are well illustrated by Kulkarni et al, includes VSD, ASD, PDA, AS, PS, CoA, TOF, TGA.[15] Shibata et al made observations and recorded 43% of accuracy of clinical diagnosis in CHDs.[16]

Mitchell SC et al correlated clinical diagnosis with echo diagnosis and found accurate correlation in only 44%.[17] However, this was derived only on clinical grounds without the aid of basic investigational tools like X-ray, ECG etc.

Onishiet al arrived at a 50% diagnostic accuracy in CHDs by only clinical means.[18] Pestana C et al studied the clinico echo correlation in CHDs in Mayo Clinic, Rochester, USA and were able to clock 80% accuracy.[19] The study was conducted by experienced paediatric cardiologist. Kleweret al in a study of 154 subjects derived a clinico-echo correlation of 81% in USA.[20]

Swenson et al categorized patients on basis of clinical acumen into no CHDs.[21] Possible and definite CHDs and of the 21 definite CHDs cases 19 correlated with echo. Among eight possible CHDs cases five correlated, hence taking the clinico echo correlation to (19/21+5/8=24/29)=82%.

Tandon R from AIIMS, New Delhi in a review of work from CHDs highlighted the fact that in spite of fully being aware that a 100% diagnosis in CHDs was not possible.[22] A reasonable accuracy of 80-85% is achievable with clinical history, examination, X-ray and ECG and must be tried.

Attenhofer C et al compared diagnostic accuracies of diagnosis in hands of pediatric cardiologist against echocardiography and found on accuracy of 70-97% (study in adult population).[23]

CONCLUSION

The incidence of CHDs was 1.56% of the patients attending the pediatric department. Infancy is the most common age of presentation of CHDs. Incidence of CHDs was more in Males than in females. ACHDs were far more than CCHDs and VSD was the commonest ACHDs. Among CCHDs, TOF is the most common. Incidence of CHDs were more in rural areas than urban areas. In evaluating a CHDs case consanguinity was observed as a predisposing factor for causing CHD in 26%. Negative family history of CHDs was noted in majority of cases and Positive family history was noted in few cases. ECA were noted in 9.23%. The most common presenting symptoms in CHDs were chest retraction, breathlessness. Among CCHDs, the common symptoms were cyanosis, chest retractions and breathlessness.

Nearly 31% of CHDs cases presented in CCF. One third of cases of CHDs presented with recurrent respiratory tract infection (RRTI). Chest X-ray is an important corroborative evidence in the diagnosis of CHDs and was abnormal in 80% of cases with presence of increased cardiothoracic ratio in 58.3% of all cases. ECG is another important invaluable tool and was abnormal in 73.3% of cases and majority of them had left axis deviation / left ventricular hypertrophy. 95% of the cases of CHDs, including both ACHDs and CCHDs which were diagnosed clinically include ASD, VSD, PDA, AS, PS, CoA, TOF, TGA. Most common clinically diagnosed CHD was VSD. Clinico echocardiographic correlation was accurate in 75% of cases and couldnot establish the clinical diagnosis in 25% of cases.

Clinico echo correlation was highest with isolated lesions like VSD, ASD and in cases with typical findings like CoA, PDA, AS, TOF and least with multiple lesions or atypical findings. Properly carried out clinical examination, X-ray and ECG evaluation are important tools in arriving at a near accurate diagnosis in CHDs, however before undertaking surgical intervention echocardiographic confirmation of diagnosis is required. X-ray chest and ECG are complimentary to the clinical examination.

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