Original Article

Pattern and Frequency of Congenital Heart Diseases in Children with Down's Syndrome

Farman Ullah Burki, Taj Muhammad and Fazal Rehman

ABSTRACT

Objective: Congenital heart disease is a significant morbidity in children with Down's syndrome, the most prevalent chromosomal anomaly. The aim of this study is to determine the frequency and pattern of congenial great diseases in children with Down's syndrome in District Head Quarter teaching Hospital D.I.Khan.

Study Design: Descriptive Hospital based study

Place and Duration of Study: This study was conducted at the Department of Pediatrics, DHQ Teaching Hospital D.I.Khan from July 2016 to June 2017.

Materials and Methods: Fifty Eight(58) phenotypically Down's syndrome children That Presented to pediatrics department age 2 days to 14 years were included in study. Children more than 14-years age, Children with Congenital heart Disease without Down's Syndrome and normal children were excluded from study. Detailed data analysis including History, physical examination was done and there were subjected to 2 dimensional echocardiography.

Results:-Congenital heart diseases were found in 34 (58%) out f 58 patients. Among the affected patients, 21 (61.76%) were male, 13(38.2%) were female with male to female ratio 1.5: 1. Atrioventricular canal defect was the most common heart disease 12 (35.7). Followed by Fallots Tetralogy in 8 patient (23.52%),Ventricular septal defect 5 patient (14.7%) while combination of ventricular septal defect with pulmonary stenosis and patent Ductus Arteriosisin 5(14.71%) and least common disease was Isolated patent Ductus Arteriosis 4 patients (11.7%).

Conclusion: Congenital heart diseases are common in Down's syndrome. The commonest one is Atrioventricular canal defect followed by Fallots Tetralogy. All children with Down's syndrome should have a cardiac evaluation at birth.

Key Words: Down's syndrome, Congenital Heart Disease, Atrio Ventricularcanal defect.

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INTRODUCTION

Down's syndrome is the most prevalent autosomal chromosomeanomaly in live born neonates. Down's syndrome is defined by Trisomy of chromosome 21 in 95% cases and translocation or mosaic in 5% cases.[1,2,3] It is the most common cause of moderatemental retardation and almost all patients have cognitive impairment with wide range of severity.[4]

Approximate incidence of Downs syndrome is 1 in 700-750 live birth while its prevalence rate is of 0.7 per 1000 births.[5,6]

Down's syndrome is responsible for 4.9% of all congenital heart Diseases.[7]

Department of Pediatrics Gomal Medical College D.I.Khan.

Correspondence: Dr: Farman Ullah Burk, Assistant professor of Pediatrics Gomal Medical College D.I.Khan.
Contact No: 0333-9186778
Email: ufarman310@gmail.com

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Down's Syndrome is characterized by Several clinical features including upslanting palpebral fissure, Epicanthic folds, depressed nasal bridge and cardiac anomalies as the most common congenital defects which occurs in 40-60% of patients and carries poor prognosis if not treated properly and timely.[2,8,9]

The most common congenial heart diseases in Down's syndrome include atrial ventricularseptal, Atrialseptal defect, Tetralogy of Fallot, and Atrio Ventricular canal defect which is regarded as a characteristic cardiac anomaly in such patients.[10,11] In Down's syndrome as Congenital heart diseases impose greater risk on patients, parents and health care system, knowledge of its epidemiology and prevalence in every geographical region is important as there is evidence that cardiovascular malformations may vary Based upon geography, ethnicity, and environmental factors.[9,11,12]

This study was conducted to evaluate frequency and pattern of congenial heart disease in Down's syndrome district head Quarter Teaching Hospital Dera Ismail Khan of Khyber Pakhtunkhwa province

MATERIALS AND METHODS

This Hospital based descriptive study was carried out in Department of pediatrics, District Headquarter Hospital

Dera Ismail Khan from July 2016 to June 2017 over a period of one year. 58 (fifty eight) Down's Syndrome children aged 2-days to 14 years, diagnosed on basis of phenotypic appearance were randomly selected over the above mentioned period, irrespective of clinical or X-rays, findings of CHD. Detailed history and examination was performed in all patients and all were subjected to 2 dimensional echocardiography. In addition routine tests like CBC X-rays chest and ECG after written consent was obtained from the parents. Children more than fourteen years age, Children with Congenital heart Disease without Down's syndrome and normal children without Clinical features of Down's Syndrome were excluded from study.

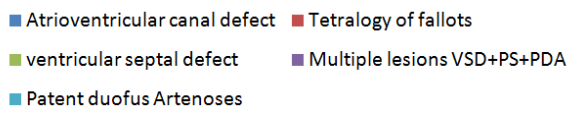
All children were diagnosed as Down's syndrome on basis of their phonotypic feature like mongoloid faces, depressed nasal bridge, Low set ears, Upslanting eyes with epicanthic folds, transvers single Palmar crease, hypotonia and delayed milestones.

RESULTS

In 58 patients with Down's syndrome, 37(63.8%) were male and 21(36.2%) ware female with male to female ratio of 1.6:1.The youngest patient was 15 days and oldest was 14 years old.Congenital heart diseases were found in 34 (58%) out of 58 patients. Among the affected patients, 21(61.76%) were male, 13 (38.2%) were female with male to female ratio of 1.5:1.The most common CHD was AV canal defect found in 12(35.7%) out of 34 patients followed by Tetralogy of Fallots in 8 (23.52%) out of 34 patients. Ventricular septal defect in 5% (14.5) and combination of Ventricular septal defect with pulmonary stenosis and Patent DuctusArteriosus in 5(14.5%) and of 34 patients,and the least common was Isolated Patent DuctusArteriosus in 4 (11.7%).

Table No.1: Type and Frequency of Congenital heart decrease in Children and Down's syndrome.

S.No	Types of cardiac defect	No	Percentage
1	Atrioventricular canal defect	12	35.7%
2	Tetralogy of fallots	8	23.52%
3	ventricular septal defect	5	14.7%
4	Multiple lesions VSD+PS+PDA	5	14.7%
5	Patent duofus Artenoses	4	11.4%
	Total	34	100%



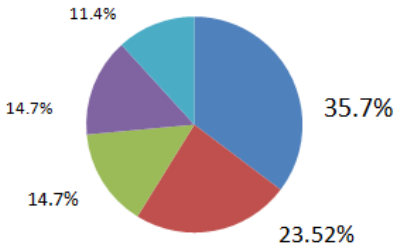


Figure No.1: Pie Diagrame Showing Percentages of Congenital Heart Diseases

DISCUSSION

The presence of high incidence of CHD in Down's syndrome is well known fact. Many authors and literatures have published figures on frequency of congenital heart disease in Down's syndrome. These figures very from 35 to 80% in different studies.[13-17] The frequency of CHD in our study is 58 percent which is quite comparable to these studies.

The most common type of congenital heart disease in our study is Arterio ventricular canal defect (35.7%). This finding is similar to Irani, Indian, Lebanese, Caucasians and Sudanies children, where Arterio ventricular canal defect is the most common CHD accounting for 35 to 50 percent.[17,18,20]. But is different from findings in Saudi Arabian, Chinese and some local children's [21,22].

The frequency of Arterio ventricular canal defect in our study is very close that reported in India (28%),Nigeria (50%).[11, 15] In our study Atrio ventricular canal defect was present in 35.7% children with Down'ssyndrome, this is similar to that reported by Ali et al (48.1%),[21] Freeman et al, (45%)[24], Ashraf etal (28%)[25], Asim etal (50%)[26], and Okeniyietal (54.7%)[27].but it was different from that reported by Khan et al (19.4%)[28].

The frequency of ventricular septal defect in Down's syndrome in our study was 5 (14.7%) and it was second most common congenital heart disease in our study. It is lower that reported by other workers[28.29].

In our study Patent Ductus Arteriosus was least common congenital heart disease that was present in 4 patient (11.5%) which is quite different from studies of other workers from Guatemala and Saudi Arabia where Patent Ductus Arteriosus is the most common congenital heart disease with frequency 28.6 and 47.8% respectively[30-31].

The most striking feature of our study was the high frequency of cyanotic congenital heart disease that is Fallots Tetralogy in Down's syndrome in 8 patient (23%), this frequency is quite high from that in Caucasians (6%),[24,28]Saudi Arabians (5.3%),[23] but was somewhat similar to that reported by other workers Lo NS et al (6%),[22]El Elmagrpy Z, el al(6.2%).[32]

Congenital Heart Diseases in Down's syndrometend to be single but may bemultiple. In Our study isolated

Congenital Heart Diseases represented 85% of all the Congenital Heart Diseases in Down's syndrome Compared with 65% in Libya, 80% in Guatemala, 74% in Mexico and 78% in Turkey.[32]

Our study has some limitations. This was single center study and not population based and diagnosis was made on clinical ground and no cytogenetic studies werecarried out. So we could not comment on a frequency of CHD in different chromosomal alterations of Down's Syndrome.

CONCLUSION

Congenital Heart disease is common in Down's Syndrome Children. The most common congenital heart disease in Down's Syndromeis AVSD and VSD respectively. All children with Down's Syndrome should have cardiac evaluation with Echocardiograph in first week of life.

Author's Contribution:

Concept & Design of Study:	Farman Ullah Burki
Drafting:	Taj Muhammad
Data Analysis:	Fazal Rehman
Revisiting Critically:	Farman Ullah Burki, Taj Muhammad
Final Approval of version:	Farman Ullah Burki

Conflict of Interest: The study has no conflict of interest to declare by any author.

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