

The Spectrum of Congenital Heart Diseases Operated at Shahid Gangalal National Heart Center in the last 5 Years.

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Abstract

Background and Aims: The spectrum of congenital heart diseases (CHD) presenting for surgery to any cardiac center varies depending upon the geographical location of the center. This study is aimed to reveal the spectrum of CHD operated at a tertiary level cardiac center of Nepal.

Methods: This is a retrospective study conducted at the Department of Cardiovascular Surgery at Shahid Gangalal National Heart Center (SGNHC), Kathmandu, over a period of five years, from April 14, 2015 to April 13, 2020. All patients with a confirmed diagnosis of congenital heart disease who had undergone palliative or definitive cardiac surgery were included.

Results: Out of the total 2698 patients, there were 1374 (50.93%) females. The total number of acyanotic congenital heart disease was 1919 (71.13%), and that of cyanotic was 779 (28.87%). Male to female ratio among acyanotic and cyanotic were 0.85:1 and 1.31:1 respectively. Among all CHDs, atrial septal defect (ASD) (38.13%) followed by ventricular septal defect (VSD) (21.68%) and atrioventricular septal defect (AVSD) (5.04%) respectively were the commonest acyanotic heart lesions. Considering only the pediatric population (age 0-18 years), the commonest was VSD (27.06%) followed by ASD (22.3%) respectively. The commonest cyanotic was tetralogy of Fallot (TOF) (14.15%) followed by double outlet right ventricle (DORV) (4.44%) and total anomalous pulmonary venous connection (TAPVC) (2.89%) respectively.

Conclusion: Female population was slightly more in number amid acyanotic lesion whereas male population was dominant in cyanotic lesion. The commonest acyanotic CHD was ASD and, TOF was the commonest cyanotic lesion.

Keywords: Atrial Septal Defect; Cyanotic and Acyanotic Congenital Heart Disease; Tetralogy of Fallot; Ventricular Septal Defect.

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Introduction

Congenital heart disease (CHD), in definition proposed by Mitchell et al, is "...a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance".¹ The global prevalence of CHD at birth, in 2017, is 1.8 cases per 100 live births.² Prevalence of CHD among school children in Kathmandu, Nepal is 1 per thousand and Atrial Septal Defect (ASD) was found to be the most common CHD.^{3,4}

Different genetic and environmental factors affect the spectrum and prevalence of CHD worldwide.⁵ Right ventricle outflow tract obstruction lesions were found more in number in Asian population than in European and American population.⁶ Population-based study regarding spectrum of CHD in Nepal is unavailable. The

hospitals-based study done at pediatric outpatient clinic had found Ventricular Septal Defect (VSD) the most common acyanotic and Tetralogy of Fallot (TOF) the most common cyanotic CHD.^{7,8} In our part of world, where the health care system is not optimized, early diagnosis and timely treatment of CHD, an important factor for reducing morbidity and mortality with CHD, is still uncommon.

To our knowledge, the spectrum of CHD undergoing surgery in any cardiac center of Nepal was not reported previously. The purpose of the study is to present a single-center evaluation of the spectrum of CHD operated at Shahid Gangalal National Heart Center (SGNHC), which is a tertiary level cardiac center performing yearly considerable portion of the total cardiac operations done in Nepal.

Considering the portion of cardiac operation done in this center, it can be said that the data represent the national level data till this date.

METHOD

This is a retrospective observational study conducted at the department of cardiovascular surgery at SGNHC, Kathmandu, Nepal, from April 14, 2015, to April 13, 2020. All patients with the confirmed diagnosis of CHD who had undergone definitive or palliative cardiac surgery were included. All age groups were included. Patients with multiple cardiac anomalies were also included. Patients with CHD associated with acquired heart disease were excluded.

Clinical data were reviewed from the patients' record file, intensive care unit record, perfusion sheet, and operation theater data. Consideration was given to the total number of CHD patients with age, sex and type of CHD. Age ranges were divided as neonates (0-28 days), infant (1 month to 1 year), toddler and preschool (above 1 year to 6 years), older children (above 6 years to 12 years), adolescent (above 12 years up-to 18 years) and adults (18 years and older). Cyanotic and acyanotic CHDs were classified according to standard practice.⁹

RESULTS

There were a total of 2698 Congenital heart disease (CHD) diagnosed patients who had undergone palliative or definitive surgical procedures during the study duration. The total number of males were 1324 and female were 1374. Male to female ratio among acyanotic and cyanotic were 0.85:1 and 1.31:1 respectively. The age ranged from 11 days of neonate to 62 years among operated patients for CHD. The baseline distribution of CHD presented for surgery is summarized in table no 1.

Table 1: Baseline distribution of CHD presented for surgery.

	Acyanotic	Cyanotic
Male	882	442
Female	1037	337
Total	1919	779
0-28 Days	2	32
1 Month-1 Year	215	135
1 yr-6 yrs	430	339
6 yrs-12 yrs	341	138
12 yrs-18 yrs	254	87
> 18 yrs	677	48

The commonest acyanotic CHDs were ASD followed by VSD and AVSD with 38.3%, 21.7%, and 5.04% respectively, among all CHDs. Among the pediatric age group (that is 0-18 years), VSD was the commonest acyanotic CHD (27.06%). ASD diagnosed after 18 years of age comprises 589 (21.8%) of all CHDs. There were two cases of supra-valvular aortic stenosis and one case of the dysplastic aortic valve. An eleven-day-old neonate with Coarctation of aorta was operated. The commonest age range for acyanotic operated cases was above 18 years of age (35.3%), followed by 1-6 years of age (22.4%). Age-wise distribution of acyanotic CHDs, along with total individual disease and percentage are shown in Table 2.

Table 2: Age-wise distribution of acyanotic CHDs along with total individual disease and percentage from all CHDs.

Diagnosis	0-28 Days	1 month-1 yr	1 yrs-6 yrs	6 yrs-12 yrs	12-18 yrs	>18 yrs	Total no of Individual	Percentage Among all CHD
Ventricular Septal Defect (VSD)		143	228	84	79	51	585	21.68
Atrial Septal Defect (ASD)		7	120	179	134	589	1029	38.13
Patent Ductus Arteriosus (PDA)		6	29	19	10	13	77	2.85
Atrioventricular Septal Defect (AVSD)		33	30	49	13	11	136	5.04
Coarctation of Aorta (COA)	1	7	10	4	6	7	35	1.29
Congenitally Corrected Transposition of the Great Arteries (CCTGA)/VSD		2	2	4			8	0.296
Aortopulmonary (AP) Window		2	1				3	0.11
Interrupted Aortic Arch (IAA)	1	1					2	0.074
VSD, Right Ventricular Outflow Tract Obstruction/Pulmonary Stenosis (RVOTO/PS)		7	3		12	6	28	1.03
Subaortic Membrane		2	1	1			4	0.14
Partial Anomalous Pulmonary Venous Connection (PAPVC)		2	2				4	0.14
Cortriatriatum		2	3				5	0.18
Dysplastic Aortic Valve			1				1	0.037
Supra Aortic, Aortic Stenosis (AS)		1		1			2	0.07
Total	2	215	430	341	254	677	1919	

In cyanotic heart disease, TOF was the commonest with a total of 382 (14.15%), followed by Double outlet right ventricle (DORV) 120 (4.44%) and Total Anomalous Pulmonary Venous Connection (TAPVC) 78 (2.89%). The commonest age range for presentation of cyanotic was 1-6 years of age (43.5%). Among all Cyanotic CHD, TOF was 49%. Age-wise distribution of cyanotic CHD, along with total of individual disease and percentage are presented in table 3.

Table 3: Age-wise distribution of cyanotic CHD, along with total of individual disease and percentage.

Diagnosis	0-28 Days	1 Month- 1 yr	1 yr- 6 yrs	6 yrs- 12 yrs	12-18 yrs	>18 yrs	Total of Individual Disease	Percentage Among all CHD
Tetralogy of Fallot (TOF)		17	212	72	55	26	382	14.15
Total Anomalous Pulmonary Venous Connection (TAPVC)	3	33	21	17	3	1	78	2.89
dextro-Transposition of the Great Arteries (d-TGA)	22	30	6	6			64	2.3
Tricuspid Atresia (TA)/VSD		13	13	13	1	1	41	1.51
Truncus Arterious		1					1	0.03
Pulmonary Atresia	7	7	7	1	1		23	0.85
Ebstein Anomaly		1	6	13	11	7	38	1.40
Double Inlet Left Ventricle (DILV)/DIRV/ Double-Outlet Left Ventricle (DOLV)		4	7	3	2	1	17	0.63
Double Outlet Right Ventricle (DORV)		25	61	9	14	11	120	4.44
VSD Critical RVOTO/PS		4	6	4		1	15	0.55
Total	32	135	339	138	87	48	779	

DISCUSSION

This is the first review on the spectrum of CHD operated in Nepal, though this is from a single cardiac center, which is basically one of the only two major cardiac centers in the country for 30 million populations.

In the study male and female patients were almost equally distributed. When we analyzed only the pediatric age group (0-18 years), there were a total of 1973 cases of CHD: male 1058 (53.6%) and female 915 (46.4%). The male predominance among pediatric age group in our study is similar to that done by Zahid et al. (study population included up to the age of 16 years) at Rehman Medical Institute, Pakistan where males were 57.7% and females 42.2%.¹⁰ The study done by Mir AB et al, at tertiary care center of north India (study population <14 years of age) showed male 60% and female 40% with male dominance.¹¹ In contradictory population based Chinese study (0-18 years included), about spectrum of CHD, the girl population (56.3%) dominated the male population (43.8%).¹²

Our study is a hospital-based study. When we consider only pediatric population (0-18 years) male dominance pattern may be due to our socio-economic status where male child is given more preference to female child. So, male patients were timely diagnosed and put to surgery. Maximum number of CHD among pediatric age (0 to 18 years) is operated between 1-6 years of age in both cyanotic (46.3%) and acyanotic (22.4%) CHD. The same was observed in the study done by Zahid et al.¹⁰

ASD was the most common CHD in this study with 38% of all CHDs. This result is consistent with the study conducted by Lakhotia et al. at the Institute of Medical Sciences, Banaras Hindu University, India where ASD comprised 40% of all CHD operated population, and it also included all age group.¹³

Among pediatric age group, VSD was the commonest type with 27% of CHD operated in our study which is consistent with the results

of study from Pakistan done by Zahid et al.¹⁰ In his study he included pediatric population (but up to the age of 16 years) and VSD was the commonest CHD operated with 33.8%. That VSD remains the commonest cardiac operation for pediatric population is supported by a publication of Hoffman et al.⁹

In our study, AVSD accounts for 5% of total CHD. Regarding AVSD, our number is equal to that of a study conducted in newborn echo evaluation at Oman with 5.9%.¹⁴ We included all form of AVSD in one category. Incidence of AVSD in our study was double that of an Indian study (2.7%),¹¹ which also did not categorize the type of AVSD. Pakistani study with prevalence of AVSD (10.3%),¹⁰ included only complete type of AVSD. In general, the frequency of AVSD in studies from developing countries is reported between 3.5 and 8.8%.¹⁵ There is high association of Down's Syndromes with AVSD. Nearly 30-40% of Down's syndrome babies have associated AVSD.¹⁶ We do not know the incidence of down's syndrome in Nepal. Each year babies born with Down's syndrome are in increasing trend throughout the world.

In our study, the total number of PDA was 71 (2.6%) among all age groups and 64 (3.2%) among pediatric age group. This is far less than in the study done by Zahid et al. where PDA was 23.0% among pediatric age group.¹⁰ Reason behind the low number of PDA closure surgically in our study was that, in the last 5 years, our cardiologists did significant number of device closure for PDA.¹⁷ Cardiologists, at our center are performing significant number of transcatheter closure of ASD as well as VSD.^{18,19} Overall, the impact of transcatheter closure of isolated cardiac lesion to the number of surgical closure in our center is not studied yet.

TOF being the commonest cyanotic occupied nearly 14.1% of all CHD and 49.3% of cyanotic CHD. This finding is comparable with the study (at surgical center with inclusion of 0-16 years) done by Zahid et al., where TOF comprised 51.7% among cyanotic CHD.¹⁰ The study

finding is similar to that of the study done at North India by Mir AB et al. (0-14 years) echocardiography based on suspected cardiac patient screening where the prevalence of TOF was 12.2% among all CHD.¹¹

In our study, the right-side obstruction lesion (TOF, PS) has higher prevalence than the left side obstruction (CoA and AS) which is similar to descriptions made by Jacob et al. in the study done at Hong Kong, explaining white children seem to have more left ventricular obstructive lesions, whereas Asian children have more right ventricular outflow lesion.⁶

After TOF, DORV was the second commonest CHD with 4.4% of all CHD in this study. In general, DORV affects between 1% to 3% of people born with CHD.²⁰ Akin to our finding Mir AB et al. found DORV, 3.2% of total CHD in his study in a tertiary cardiac center of north India.¹¹

Third commonest cyanotic CHD, in our study was TAPVC. TAPVC was 2.9% of all CHD. This finding is closer to that of a hospital-based study conducted at one of the children hospitals of Nepal by Shah GS et al., and TAPVC noted as the third commonest was 3.6%.²¹

In our series, d-TGA comprises the fourth commonest cyanotic at 2.4%, among all CHD. A study from north India noted 3.3% in the hospital-based (<14 years aged) screening suspected patient of CHD.¹¹ Overall prevalence of d-TGA accounts for 5–7% of all congenital heart defects (CHD) with a prevalence of 0.2 per 1000 live births.²² This suggests that our d-TGA born children may not have reached hospital for diagnosis and further management.

Limitation of the Study

This is a single-center hospital-based review. Therefore, this study does not provide information on the prevalence of heart diseases in the general population of Nepal. Furthermore, this is a retrospective study; so, review of the hospital data, possibility of missing data, incomplete information may be there. While comparing with different publications, we even had to compare with papers studying echocardiography-based publication, some papers had included their study population up to 14 years, and some had included echocardiography based up to 18 years of age. We could not include data regarding the prevalence of Dextrocardia, Bicuspid aortic valve, and syndromic conditions. We could not categorize certain diseases though necessary, like DORV, Single Ventricle physiology. We included pentalogy under TOF. There is no definite definition of complex congenital heart disease; we did not use this terminology in our study for categorization of disease.

CONCLUSION

The commonest acyanotic CHD was ASD and, TOF was the commonest cyanotic lesion. Among pediatric age group, VSD was the commonest type of CHD. The findings of the study on spectrum of congenital heart diseases diagnosed at the center for surgery during the last five years corresponds well to the findings reported in other South-East-Asian countries. The study gives us an overview of the pattern of distribution of diseases operated for congenital heart surgery at Shahid Gangaal National Heart Center, Kathmandu, Nepal.

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