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Recent Burden of Congenital Heart Diseases and Their Impact on Patients

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Abstract

Congenital heart disease (CHD) is known as series of structural and functional lesions of the major blood vessels and the heart itself during the time period of embryogenesis. CHD is responsible for the 1/3 of congenital aberrations and is the most prevalent type of congenital anomalies. The most commonly stated incidences of CHDs in the United States range between 4-10/1,000 with a clustering of approximately 8/1,000 live births. Global disparities in CHD incidence have been reported from 6.9/1000 to 9.3/1000 in Europe and Asia, respectively. International Classification of Diseases (ICD)-9 has enlisted 25 CHD subgroups, out of which 21 specify particular structural or hemodynamic defects. CHD is usually categorized into isolated type and syndrome type. Antenatal screening for CHDs comprises of ultrasonography during the second trimester of gravidity and postpartum clinical examination; although, detection rates are not high. Prior detection of CHDs is vital because clinical manifestation and worsening of symptoms may be abrupt and few responsive to treatment CHDs may lead to death before the proper diagnosis. The medical and surgical treatment of CHDs has remarkably ameliorated over the previous 50 years. Unfortunately, long-term survival has been attained at a high price, because patients endure late health issues, of which arrhythmias and heart failure are the most conspicuous. The purpose of this article is to present the latest information on etiology, incidence, and morphology, findings on physical examination, diagnostic imaging modalities, medical and surgical treatment approaches with their pros and cons, and survival rates of CHDs in patients.

Introduction

Congenital heart disease (CHD) is a prevalent congenital anomaly in the newborns that exists at birth. CHDs are a well-known risk factor of infant demise from congenital anomalies over the very first year of life. CHD is responsible for the 1/3 of congenital aberrations and is the most prevalent type of congenital anomalies [1]. Global disparities in CHD incidence have been reported, from 6.9/1000 to 9.3/1000 in Europe and Asia, respectively [2]. The most commonly stated incidences of CHDs in the United States range between 4-10/1,000; with a clustering of approximately 8/1,000 live births. During 2013, 3,051 deaths were related to CHDs in the United States. International Classification of Diseases (ICD)-9 has enlisted 25 CHD subgroups, out of which 21 specify particular anatomic or hemodynamic defects. CHDs vary in asperity from insignificant defects between heart chambers that get fixed immediately to considerable aberrations that may need several surgical procedures prior to school age and may lead to in utero fetal demise, during infancy, or during childhood period. In the United States, the most prevalent CHDs in children are ventricular septal defects (VSD), atrial septal defects (ASD), valvular pulmonary stenosis, and patent ductus arteriosus (PDA), while most common complex CHDS include; tetralogy of Fallot (TOF), transposition of great arteries (TGA), atrioventricular septal defects (AVSD), coarctation of aorta, and hypoplastic left heart syndrome (HLHS). CHD is usually categorized into isolated type and syndrome type. Isolated CHD establishes as a solitary anomaly (single or complex lesion limited to the heart and major blood vessels), while in syndrome category; a combination of heart lesion and aberrations of various organ systems exists. Down syndrome is the most prevalent syndrome, which is typically associated with CHDs; especially complete atrioventricular canal defect (CAVC), which is present in 40-50% of CHD patients. Current studies have illustrated that isolated and syndromic types of CHD are caused by various genetic and epigenetic inheritance modes, respectively. Several risk factors of nongenetic nature contribute to CHDs such as higher fertility rates in lower income countries, folate deficiency, maternal smoking in the first trimester of gravidity, gestational diabetes mellitus, maternal binge alcohol consumption during gravidity and increased Body Mass Index (BMI) [3]. CHDs are commonly pathophysiologically divided into following categories: (1) Acyanotic CHDs; (2) cyanotic CHDs and (3) obstructive valvular and non-valvular CHDs.

Acyanotic CHDs

Defects that permit the shunting of blood from the left to right side of the heart. They are related to alternating intensities of augmented pulmonary flow of blood and are generally acyanotic. In few lesions, the location of the shunt may not be present in the heart itself. Cyanosis appears only if the defects are considerable in size and not corrected in the early childhood, and if the patient establishes increased pulmonary vascular obstructive disease (PVOD) (Eisenmenger's phenomenon). In the present era, echocardiography is the initial imaging technique, and cardiac catheterization is generally used for interventional procedures [4]. Examples include (1) Atrial septal defect (ASD), (2) atrioventricular septal defect (AVSD), (3) patent ductus arteriosus (PDA), (4) partial anomalous pulmonary venous connection (PAPVC), and (5) ventricular septal defect (VSD).

Atrial septal defect (ASD)

ASDs exhibit 6-10% of total CHDs (Fig. 1A) and female to male predominance is 2:1 [5, 6]. There are four subtypes of ASDs: ostium secundum defect (60-70 % of ASDs are ostium secundum defect, located in the area of the fossa ovalis); ostium primum defect, a subtype of AVSD, located in the inferior aspect of the atrial septum); sinus venosus defect (an AVSD that is not an actual ASD, usually arises due to failure of the normal fusion of the right pulmonary veins into the left atrium at the entry point of the superior vena cava into the right atrium); and unroofed coronary sinus, the least prevalent type [7-8]. Unroofed coronary sinus or coronary sinus ASD is not an actual ASD [9]. It arises when an aperture in the roof of the coronary sinus permits the coronary sinus and left atrium to connect with each other. Patients with solitary ASDs are usually asymptomatic [10].

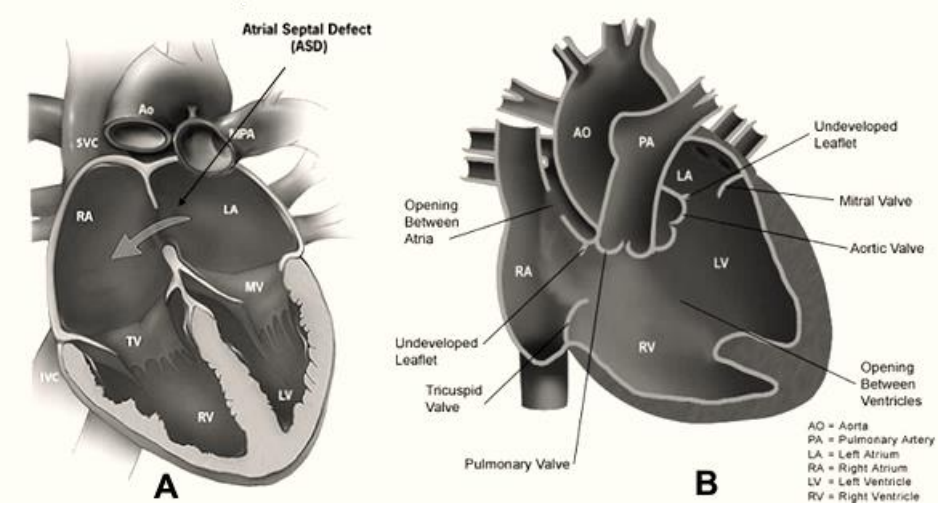

Fig. 1 An illustration of atrial septal defect (A) and atrioventricular septal defect (B) of the human heart.

ASD is the most frequently undetected CHD in childhood, usually not discovered until adolescence. ASD without treatment leads to atrial arrhythmias, exercise intolerance, increased pulmonary blood flow and PVOD in the third or fourth decade of life. The untreated patient may be on the verge of developing stroke secondary to paradoxical embolization through the defect. In moderate to large ASDs, physical examination of moderate-large ASDS reveals a widely split fixed second heart sound, an increased right ventricular impulse, and a soft systolic ejection murmur best audible at the upper left Sternal border. The murmur is produced due to the augmented flow of blood through the pulmonary valve, not through the defect. In large ASDs, excessive blood flow through the tricuspid valve produces a diastolic murmur at the lower Sternal border [11]. CXRs are not useful in assessing the sub-type and may appear normal with an ASD of a small size. In the very same way, an ECG can also appear normal in the unroofed coronary sinus, sinus venosus, and small secundum. With an ASD of a larger shunt, there may be right axis deviation, right ventricular hypertrophy or right atrial enlargement. ASDs of ostium primum type are identified by left axis deviation and an initial counterclockwise frontal plane loop. Treatment of ASDs usually requires either percutaneous device closure or an operation. Device closure is the popular method reserved for ostium secundum defects if the septal margins are appropriate enough to secure a device, but may not be used in other ASDs sub-types due to the close intimacy of the defects to other heart structures [12]. Repair of partial AVSD (cleft mitral valve and an ASD of ostium primum subtype) can be done later at the age of 18-24 months in the majority of patients and usually comprises mitral cleft closure in addition to the ASD [13].

Atrioventricular septal defect (AVSD)

AVSDs exhibit 4-5% of total CHDs and 40% of Down's syndrome patients present with AVSDs [14]. These defects are also known as AV canal defects or endocardial cushion defects (Fig. 1B). Endocardial cushions proximate the ostium primum and configure segments of the ventricular septum and the AV valves. AVSDs are classified into partial or complete; primum ASD is one type of a partial AVSD. A primum ASD and a contiguous inlet VSD constitute a complete AVSD. Signs and symptoms on physical examination of patients with partial and complete AVSDs are identical to patients with an ASD or VSD, respectively. The severity of AVSD is also estimated by the shunt's volume, associated cardiac and extracardiac abnormalities, the related size of the two ventricles, and intensity of the AV valve anomalies. ECG manifests an initial frontal plane loop of counter-clockwise consistency and a superior QRS axis. Usually, cardiomegaly and increased pulmonary vascular markings and cardiomegaly are present on CXR in patients of complete AVSDS. Patients having a complete AVSD need surgical repair,

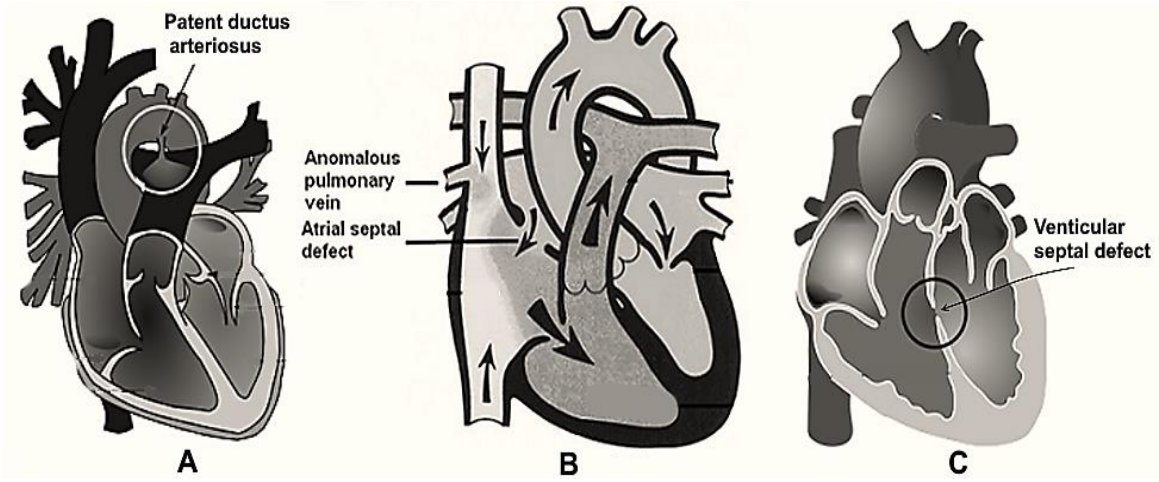

Fig. 2 An illustration of partial anomalous patent ductus arteriosus (A), pulmonary venous connection (B) and ventricular septal defect (C) of the human heart.

mostly done at the age of 3-6 months. Patients need lifetime follow-up and around 15% of patients establish progressive AV valve regurgitation or obstruction of the left ventricle outflow tract (LVOT) [15].

Patent ductus arteriosus (PDA)

PDA exhibits 9-12% of total CHDs (Fig. 2A). Usually, the ductus arteriosus closes in a period of a few days of birth and is an indispensable element of typical in-utero blood circulation. PDA develops in 1/5000 live births in full-term neonates, but is considerably more prevalent in pre-term infants [16]. In preterm neonates, a PDA has been recognized during hospital admission in 42% of infants having the weight of 500 to 999 g, 21% having the weight of 1000 to 1499 g, and 7% having the weight of 1500 to 1750 g [17-20]. Infants born at higher altitudes carry 30-fold higher prevalence of PDA [21]. Patients usually manifest no symptoms when the diameter of ductus is the minute [22].

With increasing size, infants manifest symptoms of elevated pulmonary blood flow, bounding pulses, and wide pulse pressure. Eisenmenger's phenomenon, secondary to (PVOD) and reversal of shunt, may precipitate if the PDA is of large size and duration, and causes cyanosis exclusively in the lower half of the body. Commonly, a PDA manifests a continuous murmur that is best heard at the left infraclavicular area. The nature of murmur is continuous because the aortic pressure is greater than the pulmonary artery pressure during both systolic and diastolic components. In a case of the large shunt; audible diastolic mitral flow murmur may also be present. In few newborn and preterm neonates, the murmur heard only in systole, may be confused with the murmur of a VSD. CXR and ECG usually are normal. In a PDA of large size, cardiomegaly on CXR, and biventricular hypertrophy on the surface ECG and increased pulmonary blood flow may be present [23, 24]. Echocardiography provides the outline of the PDA anatomy, and shunt's volume and direction [25-27]. In pre-term neonates, the ductus can often be closed with ibuprofen or indomethacin, or surgically ligated. In adolescents and adults, PDAs are usually closed percutaneously using devices or coils. Patients with manifestations of excess pulmonary blood flow can be treated with diuretics until definitive closure [28-48].

Partial anomalous pulmonary venous connection (PAPVC)

An aberration where ≥1 (though not all) pulmonary veins join up with the right atrium or systemic veins, rather than joining with the left atrium (Fig. 2B). This CHD exhibits less than 1% of the total CHDs. The aberrated joining of the right middle or upper pulmonary vein(s) is related with an ASD of sinus venosus type and joining of the right lower pulmonary vein (s) to the inferior vena cava(IVC) may be the component of the "Scimitar's syndrome" [49]. Presentation of PAPVC on CXR and ECG

findings are similar with ASDs presentation; however, patients with just one anomalous vein may be asymptomatic clinically. The clinical manifestation of children with the Scimitar's syndrome ranges from acutely sick neonates with pulmonary HTN to nearly clinically asymptomatic adults. CT, transesophageal echocardiography, and MRI may be mandatory for the diagnosis of PAPVC, because transthoracic echocardiography may not be sufficient to make the accurate diagnosis of PAPVC. Surgical intervention is indicated if the In some cases, shunt volume causes consequential volume overload of the right side, which is related with a murmur of pulmonary outflow and infrequently with a murmur of diastolic tricuspid inflow [50].

Ventricular septal defect (VSD)

VSD is the most prevalent type of CHD (Fig. 2C), responsible for 20% of total cases of CHDs, with the exclusion of mitral valve and bicuspid aortic valve prolapse [51]. Sub-types of VSDs are dependent on the basis of their location include: muscular (in the trabeculated or muscular segment of the ventricular septum); outlet (either under one or both semilunar valves); inlet (inferior to the atrioventricular [AV] valves); and perimembranous (in the area of the membranous septum). Sometimes VSDs of perimembranous type may close immediately by the apposition of the tricuspid valve septal leaflet to the defect. VSDs of outlet type may have large caliber and related to CHDs of relatively complex types. VSDs of the outlet and perimembranous types are in close vicinity to the aortic valve right cusp. Due to the Venturi effect, VSDs outlet and perimembranous ECG and CXR can be normal in VSDs of small caliber. In VSDs of large caliber, excessive pulmonary vascular markings and heart enlargement on CXR, and left ventricle hypertrophy of the left ventricle on ECG can be seen; hypertrophy of the right ventricle can occur with large VSDs. Left axis deviation is associated with VSDs of inlet type. Echocardiography gives significant details regarding the anatomical outline of the defect, right ventricular pressure and the volume of the shunt [59]. The objective of treatment is to assure appropriate somatic development and avoid PVOD. VSDs of small diameter and VSDs unassociated with excess pulmonary blood flow usually do not need closure. VSDs of large diameter and VSDs related with aortic regurgitation due to aortic valve cusp prolapse or excessive pulmonary blood flow may require diuretics and an increased caloric formula, followed by closure of the defect (either surgical closure or transcatheter device closure). VSDs manifesting failure to thrive and excessive pulmonary blood flow are surgically closed between the ages of 1-4 months; usually, large VSDs are mostly closed by the age of 6-9 months [60-63].

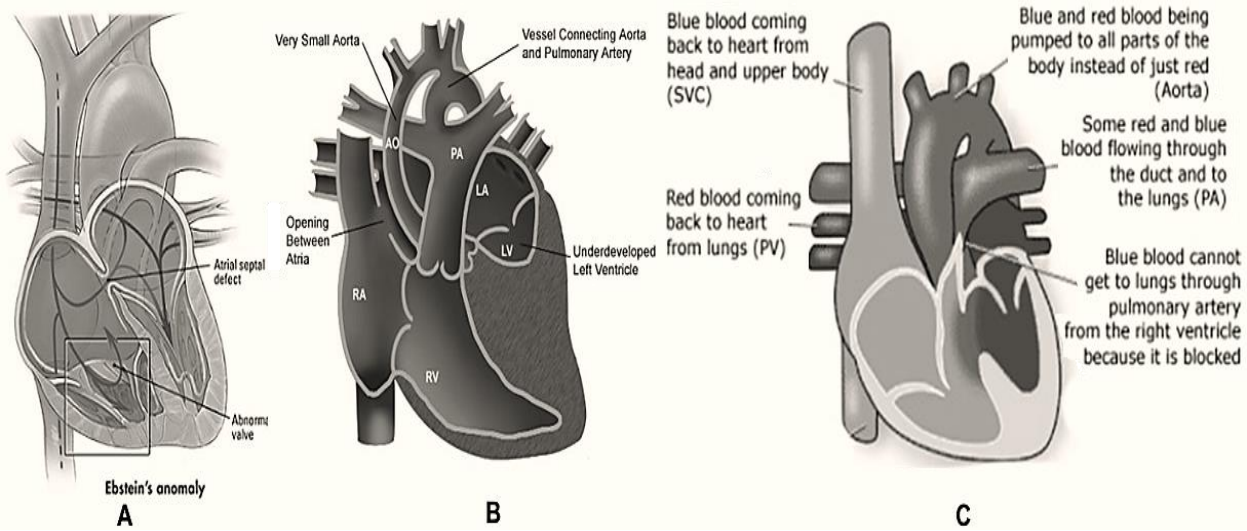

Fig. 3 An illustration of Ebstein's anomaly (A), hypoplastic left heart syndrome (B) and pulmonary valve atresia with ventricular septal defects (C) of the human heart.

Cyanotic CHDs

Heart defects that cause de-oxygenated blood entering into the aorta and would be related with a raised or diminished pulmonary blood flow. Examples of such lesions are discussed below.

Ebstein's anomaly

An unusual anomaly of the tricuspid valve in which aberrant valve leaflets delamination results into extreme valvular regurgitation [64]. Additionally, this anomaly manifests itself in a form of right ventricular and occasionally biventricular cardiomyopathy. The majorities of the patient also possess an ASD and manifest cyanosis due to shunting of blood from right to left side via the ASD (Fig. 3A). Auscultation reveals splitting of first and second heart sounds. CXR shows cardiomegaly with an obvious right sided border and diminished pulmonary vasculature. In limb lead II tall "P" waves and a right bundle branch block (RBBB) pattern is seen on ECG [65]. Around 15% of patients' manifest Wolff Parkinson White syndrome. Echocardiography is the best modality for assessing the valvular structural aberrations that carry a broad range of physiological and anatomical severity. Surgery is required for minimal cardiomegaly and tricuspid regurgitation. Prostaglandin E1is mandatory in newborns having extreme forms of this anomaly to increase pulmonary blood flow until pulmonary vascular resistance (PVR) decreases. Few newborns do well, even without having a corrective repair. However, massive cardiomegaly is difficult to manage in a subgroup of patients [66].

Hypoplastic left heart syndrome (HLHS)

It is ascribed to a range of defects that consists of an unimpaired ventricular septum with an insufficient growth of the left ventricle, mitral valve, and aortic valve (Fig. 3B). Patients usually exhibit coarctation of the aorta. Patients can stay acyanotic for certain days after their birth, but are reliant on the maintenance of a PDA. Patients manifest shock as soon as the ductus arteriosus closes. A physical examination shows an augmented right ventricular impulse and weakened pulses in the lower extremity. Mortality rates of HLHS have been considerably decreased during the last decade, but still, it is lethal if left untreated. Patients need a surgical intervention in a staged manner [67]. Firstly, a Norwood procedure (stage 1) is carried out which consists of the aortic arch by using the main pulmonary artery and secondly, a modified Blalock-Taussig procedure (stage 2) is carried out, in which a shunt providing blood flow to the branches of the pulmonary artery is established. Currently, a modified Blalock-Taussig shunt also known as a Sanaa shunt (a shunt between the right ventricle and the pulmonary artery) has been utilized as a substitute to a Blalock-Taussig shunt which is placed for palliation during stage 1 [68]. Stage 2 of surgical intervention includes replacement of the modified Blalock-Taussig shunt with Glenn shunt (bidirectional cavopulmonary anastomosis). Stage 3 comprises of a modified Fontan's procedure, which shifts venous return of lower extremities to the pulmonary arteries. During recent years, interventional catheterization/ hybrid surgical operations have been availed to diminish pulmonary blood flow with branch pulmonary artery bands, and stent placement to preserve ductal patency; however, the advantage of this intervention remains vague. A heart transplant is sometimes sought as a substitute to staged surgical operation [69].

Pulmonary valve atresia with ventricular septal defects

This anomaly is mostly regarded as the most severe type of TOF, in which the pulmonary valve does not exist (Fig. 3C). Around 70% patients of this defect have a PDA that preserves pulmonary blood flow, and the rest of the patients have several systemic to pulmonary ancillary vessels. Pulmonary artery aberrations are frequent in this defect and include abnormal distribution, hypoplasia, and non-confluence. Patients mostly appear cyanotic at the time of birth. A murmur of continuous nature from the systemic to pulmonary artery or PDA collaterals may be detectable. Single second heart sound, right axis deviation on ECG, and right ventricular hypertrophy are present. Considerably decreased pulmonary vasculature is usually presented on CXR. Ductus arteriosus is kept patent by the commencement of prostaglandin E1. The majority of patients will probably require a systemic

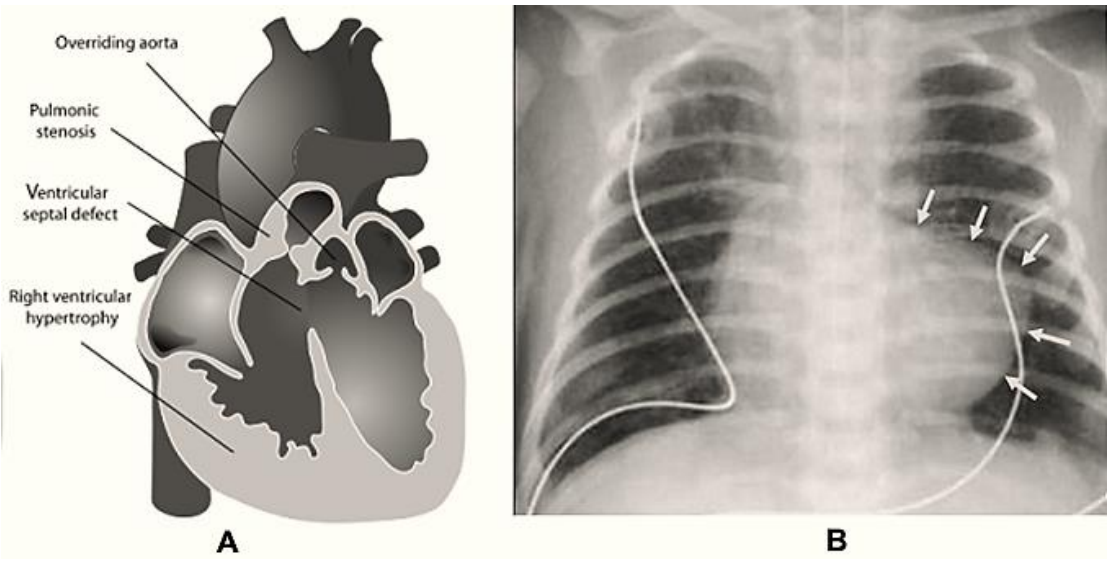

Fig. 4 An illustration of tetralogy of fallot (TOF) (A) and chest X-ray showing "boot shaped" human heart of TOF (B) as indicated by arrows.

to pulmonary shunt to preserve appropriate pulmonary blood flow. Patients having large, confluent pulmonary arteries might undergo just one-stage repair; but other patients need a staged repair [70].

Pulmonary atresia with an intact ventricular septum

This CHD is uncommon and asperity relies upon usually associated hypoplasia of tricuspid valve hypoplasia and the right ventricle. Fistula of the coronary artery to the right ventricle is also associated with this CHD [71]. An unrestrictive, wide ASD is mandatory for continuity of life as there is no communicating channel between the right ventricle to the pulmonary arteries. On physical examination, there are no distinguishing findings of this CHD. There is cardiomegaly with an accompanying prominent right atrium on CXR, and ECG reveals an enlargement of the right atrium. Echocardiography is used to evaluate tricuspid valve hypoplasia and right ventricular size.

An ASD of restrictive type is an imminent medical emergency with patients of this CHD and an immediate balloon atrial sept Ostomy is required. Ductus arteriosus is kept patent by the commencement of prostaglandin E1 because pulmonary blood flow is only maintained by the PDA. The majority of the patient need staged surgeries that probably permit the right ventricle and tricuspid valve to increase in diameter. A 1.5 ventricle repair or a modified Fontan's operation may be indispensable if the tricuspid valve and right ventricle do not gain an appropriate diameter to permit a second ventricle repair. A 1.5 ventricle repair consists of a bidirectional cavopulmonary shunt to switch superior vena caval (SVC) blood flow in the pulmonary arteries to decrease right ventricular preload, closing the ASD and creation of continuity between the right ventricle and the pulmonary artery. Modified Fontan's operation is required by patients having coronary artery to right ventricle fistula [72].

Tetralogy of fallot (TOF)

Tetralogy of fallot (TOF) is the most trivial cyanotic CHD, exhibiting 4-8% of total CHDs [73]. TOF comprises of 4 anomalies: Right ventricular outflow tract (RVOT) obstruction, VSD of outlet type, an overriding aorta, and hypertrophy of RV, due to obstruction of RVOT (Fig. 4A). If obstruction of RVOT is mild and there is no appearance of cyanosis, then it is sometimes ascribed as "pink tetralogy". The majority of patients having TOF manifest gradually advancing cyanosis after birth with subsequent dyspnea on moderate physical activity as a young child. In order to, compensate chronic hypoxemia, development of secondary erythrocytosis results. Poor physical development might be present. During

"Tet" or hyper cyanotic spells, which are hypoxic episodes consisting of a sudden onset of accelerated shallow breathing, cyanosis, increased agitation, and a murmur of decreased intensity due to reduced flow of blood via the RVOT. Without proper treatment, these hypoxic spells can lead to fatal seizures, hypoxemia, and patient's demise. Children of older age, usually squat during episodes of hypoxia spell to increase systemic vascular resistance (SVR) and by that decrease hypoxemia [74].

An ejection systolic murmur of loud intensity is heard at the left upper sternal border and radiates to bilateral axillae due to obstruction of RVOT. Seldom, patients with nominal obstruction of RVOT present with isolated mild cyanosis and due to a left to right shunt may establish signs of increased pulmonary blood flow through the VSD. A right ventricular impulse may be felt. Mostly, CXR appears normal with the exception of diminished markings of the pulmonary vasculature. Less than 25% of neonates have the typical cardiac silhouette of "boot shape" (Fig. 4B). Prostaglandin E1 is usually given to markedly hypoxic neonates to resume or preserve the patency of the ductus arteriosus and raise pulmonary blood flow. If hypoxemia does not improve, the patient may need a Blalock-Taussig shunt, until the ultimate repair (which comprises of alleviation of RVOT and obstruction of the VSD by patch closure) [75-78]. Absolute repair may be done in a few centers in the newborn period, although most physicians favor it at the age of 3 months or when gradual cyanosis establishes. In the recent decades, excellent surgical results have been seen, though the majority of patients got "repaired" as young children need re-surgery for placing an efficient pulmonary valve when older [79].

Truncus arteriosus

It exhibits <1 % of total CHDs. This defect consists of a wide VSD outlet and a single great artery, known as truncus, originating from the heart, which results into the pulmonary arteries, the coronary arteries, and the aorta (Fig. 5A). Associated aberrations include: truncal valve insufficiency, interrupted aortic arch, and DiGeorge syndrome (22q11.2's micro-deletion). Cyanosis or congestive heart failure (CHF) is the common manifestations. Auscultation of systolic ejection murmur at the left sternal border and pronounced right ventricle impulse, are the findings on physical examination. Ejection click of the truncal valve might be heard. Regurgitation of the truncal valve gives rise to a diastolic murmur of decrescendo nature. A restrictive type of PDA and interruption of the aorta causes weak femoral pulses. The dominance of right ventricular on ECG and cardiomegaly with accompanying augmented pulmonary markings are visible on CXR. Echocardiography precisely describes different features of this defect [80]. Diuretics are used to treat CHF. Surgical correction requires segregation of the pulmonary and systemic blood circulations, VSD closure and placement of a conduit between the right ventricle and pulmonary artery. Surgery is usually carried out by the age of 2-3 months to avoid PVOD. Dysfunction of the conduit between the right ventricle and pulmonary conduit requires re-operations and various transcatheter interventions. Aortic arch's interruption increases the risk of operative mortality [81].

Total anomalous pulmonary venous connection (TAPVC)

Total anomalous pulmonary venous connection (TAPVC) consists of <1% of total CHDs, TAPVC is an aberration where all the pulmonary veins join with the right atrium or the systemic venous system (Fig. 5B). It is further subdivided into infra cardiac, cardiac, supra cardiac, and mixed types, among all subtypes the supra cardiac is the usually more common. In this defect, entirely oxygenated blood coming from the lungs is going back to the right atrium and without ASD survival is impossible, because blood cannot enter into the aorta. Congestion and pulmonary HTN due to the impedance of the veins draining the pulmonary venous convergence, make TAPVC more complicated. A manifestation of elevated pulmonary blood flow and minimal cyanosis is usually seen in patients with a wide intraatrial connection and pulmonary veins without any obstruction. An obvious murmur of systolic ejection and right ventricular impulse is present on physical examination. Augmented pulmonary vascular outline and considerate cardiomegaly are present on CXR. Typical "snowman" shape is manifested in TAPVC of supra cardiac type. Patients

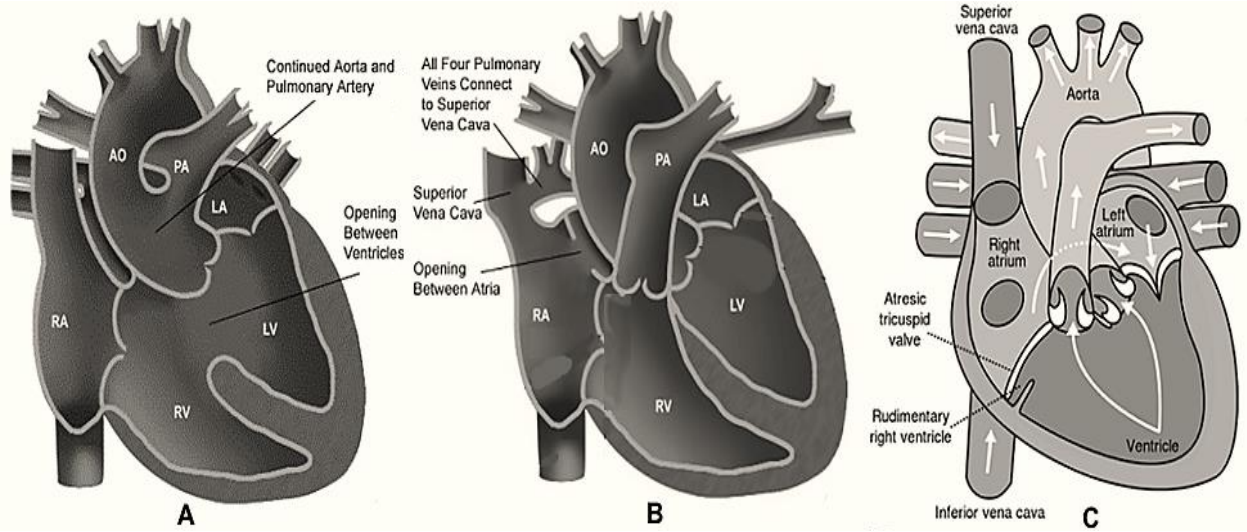

Fig. 5 An illustration of Truncus arteriosus (A) and total anomalous pulmonary venous connection (TAPVC) (B) and tricuspid valve atresia (C) of the human heart.

usually, need surgery but can achieve outstanding survival of long-term. Patients having impeded pulmonary venous blood flow suffer from respiratory distress due to the pulmonary edema. This is usually more common in newborns having an infra-cardiac TAPVC are usually having more respiratory distress due to the pulmonary edema and may require immediate surgical correction. Mortality is 40%, although urgent surgeries [82].

Tricuspid valve atresia

This aberration comprises of near about 3% of total CHDs. It occurs due to the developmental failure of an absolute communication within the right atrium and right ventricle. Preliminary survival is based on a communication between the right atrium and the left atrium (Fig. 5C). Clinical manifestation and therapeutics are based on the association of the great arteries (transposed or normal), the existence of a VSD, and stenotic asperity of the pulmonary valve. In this CHD, patients typically manifest cyanosis. Apical impulse might be obvious. On auscultation, second heart sound might be split or single, systolic murmur due to a pulmonary stenosis or VSD, while an apical mid-diastolic murmur might be heard due to CHF and augmented pulmonary blood flow. CXR exhibits cardiomegaly with an obvious right atrial margin and pulmonary vascular outline differs based on the severity of pulmonary stenosis. ECG shows a deviation of the left axis and an early frontal plane loop of counter-clockwise direction. LVH might be present [83]. If pulmonary blood flow source is not constant; prostaglandin E1is usually commenced until the establishment of a shunt between systemic to the pulmonary artery. Patients having transposition of great arteries have an unimpeded flow of pulmonary blood and need banding of the pulmonary artery to avoid PVOD or pulmonary edema, with medicines preventing pulmonary congestion. Ultimately all patients require a modified Fontan's surgical intervention.10 years follow-up of post-surgery showed >85% long-term survival. Fontan's operation has few long-term problems such as left ventricular dysfunction include, atrial arrhythmia and protein-losing enteropathy [84].

Obstructive valvular and non-valvular CHDs

Obstructive valvular and non-valvular lesions include: (1) Aortic valve stenosis (AS), coarctation of the aorta, (3) left ventricular outflow tract (LVOT) obstruction and (4) pulmonary valve stenosis (PS). However, here only 3 out of 4 will be discussed.

Coarctation of the aorta

It comprises of 5% of the total CHDs and is generally more prevalent among males; in females, this defect is related to Turner's syndrome [85-87].

It comprises of a posterior ridge of tissue that bulges into the aorta and is usually characterized as juxta- ductal because it develops beyond the ductus arteriosus (Fig. 6A). In 70% of the patients, it is generally related to a bicuspid aortic valve and less frequently with an aortic stenosis of subvalvular type or VSD. Patients having mild obstruction might not seek medical assistance until pre-adulthood with systemic HTN or a heart murmur. In patients having a severe type of obstruction, the PDA is the main supply of systemic blood flow beyond the obstruction is provided by the PDA. The neonates establish shock and metabolic acidosis with the closure of the ductus. Patients having severe obstruction establish CHF, weakened pulses and decreased blood oxygen saturation in the lower extremities [88, 89]. It is very imperative to measure the blood pressure in both arms and leg before establishing the diagnosis of aortic coarctation due to the possibility of overlooking the diagnosis in those patients having a coarctation near to the left subclavian artery or those having an aberrated root of the right subclavian artery. A systolic murmur is produced by turbulent flow of blood through the segment of coarctation. Patients having an accompanied bicuspid aortic valve exhibit an ejection click. Paradoxically, infants have RVH on echocardiography and ECG due to the PDA providing the aorta beyond to the obstruction [90].

Older patients manifest signs of LVH. MRI and CT might be helpful in delineating the anatomical features of the coarctation in older and adult patients [91-94]. Native coarctation is pliable to balloon dilation or surgical correction. Repair in the newborn period is associated with an increased incidence of recurrence of coarctation increases with repair done during the newborn interval. Recurrence is excellently dealt by balloon dilation either with/without placement of a stent [95-110]. In comparison with the general population, all patient undergone repairs are at high risk of early-onset systemic HTN, MI, CAD, and stroke [111-118]. Follow-up of life-long duration is required, albeit in patients who are free from residual obstruction [119-121].

Left ventricular outflow tract (LVOT) obstruction

It includes four sub-types: tunnel subaortic stenosis, aortic valve stenosis, discrete subvalvular stenosis, and supra valvular stenosis. Most common form of LVOT is aortic valve stenosis and exhibits 5% of total CHDs [122]. The cusps or leaflets are mostly thickened or malformed. Nearly 2% of the common population carries a bicuspid aortic valve, but the majority of these personnel never establish clinically evident aortic valve regurgitation or stenosis. Supravalvular stenosis develops as a region of diffuse or discrete narrowing exactly distal to the sinotubular intersection in the ascending aorta. It is mostly correlated with an error in the elastin gene on chromosome 7 (William's syndrome). Individual subvalvular stenosis develops when there is a fibromuscular rim that establishes obstruction exactly below the aortic valve. Turbulent blood flow might damage aortic valve leaflets, producing a gradual regurgitation of the aortic valve. Tunnel subaortic narrowing/stenosis are ascribed to a longer narrowed/stenotic section of the outflow tract which is distinguished from the distinct subvalvular stenosis. Manifestation relies on the asperity of the obstruction. The majority of patients are asymptomatic. Older patients may present with Syncope or chest pain may be manifested by older patients. A subgroup of patients manifest low cardiac output, poor left ventricular function, and signs of CHF (usually in severe aortic stenosis) and shock. Physical examination exhibits a systolic murmur of crescendo-decrescendo pattern, which is best heard at the left sternal margin that radiates to the upper right sternal margin. A detectable thrill and late pulses are the results of the medium or severe type of stenosis. In the patients with aortic valve stenosis, an ejection click might be auscultated right after the S1. In comparison to the ejection click of the pulmonary valve, ejection click in aortic stenosis does not alter with respiration. Patients having coexistent aortic regurgitation also have a wide pulse pressure and decrescendo diastolic murmur. ECG eccentrically exhibits evidence of LVH. Inversion or flattening of the T waves in chest leads V5 and V6 is associated with critical stenosis of LVOT [123].

CXR might show an obvious aorta due to post-

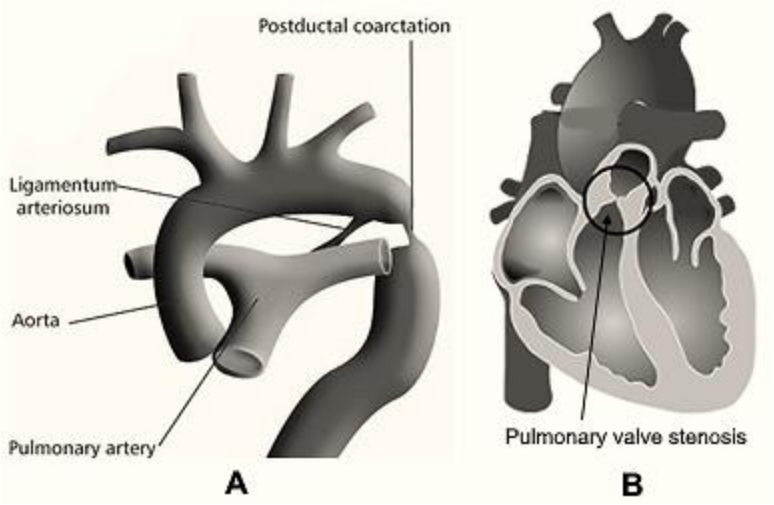

Fig. 6 An illustration of coarctation the aorta (A) and pulmonary valve stenosis (B) of the human heart.

stenotic dilation. Echocardiography precisely assesses the morphology of aortic valve and calculates the pressure gradient across the LVOT. In aortic stenosis of valvular type, the severity of the stenosis is assessed by mean gradient estimated by Doppler. The gradient varies according to the cardiac output. Emergency relief is required for the patients having severe aortic stenosis and patients who are symptomatic also need relief for the obstruction, irrespective of the severity of the stenosis. Balloon valvuloplasty is required in infants having critical valvular stenosis; however co-existing aortic regurgitation may require surgery. Valve replacement or surgical valvotomy is contemplated for adults and older children. Patients having severe stenosis followed up by (Second Natural History Study of CHDs) revealed 81 % long-term survival of 25-year [124].

Pulmonary valve stenosis

It exhibits around 8% of total CHDs (Fig. 6B) and is usually present in patients having Noonan's syndrome. Asymptomatic murmur is the only entity in the majority of the children at the time of presentation [125]. However, neonates having censorious pulmonary valve stenosis exhibit cyanosis due to shunting of blood from right side to left side at the level of atria. Physical examination detects an augmented right ventricular impulse, an ejection click after the S1 that alters with respiration, a normal to widely split S2 based on the degree of severity, and an ejection murmur of a crescendo-decrescendo pattern. With augmented level of stenotic severity, the pulmonary ejection click appears prior to systole; but in the majority of severe cases, the click may mix with the S1 and become imperceptible [126-128]. Conversely, with the augmented asperity of stenosis, the S2 splits more widely and might exhibit fixed splitting in severe stenotic cases. In more severe stenotic cases, the pulmonary part of the S2 might become more challenging to be auscultated as a result of a loud murmur that comes during diastole. A fourth heart sound (S4) may be heard in patients with right ventricular failure. ECG may show the deviation of the right axis and a CXR may reveal the manifestation of right ventricular enlargement. Generally, the pulmonary arteries have post-stenotic dilation. The most popular treatment option is balloon valvuloplasty [129, 130], and additional intervention is not required in 85% of patients. The long-term results are outstanding [131].

Recommendations

CHDs have several extrinsic and intrinsic risk factors but only some of them are recognized till date. However, there are few steps that can help in the prevention of having a newborn with CHD. Identification of some preventable causes; such as investigating the potential effects of additional factors during gravidity, such as infection, medicines,

fever, uncontrolled diabetes, smoking, alcohol consumption, and nutritional deficiency (folate and iodine deficiency) on the risk for CHDs. Antenatal ultrasonography during the second trimester is mandatory for the early detection of a fetus with CHD and termination of pregnancy may be offered to the mothers carrying a fetus with complex CHD.

Conclusions

CHD is the most trivial congenital anomaly in newborns. The medical and surgical treatment of CHDs has remarkably ameliorated over the previous 50 years, which has made the majority of patients to attain adulthood. Unfortunately, long-term survival has been attained at a high price, because patients endure late health issues, of which arrhythmias and heart failure are the most conspicuous. Consequently, these patients require constant follow-up by cardiologists with explicit knowledge in the specialty of CHDs. Birth incidence measurements vary extensively according to particular regions, and survival estimates have not been well documented. Antenatal detection has led to augmented rates of abortions. Ameliorated treatment of complications caused by CHDs has changed the mode and time of death. Different environmental and genetic risk factors have been identified to be involved in the causation of CHDs; however, this knowledge has not yet led to the significant application of preventative strategies, except for the prevention of few CHDs of environmental origin. Further research in the field of CHDs to identify the culprit risk factors involved in their etiology, and development of new medical and surgical modalities to enhance long-term survival rates with patient compliance is required.

Conflict of Interest

The authors declare that they have no conflict of interest.

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