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An understanding of congenital heart disease in children: A review article

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ABSTRACT

Heart development, is early in the embryo leading to significant mortality and morbidity affecting the clinical outcome of the affected individuals. Congenital heart defect is an anatomic malformation of the heart and/or great vessel, which occurs during intrauterine development. It is the commonest of all congenital lesions and is the most common type of heart disease among children. The incidence of congenital heart disease is approximately 8 per 1000 live birth, with a higher rate in stillbirth, spontaneous abortion and prematurity.3,4 It is believed that this incidence has remained constant worldwide. The CHD may be classified as acyanotic and cyanotic defects and the former is further divided into obstructive and left-to-right shunt lesions. The exact etiology of CHD is not known and the majority of cardiac defects can be explained by multifactorial inheritance hypothesis. Based on this review, it appears that while the etiology of CHD is not clearly identified, their recognition by clinical evaluation and non-invasive laboratory tests is possible and their treatment with currently available transcatheter and surgical methods is feasible, effective and safe.

KEYWORDS Congenital, disease, defect, heart

Introduction

Congenital heart disease (CHD) is the commonest of all congenital lesions and is the most common type of heart disease among children.[1] Congenital heart disease, in a 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." [2] The incidence of congenital heart disease is approximately 8 per 1000 live birth, with a higher rate in stillbirth, spontaneous abortion and prematurity.[3] It is believed that this incidence has remained constant worldwide. From population survey in Bangladesh, the prevalence of congenital heart disease was found 0.18%. World Health Organization (WHO) reports, among all cardiovascular disease, the incidence of Congenital heart disease in Bangladesh is 6%, 15% in India, 6% in Burma, 10% in Srilanka. The relative frequency of the most common lesions varies with different reports but nine common lesions form 80% of congenital heart disease.[4]

PREVALENCE OF CHD IN INDIA

We have no community-based data for incidence of congenital heart disease at birth in India. Since a large number of births in our country take place at home, mostly unsupervised by a qualified doctor, hospital statistics are unlikely to be truly representative. The incidence of congenital heart disease varies from as low as 2.25 to 5.2/1000 live births in different studies. There are a few studies of prevalence of congenital heart defects in school children; these are mainly offshoots of prevalence studies for rheumatic fever and rheumatic heart disease[5, 6,7]. Since a large number of such defects are critical, leading to death in early life itself, the studies on school children have limited value. Going by the crude birth rate of 27.2/1000 [8], the total live births are estimated at nearly 28 million per year. With a believed incidence rate of 6-8 per 1000 live births; nearly 180,000 children are born with heart defects each year in India. Of these, nearly 60,000 to 90,000 suffer from critical cardiac lesions requiring early intervention. Approximately 10% of present infant mortality in India may be accounted for by congenital heart diseases alone. In this way, every year a large no of children are added to the total pool of cases with congenital heart disease. We also have a large number of adult patients with uncorrected congenital cardiac defects, primarily because of lack of health awareness and inadequate health care facilities.

Congenital heart disease as a whole occurs with equal frequency in male and females but some lesions such as aortic stenosis, coarctation of aorta, transposition of great vessels and tetralogy of Fallot are more common in males whereas atrial septal defects are more common in females.The cause of most Congenital heart defects is unknown. Most cases of congenital heart disease are thought to be multifactorial and result from a combination of genetic predisposition and environmental stimulus.

Clinical Presentation

The clinical presentation of congenital heart disease varies according to the type and severity of the defect.[9] In neonatal period the presenting feature of congenital heart disease are cyanosis , heart failure, collapse, an abnormal clinical sign detected on routine examination.10 In infancy and childhood the usual presenting features are cyanosis, digital clubbing, murmur, syncope, squatting, heart failure, arrhythmia, failure to thrive.[10] The adolescent and adults present with heart failure, murmur, arrhythmia, cyanosis, hypertension, late consequences of previous cardiac surgery.[11] As a common congenital anomaly, CHD not only contribute to a significant morbidity and mortality but also causes a tremendous psychological stress and economical burden to the whole family. However, if the problems are recognized at earlier age, the chance of long term complications are less and the outcome is better. As a result of improved medical and surgical management, more children with CHD are surviving into adolescence and adulthood. Thus there is a need for an increased awareness amongst general physicians and cardiologists of the problem posed by these individuals. Except a few scattered observations, the incidence and detail clinical profile of CHD in Bangladeshi children are not well documented. This study was undertaken to find out the pattern and clinical profile of congenital heart disease among the admitted children in Rajshahi Medical College Hospital. It may help to detect and treat congenital heart disease at an earlier age and thus give the affected children and their parents hope of a better life.

Classification of CHDs

The CHDs could take place in any side of the heart: Atrial, ventricular or vascular. The common defects are classified according to: a) side of the affected heart, b) communication or short circuit between both hearts chambers and c) Presence or absence of cyanosis.[12]

(1) Atrial Septal Defect (ASD): A septal defect is a hole in different part of the atrial septum which lets some amount of blood from the left atrium to right atrium instead of flowing into left ventricle. It may be single or multiple and can be located anywhere in the atrial septum. Based on this, the ASDs are classified into 3 major types depending on the different part of the septum: a) Ostium secundum (Fossa ovalis), b) Ostium primum and c) Sinus venosum defect.

(2) Ventricular Septal Defect (VSD): A septal defect is a hole, existing between the lower chambers of the heart. Oxygen rich blood from the lungs is pumped into the aorta from the left ventricle. During this process with VSD some amount of blood is passed into the right ventricle and into the pulmonary artery back to the lungs.

(3) Patent Ductus Arteriosus (PDA): Ductus arteriosus, the temporary duct connecting the left pulmonary artery to the aorta in the fetal heart, fails to close after birth. This allows blood to mix between the pulmonary artery and the aorta, which results in too much blood traveling to the lungs.

(4) Pulmonary Stenosis (PS): Narrowing of the pulmonary valve between right ventricle and the pulmonary artery is called pulmonary stenosis. This results in the right ventricle pumping harder than normal to overcome the obstruction.

(5) Aortic Stenosis (AS): Narrowing of the aortic valve between the left ventricle and the aorta is called aortic stenosis. Normally there are 3 leaflets or cups in a valve, but in a stenotic valve there is one (unicuspid) or two (bicuspid). Obstruction may be valvular, subvalvular (sub aortic) or supra valvular.

(6) Coarctation of the aorta (COA): It is a constrictedsegment of the aorta that obstructs blood flow to the lower part of the body and increases blood pressure above the constriction. It usually occurs as isolated disease, but may occur with a VSD, sub aortic stenosis or complex CHDs.

(7) Tetralogy of Fallot (TOF): TOF is made up of 4 separate components: a) VSD, that lets blood pass from the right to the left ventricle without going through the lungs and b) a narrowing (stenosis) at or just beneath the pulmonary valve. This narrowing partially blocks the blood flow from the right side of the heart to the lungs. c) The right ventricle is more muscular than normal, and d) the aorta lies directly over the VSD. Collectively, this results in cyanosis or blue baby, which may appear soon after birth, in infancy or later in childhood.

(8) Transposition of the great arteries (TOA): The great arteries are pulmonary artery and the aorta. The normal positions of the arteries are reversed in this type of defect. The aorta is connected to the right ventricle, while the pulmonary artery is connected to the left ventricle. This results in the right ventricle pumping oxygen poor blood to different parts of the body and the left ventricle pumping oxygen rich blood to the lungs.

(9) Atrioventricular Septal Defect (AVSD): A large hole in the centre of the heart exists where the wall between the upper chambers joins the wall between the lower chambers. This is called as a complete AVSD. In case of partial AVSD, either the upper or the lower part of the septum is affected.

(10) Persistent truncus arteriosus: It is a complex malformation where only one artery arises from the heart and forms the aorta and pulmonary artery. That means the pulmonary arteries then branch off this common artery. This defect is found in association with VSD.

(11) Tricuspid Artesia (TA): The valve between the right atrium and the right ventricle is missing. As a result, oxygen-poor blood is pumped into the body along with the oxygen-rich blood. This results in cyanosis or blue baby. This defect is found in association with ASD, VSD and PDA.

(12) Pulmonary Artesia (PA): In this case no pulmonary valve exists; therefore blood cannot flow from the right ventricle into the pulmonary artery and on to the lungs. The only way for the blood to reach the lungs is the ductus arteriosus which is found during the fetal condition which closes after birth. The mixing of oxygen rich blood and oxygen poor blood results in cyanosis.

(13) Total anomalous pulmonary venous connection (TAPVC): In this case, all the pulmonary veins drain into the right atrium instead of left atrium, which brings the mixing of the blood. In addition to this, there is also presence of ASD and VSD, which results in cyanosis. There are three main types of TAPVC, depending on where the pulmonary veins drain. They are referred to as supracardiac, intracardiac, and infracardiac.

(14) Hypoplastic left heart syndrome (HLHS): In this condition the left ventricle and the aorta are small and underdeveloped. Therefore, the mitral and aortic valves are usually tiny or absent. It is one of the top three heart abnormalities to cause problems in the newborn.

(15) Double outlet right ventricle (DORV): It is a most uncommon defect in which both the pulmonary artery and aorta arises from the right ventricle, each with its own outflow tract and valve.

(16) Single ventricle / univentricular heart: It refers to a congenital malformation in which two atria are related to one ventricle that qualifies as left, right or indeterminate ventricle on purely morphologic ground.

(17) Ebstein's anomaly (EA): In this case there is a downward displacement of the tricuspid valve into the right ventricle. It is usually associated with an ASD.

(18) Dextrocardia (heart on the right): If the developing heart tube bends to the left instead of the right, then the heart is displaced to the right and develops in a mirror image of its normal state. This is a condition called situs inversus. In many a cases Dextrocardia heart functions normally unless there are no associated vascular abnormalities. In cases where the heart is the only organ, which is transposed, known as isolated Dextro-cardia, there are usually other severe cardiac abnormalities associated with it.

Conclusion

Heart development, is early in the embryo leading to significant mortality and morbidity affecting the clinical outcome of the affected individuals. Congenital heart defect is an anatomic malformation of the heart and/or great vessel, which occurs during intrauterine development. The incidence of CHD is 0.6 to 0.8% of live-births. The exact etiology of CHD is not known and the majority of cardiac defects can be explained by multifactorial inheritance hypothesis. Based on this review, it appears that while the etiology of CHD is not clearly identified, their recognition by clinical evaluation and non-invasive laboratory tests is possible and their treatment with currently available transcatheter and surgical methods is feasible, effective and safe.

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