Review Article

Pediatric Education and Research
Volume 5 Number 2, May - August 2017
DOI: https://dx.doi.org/10.21088/per.2321.1644.5217.17

Critical Congenital Heart Disease in Newborns

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Abstract

A congenital heart defect (CHD) is a structural or functional abnormality of heart that is present at birth. CHD accounts for nearly one-third of all major congenital anomalies. Screening infant with non-invasive measurement ofoxygen saturation bypulse oximetry has been proposed as an aid for early detection of duct dependentcirculation. Critical congenital heart defect (CCHD) is associated with hypoxemia among infants during the newborn period, and hypoxemia represents 17-31 percent of all CHDs. The CCHD screening include: hypoplastic left heart syndrome, pulmonary atresia (with intact ventricular septum), tetralogy of Fallot, total anomalous pulmonary venous return, and transposition of the great arteries with Intact Ventricular Septum. The risk of morbidity and mortality in CCHD increases when there is a delay in diagnosis and referral to a tertiary center. Babies with CCHDs usually require surgery or catheter intervention in the first year of life and represent more than one third of all CHD.

Keywords: Critical congenital heart defect; Hypoxemia; Neonates.

Introduction

Congenital heart disease (CHD) occurs in about 8 – 9 of every 1000 live births [1,2]. India has a high birth rate leading to high burden of CHD with over 1,80,000 children being born with a CHD [3]. Critical congenital heart diseases (CCHD) are CHD which require surgery or catheter intervention in the first year of life [1]. CCHD account for approximately 20 to 25% of the CHDs. CCHD is responsible for more deaths than any other type of malformation [1].

CCHD may present with -

1. Decreased pulmonary blood flow.
2. Decreased systemic blood flow.
3. Abnormal mixing.
4. Increased pulmonary blood flow.

Screening with pulse oximetry at birth can diagnose many asymptomatic babies with CCHD. These babies require an extensive workup for the diagnosis of the heart condition which includes Chest X ray, ECG, 2 D Echocardiogram, catheterization studies, CT scan and MRI studies. The main critical congenital heart diseases are Hypoplastic left heart syndrome, Pulmonary atresia with intact ventricular septum, Tetralogy of Fallot, Total anomalous pulmonary venous return, Transposition of the great arteries, Tricuspid atresia, Truncus arteriosus

Hypoplastic Left Heart Syndrome (HLHS)

Hypoplastic left heart syndrome refers to the abnormal development of the left-sided cardiac

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Received on 19.04.2017, **Accepted on** 09.05.2017

structures, resulting in obstruction to blood flow from the left ventricular outflow tract. It includes various degrees of underdevelopment of the left side of the heart: stenosis or atresia of the aortic and mitral valves and hypoplasia of the left ventricular cavity and ascending aorta. The severity of outflow obstruction, the left heart structures involved, and the degree of left ventricular and aortic hypoplasia, may vary among patients, resulting in a spectrum of patients with varying levels of severity [4]. HLHS has been reported to occur in approximately 0.016 to 0.036% of all live births.

Pathophysiology

Pulmonary venous blood enters the left atrium (LA). As the left outflow tract is obstructed, the blood passes through an atrial septal defect (ASD) or dilated foramen ovale from the left to the right side of the heart, where it mixes with systemic venous blood.The right ventricular blood is ejected into the main pulmonary artery; the descending aorta is supplied via the ductus arteriosus, and flow from the ductus also fills the ascending aorta and coronary arteries in a retrograde fashion. As the normal postnatal drop in pulmonary vascular resistance occurs, pulmonary flow increases at the expense of systemic flow. Thus inadequate maintenance of the systemic circulation and either pulmonary venous hypertension (restrictive foramen ovale) or pulmonary over circulation (moderate or large ASD) is seen.

Clinical Manifestations

Newborn infants generally are born at full term, and initially appear healthy. As the ductus begins to close, the systemic perfusion becomes decreased, resulting in hypoxemia, acidosis, and shock. Usually, no heart murmur, or a non-specific heart murmur, may be detected. The second heart sound (S2) is loud and single because of aortic atresia. Often the liver is enlarged secondary to congestive heart failure. It can be diagnosed by fetal echocardiography between 18 and 22 weeks of gestation.

Management

Patency of ductus is maintained by prostaglandin infusion. These children require surgical intervention soon. There are two major treatment modalities. These are primary cardiac transplantation, or a series of staged functionally univentricular palliations [5]. The functionally univentricular palliation typically includes three operations. The first stage of palliation, or the Norwood operation, is performed at birth. The second stage is the Glenn anastomosis which connects the superior vena cava to the pulmonary arteries, usually undertaken at 6 to 8 months of age. The third, and final, stage is the modified Fontan procedure, which can be performed between the ages of 18 months and 4 years.

- *Pulmonary Atresia with Intact Ventricular Septum*

In this condition, the pulmonary valves are completely fused or they fail to form with the result there is complete obstruction of right ventricular outflow with an intact ventricular septum. The condition is morphologically heterogeneous, with varying degrees of right ventricular and tricuspid valve hypoplasia. Incidence is 1% of all CHDs.

Pathophysiology

Systemic venous blood drains into the right atrium passes through the tricuspid valves to the right ventricle (RV). As there is no outflow path from the right ventricle it regurgitates back to the right atrium increasing the right atrial pressure and causing blood to pass through the foramen ovale or ASD to the left atrium where mixing takes place. This mixed blood now enters the left ventricle (LV) which is the only ventricle for both systemic and pulmonary circulation. The ductus arteriosus is the only channel for the pulmonary circulation. The main and branch pulmonary arteries are usually of good size; this is in contradistinction to pulmonary atresia with ventricular septal defect (VSD) where the pulmonary arteries are usually small and hypoplastic.

Dilated coronary sinusoids connecting to right or left coronary arteries usually have continuity with RV cavity from embryonic period. Coronary arteries that communicate with coronary sinusoids may be associated with either stenosis or atresia of their origins from the aortic root. This arrangement creates the so-called RV-dependent coronary circulation (RV-DCC), in which coronary circulation depends on the perfusion from the right ventricle and is driven by RV pressure. RV-DCC precludes decompression of the RV by opening the atretic pulmonary valve because this may lead to myocardial ischemia from reduction in RV pressure.

Clinical Manifestations

Infants are cyanotic at birth. As the pulmonary circulation is totally dependent on the ductus arteriosus which begins to close after birth these infants develop severe cyanosis and respiratory distress. Untreated, most patients die within the 1st wk of life. On auscultation there is a single and loud

S2. A continuous murmur of ductus may be present; also a holosystolic murmur of tricuspid regurgitation may be heard. Often, no murmurs are audible.

Investigations

- *Chest x ray:* The heart size is normal or mildly enlarged with prominent right atrial shadow. The pulmonary vascular markings are decreased.
- **ECG**: ECG shows tall and peaked P wave of right atrial enlargement. Due to lack of right sided forces, the frontal QRS axis is between 0 and +90 degrees; which in the setting of neonatal cyanosis with pulmonary oligemia is virtually diagnostic of pulmonary atresia with intact ventricular septum.

Treatment

Initially, prostaglandin E1 (PGE1) is used to maintain ductal patency until a more permanent source of pulmonary blood flow is provided.

Surgical Management [6]

The prognosis for patients with pulmonary atresia with intact ventricular septum is poor with and without conventional surgical treatment. The choice of surgical procedure depends on whether there is an RV dependent coronary circulation and on the size of the right ventricular cavity.

In patients with only mild to moderate right ventricular hypoplasia without RV dependent coronary circulation, a surgical pulmonary valvotomy is carried out to relieve outflow obstruction. The aim of surgery or interventional catheterization is to encourage growth of the right ventricular chamber by allowing some forward flow through the pulmonary valve while using the shunt to ensure adequate pulmonary blood flow. If the right ventricular chamber remains too small for use as a pulmonary ventricle, then the patient is treated as a single ventricle circulation, with a Glenn procedure followed by a modified Fontan procedure, allowing blood to bypass the hypoplastic right ventricle by flowing to the pulmonary arteries directly from the venae cavae. Few children are candidates for heart transplant surgery.

Tetralogy of Fallot (TOF)

It is the most common cyanotic cardiac defect, accounts for 7–10% of all congenital cardiac malformations.

Pathophysiology

Tetralogy of Fallot is one of the conotruncal family of heart lesions in which the primary defect is an anterior deviation of the infundibular septum along with abnormal morphology of the septoparietal trabeculations that encircle the subpulmonary outflow tract [7], the combination of this produces right ventricular outflow tract obstruction, malalignment type of ventricular septal defect (VSD), dextroposition of the aorta so that it overrides the ventricular septum and right ventricular hypertrophy.

The ventricular septal defect is typically large, unrestrictive, and subaortic, involving membranous septum. Obstruction to pulmonary arterial blood flow is usually at both the right ventricular infundibulum (subpulmonic area) and the pulmonary valve and it often increases with age in unrepaired patients. The main pulmonary artery may be small, and various degrees of branch pulmonary artery stenosis may be present.

Complete obstruction of right ventricular outflow (tetralogy with pulmonary atresia) is classified as an extreme form of tetralogy of Fallot. In about half of these patients, the blood to the pulmonary arteries is through the persistently patent arterial duct while in the other half it is from multiple aorto-pulmonary collateral arteries. When TOF is associated with atrial septal defects (15%) it is called pentology of Fallot. Right aortic arch is present in 25% of TOF cases.

Clinical Manifestations

The clinical presentation depends upon the degree of right ventricular outflow tract obstruction. Typically the infant may be not cyanotic as a neonate and develops cyanosis between 2 to 6 months of age. The modes of presentation are asymptomatic murmur, cyanosis observed by the parent; hypercyanotic spells, and decreased exercise tolerance.

Hypercyanotic spells are variously described as anoxic spells, hypoxic spells, blue spells. The peak incidence is in the first year of life. They are commonly seen after awakening from sleep; crying, defecation and feeding are the common precipitating factors. Spells are characterized by increasing rate and depth of respiration (hyperpnea) with increasing cyanosis, progressing to limpness and syncope, occasionally terminating in convulsions, cerebrovascular accident or death. The mechanism of onset of spells is still being debated. Right ventricular infundibular spasm, precipitated by acute increase in endogenous catecholamines has been proposed as a mechanism.

Another mechanism proposed is paroxysmal hyperpnea [8]. During sleep oxygen consumption is reduced and there is a normal acid base balance. When the infant awakens the O2 consumption increases and there is a slight acid base imbalance. There are adjustments made by the respiratory center to bring the imbalance back to normal. But, if there is a sudden increase in activity and consequent increase in oxygen consumption before the above described adjustments occur, decrease in PO2 and pH and increase in PCO2 take place triggering a hyperpnea response from the respiratory center and enter a vicious cycle. The hyperpnea will increase cardiac output and decrease pulmonary blood flow, resulting in further right-to-left shunting and greater arterial hypoxemia and thus a vicious cycle.

Clinical Examination

Central cyanosis is observed in most cases with clubbing of fingers and toes observed beyond the first few months of life. Prominent right ventricular impulse or heave may be present. The S2 is single without an audible pulmonary component. Blood flow through the right ventricular outflow tract gives rise to a long, ejection, systolic murmur, heard at the left upper sternal border. During hypercyanotic spell the murmur disappears or becomes very soft. Older children may have an audible continuous murmur of bronchial collateral flow into the lungs, usually heard on the back.

Investigations

Chest X ray: Demonstrate a boot-shaped cardiac silhouette and pulmonary oligemia. The boot shape is due to upward displacement of the right ventricular apex as a consequence of the right ventricular hypertrophy, and a narrowing of the mediastinal shadow due to the hypoplastic pulmonary outflow tract (Figure 1).

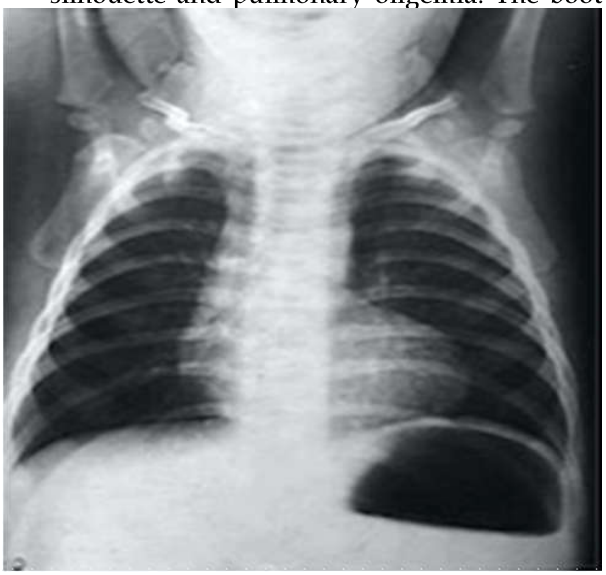

Fig. 1: Chest X ray in TOF showing boot-shaped cardiac silhouette and pulmonary oligemia

ECG: Right axis deviation and right ventricular hypertrophy.

Treatment

1. *Cyanotic spell:*

a. The infant should be placed in a knee-chest position which increases the systemic vascular resistance.

b. Correction of metabolic acidosis (with sodium bicarbonate), anemia (by blood transfusion), and dehydration (by appropriate fluids).

c. If the spell continues, vasopressors like methoxamine an alpha agonist or phenylephrine to increase the systemic vascular resistance has been most helpful.

d. Propranolol, 0.1 mg/kg body weight, can be used.

e. Rarely, general anesthesia may be necessary to break the spell.

f. Lastly, an emergency systemic-to pulmonary artery shunt (modified Blalock-Taussig anastomosis) may be performed if the infant continues to be in spell.

2. *Palliative Surgery:* Augments pulmonary blood flow to allow the patients to grow into an age, size and anatomy that are more likely suitable for complete correction. Classic or modified Blalock-Taussig shunt are options. Most surgeons prefer modified Blalock-Taussig shunt using an interposition Gore-Tex graft between right or left subclavian arteries to the ipsilateral pulmonary artery.

3. *Corrective Surgery:* Total surgical correction to include closure of VSD in such a manner as to direct left ventricular output into the aorta and resection of the infundibulum and/or relief of pulmonary valvar obstruction. Enlargement of the right ventricular outflow tract with a pericardial patch may be necessary in some cases.

4. *Transposition of Great Arteries (TGA)*

In this congenital cardiac malformation the morphological right atrium is connected to the morphological right ventricle which gives rise entirely to or most of the aorta; the morphological left atrium is connected to the morphological left ventricle from where the pulmonary trunk emerges, thus it is atrioventricular concordance and ventriculoarterial discordance. In d-

transposition, the aortic valve lies to the right of the pulmonary valve. It accounts for 5% of all CHD and 10% of all neonatal cyanotic CHD.

The term **congenitally corrected transposition** of the great arteries describes a different entity that conjugates atrioventricular and ventriculoarterial discordance [9]. In congenitally corrected hearts, the aorta usually lies on the left (l-transposition). The a-transposition refers to the anterior position of the aortic valve in relation to the pulmonary trunk.

Pathophysiology

In 50% of the cases, the ventriculoarterial discordance is an isolated finding. This condition is designated as simple transposition. Complex transposition includes all the cases with coexisting malformations, such as ventricular septal defects, left ventricular outflow tract obstruction, aortic arch anomalies, and anomalous venous systemic return.

TGA is divided in three anatomic types, namely Group I, TGA with intact ventricular septum; Group II, TGA with VSD, and Group III, TGA with VSD and PS. The coronary anatomy in transposition of the great arteries is important since the location of the coronary arteries influences the outcome of the arterial switch procedure [10].

The systemic and pulmonary circulations run in parallel, rather than in series. Deoxygenated blood returning to the right atrium flows to the right ventricle and is circulated to the body and it comes back to the right atrium, while the oxygenated blood flows from left atrium to the left ventricle to the lungs and back to the left atrium. Survival is only possible, if there is adequate mixing between the two circulations via the septal defects or through the arterial duct.

Clinical Manifestations

Clinical features depend upon the anatomic type, namely Group I, TGA with intact ventricular septum; Group II, TGA with VSD, and Group III, TGA with VSD and PS (11).

1. In group I, the infants usually present with cyanosis within the first week of life. They may otherwise be asymptomatic. However, they will, with time, become tachypnoeic and develop respiratory distress. If they are not appropriately treated, they become acidotic and go on to become lethargic without lack of spontaneous movement, and eventually die. Clubbing may develop at 3 to 6 months. The right ventricular impulse is increased and the second heart sound is single. Either no murmur or a soft ejection systolic murmur may be heard.
2. Group II present with symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) between 4 to 8 weeks of life, but the cyanosis is minimal. Examination reveals increased right and left ventricular impulses, single second sound, a grade III-IV holosystolic murmur at the left lower sternal border and a mid-diastolic flow rumble (murmur) at the apex may be present.
3. Group III patients have variable presentation, depending upon the severity of PS. If there is poor mixing, they may present early in life and mimic TGA with intact septum. If the PS is severe, the presentation is essentially similar to that described in the TOF. With moderate PS the presentation is late with longer survival. With mild PS, congestive heart failure signs may be present, similar to group II patients.

Investigations

Chest X-ray

1. In the intact septum group normal to minimal cardiomegaly and normal to slightly increased pulmonary vascular marking. The thymic shadow may rapidly involute and a narrow pedicle (superior mediastinum) may be seen giving rise to characteristic "egg on a string" appearance (Figure 2).

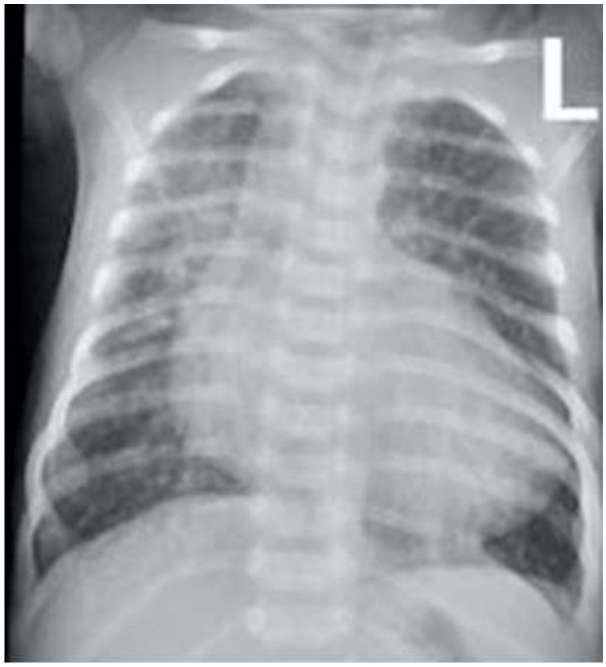

Fig. 2: Chest X ray in TGA showing egg on a string appearance.

2. In group II patients with VSD, moderate to severe cardiomegaly and increased pulmonary vascular markings are usually seen.
3. In group III patients, mild to moderate cardiomegaly may be observed. The pulmonary vascular marking may be increased, normal or decreased, dependent upon the severity of PS.

ECG:

1. In group I, ECG may be normal in early infancy but over time right ventricular hypertrophy and right atrial enlargement may be seen.
2. In group II patients, biventricular hypertrophy and left atrial enlargement are usual.
3. In group III, right ventricular or biventricular enlargement is seen.

Treatment

Palliative treatment with prostaglandin E1 and balloon atrial septostomy are usually required soon after birth. The Jatene arterial switch with relocation of the coronary arteries is the procedure of choice. In this surgery, the aorta and pulmonary trunks are sectioned and their distal extremities are transposed and anastomosed; coronary arteries are then translocated to the neo-aorta.

In cases with TGA with intact septum, this surgery should be preferably performed within two weeks of life. If Jantene arterial switch is not feasible atrial switch (Mustard or Senning procedure) or the REV procedure or its modification and the Rastelli operation [12].

TGA patients with VSD and PS may have varying types of presentation. If the reason for hypoxemia is poor mixing, balloon atrial septostomy is the treatment of choice. If the hypoxemia is secondary to decreased pulmonary flow, a Blalock-Taussig type of shunt may be needed. Sometimes both balloon septostomy and balloon dilatation of pulmonary valve may be performed via catheters in some of these children. Eventually these patients require a Rastelli type of repair.

- *Total Anomalous Pulmonary Venous Return (TAPVR)*

In total anomalous pulmonary venous return (TAPVR) the pulmonary veins return to a systemic vein or directly to the right atrium, rather than the left atrium. It accounts for 1 -2 % of congenital heart disease.

Types of TAPVR [13]

1. Supracardiac total anomalous pulmonary venous return 50% - All the pulmonary veins collect in a common channel behind the left atrium. From there, blood courses upward to the left innominate vein, then to the superior vena cava, and reenters the heart at the right atrium. All blood entering the left atrium passes from the right to the left atrium. Obstructed pulmonary venous return occurs in about 50% of the cases.
2. Infradiaphragmatic total anomalous veins 20% - All the pulmonary veins collect behind the left atrium and, through a common channel, proceed through the diaphragm to the portal or hepatic vein and ductus venosus, and from there to the inferior vena cava and to the right atrium. Obstruction of the pulmonary venous return is seen in more than 95% of the cases
3. Total anomalous pulmonary venous returns to the coronary sinus 25% - All the pulmonary veins collect behind the left atrium and enter the coronary sinus. The pulmonary venous blood with coronary sinus blood drains into the right atrium. Chances of pulmonary venous return obstruction are the least in this group of about 5 -10%.
4. Mixed – accounts for about 5% of cases.

In all forms of TAPVR, the entire oxygenated blood from the lungs through the pulmonary veins is completely mixed with the systemic blood before or at the level of the right atrium. The left atrium receives blood only through the ASD or patent foramen ovale. The clinical manifestations of TAPVR depend on the presence or absence of obstruction of the venous channels. If pulmonary venous return is obstructed, severe pulmonary congestion and pulmonary hypertension develop; rapid deterioration occurs without surgical intervention. Obstructed TAPVR is a pediatric cardiac surgical emergency because prostaglandin therapy is usually not effective.

Clinical Manifestations

The clinical presentation in TAPVR varies, depending on the presence or absence of obstruction. In cases with severe pulmonary venous obstruction, the presentation is in the neonatal period with marked cyanosis and severe respiratory distress. These are severely ill and fail to respond to mechanical ventilation.

On auscultation usually no murmur is present and the S2 is single and loud. Children without pulmonary venous obstruction have minimal

symptoms in the neonatal period. Cyanosis is usually not apparent. Overtime they present with right ventricular failure due to volume overload. Physical examination may reveal a right ventricular heave, and features of similar to those seen in secundum atrial septal defect i.e. fixed split S2 and a soft systolic ejection murmur in the pulmonic area.

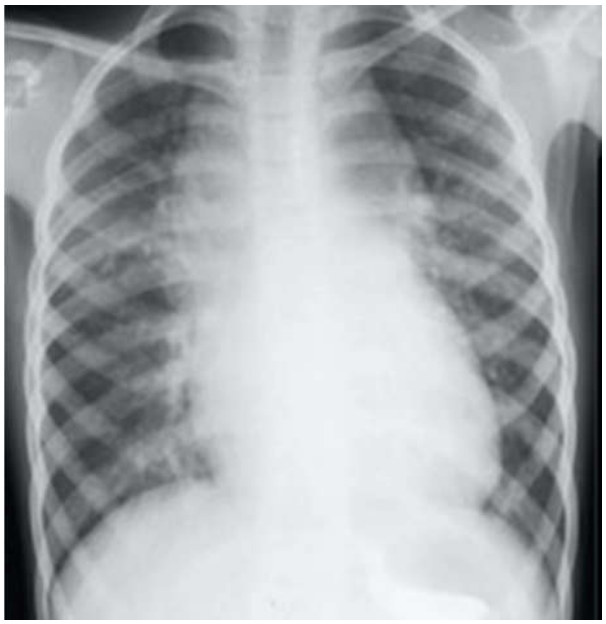

Fig. 3: Chest X ray in TAPVR showing "snowman" appearance

Investigations

- *Chest X ray:* In neonates with marked pulmonary venous obstruction, the heart shadow is small and pulmonary edema is present. In older children, **"snowman"** appearance may be seen if the anomalous pulmonary veins enter the innominate vein and persistent left superior vena cava which form a large supracardiac shadow (Figure 3). In most cases without obstruction, the heart is enlarged, the pulmonary artery and right ventricle are prominent, and pulmonary vascularity is increased.
- **ECG**: Right ventricular hypertrophy.

Treatment

Emergency repair surgery is performed in cases with pulmonary venous obstruction. Surgical correction is by anastomosis of the common pulmonary vein with the left atrium and closure of ASD.

Tricuspid Atresia

Tricuspid atresia is characterized by absence or agenesis of the morphologic tricuspid valve of and hypoplasia of the right ventricle. It accounts for 1% of subjects with CHD.

Unified Classification of Tricuspid Atresia (14):

Type I: Normally related great arteries

Type II: D-transposition of the great arteries

Type III: Malpositions of the great arteries other than D-transposition

Type IV: Persistent truncus arteriosus

Pathophysiology

In patients with tricuspid atresia, systemic blood enters the right atrium. As there is no connection between the right atrium and right ventricle, the entire blood flows from the right atrium into the left atrium in most cases through an atrial septal defect or sometimes via foramen ovale. In patients with normally related great arteries (type I: about 80%) entry to the pulmonary circulation occurs through a ventricular defect and a hypoplastic right ventricle. Pulmonary blood flow may be augmented by or be totally dependent on a PDA in absence of a ventricular defect. The ventricular septal defect is usually perimembranous, less frequently muscular (single or multiple) in location. Usually, the ventricular defect is small, tends to get smaller with time, and may ultimately close. The ductus arteriosus usually has a small diameter and closes on schedule.When great arteries are transposed (type II), the ventricular defect carries blood to the aorta: thus restriction at this level results in subaortic stenosis. Aortic coarctation is more common in this group. Excessive pulmonary blood is more likely in these patients than in those with normally related great arteries.

Clinical Manifestations

About 50% of patients with TA present on the first day of life and around 75 - 80% will have symptoms in the neonatal period. Infants with pulmonary oligemia present with symptoms of cyanosis within the first few days of life. Infants with increased pulmonary blood flow usually present with minimal cyanosis and signs of heart failure within the first few weeks of life.

Recurrent respiratory tract infections and failure to thrive are other modes of presentation. Clinical examination may reveal an increased left ventricular impulse. This is in contrast to most other causes of

cyanotic heart disease, in which an increased right ventricular impulse is usually present. The majority of patients have holosystolic murmurs audible along the left sternal border; the 2nd heart sound is usually single.

Investigations

- *Chest X ray*: Pulmonary oligemia or plethora.
- *ECG*: Left axis deviation and left ventricular hypertrophy are generally noted on the electrocardiogram (except in those patients with transposition of the great arteries), and these features distinguish tricuspid atresia from most other cyanotic heart lesions.

Treatment

PGE1 is used initially to maintain a PDA. If there is restriction to flow at the atrial level, a balloon septostomy is performed. Patients with ductal dependent pulmonary blood flow have a systemic-to-pulmonary artery shunt placed (modified Blalock-Taussig shunt). Those with adequate pulmonary blood flow undergo a superior vena cava–to–pulmonary artery anastomosis (bidirectional Glenn) at approximately 6 months of age, with completion of the Fontan procedure around 3 years of age.

Truncus Arteriosus

Truncus arteriosus is characterized by a single arterial vessel that originates from the heart, overrides the ventricular septum, and supplies the systemic, coronary, and pulmonary circulations.

The pulmonary artery arises from the truncus and forms the basis of classification [15]:

Type I (50 – 70 %): The main pulmonary artery (usually short) arises from the side of the Truncus (ascending aorta) and divides into right and left pulmonary arteries.

Type II (30 to 50%): The right and left pulmonary arteries arise from the posterior aspect of the truncus, most commonly as separate vessels.

Type III (6 to 10%): The pulmonary arteries arise from the lateral aspect of the truncus separately.

Type IV: Is no longer considered as a part of truncus arteriosus. Pulmonary blood flow is derived from the ductus arteriosus and/or multiple aorto-pulmonary collateral vessels from the descending aorta are described as pulmonary atresia with VSD.

The number of truncal valve cusps varies from 2 to as many as 6 and the valve may be stenotic, regurgitated, or both. In rare cases, one pulmonary artery arises from the ascending aorta and the other comes off the right ventricle, the so called hemi-truncus. Right aortic arch is present in nearly 40% of cases.

Clinical Manifestations

Systemic venous blood from right atrium through right ventricle and pulmonary venous blood from the left atrium through the left ventricle is ejected into the truncus where complete mixing takes place. Initially the pulmonary vascular resistance is high and pulmonary blood flow is adequate. Clinical examination reveals mild cyanosis, hyperdynamic precordium and a loud single S2. Slowly as the pulmonary vascular resistance begins to fall, pulmonary blood flow increases and signs of heart failure develops. The pulses now become bounding and the pulse pressure widens because of runoff into the pulmonary arteries. A systolic ejection murmur sometimes accompanied with a thrill is heard along the left sternal border. A low-pitched diastolic inflow rumble may be heard at the apex because of increased blood flow across the mitral valve. If the truncal valves are regurgitant a medium- to high-pitched diastolic murmur may be heard from truncal valve insufficiency.

Investigations

- *Chest x-ray*: Initially normal, but over the first few weeks of life, increase in cardiac shadow and pulmonary vascularity is seen.
- *ECG*: Right, left, or combined ventricular hypertrophy.

Treatment

Surgery is performed in the first two months of life. The usual repair involves closure of the ventricular septal defect so that the arterial trunk arises from the left ventricle. A right ventricle to pulmonary artery conduit is then placed. This conduit needs to be replaced several times over the course of the patient's life.

Double outlet Right Ventricle (DORV)

DORV is a type of ventriculoarterial connection in which both great vessels arise either entirely or predominantly from the right ventricle [16].

Pathophysiology

The only exit for the left ventricular blood is through a large VSD. The position of this VSD determines the pathophysiology. If the VSD is perimembraneous and subaortic, the left ventricular blood is directed to the aorta. This is the most common form of DORV (50%). In absence of pulmonary stenosis, the clinical manifestations are similar to those of VSD and if significant pulmonary stenosis is present the features are those of tetralogy of Fallot. In 25% of cases of DORV, Taussig-Bing malformation is present which refers to cases where the VSD is subpulmonary, so the left ventricular blood is directed mainly to the pulmonary trunk. The clinical picture is now similar to that of transposition of the great arteries. In the remaining 25% cases, the VSD is either doubly committed to both vessels or noncommitted to either vessel.

Management

The management depends on the type of DORV. Surgical correction depends on the relationship of the great vessels to the VSD. If the VSD is subaortic, the repair may be similar to that used for tetralogy of Fallot, or consist of creating an intraventricular tunnel so that the left ventricle ejects blood through the VSD, into the tunnel, and into the aorta. The pulmonary obstruction is relieved either with an outflow patch or with a right ventricular to pulmonary artery homograft conduit (Rastelli operation). If the VSD is subpulmonic, the great vessels can be switched and the Rastelli operation performed

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