Comorbidities with Congenital Heart Disease among Hospitalized Children in a Specialized Cardiac Hospital in Bangladesh

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Key word:
Congenital heart disease, Comorbidities, Down syndrome

Abstract:

Background: *Congenital heart diseases (CHD) when associated with any comorbid condition may complicate the outcome of management. So to find out and treat comorbidities before definite treatment is essential for better outcome.*

Methods: *This observational cross sectional study was conducted in the inpatient Department of Paediatric Cardiology at National Institute of Cardiovascular Diseases, Dhaka, during the period of January 2014 to January 2015 to identify the comorbid conditions of congenital heart disease.*

Results: *A total of 794 children with different types of congenital heart disease were enrolled during the study period. Among them, 48.6% were simple CHD and 51.4% were complex CHD. Among simple CHD, ventricular septal defect (VSD) was commonest (27.2%), Tetralogy of Fallot (TOF) occupied the second position (20.5%). One child presented with Ectopia cardis. Among complex CHD most common anomalies were VSD with either atrial septal defect (ASD) or patent ductus arteriosus (PDA) or valvular lesion. Next common anomalies were TOF with PDA or ASD &/or pulmonary valvular abnormalities. Double outlet of right ventricle (DORV) with shunt and transposition of great arteries (TGA) with shunt also occur frequently. AV canal defect with or without valvular lesion presented in significant number. Among them 616 (77.6%) had different types of comorbid conditions. Among comorbidities, respiratory and acquired cardiac comorbidities were common (37.9% & 34.1% respectively). Other congenital or genetic comorbidities were in 11.2%. Multisystem involvement was in 9.8% cases.*

Conclusion: *Pneumonia and heart failure were the most frequent comorbid condition among both simple and complex congenital heart disease. They were common among acyanotic heart disease. But cyanotic spell, acute stroke syndrome and brain abscess were common among cyanotic heart disease. Among genetic comorbidities Down's syndrome occupied the major part.*

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Introduction:

Congenital heart disease is the most common cause of major congenital anomalies, representing a major global health problem. Twenty eight percent of all major congenital anomalies consist of heart defect.[1] Despite remarkable progress in clinical care for affected individuals, CHD remains the leading cause of infant mortality among birth defects.[2] For those that survive infancy, there is a high rate of comorbidities, both cardiac and extra cardiac, and expected lifespan is still diminished.[3] These issues have become increasingly important, as the number of adults with CHD now exceeds the number of children with CHD.[4] Between 25% to 30% of children with CHD have some form of additional congenital lesion, a comorbidity or structural extra cardiac anomaly (ECA) that may or may not be immediately apparent. However, comorbidity may have an important, even crucial, bearing on the course and outcome of the management of a child with a congenital heart lesion.[5] It is known that in recent years there has been an increase in the number of surgery on correction of congenital heart defects in children

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during the first years of life due to advances in Pediatric Cardiac Surgery and it is associated with the expansion of early surgical care.[6,7] The state of the operated child, as well as the duration of rehabilitation therapy, is not only determined by the peculiarities of cardio-vascular system, the volume and specific features of interven-tion, but by the presence of concomitant pathology. So, now a day, a great attention is paid to comorbid diseases in association with congenital heart disease.[8] There are insufficient studies available regarding children with congenital heart defect with other comorbid conditions in our Country. National Institute of Cardiovascular disease (NICVD), Dhaka provides the highest service to the patients with CHD in Bangladesh. So this study was designed to observe the situation of CHD and their comorbid conditions in this hospital where patients are being referred from different corners of the country, and it will provide the exact presentation of comorbidities among children with CHD in our country.

Methods:

This observational cross sectional study was conducted in the inpatient Department of Paediatric Cardiology at NICVD, Dhàkà, during the period of January 2014 to January 2015. All children aged < 15 years with known CHD or suspected to have heart disease who were admitted in this hospital due to different co-morbid conditions were enrolled in this study. But the children with acquired heart disease like rheumatic valvular disease or viral myocarditis or isolated pericardial effusion with co-morbidities were excluded from this study. Samples were selected purposively. A total of 987 children were admitted during this period. All new and old cases were diagnosed by color Doppler echocardiography done by expert pediatric cardiologist in this hospital. Finally 794 children with different types of CHD were identified and enrolled in this study. After registration, a detailed history and clinical examinations were done to find out the comorbidities along with congenital heart disease. Then laboratory investigations, both routine and special were done to reach a final diagnosis. Comorbidities were categorized as congenital/genetic, respiratory, cardiac, gastrointestinal, renal, haematological, neurological, endocrine and multisystem comorbidities. Data were collected by using a semi structured questionnaire. Verbal consent was taken from patient and their guardian before registration. SPSS 11.5 were used for analysis the data.

Results:

A total of 794 children were studied. Among them 4.3% were neonates, 27.2% were infants and most of the children were in between 1-5 years (32.9%) and 5-10 years (27.8%) age group. Only 7.8% children were enrolled who reached puberty and adolescent in >10 years age group. Among studied children 51.3% were male and 48.7% were female. In this study, 386 (48.6%) children were presented with simple CHD having single defect and 408 (51.4%) children had complex CHD having multiple defects. Among simple CHD, VSD were commonest (29.0%), TOF occupied the second position (23.3%). Among valvular lesion (3.4%) bicuspid aortic valve (BAV) with aortic stenosis (AS) or aortic regurgitation (AR), congenital mitral stenosis (MS) or mitral regurgitation (MR), pulmonary stenosis (PS) & tricuspid regurgitation (TR) were present. There were 4.1% children had Coarctation of Aorta (CoA). One child presented with Ectopia cordis in this study.

Table-I

Distribution of different simple congenital heart disease (n=386).

CHD	Frequency	Percent
VSD	112	29.0
ASD	71	18.4
PDA	83	21.5
TOF	90	23.3
Valvular Lesion	13	3.4
CoA	16	4.1
Other (Ectopia Cordis)	1	0.3
Total	386	100.0

Among complex CHD multiple defect were identified. Most common anomalies were VSD with either ASD or PDA or any valvular lesion. Next common anomalies were TOF with PDA or ASD &/or pulmonary valvular abnormalities. DORV with shunt and TGA with shunt also occur frequently. A-V canal defect with or without valvular lesion occur in significant number.

Table-II

Distribution of different complex congenital heart disease (n=408).

Complex CHD	Frequency	Percent
VSD+ASD	49	12.0
VSD+PDA	34	8.3
VSD+ASD+PDA	24	5.9
VSD+Valvualr lesion	22	5.4
ASD+Valvular lesion	13	3.2
PDA+ valvular lesion	16	3.9
VSD+ASD/PDA+ valvular lesion	23	5.6
TOF + ASD	9	2.2
TOF+PDA	31	7.6
TOF+Hypoplastic PA	24	5.9
Single Ventricle	5	1.2
DORV+VSD/ASD/PDA	23	5.6
DORV+VSD/ASD/PDA+ Valvular lesion	8	2.0
DORV+ PS	18	4.4
TGA+VSD/ASD/PDA	30	7.4
TGA+VSD/ASD/PDA+ Valvular lesion	8	2.0
A-V Canal Defect	22	5.4
A-V Canal defect + Valvular lesion	6	1.5
Ebstein anomalies+ASD/PDA	13	3.2
EA + ASD/PDA+ valvular lesion	3	0.7
Truncus+ VSD/ASD/PDA	8	2.0
TA+ASD/VSD/PDA+ valvular lesion	3	0.7
TAPVC/PAPVC+ ASD	16	3.9
Total	408	100.0

Comorbidities:

Among 794 children with different type of CHD, 616 (77.6%) children were admitted due to some associated comorbid condition but 178 children had no comorbidities. These 178 children were admitted for preoperative investigation (Cardiac catheterization) or cardiac interventional purpose (device/coil closure of defect or valvuloplasty). Among comorbidities, respiratory and cardiac comorbidities were common (37.9% & 34.1% respectively). Congenital or genetic comorbidities were 11.2%. Multisystem involvement occurs in 9.8% cases.

Congenital/Genetic Comorbidities

Among congenital/Genetic abnormalities, Children with Down's syndrome were commonest (47.9%), next frequent abnormalities were congenital rubella syndrome (16.9%). Cleft lip & or Cleft palate were found in 11.3% cases. Turner, Noonan's, Edward & Marfans syndrome were identified, two in number of each syndrome. Five cases were identified with Hirschprung disease along with CHD. One Carvezal syndrome was diagnosed along with DCM with MVP with MR.

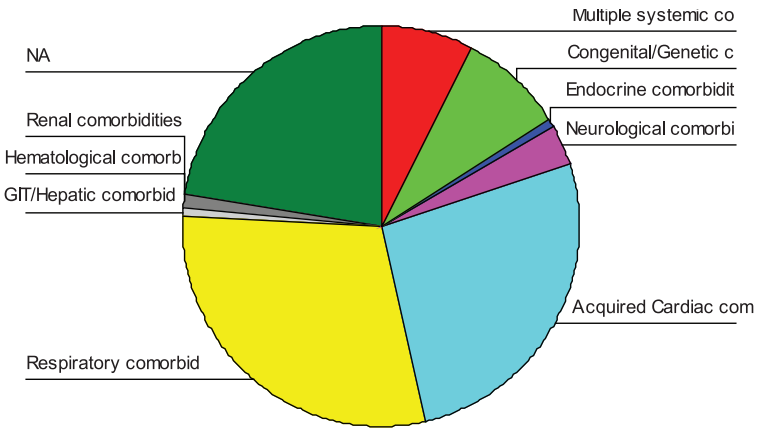

Fig.-1: *Comorbidities associated with CHD.*

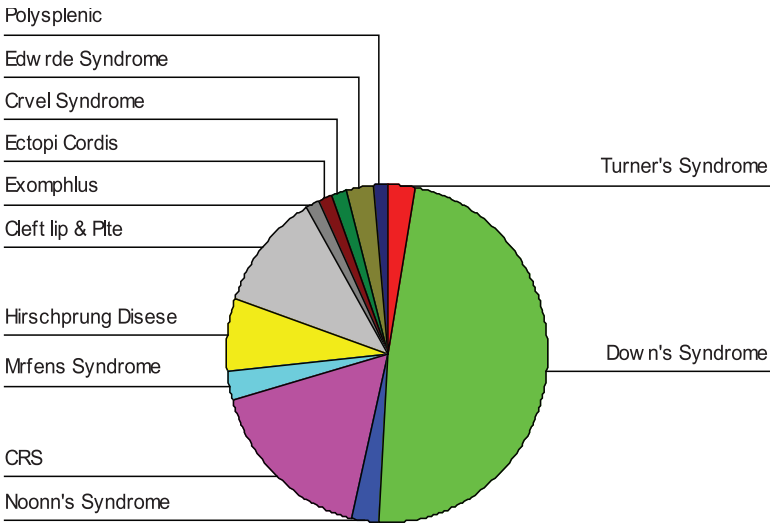

Fig.-2: *Congenital/Genetic abnormalities.*

Respiratory Comorbidities

In this study 309 (37.9%) children were admitted due to respiratory problems along with congenital heart diseases. Among them pneumonia accounted for 64.7%, Bronchiolitis 25.9% and pulmonary TB 2.1%. Others were Bronchial asthma and pleural effusion. Pneumonia & bronchiolitis were more commonly associated with complex CHD and bronchial asthma, pleural effusion and pulmonary TB were more common in simple CHD.

Cardiac comorbidities

Among cardiac comorbidities, 154 patients with simple CHD and 143 patients with complex CHD presented with different types of presentation. Heart failure, infective endocarditis and cyanotic spell were common presentation in both simple and complex CHD. Cyanotic spell were associated with RVOT obstruction. Patients present with shock were more common in complex CHD. Arrhythmia and pericardial effusion along with congenital heart disease also presented as a cardiac comorbidities.

Other comorbidities:

Among neurological comorbidities 24 patients in simple CHD and 12 patients in complex CHD presented with different presentation. Brain abscess and acute stroke syndrome were more common. Both were associated with TOF with or without other anomalies. Among other comorbidities, 6 patients presented with

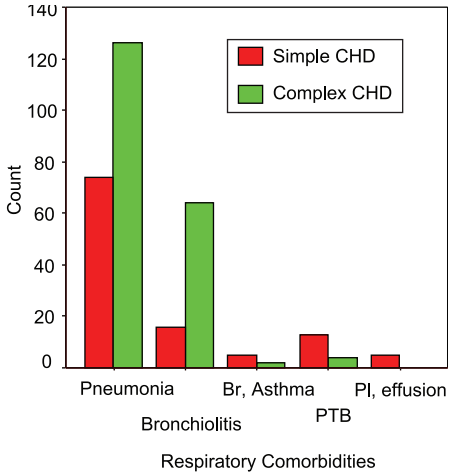

Fig.-3: *Respiratory comorbidities with CHD*

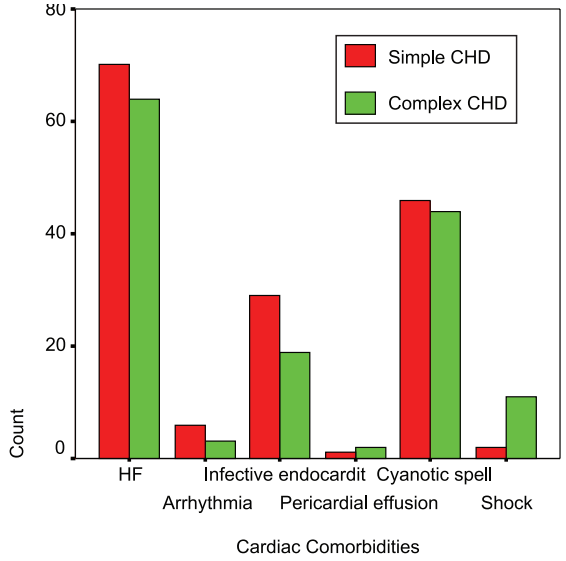

Fig.-4: *Cardiac comorbidities with CHD.*

hypothyroidism, 6 with hepatitis (Acute hepatitis 5 and CLD 1) 2 with chronic diarrhea, 31 patients with septicemia, 3 with aplastic anaemia, 4 with bleeding manifestations, 12 patients with kidney diseases (AKI 7, CKD 2 & AGN 3). Multisystem involvement occur in 9.8% cases in the form of concurrent involvement of two or more system, like respiratory (e.g. Pneumonia), cardiac (e.g. heart failure, arrhythmias, cardiogenic shock etc), Renal (e.g. AKI), hematological (e.g. anemia, bleeding manifestations or shock) or metabolic (hypoglycemia, hypocalcaemia, acidosis or alkalosis).

Discussion:

CHDs are an important subgroup of the congenital malformations since they carry a high risk of mortality and morbidity when it is associated with comorbid condition.[9] In this study most patients admitted with congenital heart defects were infants, followed by toddlers and preschoolers. Only 7.8% children were enrolled who reached puberty and adolescent in >10 years age group. Adrian Hruºcã et al. also showed that most of the children in his study were toddler to young children below 10 years of age.[10] However, it should be noted that, in developed countries, congenital heart defects are often caught during the neonatal phase or at birth.[11] But due to inadequate facilities, neonatal diagnosis could not be possible in our country. In this study, 48.6% children were presented with simple CHD and 51.4% children had complex CHD having multiple defects. Among simple CHD, VSD were commonest (29.0%), TOF occupied the second position (23.3%). There were 4.1% children had CoA. One child presented with Ectopia cordis in this study. Among complex CHD most common anomalies were VSD with either ASD or PDA or any valvular lesion. Next common anomalies were TOF with PDA or ASD &/or pulmonary valvular abnormalities. DORV and TGA with shunt also occur frequently. AV canal defect with or without valvular lesion occur in significant number. This result correlates with other studies, where they showed more than half of the children presented with complex CHD and VSD were the commonest among simple CHD.[12-14] But Islam MN et *al.* showed that, most (92%) CHD were simple.[15] Among simple CHD, VSD was the commonest followed by ASD then PDA & TOF. Among Complex CHD, TGA with shunt were common. Rashmi K & Shipra G also showed the similar results.[16] But our study differs from some other studies where they found ASD was the commonest lesion.[17-19] This difference in observation might be due to that Rahman et al. and Siddique et al. included many adult patients in their study. A significant proportion of VSD close spontaneously before adulthood and some untreated patients with large VSD die in childhood from heart failure. On the other hand ASD patients may remain asymptomatic in childhood and are diagnosed for the first time when they are adult. The study subjects of Fatema et al. were all newborn and many small sized VSD and most of the child with TOF may not manifest by that time.[19]

In this series 77.6% children were admitted due to some associated comorbid condition along with CHD. Among comorbidities, respiratory and cardiac comorbidities were common (37.9% & 34.1% respectively). Congenital or genetic comorbidities were 11.2%. Multisystem involvement occurred in 9.8% cases. Among congenital/Genetic abnormalities, children with Down's syndrome were commonest (47.9%), next frequent abnormalities were congenital rubella syndrome (16.9%). Cleft lip & or Cleft pàlàte were found 11.3%. Turner, Noonan's, Edward & Marfan's syndrome were also identified. Five cases were identified with Hirschprung disease along with CHD. One Carvezal syndrome was diagnosed along with DCM with MVP with MR. Extra cardiac abnormalities are frequently detected in children with CHD. Skeletal abnormalities, especially those of the hand and arm, are often associated with cardiac malformations. CHD may be a component of many specific syndromes and chromosomal disorders.[20] In a review of the population-based surveillance data from the Metropolitan Atlanta Congenital Defects Program, 12.3 percent of infants with CHD had a chromosomal abnormality. Infants with genetic disorders associated with cardiovascular malformations should be evaluated for possible cardiac abnormalities.[21] Hussain at *el.* showed the co-morbid problem in CHD among 224 children in Bangladesh in their study, 21.2% children had co-morbidities.[22] Among them 11.2% had genetic or syndromic conditions, of which mostly had Trisomy 21. The most frequent associated conditions were mental retardation, asthma, epilepsy and scoliosis. Islam MN et al. showed the associated non-cardiac anomalies.[15] Down's syndrome was noticed in 6% cases, polydactyly and syndactyly were detected in 4% newborn. Cleft lip with palate, cataract and renal anomaly were also found in 2% cases respectively. Comorbidities along with CHD in our study was also almost similar to other studies.[23,24]

Danford DA & McNamara DG showed in neonates with critical CHD may present during their birth, hospitalization with serious and life-threatening manifestations including shock, cyanosis, or respiratory signs and symptoms of pulmonary edema.[25] However, some infants with CHD may appear normal on routine examination and signs of critical CHD may not be apparent.

Adrian Hruºcã et al. study showed the largest part of congenital heart defects do not associate with other types of abnormalities, but when they do, the most frequent associations are made with: craniofacial deformities, bone and muscle defects, gastrointestinal, renal, genital abnormalities, hemangiomas, ophthalmological abnormalities and lung malformations.[10] As far as syndromes are concerned, the highest incidence rate is recorded for Down syndrome, followed by the Turner syndrome and the Noonan, Marfan's and Pierre Robin syndromes. Aneuploidies such as trisomy 21 (causing Down's syndrome) are strongly associated with CHD.[26] Our study findings are also consistent with this study.

Conclusion:

Pneumonia and heart failure were the most frequent comorbid conditions among both simple and complex congenital heart disease. These were common among acyanotic heart disease. But cyanotic spell, acute stroke syndrome and brain abscess were common among cyanotic heart disease. Among genetic comorbidities Down's syndrome occupied the major part.

Conflict of Interest - None.

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