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Original Research Article

Clinical profile and outcome of children with congenital heart diseases admitted with acute events in a paediatric tertiary care unit in North Karnataka

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ABSTRACT

Background: Congenital heart diseases are the predominant causes of paediatric morbidity and mortality. This study was done to know the clinical profile, various acute presentations, and risk factors for repeated hospitalizations and their outcome in children with congenital heart diseases.
Methods: This prospective observational study of children in the age group of 0 hour-12 years, who were previously diagnosed and or newly diagnosed with congenital heart diseases.
Results: A total of 102 children were present during the study period. The most common age group for congenital heart disease was infancy with 46.1% (47 cases) of children. Acyanotic heart diseases were observed in 73.5% (75 cases) of the total, followed by cyanotic CHD with 14.7% (15 cases), complex CHD with 9.8% (10 cases), and the least, valvular diseases with 2% (2 cases). The most usual presenting symptoms in children with CHDs were breathlessness with 66.7% (67 cases), followed by fever with 54.9% (56 cases) and cough with 48% (49 cases. In this study, 23.5% (24 cases) of the children were completely immunized. The protein-energy malnutrition was present in 51% of cases (52 cases). The 16.7% of them (17 cases) have got the motor developmental delay predominantly. The dysmorphic features were present in 19 cases (18.6%) of total cases. The chest X-ray findings were abnormal in 58.8% (60 cases). The most common risk factor(co-morbidity) in children with CHD was noticed to be anaemia in 43 cases (42.2%).
Conclusions: The infancy and early childhood are the most common age group for the presentation of CHDs. Most of them had the associated risk factors (co-morbid) like nutritional anaemia, pneumonia, and failure to thrive.

Keywords: Congenital heart disease, Children, Profile

INTRODUCTION

Congenital heart disease (CHD) comprises one of the major diseases in the paediatric age group. Congenital heart disease is the leading cause of morbidity and mortality in children.[1] CHD by definition is the structural abnormalities of the heart or intra-thoracic great vessels present since birth that is actually or potentially of functional significance regardless of the age of detection.[1] The reported prevalence of CHD ranges from 1.01 to 17.5 per 1000 live births according to various studies over the world.[2-4] In India, the incidence of CHD is 3.9/1000 live

births, as reported by Khalil et al in a hospital-based study.[5] In community-based studies from India observed the prevalence of CHD ranges from 0.8-5.2/1000 children.[5,6] Most of the CHD present in the age group of 1 year to 6 years and are associated with multiple risk factors. Children with congenital heart diseases present with generalized symptoms such as fever, cough, etc. such symptoms can often be misleading and often leads to a lot of misdiagnoses.[7]

METHODS

This study was a prospective observational study carried out in a tertiary care centre in north Karnataka (Dr. Bidari's Ashwini Institute of Child Health and Research centre) over 16 months (April 2017 to November 2018).

Objective

The objective of this study was to determine various clinical presentations and causative factors for hospitalization and various risk factors present during admission, the clinical course, and the outcome of these children.

Inclusion criteria

All children with CHD newly or previously were included. CHD with the following clinical basis (a) hurried breathing; (b) refusal of feeding; (c) excessive sweating; (d) cyanosis; (e) cough; (d) fever; and (e) decreased activity

Exclusion criteria

Children's with (a) post-operative cardiac cases; and (b) acquired heart diseases were excluded.

All the eligible children were screened for congenital heart disease and probable risk factors using history, physical examination, and investigations. The eligibility was cross-checked and 'confirmed' by the co-investigator and a third independent party/person, who is not a part of the study team and will be blinded to the study details.

All children, including neonates who have been detected to have congenital heart diseases, were studied. Various risk factors, associated comorbidities, reasons for current admission as well as previous admissions, current clinical course, and outcome have been studied. A sample size of the study was calculated with a 95% confidence level and margin of error of ±5%, a sample size of 65(~70) subjects will allow the study to determine the risk factors and outcome of children with CHD with finite population correction.

By using the formula,

$$n = \frac{Z^2 p(1-p)}{d^2}$$

where, Z =z statistic at 5% level of significance, d is margin of error, p is anticipated prevalence rate. However, a total of 102 children were studied during the study period and the results are as follows. Data entry and tabulation was done using Microsoft excel 2013 and analysis using SPSS 16. For qualitative data, frequency and percentages were estimated.

RESULTS

The total number of children admitted with CHD was 102. CHD in our study showed the highest prevalence 77.5% (79 cases) in the 1st year of life, followed by 31.4% (32 cases) of CHD's in the neonatal period, 16.7% (17 cases) in the age group of 1 year to 5 years and the least with 5.8% (6 cases) in the age group of 5 years to 12 years (Table 1). Males accounted for 69.6% (71) of the total cases as compared to 30.4% (31) of the females. The prevalence of CHD is 2 times more common in males as compared to females (Table 2).

Table 1: Table showing the age wise distribution of children with congenital heart diseases.

Age	Number	Percentage (%)
<1 month	32	31.4
1 month to 1 year	47	46.1
1-5 years	17	16.7
5-12 years	6	5.8
Total	102	100

Table 2: Sex distribution of children with congenital heart disease.

Sex	Number	Percentage (%)
Male	71	69.6
Females	31	30.4
Total	102	100

Acyanotic heart diseases accounted for 73.5% (75 cases), followed by cyanotic CHD with 14.7% (15 cases), complex CHD with 9.8% (10 cases), and the least, valvular diseases with 2% (2 cases) (Figure 1). VSD was the most common cyanotic CHD with 53.3% (40 cases), followed by ASD with 22.7% (17 cases), PDA with 21.4% (16 cases), and the lowest with bicuspid aortic valve and coarctation of aorta with 1.3% each (1 case each) (Table 3). Among cyanotic CHD, the incidence of TAPVC category was the highest with 33% (5 cases), followed by TOF with 20% (3 cases), PAPVC and TGA with 13% each (2 cases each) respectively while truncus arteriosus, Ebstein's anomaly and tricuspid atresia with 7% each (1 case each) respectively (Figure 2).

In complex congenital heart diseases, double outlet right ventricle is the most common complex CHD with 30% (3 cases), followed by VSD+PS physiology and combination of VSD, ASD, PDA with 20% each (2 cases each), followed by lowest with 1 case each of single ventricle

physiology, common AV valve defect and a case of myxoma of the right ventricle with ASD (Table 4).

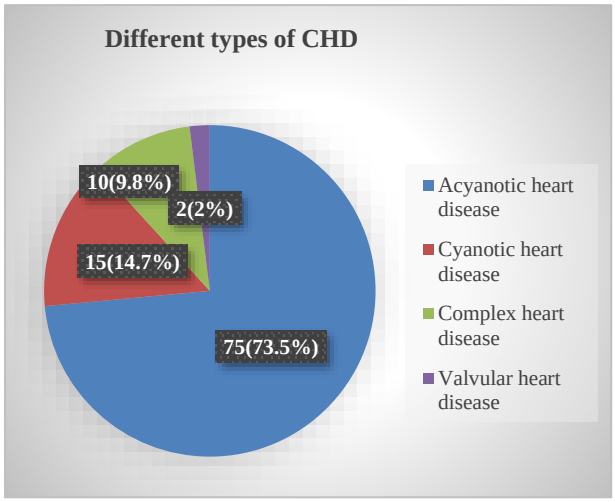

Figure 1: Different classes of CHD.

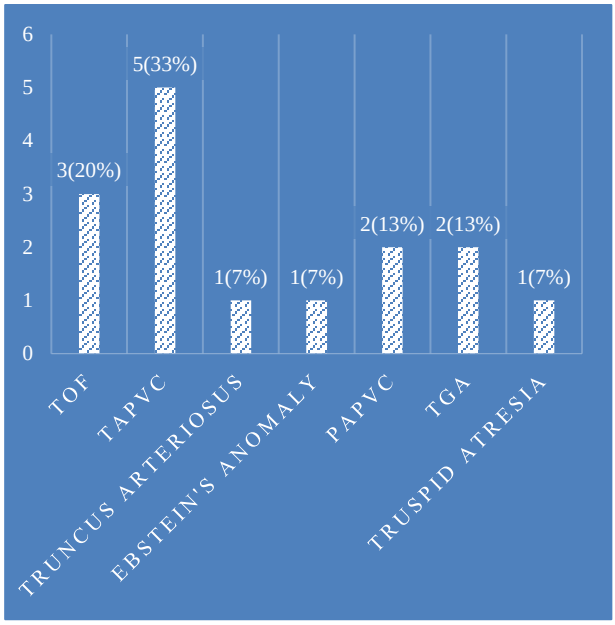

Figure 2: The incidents various types of cyanotic CHD.

Note: TAPVC- total anomalous pulmonary venous connection, PAPVC- partial anomalous pulmonary venous connection.

Table 3: Incidence of various types of acyanotic CHDs.

CHD	Number	Percentage (%)
VSD	40	53.3
ASD	17	22.7
PDA	16	21.4
Bicuspid aortic valve	1	1.3
Coarctation of aorta	1	1.3
Total	75	100

Note: VSD- ventricular septal defect, ASD-atrial septal defect, PDA- patent doctus arterious.

Table 4: Incidence of various types of complex CHDs.

CHD	Number	Percentage (%)
DORV	3	30
Single ventricle physiology	1	10
VSD+PS	2	20
VSD+ASD+PDA	2	20
Common AV valve	1	10
ASD+myxoma	1	10
Total	10	100

Note: DORV- double outlet right ventricle, VSD- ventricular septal defect, ASD-atrial septal defect, PDA- patent doctus arterious.

The most common presenting symptoms in children with CHDs were breathlessness with 66.7% (67 cases), followed by fever with 54.9% (56 cases), and cough with 48% (49 cases) (Figure 3). There was a constitution of other symptoms such as vomiting, rash, convulsions, and pin abdomen with 55.8% (57 cases). In our study, 23.5% (24) of children with CHD's were immunized completely for age as per IAP schedule, followed by partially immunized status in 14.7% of children (15 cases), whereas the majority of children have not started the immunization observed in 61.8% of children (63 cases) (Table 5). In our study, 17 children have got developmental delay accounting for 16.7% of total cases with most of them being isolated motor developmental delay while 2 cases accounted for global developmental delay.

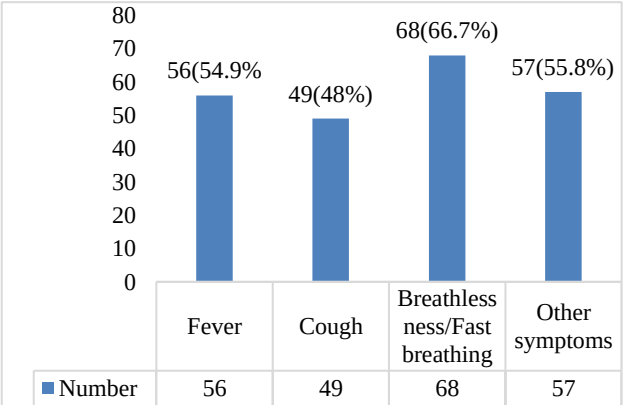

Figure 3: Most common presenting symptoms of CHDs.

Table 5: Immunization status in children with CHDs.

Immunisation	Number	Percentage (%)
Immunized	24	23.5
Partially immunized	15	14.7
Unimmunized/not started immunization	63	61.8
Total	102	100

In our study, most of the children with CHDs 51% (52 cases) had features of protein-energy malnutrition as compared to 49% (50 cases) of children who were normal for age. Of which, PEM grade 1 and grade 2 were noticed in 35.3% (36 cases) (Figure 4).

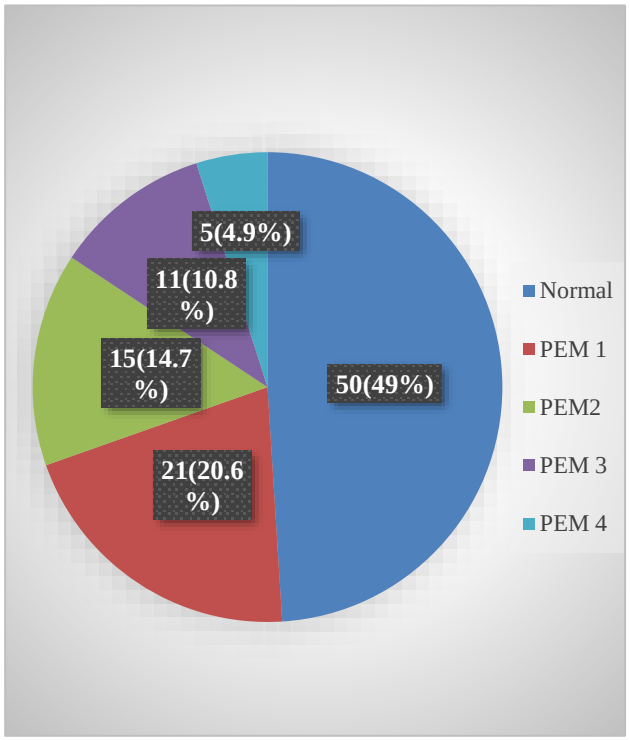

Figure 4: IAP classification of nutritional status in children with CHD.

In our study, the dysmorphic features were present in 19 cases (18.6%) with Down's syndrome phenotype was the commonest with 4.9% (5 cases), followed by one diagnosed case of Down's syndrome by karyotyping. Other dysmorphic findings such as microcephaly, low set ears, microtia, syndactyly, etc. were found in 13 cases (18.6%) (Figure 5). The chest X-ray findings were abnormal in 58.8% (60 cases) in our study. The most common chest X-ray findings include cardiomegaly in 24 cases (23.5%), features of consolidation (pneumonia) in 14 cases (13.7%), and a combination of both cardiomegaly and pneumonia in 16 cases (15.7%), and some special findings of the heart such as boot-shaped heart, egg on end appearance and figure of 8 have been noticed few cases (Table 6).

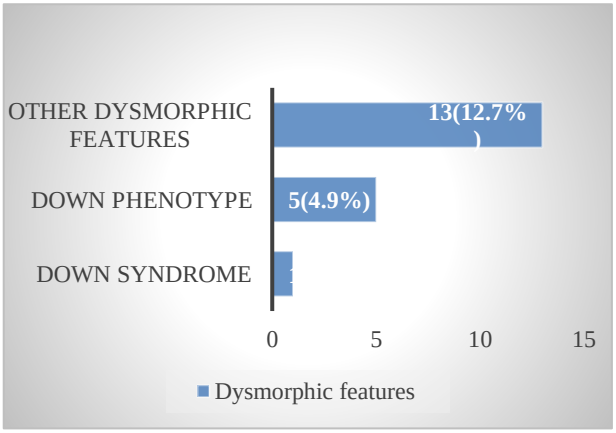

Figure 5: The presence of various dysmorphic features in children with CHD's.

Table 6: Various findings on chest X-ray in children with CHD's.

CHD	Findings								Total
	Car en sa boot	Cardi-omegaly	Egg on slide appearance	Emp-yema	Figure of 8	Pneumonia	Pneumonia, cardiomegaly	Situs inver-sus	
Acyaotic	0	17	0	1	0	12	14	1	45
Cyanotic	1	5	2	0	1	2	0	0	11
Complex	0	2	0	0	0	0	2	0	4
Total	1	24	2	1	1	14	16	1	60

The most common co-morbidity in children with CHD was noticed to be anaemia in 43 cases (42.2%), followed by pneumonia in 34 cases (33.3%), FTT in 19 cases (18.6%), CCF in 17 cases (16.7%), and PAH in 11 cases (10.8% and others with fewer cases include dehydration, septic shock, dys-electrolytemia, neonatal jaundice, tet spells, etc. (Table 7). Most of the children with CHD`s 68.6% (70 cases) in our study have improved with conservative treatment, 18 (17.6%) of them were referred to higher centre for surgery and 10 children (9.8%) have discontinued treatment and left against medical advice, the crude mortality rate in our study was 4% (4 cases expired) (Table 8).

Table 7: Table showing various risk factors in CHD.

Risk factors	Total	
	N	Percentage (%)
Anemia	43	42.2
Pneumonia	34	33.3
Family history of CHD	13	12.7

Continued.

Risk factors	Total	
	N	Percentage (%)
PAH	11	10.8
FTT	19	18.6
CCF	17	16.7
Sepsis with septic shock	7	6.9
Dehydration	6	5.9
Dyselectrolytemia	5	4.9
Sepsis	4	3.9
NNHB	4	3.9
Tet spell	4	3.9

Note: PAH- pulmonary hypertension, FTT- failure to thrive, CCF- congestive cardiac failure.

Table 8: Outcome of our study.

Outcome	Total	
	N	Percentage (%)
LAMA (left against medical advice)	10	9.8
Death	4	4
Improved	70	68.6
Referred for surgery	18	17.6
Total	102	100.0

DISCUSSION

In this study, most children with CHD's presented during infancy with 77.5% as compared to the study done by Shah et al in which 55.9% of children with CHDs presented in the infancy period. CHD's were observed nearly twice more common in males with 69.6% than in females with 30.4%, similar observations noticed in the studies done by Arodiwe et al which had reported that out of the total 50 children with CHD's, 31 (62.0%) were males and 19 (38.0%) were females and a similar observation was noted in a study done by Begum et al in Assam.[8-10]

In our study, acyanotic CHD's were commoner (73.5%) than cyanotic CHD's (14.7%), this observation was similar to studies done by Shah et al in which acyanotic CHD's were 69% and cyanotic CHD's were 31%.[8] In another study by Harshangi et al, similar observations like 68% were acyanotic CHD's and 32% were cyanotic.[11] The most frequent acyanotic heart disease in our study was VSD (53%) followed by ASD (22%) and PDA (21%) both in the form of isolated or mixed forms causing severe pulmonary hypertension. This observation was similar to a study done by Arodiwe et al, in which, VSD was the commonest with 40%, followed by PDA with 18% and least with ASD (4%).[9]

The frequent cyanotic congenital heart disease was TAPVC group with 33% (5 cases). TAPVC and PAPVC accounted for 46% of the cases (7 cases), followed by TOF with 20% (3 cases), TGA with 13% (2 cases), least with Tricuspid atresia (7%), Ebstein's anomaly (7%), and Truncus arteriosus with 7%. This was a different observation from other studies done by Shah et al, in which the commonest among the cyanotic CHD's was TOF 13.1% whereas the TAPVC group accounted for 3.6%.[8] Another observation was noted from Harshinagi et al study in which 33.3% of the cyanotic CHD's were TOF, the next commonest was TGA 16.6% and the TAPVC group accounted for 11.1%.[11]

The incidence of complex congenital heart diseases in our study was more diverse and was more in number when compared to any other studies. Most common amongst the complex CHDs was DORV 30% (3 cases), next commonest was VSD+PS 20% (2 cases), VSD with ASD with PDA with 20% (2 cases) followed by lowest CHD`s such as single ventricle physiology, common AV valve and combination of ASD and atrial myxoma accounted for 10% each (1 case). Most of the children in our study have presented to us with breathlessness as a common complaint, the reason being either pneumonia, CCF, or a combination of both. Breathlessness was seen in 66.7% (68 cases) of children. The next common symptoms were fever with 54.9% (56 cases) and cough with 48% (49 cases). Breathlessness was also the commonest presenting complaint in studies done by Harshinagi et al study and Yun et al study.[7,11] 56% of cases have presented with breathlessness in Harshinagi et al study and 41% of cases were found to be presenting with breathlessness in Yun et al study.[7]

Another important factor studied here was immunization status. Only 23.5% (24) children were immunized completely as per the Indian academy of the paediatrics immunization schedule. 14.7% of children (15 cases) were incompletely immunized, whereas 61.8% (63 children) were not started immunization at all. The study done by Harshinagi et al showed 30% incomplete immunization status.[11] Children with CHD's are very often deprived of

their basic physical needs such as immunization due to multiple reasons like recurrent chest infections, poor weight gain, etc. Children with CHD's are more prone to malnutrition thereby leading to delayed physical/motor developmental milestones. Our study has observed that 17 cases (16.7%) of them had delayed motor development except for 2 cases with global developmental delay, these results were similar to other studies. A study was done by Latha et al has reported 25% of children with developmental delay, whereas the study by Shah et al have reported a large percentage of children with developmental delay accounting for 86.9% cases (73 cases).[8,12]

In our study, most of the children had protein-energy malnutrition as per the IAP classification of PEM and about 51% of children (52 children) with CHD's have got features of protein-energy malnutrition. The finding of PEM in our study were comparable to other studies. Harshangi et al has shown the incidence of malnutrition to be 40% in their study as failure to thrive, Shah et al have reported 11.9% of children in their study to have presented with FTT, Ijeoma et al have reported maximum prevalence of malnutrition in children with CHD's accounting to 92% of children which was more than any other study.[8,9,11] The study by Ijeoma et al has also concluded that Children with CHD develop severe malnutrition and growth failure. The significant contributing factors are mean age at presentation and age-appropriate dietary adequacy. The causes could be increased basal metabolism due to the disease, recurrent infections, decreased nutritional intake and chronic hypoxia in case of cyanotic heart diseases and another reason could be due to poverty in developing countries.

The presence of other morphological and systemic feature in our study was minimal when compared to other studies, we had a genetically proven case of Down's syndrome and 5 children with clinical features of Down's syndrome and there were other features such as cleft palate, micrognathia, low set ears, polydactyly, microcephaly, microtia, etc. Our study noticed 28.7% of children (18 cases) as above features. Other studies done by Harshangi et al have reported more associated morphological and systemic involvement in their studies 41.6% (5 cases) of Down's syndrome and other features such as polydactyly to be 42.8% (3 cases), another 42.8% (3 cases) with CTEV and 14.28% (1 case) with a webbed neck.[11] Prevalence of Down's phenotype and the syndrome were contributing to 6% (6 cases). In our study, other features such as polydactyly, microtia, etc, contributed to 28.7% of cases. More prevalence of dysmorphism was seen in the other study when compared to our study.

The chest X-ray findings were abnormal in 58.8% (60 cases) in our study. The most common chest X-ray findings include cardiomegaly in 24 cases (23.5%), features of consolidation (pneumonia) in 14 cases (13.7%), and a combination of both cardiomegaly and pneumonia in 16 cases (15.7%), and some special findings of the heart such as boot-shaped heart, egg on end appearance and figure of 8 have been noticed few cases. These findings were similar to Harshnagi et al study, which had 29 cases with cardiomegaly and pneumonia being the next commonest with 12 cases diagnosed based on chest X-ray.[11]

The most common comorbidity in children with CHD was noticed to be nutrional anaemia in 43 cases (42.2%), followed by pneumonia in 34 cases (33.3%), FTT in 19 cases (18.6%), CCF in 17 cases (16.7%), and PAH in 11 cases (10.8% and others with fewer cases include dehydration, septic shock, dys-electrolytemia, neonatal jaundice, tet spells, etc. This was comparable to the study done by Mukherjee et al, where they reported 24 of 51 children (47.06%) with iron deficiency anaemia and this group also had a higher prevalence of cyanotic spells.[13] A study by Esekowitz et al, has reported the prevalence of iron deficiency anaemia in 17% of all patients suffering from congestive heart failure in adults.[14]

The crude mortality rate in our study was less than in other studies. Mortality was 4% in our study whereas Yun et al have reported an 8.6% mortality rate.[7] Another study by Harshangi et al has reported 18% mortality in their study.[11] Reasons for mortality were also different in all the studies, the main cause for mortality in Yun et al study was pneumonia which accounted for 4.8% and the second cause was CCF which accounted for 2.8%.[7] Harshangi et al study has shown refractory CCF as the most common cause 66.6% (6 cases), the second common cause was complex CHD 22.2% (2 cases) and the third common cause was septicaemia with infective endocarditis 11.1% (1 case).[11]

In our study, 70% (70) of children have improved with conservative/medical management, and 17.6% of them (18) were referred to a higher centre for surgical intervention. 9.8% (10) of them have discontinued treatment and went home against medical advice due to various reasons such as unaffordability, poor prognosis, etc. The causes of death in our study were different in each one of them. One case had TOF with severe pulmonary atresia with cyanotic spell which developed severe hypoxia, the second one was severe pneumonia with a large VSD with CCF with acute fulminant myocarditis with cardiorespiratory failure, 3rd one with VSD with sepsis, AKI, VAP and last one with large ASD and septic shock with pulmonary haemorrhage with disseminated intravascular coagulation.

Limitations

The limitations of the study were some of the children could not receive proper treatment as they went LAMA due to financial issues and poor prognosis; proper documentation in children with previously diagnosed CHD`s was missing; ECG changes and axis deviation were not included; follow up of children who were referred to higher center for surgery could not be done due to high patient load; and this study being a prospective

observational study, follow up after discharge was not included.

CONCLUSION

Infancy and early childhood are the most common age group for the presentation of CHDs. Most of them had the associated risk factors (co-morbid) like nutritional anaemia, pneumonia, and failure to thrive.

Recommendations

The prevention of risk factors associated with congenital heart diseases, can be done by dietary counselling and appropriate nutritional intervention, regular immunization, and follow-up and safe hygienic environment are required to prevent recurrent infections and hospitalizations thereby reducing the morbidity and mortality in children.

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