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Commonest Type of Congenital Heart Disease (CHD) Among Children in Maternity and Children Hospital (MCH)

Nasser Mesfer Rashed AL-Abataheen[1], Abdullah Saleh Alyami[2], Mansour Ahmed Mohammed Almakrami[3], Ibrahim Awad Mohdi Alyami[4], Khalid Hamad Hussein Alsalem[5], Mohammed Yahia Hassan Alyami[6], Mahdi Yahia Alzamanan[7], Fahad Ahmed Ali Alkanfari[8], Abdulrhman Ali Alyami[9], Obaid Mahdi Mohammad Al Abbas[10]

Abstract

CHD means a child is born with an abnormally structured heart and/or large vessels. Such hearts may have incomplete or missing parts, may be put together the wrong way, may have holes between chamber partitions or may have narrow or leaky valves or narrow vessels(1). It has two types cyanotic and acyanotic. (2)

facility- based descriptive case series study including qualitative research approaches study will be conducted in Maternity and Children's Hospital in Najran to assess patients with CHD from neonatal period to two years old from (1)- 1436 H to (9) – 1436 H. sampling non-probability sampling by using the purposive and convenience techniques. The data will be collected by structured the check lists. Data processing and analysis will be done by entering the data into SPSS for Windows.

After analyze of data the commonest type of congenital heart disease is atrial septal defect as percent 27.27% followed by ventricular septal defect as percent 18.18%. The consanguinity marriage has a relationship with congenital heart disease as it is effect 68.2% and just 31.8% are not affected.

(INTRODUCTION)

1.1) Definition of CHD:-

CHD means a child is born with an abnormally structured heart and/or large vessels. Such hearts may have incomplete or missing parts, may be put together the wrong way, may have holes between chamber partitions or may have narrow or leaky valves or narrow vessels.[(1)]

1.2) Statement and analysis:

(Maternal disorders like rubella infection and diabetes mellitus can lead to patent ductus arterioses and increase the incidence of CHD overall . In addition the drugs that taken by mother during gestational period like warfarin therapy and valporic acid can share in malformation of the heart such as PDA ,pulmonary stenosis, atrioventricular septal defect, ventricular septal defect and tetralogy of fallot And the chromosomal abnormality like Down syndrome, Edwards syndrome and Patau syndrome can cause AVSD and VSD. Family history and consanguous marriage of first degree relative increase the susceptibility of CHD development. All these risk factors help to identify the magnitude of CHD). [(2, 3)]

Table (1) shows the classification of congenital cardiac defects [(3)]

Table 143-1 Classification of Congenital Cardiac Defects

	SHUNTING		
STENOTIC	RIGHT → LEFT	LEFT → RIGHT	MIXING
Aortic stenosis	Tetralogy	Patent ductus arteriosus	Truncus
Pulmonary stenosis	Transposition	Ventricular septal defect	TAPVR
Coarctation of the aorta	Tricuspid atresia	Atrial septal defect	HLH

HLH, Hypoplastic left heart syndrome; *TAPVR*, total anomalous pulmonary venous return.

Figure (I) shows the common congenital heart diseases [(4)]

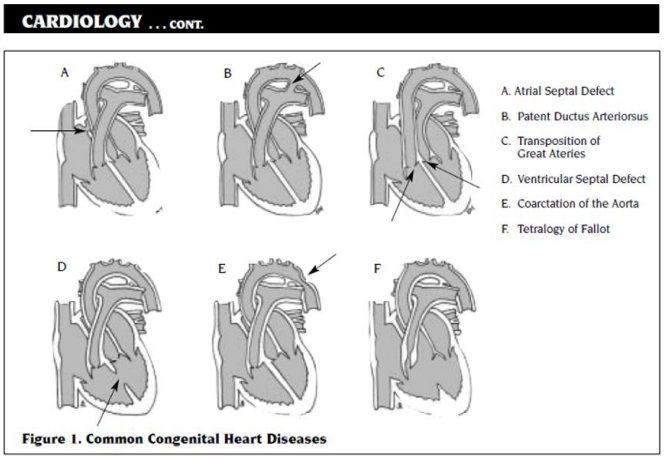

Figure 1. Common Congenital Heart Diseases

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1.3) Background information global and local:

A. Global:

The reported prevalence of congenital heart disease (CHD) at birth ranges from 6 to 13 per 1000 live births. Variation is primarily due to the use of different methods to detect CHD, such as referral to a cardiac center or fetal echocardiographic data [(5)].

The following studies provide a global perspective on the incidence of neonatal CHD:

- In one English health region, reported prevalence of cardiovascular malformations was 6.5 per 1000 live births.
- In a population-based study from Atlanta, the prevalence of CHD was 8.1 per 1000 live births from 1998 to 2005. The most common diagnosis was muscular and perimembranous ventricular septal defect (VSD), followed by secundum atrial septal defect (ASD) (prevalence of 2.7, 1.1, and 1 per 1000 live births, respectively). Tetralogy of Fallot was the most common cyanotic CHD (0.5 per 1000 births) [(5)].
- In a population-based study of all Danish live births from 1977 to 2005, the prevalence of CHD was 10.3 per 1000 live births. Chromosomal defects were detected in 7 percent of those patients, and extracardiac anomalies in 22 percent. In a population-based study, the prevalence of CHD in Greater Paris was 9 per 1000 live births. With the exclusion of VSD, 40 percent of the patients were diagnosed prenatally.
- The highest prevalence for CHD was observed in a population-based study from Taiwan with a prevalence of 13.1 per 1000 live births between 2000 and 20. The most common defect was VSD, followed by secundum ASD and patent ductus arteriosus (prevalence of 4, 3.2, and 2 per 1000 live births, respectively) [(5)].

In preterm infants (gestational age <37 weeks), CHD is two to three times that found in term infants. In addition, reported maternal conditions that increase the risk of CHD include multifetal pregnancy, diabetes mellitus, hypertension, maternal CHD, thyroid disorders, systemic connective tissue disorders, and epilepsy and mood disorders [(5)].

B. Local:

The results of epidemiological studies done in 4 regions of Saudi Arabia (August 1988-February 2000) and 2604 individuals with congenital heart disease were evaluated. Ventricular septal defect was the commonest lesion (33.9%) followed by atrial septal defect (18.1%). Overall, sex distribution was similar; for 3 conditions, more males than females were affected. Of 2269 (59%) presenting in the first year of life, 566 (24.9%) had neonatal congenital heart disease. Down syndrome was the commonest cause. Distribution of specific lesions and sex distribution was similar to findings from other parts of the world; however, the overall detection rate at 1 year of age was lower [(6)].

1.4) Rationale:

The CHD they are increased globally and locally depend on global statistics study.(increasing in number of CHD as shown by previous global study).

- Knowledge about the most common type of CHD in MCH.
- Identification of CHD among patient in MCH.
- The risk factors that leading to congenital heart diseases are many, and society education is one of our concerns.
- There is no previous published data about CHD in Najran area.
- We are interested of this subject.

(OBJECTIVES)

2.1) General objectives:

To assess patients with CHD from neonatal period to two years old from (1) – 1436H to (9) – 1436H in MCH from May to July 2015 to identify the magnitude and common lesions of CHD.

2.2) Specific objectives:

1. To identify the relationship between CHD and syndromes, like Down syndrome, Edwards syndrome and Patau syndrome.
2. To describe the presenting symptom and time of presentation of CHD.
3. To estimate the prevalence rates of the different common types of CHD.
4. To assess the relation between CHD and consanguineous marriages.
5. To review the prenatal detection in (MCH) for CHD.

(LITERATURE REVIEW)

3.1) Definition of CHD:-

CHD means a child is born with an abnormally structured heart and/or large vessels. Such hearts may have incomplete or missing parts, may be put together the wrong way, may have holes between chamber partitions or may have narrow or leaky valves or narrow vessels.[(1)]

3.2) ETIOLOGY:-

The cause of most congenital heart defects is unknown. Most cases of congenital heart disease were thought to be

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multifactorial and result from a combination of genetic predisposition and environmental stimulus. A small percentage of congenital heart lesions are related to chromosomal abnormalities, in particular, trisomy 21, 13, and 18 and Turner syndrome; heart disease is found in more than 90% of patients with trisomy 18, 50% of patients with trisomy 21, and 40% of those with Turner syndrome.[2]

3.3) Types of CHD:-

3.3.1) ACYANOTIC CONGENITAL HEART LESIONS

Acyanotic congenital heart lesions can be classified according to the predominant physiologic load that they place on the heart. The most common lesions are those that produce a volume load, and the most common of these are left-to-right shunt lesions. The chest radiograph and electrocardiogram are useful tools for differentiating between these major classes of volume and pressure overload lesions. [2]

The most common lesions in this group are those that cause leftto- right shunting: atrial septal defect, ventricular septal defect (VSD), AV septal defects (AV canal), and patent ductus arteriosus. The pathophysiologic common denominator in this group is communication between the systemic and pulmonary sides of the circulation, which results in shunting of fully oxygenated blood back into the lungs. This shunt can be quantitatedby calculating the ratio of pulmonary to systemic blood flow. [2]

Lesions Resulting in Increased Pressure Load

The most frequent are obstructions to ventricular outflow: valvular pulmonic stenosis, valvular aortic stenosis, and coarctation of the aorta. Less common are obstruction to ventricular inflow: tricuspid or mitral stenosis, cor triatriatum and obstruction of the pulmonary veins. Ventricular outflow obstruction can occur at the valve, below the valve (double-chambered right ventricle, subaortic membrane), or above it (branch pulmonary stenosis or supravalvular aortic stenosis .The infant may become critically ill within several hours of birth. [2]

3.3.2) CYANOTIC CONGENITAL HEART LESIONS

This group of congenital heart lesions can also be further divided according to pathophysiology: whether pulmonary blood flow is decreased (tetralogy of Fallot, pulmonary atresia with an intact septum, tricuspid atresia, and total anomalous pulmonary venous return with obstruction) or increased (transposition of the great vessels, single ventricle, truncus arteriosus, total anomalous pulmonary venous return without obstruction). The chest radiograph is a valuable tool for initial differentiation between these two categories. [2]

Cyanotic Lesions with Decreased Pulmonary Blood Flow

These lesions must include both an obstruction to pulmonary blood flow (at the tricuspid valve or right ventricular or pulmonary valve level) and a pathway by which systemic venous blood can shunt from right to left and enter the systemic circulation (via a patent foramen ovale, atrial septal defect, or VSD). These patients may have hypercyanotic ("tet") spells during conditions of stress. In contrast, if the obstruction is severe, pulmonary blood flow may be totally dependent on patency of the ductus arteriosus.

Cyanotic Lesions with Increased Pulmonary Blood Flow

This group of lesions is not associated with obstruction to pulmonary blood flow. Cyanosis is caused by either abnormal ventricular-arterial connections or total mixing of systemic venous and pulmonary venous blood within the heart. Transposition of the great vessels is the most common of the former group of lesions. The persistence of fetal pathways (foramen ovale and ductus arteriosus) allows for a small degree of mixing in the immediate newborn period; when the ductus begins to close, these infants become extremely cyanotic. Total mixing lesions include cardiac defects with a common atrium or ventricle, total anomalous pulmonary venous return, and truncus arteriosus. In contrast, if pulmonary stenosis is present, these infants may have cyanosis alone, similar to patients with tetralogy of Fallot. [2]

3.3.1.1) Acyanotic Congenital Heart Disease: *The Left-to-Right Shunt Lesions*

Atrial Septal Defect

Atrial septal defects (ASDs) can occur in any portion of the atrial septum (secundum, primum, or sinus venosus), depending on which embryonic septal structure has failed to develop normally. The majority of cases of ASD are sporadic; autosomal dominant inheritance does occur as part of the Holt-Oram syndrome (hypoplastic or absent radii, 1st-degree heart block, ASD) or in families with secundum ASD and heart block. [2]

CLINICAL MANIFESTATIONS

A child with an ostium secundum ASD is most often asymptomatic; the lesion is often discovered inadvertently during physical examination. Even an extremely large secundum ASD rarely produces clinically evident heart failure in childhood. [2]

DIAGNOSIS

The chest roentgenogram shows varying degrees of enlargement of the right ventricle and atrium, depending on the size of the shunt. The pulmonary artery is enlarged, and pulmonary vascularity is increased. These signs vary and may not be conspicuous in mild cases. Cardiac enlargement is often best appreciated on the lateral view because the right ventricle protrudes anteriorly as its volume increases. [2]

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COMPLICATIONS

Secundum ASDs are usually isolated, although they may be associated with partial anomalous pulmonary venous return, pulmonary valvular stenosis, VSD, pulmonary artery branch stenosis, and persistent left superior vena cava, as well as mitral valve prolapse and insufficiency[(2)]

TREATMENT

Surgical or transcatheter device closure is advised for all symptomatic patients and also for asymptomatic patients with a Qp : Qs ratio of at least 2 : 1 or those with right ventricular enlargement. [(2)]

Partial Anomalous Pulmonary Venous Return*:*
One or several pulmonary veins may return anomalously to the superior or inferior vena cava, the right atrium, or the coronary sinus and produce a left-to-right shunt of oxygenated blood. Partial anomalous pulmonary venous return usually involves some or all of the veins from only one lung, more often the right one. When an associated ASD is present, it is generally of the sinus venosus type, although can be of the secundum type When an ASD is detected by echocardiography, one must always search for associated partial anomalous pulmonary venous return. [(2)]

Atrioventricular Septal Defects (Ostium Primum and Atrioventricular Canal or Endocardial Cushion Defects*:*
The abnormalities encompassed by AV septal defects are grouped together because they represent a spectrum of a basic embryologic abnormality, a deficiency of the AV septum. In most instances, a **cleft** in the **anterior leaflet** of the **mitral valve** is also noted. The tricuspid valve is usually functionally normal, although some anatomic abnormality of the septal leaflet is generally present. [2)]

CLINICAL MANIFESTATIONS

The physical signs are similar to those of the secundum ASD, but with an additional apical holosystolic murmur caused by mitral insufficiency. A history of exercise intolerance, easy fatigability, and recurrent pneumonia may be obtained, especially in infants with large left-to-right shunts and severe mitral insufficiency. [(2)]In these patients, cardiac enlargement is moderate or marked, and the precordium is hyperdynamic.

PATHOPHYSIOLOGY

The basic abnormality in patients with ostium primum defects is the combination of a left-to-right shunt across the atrial defect and mitral (or occasionally tricuspid) insufficiency, and pulmonary arterial pressure is typically normal or only mildly increased. The physiology of this lesion is therefore similar to that of an ostium secundum ASD [(2)]

DIAGNOSIS

Chest radiographs of children with complete AV septal defects often show moderate to severe cardiac enlargement caused by the prominence of both ventricles and atria. The pulmonary artery is large, and pulmonary vascularity is increased. The electrocardiogram in patients with a complete AV septal defect is distinctive. The principal abnormalities are (1) superior orientation of the mean frontal QRS axis with left axis deviation to the left upper or right upper quadrant, (2) counterclockwise inscription of the superiorly oriented QRS vector loop (often manifest by a Q wave in leads I and aVL), (3) signs of biventricular hypertrophy or isolated right ventricular hypertrophy, (4) right ventricular conduction delay (rSR□□pattern in leads V3R and V1), (5) normal or tall P waves, and (6) occasional prolongation of the P-R interval. The echocardiogram is characteristic and shows signs of right ventricular enlargement with encroachment of the mitral valve echo on the left ventricular outflow tract; the abnormally low position of the AV valves results in a "gooseneck" deformity of the left ventricular outflow tract. [(2)]

TREATMENT

Ostium primum defects are approached surgically from an incision in the right atrium. The cleft in the mitral valve is located through the atrial defect and is repaired by direct suture. The defect in the atrial septum is usually closed by insertion of a patch prosthesis. The surgical mortality rate for ostium primum defects is very low. Surgical treatment of complete AV septal defects is more difficult, especially in infants with cardiac failure and pulmonary hypertension. Because of the risk of **pulmonary vascular disease** developing as early as 6-12 mo of age, surgical intervention must be performed during infancy [(2)]

Ventricular Septal Defect
VSD is the most common cardiac malformation and accounts for 25% of congenital heart disease. Defects may occur in any portion of the ventricular septum, but most are of the membranous type. These defects are in a posteroinferior position, anterior to the septal leaflet of the tricuspid valve. VSDs between the crista supraventricularis and the papillary muscle of the conus may be associated with pulmonary stenosis and other manifestations of tetralogy of Fallot[(2)]

PATHOPHYSIOLOGY

The physical size of the VSD is a major, but not the only determinant of the size of the left-to-right shunt. The level of pulmonary vascular resistance in relation to systemic

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vascular resistance also determines the shunt's magnitude. When a small communication is present (usually □5 mm), the VSD is pressure **restrictive**, meaning that right ventricular pressure is normal. [2] The higher pressure in the left ventricle drives the shunt left to right and the size of the defect limits the magnitude of the shunt. In large **nonrestrictive VSDs** (usually □10 mm), right and left ventricular pressures are equalized. In these defects, the direction of shunting and the shunt magnitude are determined by the ratio of pulmonary to systemic vascular resistance [2]

CLINICAL MANIFESTATIONS

The clinical findings of patients with a VSD vary according to the size of the defect and pulmonary blood flow and pressure. Small VSDs with trivial left-to-right shunts and normal pulmonary arterial pressure are the most common. These patients are asymptomatic, and the cardiac lesion is usually found during routine physical examination. Characteristically, a loud, harsh, or blowing holosystolic murmur is present and heard best over the lower left sternal border, and it is frequently accompanied by a thrill. In a few instances, the murmur ends before the 2nd sound, presumably because of closure of the defect during late systole. [2]

DIAGNOSIS

The presence of right ventricular hypertrophy is a warning that the defect is not small and that the patient has pulmonary hypertension or an associated lesion such as pulmonic stenosis. In large VSDs, the chest x-ray shows gross cardiomegaly with prominenceof both ventricles, the left atrium, and the pulmonary artery .Pulmonary vascular markings are increased, and frank pulmonary edema, including pleural effusions, may be present.

TREATMENT

The vast majority of defects that close do so before the age of 4 yr, although spontaneous closure has been reported in adults. VSDs that close often have ventricular septal aneurysm (accessory tricuspid valve) tissue that limits the magnitude of the shunt. Most children with small defects remain asymptomatic, without evidence of an increase in heart size, pulmonary arterial pressure, or resistance. [2] The declining risk of open heart surgery has led others to suggest that all VSDs be closed electively by mid-childhood. [2]

Supracristal Ventricular Septal Defect with Aortic Insufficiency

A supracristal VSD is complicated by prolapse of the aortic valve into the defect and aortic insufficiency, which may eventually occur in 50-90% of patients. Although supracristal VSD accounts for □5% of all patients with VSD, the incidence is higher in Asian children. The right or, less often, the noncoronary aortic cusp prolapses into the defect and may partially or even completely occlude it. Such occlusion may limit the amount of left-to-right shunting and give the false impression that the defect is not large. Aortic insufficiency is most often not recognized until late in the 1st decade of life or beyond. Of note, aortic insufficiency is occasionally associated with VSDs located in the membranous septum [2]

PATHOPHYSIOLOGY

If the PDA is large, pulmonary artery pressure may be elevated to systemic levels during both systole and diastole. Thus, patients with a large PDA are at high risk for the development of pulmonary vascular disease if left unoperated[2]

CLINICAL MANIFESTATIONS

A small PDA is usually asymptomatic. A large PDA will result in heart failure similar to that encountered in infants with a large VSD. Retardation of physical growth may be a major manifestation in infants with large shunts.

The heart is normal in size when the ductus is small, but moderately or grossly enlarged in cases with a large communication. In these cases, the apical impulse is prominent and, with cardiac enlargement, is heaving. [2] A thrill, maximal in the 2nd left interspace, is often present and may radiate toward the left clavicle, down the left sternal border, or toward the apex. It is usually systolic but may also be palpated throughout the cardiac cycle.

Patent Ductus Arteriosus:

During fetal life, most of the pulmonary arterial blood is shunted right-to-left through the ductus arteriosus into the aorta. Functional closure of the ductus normally occurs soon after birth, but if the ductus remains patent when pulmonary vascular resistance falls, aortic blood then is shunted left-to-right into the pulmonary artery. [2]The aortic end of the ductus is just distal to the origin of the left subclavian artery, and the ductus enters the pulmonary artery at its bifurcation. Female patients with patent ductus arteriosus (PDA) outnumber males 2: 1. PDA is also associated with maternal rubella infection during early pregnancy, a now uncommon occurrence.

DIAGNOSIS

The diagnosis of an isolated, uncomplicated PDA is untenable when right ventricular hypertrophy is present. Radiographic studies in patients with a large PDA show a prominent pulmonary artery with increased pulmonary vascular markings. Cardiac size depends on the degree of left-to-right shunting; it may be normal or moderately to

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markedly enlarged. The chambers involved are the left atrium and left ventricle.

PROGNOSIS AND COMPLICATIONS

Spontaneous closure of the ductus after infancy is extremely rare. Patients with a small PDA may live a normal span with few or no cardiac symptoms, but late manifestations may occur. Rare complications include aneurismal dilatation of the pulmonary artery or the ductus, calcification of the ductus, noninfective thrombosis of the ductus with embolization, and paradoxical emboli. Pulmonary hypertension (Eisenmenger syndrome) usually develops in patients with a large PDA who do not undergo ductal closure [(2)].

TREATMENT

In patients with a moderate to large PDA, closure is accomplished to treat heart failure or prevent the development of pulmonary vascular disease, or both. Once the diagnosis of a moderate to large PDA is made, treatment should not be unduly postponed after adequate medical therapy for cardiac failure has been instituted [(2)]

Aortic regurgitation (AR) is the diastolic flow of blood from the aorta into the left ventricle (LV). Regurgitation is due to incompetence of the aortic valve or any disturbance of the valvular apparatus (eg, leaflets, annulus of the aorta) resulting in the diastolic flow of blood into the left ventricular chamber. (See Pathophysiology and Etiology.) [(2)]

Congenital causes - Bicuspid aortic valve is the most common congenital cause

Acquired causes:

- Rheumatic fever
- Infective endocarditis
- Collagen vascular diseases
- Degenerative aortic valve disease
- Traumatic
- Postsurgical (including post-trans catheter aortic valve replacement)

Abnormalities of the ascending aorta, in the absence of valve pathology, may also cause AR. [(2)] such abnormalities may occur with the following conditions:

- Longstanding, uncontrolled hypertension
- Marfan syndrome
- Idiopathic aortic dilation
- Cystic medial necrosis
- Senile aortic ectasia and dilation
- Syphilitic aortitis
- Giant cell arteritis
- Takayasu arteritis
- Ankylosing spondylitis
- Whipple disease
- Other spondyloarthropathies[(2)]

The most common cause of chronic aortic regurgitation used to be rheumatic heart disease, but presently it is most commonly caused by bacterial endocarditis. In developed countries, it is caused by dilation of the ascending aorta (eg, aortic root disease, aortoannular ectasia). [(2)]

Three fourths of patients with significant aortic regurgitation survive 5 years after diagnosis; half survive for 10 years. Patients with mild to moderate regurgitation survive 10 years in 80-95% of the cases. Average survival after the onset of congestive heart failure (CHF) is less than 2 years. (See Prognosis, Treatment, and Medication.) [(2)]

Acute aortic regurgitation is associated with significant morbidity, which can progress from pulmonary edema to refractory heart failure and cardiogenic shock. [(2)]

Pathophysiology

The pathophysiology of AR depends on whether the AR is acute or chronic. In acute AR, the LV does not have time to dilate in response to the volume load, whereas in chronic AR, the LV may undergo a series of adaptive (and maladaptive) changes. [(2)]

3.3.2.1) **Cyanotic Congenital Heart Disease*:* Tetralogy of Fallot*:***

Tetralogy of Fallot is one of the conotruncal family of heart lesions in which the primary defect is an anterior deviation of the arteries may be discontinuous. Pulmonary blood flow may be supplied by a patent ductus arteriosus (PDA) or by multiple **major aortopulmonary collateral arteries (MAPCAs)** arising from the ascending and descending aorta and supplying various lung segments. [(2)]

CLINICAL MANIFESTATIONS

Infants with mild degrees of right ventricular outflow obstruction may initially be seen with heart failure caused by a ventricular level left-to-right shunt. Often, cyanosis is not present at birth; but with increasing hypertrophy of the right ventricular infundibulum as the patient grows, cyanosis occurs later in the 1st yr of life. In infants with severe degrees of right ventricular outflow obstruction, neonatal cyanosis is noted immediately. [(2)]

DIAGNOSIS

The hypertrophied right ventricle causes the rounded apical shadow to be uptilted so that it is situated higher above the diaphragm than normal and pointing horizontally to the left chest wall. The cardiac silhouette has been likened to that of a boot or wooden shoe (“coeur en sabot”) [(2)]

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COMPLICATIONS

Thromboses occur most often in patients younger than 2 yr. These patients may have iron-deficiency anemia, frequently with hemoglobin and hematocrit levels in the normal range (but too low for cyanotic heart disease). Therapy consists of adequate hydration and supportive measures. Phlebotomy and volume replacement with albumin or saline are indicated in extremely **polycythemic** patients who are symptomatic. [2]

TREATMENT

Therapy is aimed at providing an immediate increase in pulmonary blood flow to prevent the sequelae of severe hypoxia. The infant should be transported to a medical center adequately equipped to evaluate and treat neonates with congenital heart disease under optimal conditions. Prolonged, severe hypoxia may lead to shock, respiratory failure, and intractable acidosis and will significantly reduce the chance of survival, even when surgically amenable lesions are present. [2]

Tricuspid Atresia

PATHOPHYSIOLOGY

In tricuspid atresia, no outlet from the right atrium to the right ventricle is present; the entire systemic venous return leaves the right atrium and enters the left side of the heart by means of the foramen ovale or, most often, through an atrial septal defect[2].

CLINICAL MANIFESTATIONS

The majority of patients have holosystolic murmurs audible along the left sternal border; the 2nd heart sound is usually single. Patients with tricuspid atresia are at risk for spontaneous narrowing or even closure of the VSD, which can occasionally occur rapidly and lead to a marked increase in cyanosis. [2]

DIAGNOSIS

Two-dimensional echocardiography reveals the presence of a fibro muscular membrane in place of a tricuspid valve, a variably small right ventricle, VSD, and the large left ventricle .The relationship of the great vessels (normal or transposed) can be determined. [2]

TREATMENT

The Blalock-Taussig procedure or a variation is the preferred anastomosis. Rare patients with restrictive atrial-level communications also benefit from a Rashkind balloon atrial septostomy or surgicalseptectomy. [2]

Transposition of the Great Arteries with Ventricular Septal Defect and Pulmonary Stenosis

This combination of anomalies may mimic tetralogy of Fallot in its clinical features. However, because of the transposed great vessels, the site of obstruction is in the left as opposed to the right ventricle.

The pulmonary vasculature as seen on the roentgenogram is dependent on the degree of pulmonary obstruction. The electrocardiogram usually shows right axis deviation, right and left ventricular hypertrophy, and sometimes tall, spiked P waves. Echocardiography confirms the diagnosis and is useful in sequential evaluation of the degree and progression of the left ventricular outflow tract obstruction. [2]

Ebstein Anomaly of the Tricuspid Valve

PATHOPHYSIOLOGY

In newborns, right ventricular function may be so compromised that it is unable to generate enough force to open the pulmonary valve in systole, thus producing "functional" pulmonary atresia. Some infants have true anatomic pulmonary atresia. The increased volume of right atrial blood shunts through the foramen ovale (or through an associated atrial septal defect) to the left atrium and produces cyanosis. [2]

CLINICAL MANIFESTATIONS

Newborns with severe forms of Ebstein anomaly have marked cyanosis, massive cardiomegaly, and long holosystolic murmurs. Death may result from cardiac failure, hypoxemia, and pulmonary hypoplasia. Spontaneous improvement may occur in some neonates as pulmonary vascular resistance falls and improves the ability of the right ventricle to provide pulmonary blood flow. The majority are dependent on a PDA, and thus on a prostaglandin infusion, for pulmonary blood flow [2].

DIAGNOSIS

The electrocardiogram usually shows a right bundle branch block without increased right precordial voltage, normal or tall and broad P waves, and a normal or prolonged P-R interval. **Wolff- Parkinson-White syndrome** may be present and these patients may have episodes of supraventricular tachycardia. On roentgen graphic examination, heart size varies from slightly enlarged to massive box-shaped cardiomegaly caused by enlargement of the right atrium. [2]

It may be difficult to distinguish true from functional pulmonary valve atresia. Cardiac catheterization, which is not usually necessary, confirms the presence of a large right atrium, an abnormal tricuspid valve, and any right-to-left

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shunt at the atrial level. The risk of arrhythmia is significant during catheterization and angiographic studies. (2)

PROGNOSIS AND COMPLICATIONS

The prognosis is more guarded for neonates or infants with intractable symptoms and cyanosis. Patients with milder degrees of Ebstein anomaly usually survive well into adult life. There is an association of a form of left ventricular cardiomyopathy, isolated left ventricular noncompaction, in 18% of patients with Ebstein anomaly, and the severity of the left ventricular dysfunction directly impacts the prognosis. (2)

TREATMENT

Neonates with severe hypoxia who are prostaglandin dependent have been treated with an aortopulmonary shunt alone, by repair of the tricuspid valve, or by surgical patch closure of the tricuspid valve, atrial septectomy, and placement of an aortopulmonary shunt (with eventual single ventricle repair using the Fontan procedure(2)

Total Anomalous Pulmonary Venous Return

PATHOPHYSIOLOGY

Total anomalous pulmonary venous return (TAPVR) is associated with total mixing of systemic venous and pulmonary venous blood flow within the heart and thus produces cyanosis. (2)

If surgery cannot be performed urgently, extracorporeal membrane oxygenation (ECMO) may be required to maintain oxygenation. Surgically, the pulmonary venous confluence is anastomosed directly to the left atrium, the ASD is closed, and any connection to the systemic venous circuit is interrupted. Early results are generally good, even for critically ill neonates. (2)

To date, the long-term prognosis in these patients is very guarded and in those with veno-occlusive disease **heart-lung transplantation** may be the only option (2)

Hypoplastic Left Heart Syndrome:

PATHOPHYSIOLOGY

The left ventricle may be moderately hypoplastic, very small and nonfunctional, or totally atretic; in the immediate neonatal period the right ventricle maintains both the pulmonary circulation and the systemic circulation via the ductus arteriosus. (2)

The major hemodynamic abnormalities are inadequate maintenance of the systemic circulation and, depending on the size of the atrial-level communication, either pulmonary venous hypertension (restrictive foramen ovale) or pulmonary overcirculation (moderate or large ASD). (2)

CLINICAL MANIFESTATIONS

A palpable right ventricular parasternal lift may be present along with a nondescript systolic murmur. This lesion may be isolated or associated in 5-15% of patients with known genetic syndromes, such as Turner syndrome, trisomy 13 or 18, Jacobsen syndrome (11q deletion), Holt-Oram syndrome, and Rubinstein-Taybi syndrome.

Diagnosis

The initial electrocardiogram show only the normal neonatal pattern of right ventricular dominance, but later, P waves become prominent and right ventricular hypertrophy is usual with reduced left ventricular forces. (2)

The small ascending aorta and transverse aortic arch are identified and a discrete coarctation of the aorta in the juxtaductal area may be present, although in the presence of a large ductus, it may be difficult to identify.(2)

PROGNOSIS AND COMPLICATIONS

Careful preoperative evaluation (genetic, neurologic, and ophthalmologic) should be performed in patients being considered for surgical therapy. Long-term follow up after the Norwood procedure demonstrates reduced neurodevelopmental outcomes and poor exercise tolerance (2)

TREATMENT

Primary heart transplantation, previously advocated by a few centers, is much less common due to the substantially improved survival rates with standard surgery and the limited supply of donor organs in this age group. (2)

(METHODS AND MATERIALS)

4.1) Study Design:
The research approach will be a qualitative - facility based descriptive case study with exploratory design. The aim of the study is to estimate the prevalence rates of different congenital heart diseases among Saudi children from neonatal period to two years in and MCH in Najran

4.2) Study Population:
They will be Saudis (male and female) children from neonatal period to two years old diagnosed with CHD in MCH with exclusion for those more than two years with CHD and non – Saudis children.

4.3) Study Area:

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It is located in Alathyba area and includes interior sections of the 200-bed hospital, including intensive sponsorships for children and children's surgery and care for premature and newborn, and sections of general and specialized children, also includes surgical services in the disciplines of children, brain and nerve surgery.[12]

4.4) Methods of Data Collection

A. Tools of data collection:
Data extraction sheet (checklist) will be used to collect the important information from patient's files and interview with mothers.

B. Sampling:
The study will be qualitative study based on the non-probability sampling by using the purposive and convenience techniques. Files of patients diagnosed with CHD from (1) – 1436H to (9) – 1436H will be selected for the study. The study population will be Saudis children (males and females) from neonatal period to two years old diagnosed with CHD in MCH to identify the magnitude of CHD and to determine the common lesion of CHD. Those who are more than two years old with CHD and non-Saudis children will be excluded from the study.

C. Study variables:

1. **Dependent variable:**
Types of congenital heart disease (cyanotic and Acyanotic)

2. **Independent variables:**
   a. Age (from neonatal period to two years) and Gender (male and female)
   b. consanguinity (first degree relation)
   c. Family history (pervious CHD in family members)
   d. maternal disease (rubella infection and DM)
   e. Maternal drug (warfarin and valporic acid)
   f. chromosomal abnormality (Down syndrome , Edwards syndrome and Patau syndrome)

4.5 Methods of Data Analysis:
The data will be analyzed by Master Sheet Program which is a software program Statistical Package for Social Sciences (SPSS).

(RESULTS)

As shown in table 1 the ratio of congenital heart disease between males and females children approximately equal with increase in male as:- males = 54.55% and females = 45.45%.

As shown in table 2 most of children with congenital heart disease their parents have consanguinity marriage as 68.2%.

As shown in figure I the rate of congenital heart disease is increase in age from 1 to 30 days as percent equal 40.9%

As shown in figure II the most common type of congenital heart disease is acyanotic as percent equal 72.73%.

As shown in figure III the maternal disease (diabetes mellitus) during pregnancy is cause congenital heart disease as percent equal 9.09%.

As shown in figure IV the most of congenital heart disease patients have positive chest x-ray as percent equal 54.55%.

As shown in figure V the most common lesion of congenital heart disease is atrial septal defect as percent equal 27.27%.

Table (1) Distribution of Congenital Heart Disease between males and females children in Maternity and Children Hospital:-

Gender	Frequency	Percent
Female	10	45.45
Male	12	54.55
Total	**22**	**100.0**

Table (2) The relation between consanguinity and Congenital heart disease:-

	Consanguinity	
	Frequency	Percent
No	7	31.8
Yes	15	68.2
Total	22	100.0

Figure (I):- Distribution of age among Congenital Heart Disease children in Maternity and Children Hospital:-

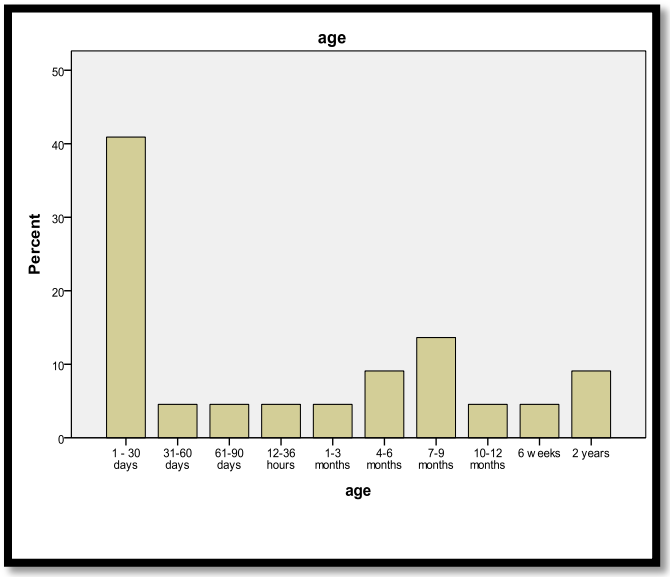

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Figure (II):- Show the distribution of Congenital Heart Disease types on Congenital Heart Disease children in Maternity and Children Hospital:-

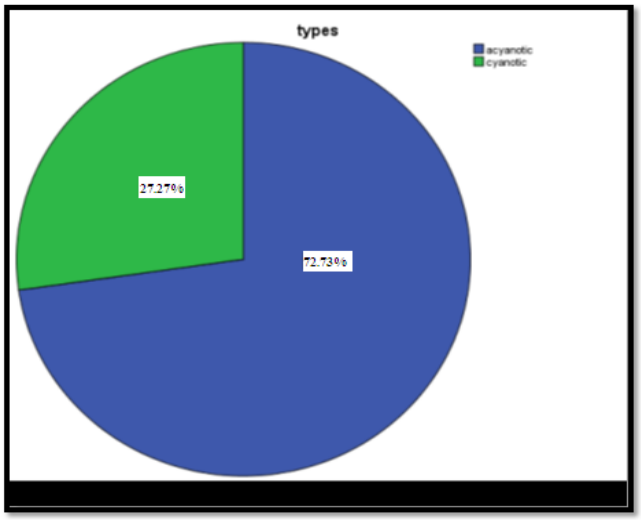


Figure (III):- The relation between maternal disease and Congenital Heart Disease children in Maternity and Children Hospital:-

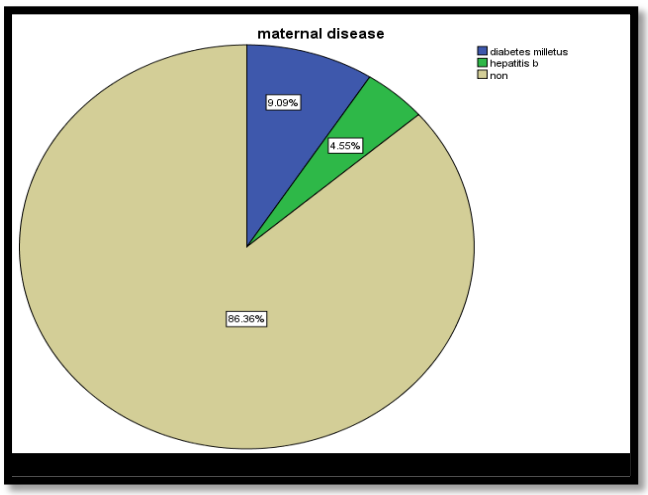


Figure (IV) :- Show the relation between Chest X ray and Congenital heart disease children in maternity and children hospital:-

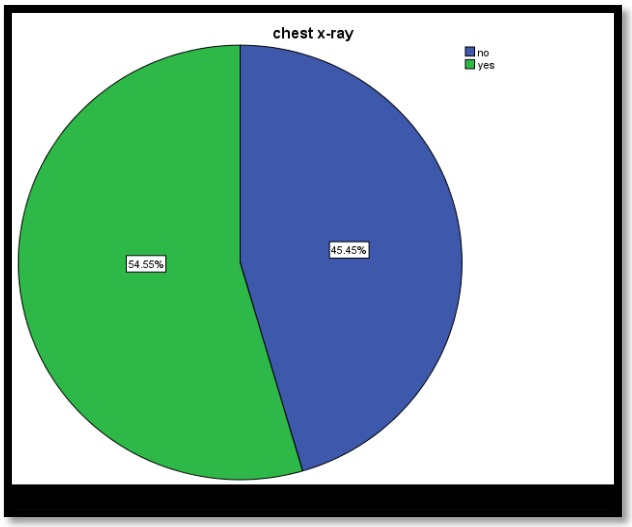
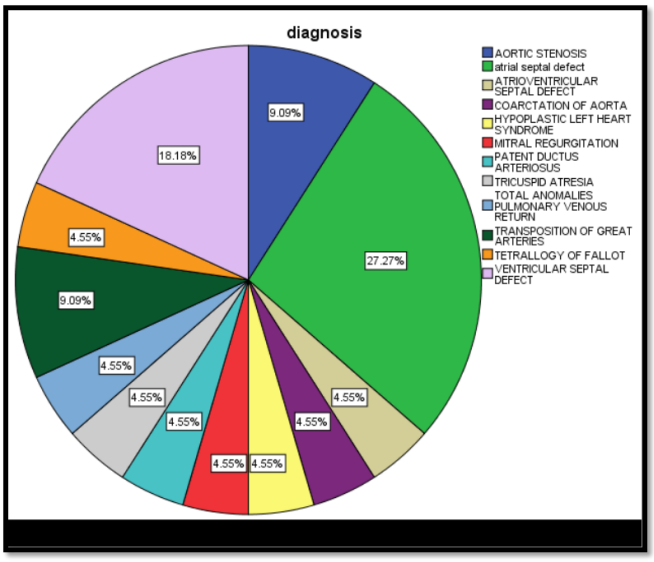


Figure (V):- Show the distribution of Congenital heart Disease lesions among children in maternity and children hospital:-



(DISCUSSION)

This study is shown the commonest lesion of congenital heart disease is atrial septal defect about 27.27% followed by ventricular septal defect 18.18%. Although studies have documented the commonest type of congenital heart disease lesion is ventricular septal defect 33.9% followed by atrial septal defect 18.1%. It is done in four regions in saudi Arabia (August 1988-February 2000).

In the other study done in Madina (May 2009-August 2009) the commonest type is ventricular septal defect 34.5% followed by atrial septal defect 8.9%.

Most of congenital heart disease children there age between from 1 to 30 days in our study. The other study show the age of congenital heart disease was less the 1 year (August 1988-February 2000).

According to the gender distribution of congenital heart disease in this study male more than female percent equal 54.55% the other study was done in (August 1988-February 2000) males are affected than females.

In this study the most common type of congenital heart disease is acyanotic as percent equal 72.73%

In this study the maternal disease (diabetes mellitus) during pregnancy is cause congenital heart disease as percent equal 9.09% followed by hepatitis BA as percent equal 4.55%.

In this study the most of congenital heart disease patients have positive chest x-ray as percent equal 54.55%.

In this study the most of children with congenital heart disease their parents have consanguinity marriage as 68.2%.

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This is the first report about congenital heart disease in Najran city.

(CONCLUSION)

Pediatric heart disease constitutes a major health problem in Najran.

The most common type of congenital heart disease in Children Hospital is acyanotic type.

The most commonest type of lesion is atrial septal defect.

The age affected by congenital heart disease is less than 1 year.

The consanguinity marriage have a major role in congenital heart disease.

(RECOMMENDATION)

Consanguinity marriage have an important role in congenital heart disease children the ministry of health should organize and awareness about the consanguinity marriage across the mass media to explain the serious problems that can occurs.

Prenatal detection program for assisting to help determine the extent of the problem and then easily handled after birth.

The majority have congenital heart disease need the pediatric cardiac center in Najran was emphasized to avoid problems of transportations.

ANNEXES:

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