

SPECTRUM OF CONGENITAL CYANOTIC HEART DISEASE IN KHYBER PAKHTOONKHTWA

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Contribution

All the authors contributed significantly to the research that resulted in the submitted manuscript.

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ABSTRACT

Objectives: To study the frequency and spectrum of congenital cyanotic heart disease among patients attending pediatric cardiology department for suspected cardiac problem.

Methodology: This was an observational study, conducted in Pediatric Cardiology Department Lady Reading Hospital Peshawar, from 1st September 2012 to 31st August 2013. Every patient with suspected cardiac problem was subjected to echocardiographic study to know frequency and spectrum of congenital cyanotic heart disease in these patient.

Results: Total number of patient studied with congenital heart disease (CHD) was 2342. Of them 434(18.53%) were found to have cyanotic congenital heart disease (CCHD). Of these CCHD patients males were 262(60.36%). Mean age was 34.42 ± 30.34 months (from 1 day to 18 years). Tetralogy of fallot (TOF) patients were 205(47.23%) which forms 8.8% of total CHD. Of these TOF patients 09 were already corrected. Transposition of great arteries (TGA) was found in 83(19.12%). Ebstein with cyanosis were found in 12(2.76%) patients. Double Outlet Right Ventricle (DORV) was found in 38 (8.75%). Total anomalous pulmonary venous drainage (TAPVD) patients were 27(6.22%). Of these supracardiac TAPVD were 15(3.45%), Cardiac TAPVD were 7(1.61%) and Infracardiac TAPVD were 5(1.15%). Pulmonary atresia with intact pulmonary septum was found in 15(3.4%). Single ventricle was found in 21 (4.84%) patients. Eisenmenger syndrome was found in 11(2.6%). Hypoplastic left heart was found in 14 (3.22%) and 8(1.84%) having truncus arteriosus.

Conclusion: About 18% of CHD in our population comprises of CCHD. TOF is the major CCHD in our population.

Key Words: Cyanotic Congenital Heart Disease (CCHD), Tetralogy of Fallot(TOF), Transposition of Great Arteries(TGA)

INTRODUCTION

Congenital heart disease (CHD) is a significant cause of morbidity and mortality amongst infants and children.¹ The optimal care of patients with CHD requires an organized Pediatric Cardiac Care program; the development of such programs requires adequate financial and human resources. Knowledge of the local spectrum of CHD provides the foundation for the rational allocation of healthcare resources. Spectrum of CHD differs in various geographical distribution.² Congenital heart disease (CHD) 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 defects.³ Prevalence of CHD varies widely among studies worldwide. The estimate of 8 per 1,000 live births is generally accepted as the best approximation.⁴ CHD, by definition, is present from birth.⁵ Over the past century, massive breakthroughs have been achieved in cardiovascular diagnostics and cardiothoracic surgery, leading to an increased survival of newborns with CHD. Consequently, more patients with CHD reach adulthood, creating a completely new and steadily growing patient population: patients with grown-up congenital heart disease (GUCH). The prevalence of CHD is estimated to be 4 per 1,000 adults.⁶ These patients with GUCH usually need long-term expert medical care and healthcare-related costs are often high.⁷ About 10 to 20 % of all the congenital heart disease are cyanotic congenital heart disease. In one study Cyanotic CHD comprises up to 25% of all causes of CHD.⁸ CCHD forms the major cause of morbidity and mortality in GUCH. Among the congenital cyanotic heart disease are Tetralogy of Fallot, Transposition of the great arteries, Total anomalous pulmonary venous return, Tricuspid atresia, Truncus arteriosus, Ebstein's anomaly (abnormal tricuspid valve), pulmonary atresia and hypoplastic left heart are the major ones. It is important to have reliable information about world wide CCHD birth prevalence because this may lead to better insight into its etiology. In addition, dedicated care could be better planned and provided after knowing the true prevalence of CCHD. The current study aims to determine the spectrum of cyanotic congenital Heart disease encountered at a referral hospital in Khyber Paktounkhwa; with a view to providing data that may contribute to the rational allocation of resources to Paediatric Cardiac Care locally. As there are a significant proportion of cases with complex cyanotic CHD, paediatric cardiologists should be familiar with the diagnosis and management of all these complex congenital malformations of the heart.

METHODOLOGY

This Echo based cross sectional study was conducted in Pediatric Cardiology Department of PGMI, LRH, Peshawar, a tertiary care hospital where patients are referred from all over Khyber Pakhtunkhwa, from 01 September 2012 to 31

August 2013. The patients were included by consecutive sampling which came for cardiac evaluation with suspected congenital heart disease referred to our unit. The diagnosis was primarily made on echocardiography. Size, number and exact location of the defects as well as magnitude of shunt were identified by two dimensional and Doppler echocardiography. Pulmonary artery pressure was estimated by using modified Bernoulli equation from Right ventricular systolic pressure and Right atrial pressure. Saturation of every patient was measured by pulseoxymetry. A definition proposed by Mitchell et al, for the diagnosis of CHD, was applied, that is, any gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance excluding the systemic great arteries and veins.⁹ By Miriam-Webster: a bluish or purplish discoloration of the skin due to deficient oxygen in the blood is called cyanosis or the appearance of a blue or purple color of the skin or mucous membranes and visually perceptible change in skin color occur when reduced hemoglobin exceeds 3g/dl. Any patient having the signs and symptoms like shortness of breath, excessive sweating, difficulty in feeding, bluish discoloration of lips and tongue, clubbing, failure to thrive, palpitation, feeling of impending doom, rapid breathing, fainting, light headedness, discrepancy in pulse, heart murmur, abnormal chest X-ray, or strong family history, high blood pressure, swelling of abdomen and feet, recurrent chest infections, chest and abdomen pain, and arrhythmias and loss of consciousness, etc. were evaluated further and those suspected of cardiac disease were subjected for chest X-ray, pulse oxymetry, electrocardiogram (ECG), followed by echocardiography. Care had been taken to avoid duplication of the cases in the hospital during the study period. Echocardiography was done as per standards laid down by the American Society of Echocardiography, using the M-mode, two-dimensional and color Doppler, pulse and continuous wave echocardiogram.¹⁰ All the data was entered to a preformed proforma and analysis was done using SPSS version 19.

RESULTS

Total number of patient studied with congenital heart disease (CHD) was 2342. Of them 434 (18.53%) were found to have cyanotic congenital heart disease (CCHD). of these CCHD patients males were 262 (60.36%). Mean age was 34.42 ± 30.34 months (from 1 day to 18 years). Tetralogy of fallot (TOF) patients were 205 (47.23%) which forms 8.8% of total CHD. Of these TOF patients 09 were already corrected. Transposition of great arteries (TGA) was found in 83 (19.12%). Ebstein with cyanosis were found in 12 (2.76%) patients. Double Outlet Right Ventricle (DORV) was found in 38 (8.75%). Total anomalous pulmonary venous drainage (TAPVD) patients were 27 (6.22%). Of these supracardiac TAPVD were 15 (3.45%), Cardiac TAPVD were 7 (1.61%) and Infracardiac TAPVD were 5 (1.15%). 01

Table 1: Spectrum of Cyanotic Congenital Heart Disease

Anomaly	Number	Percentage (%)	
Tetralogy of fallot	205	47.23	
Transposition of great arteries	83	19.12	
Double outlet Right ventricle	38	8.75	
Total anomalous pulmonary venous drainage	27	6.22	
Pulmonary atresia with intact pulmonary septum	15	3.4	
Hypoplastic left heart	14	3.22	
Eisenmenger syndrome	11	2.53	
Ebsteinanaomaly	12	2.76	
Truncusarteriosus	8	1.84	
Single ventricle	Tricuspid atresia	8	1.84
	Mitral atresia	8	1.84
	DILV	5	1.15

was already corrected. Pulmonary atresia with intact pulmonary septum was found in 15(3.4%). Single ventricle was found in 21(4.84%) patients with tricuspid atresia was found in 8(1.9%) patients, Mitral atresia in 8(1.84%) and double inlet Left ventricle in 5(1.15%). BT shunt patients with cyanosis were 10(2.3%). Eisenmenger syndrome was found in 11(2.6%). Hypoplastic left heart was found in 14 (3.22%) and 8(1.84%) having truncusarteriosus. These results are summarized in Table 1.

DISCUSSION

Pediatric Cardiology, Lady Reading hospital Peshawar, a tertiary referral centre for pediatric cardiac patients in Khyber Pakhtunkhwa catering to a population of almost 20 million. Catchment area extends to Federally administered Frontier Regions of Pakistan, parts of Punjab and Gilgit / Baltistan. To know about the estimated index of CHDs in various population groups, several studies had been carried out in past few decades and during this period a notable improvement in diagnosis of CHD was made by the introduction of echocardiography. Our study comprised all children up to 18 years, with CHD born in our hospital,

referred from other hospitals, clinics and those attended to our hospital for a variety of reasons.

From a study from Uganda shows 26 % of patients having cyanotic heart disease, of which the TOF and DORV were dominant. But in our study cyanotic heart disease were found in about 18.5% of the congenital heart disease. Of which TOF was the commonest one.¹¹

Recent studies from India and other developing countries have shown a decline in prevalence of rheumatic fever and rheumatic heart disease.¹² Congenital malformations and, in particular CHDs are likely to become important contributors to infant mortality in the near future and also to GUCH in adults. Hence, it is important to determine the exact prevalence and case burden of congenital heart disease so that appropriate changes in health policies can be recommended.¹³ Similarly here in Pakistan the rheumatic fever is decreasing so congenital heart disease will form the major bulk of adult cardiovascular structural heart disease and will replace the rheumatic heart disease in future.

In a study from India by Patra et al shows Tetralogy of Fallot and its variant was the most common congenital cyanotic heart disease with proportion of about 44%. Other common malformations were double outlet right ventricle (14%), d-transposition of the great arteries (9%), pulmonary atresia with ventricular septal defect (8%), total anomalous pulmonary venous connection (7%), truncusarteriosus, tricuspid valve anomalies, tricuspid atresia and Ebstein's anomaly, hypoplastic left-heart syndrome and complex CHD such as single ventricle.⁸ Similarly in our study in which about 18% of patients having congenital cyanotic heart disease, Tetralogy of Fallot forms the major bulk of CCHD i.e. 47%. While DORV was found in about 8.75% while in TGA was found in 19%. Isolated partial anomalous pulmonary venous connection is a rare lesion clinically, and it resembles an ASD in producing a right ventricular volume overload. Some studies, however, have shown an incidence of 0.6% to 0.7% in routine autopsies, implying that most of these are small shunts of little clinical significance.¹⁴⁻¹⁵ Cardiac defects are grossly divided into acyanotic and cyanotic heart diseases, former being more common. Ventricular septal defect (VSD) (30-35%) and tetralogy of Fallot (TOF) (5-7%) are most common among acyanotic and cyanotic CHDs respectively.¹⁶ Similarly in our study TOF was found in about 8% of total congenital heart disease patients. Over the last 15 years, stabilization occurred, corresponding to 1.35 million newborns with CHD every year. Significant geographical differences were found. Asia reported the highest CHD birth prevalence, with 9.3 per 1,000 live births, with relatively more pulmonary outflow obstructions and fewer left ventricular outflow tract obstructions.¹⁷ Similarly in our study TOF with pulmonary outflow obstruction form the major bulk of CCHD. Reported total CHD birth prevalence in Europe was significantly higher than in North America (8.2 per 1,000 live births vs. 6.9 per 1,000 live births; $p=0.001$).

Access to health care is still limited in many parts of the world, as are diagnostic facilities, probably accounting for differences in reported birth prevalence between high- and low-income countries. Observed differences may also be of genetic, environmental, socioeconomical, or ethnic origin, and there needs to be further investigation to tailor the management of this global health problem.¹⁷

LIMITATIONS

This study does not give the true incidence or prevalence of cyanotic congenital heart disease in total population as it was confined to one tertiary care hospital. Also, excluded were children not reaching a tertiary care centre due to poor access to medical facilities, yet, results are comparable with other international studies.

CONCLUSION

About 18% of CHD in our population comprises of CCHD. TOF is the major CCHD in our population.

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