CONGENITAL HEART DISEASE

INTRODUCTION:

Today, many of the congenital cardiac anomalies are being corrected surgically or by means of interventional cardiology in infancy, permitting survival to adult life. It is important for students to understand the mechanisms and hemodynamics in various types of common congenital cardiac anomalies. This lecture is to introduce the field of congenital heart disease and thereby create enthusiasm in the younger generation to enter this dynamic and evolving area in cardiology.

APPROACH TO CONGENITAL HEART DISEASE

1. Individual anomaly.

2. The complex. The term complex implies a single abnormality or a group of abnormalities that have a tendency to be associated, and the effects of that individual abnormality or group of abnormalities on the economy of the heart. Effects on economy of the heart includes those on the myocardium, endocardium, valves and the conduction system.

CLASSIFICATION

1. Shunt. A shunt is a transfer of blood from one side of the circulation to the other through an abnormal pathway.

2. Obstruction
   A right side
   B left side.

3. Shunt with obstruction.

4. Other complexes.

SHUNTS

At the atrial level - Atrial septal defect - ASD

At the ventricular level - Ventricular septal defect - VSD

At the ductal level - Patent ductus arteriosus - PDA
ATRIAL SEPTAL DEFECT

Defect in the atrial septum - types of defects:

1. Fossa ovalis or secundum type - common
2. Ostium primum type
3. Sinus venosus or proximal type of atrial septal defect - uncommon
4. Coronary sinus type of atrial septal defect - rare

LEFT TO RIGHT SHUNT AT ATRIAL LEVEL

ASD - the complex. The pressure in the left atrium is slightly higher than that of the right atrium and the compliance of the right ventricle is far greater than that of the left ventricle. Therefore, a shunt at the atrial level will produce volume hypertrophy of the right atrium and right ventricle with increased pulmonary flow and hemodynamic changes in the tricuspid and pulmonic valves.

LEFT TO RIGHT SHUNT AT VENTRICULAR LEVEL - VENTRICULAR SEPTAL DEFECT

Ventricular septal defect can occur anywhere in the ventricular septum. However, there is a predilection for the defect to occur beneath the aortic valve, confluent in part with the membranous septum and extending anteriorly to some extent. Hence, the most common type of a defect is called subaortic, in part membranous and in part perimembranous type. The complex is characterized by volume hypertrophy of the right ventricle, enlargement of the pulmonic orifice, hemodynamic changes of the tricuspid and pulmonic valves, volume hypertrophy of the left atrium and the left ventricle with enlargement of the mitral valve and hemodynamic changes in the mitral and aortic valves.

NATURAL HISTORY

Small to moderate size defects in the ventricular septum have a tendency to close spontaneously in the first few years of life. There is a tendency for infective endocarditis to occur in small or a closing ventricular septal defect.

LARGE DEFECT
When the defect is large, there is not only volume hypertrophy of the right ventricle, left atrium and left ventricle, the pressure of the left ventricle may be transmitted to the right ventricle resulting in pressure hypertrophy of the right ventricle.

**LEFT TO RIGHT SHUNT AT DUCTUS LEVEL: PATENT DUCTUS ARTERIOSUS - PDA**

In this entity, there is volume hypertrophy of the left atrium and left ventricle.

In all of the above three shunts there is increased pulmonary flow.

**PULMONARY HYPERTENSION ASSOCIATED WITH SHUNTS**

Despite the left to right shunt at the atrial, ventricular and ductal level with increased pulmonary flow, the greater distensibility of the pulmonary vascular tree is able to accommodate with increasing the pressure in the pulmonary circuit. However, pulmonary hypertension may develop for one or all of the following reasons:

1. Flow beyond the distensibility of the lung vasculature.
2. Vasoconstriction of pulmonary vascular bed.
3. Secondary pathologic changes in the intima and media of the muscular arteries and arterioles of the lungs restricting the pulmonary bed.

Thus, pulmonary hypertension may occur, most commonly in large ventricular septal defect, less commonly in patent ductus arteriosus and least commonly in atrial septal defect.

The pathologic effects of pulmonary hypertension of the heart following a left to right shunt are related to increase in pressure of the right side with or without decreased flow to the left side of the heart. This will result in pressure hypertrophy on the right side with decrease in the previously present volume hypertrophy of the left side. This may produce volume atrophy on the left side. When this occurs, there is usually a reversal of shunt from left-to-right to right-to-left at the ventricular level or at the ductal level, and least commonly at the atrial septal defect level. This is called the physiology of Eisenmenger’s complex.

**OBSTRUCTION**

The entire circulation is characterized by passage of blood through narrower and wider channels. The narrower channels are not necessarily obstructions. An obstruction is one that goes beyond the limits of physiologic narrowing. This usually occurs when anatomically the size of the orifice is narrowed more than 65%.
RIGHT SIDE
EXAMPLE: PULMONARY STENOSIS.

The pulmonic valve usually consists of a diaphragm-like structure with an attempted formation of cusps with a central opening, which may be minute or small. Uncommonly, the valve is fairly well formed, but the cusps are agglutinated at the commissures. The annulus of the pulmonic valve is quite small. There is pressure hypertrophy of the right ventricle, often associated with poststenotic dilatation of the pulmonary trunk and hemodynamic changes in the tricuspid valve.

LEFT SIDE
EXAMPLE: AORTIC STENOSIS:

1. At the valvular level.
2. Above the valve - supravalvular.
3. Below the valve - subvalvular.

The valve cusps may be bicuspid or unicuspid and may show irregular thickening termed as dysplastic valve.

SUPRAVALVULAR AORTIC STENOSIS

There are two types - one consists of thickening and accentuation at the normal supravalvular aortic ridge at the upper margins of the sinuses of Valsalva. The other consists of a ridge of thickening about a centimeter above the sinuses of Valsalva.

SUBAORTIC STENOSIS

There are several types of subaortic stenosis. Usually there is fibroelastic tissue beneath the aortic valve extending from the anterior muscular ventricular septum to the aortic leaflet of the mitral valve.

MUSCULAR TYPE

Hypertrophic cardiomyopathy is an autosomal dominant pattern of inherited genetic disease. Here there is asymmetrical septal hypertrophy of the muscular septum. This may produce subaortic obstruction and sudden death.

THE COMPLEX IN VARIOUS TYPES OF AORTIC STENOSIS
In aortic stenosis there is usually pressure hypertrophy of the left ventricle with the left ventricle being either smaller than normal or normal in size. Often the left atrium is hypertrophied with hemodynamic changes of the mitral valve.

**COARCTATION OF THE AORTA**

There is narrowing of the transverse arch in the region of the isthmus. The isthmus is the segment between the origin of the left subclavian artery and ductus or ligamentum arteriosus.

**PARADUCTAL OR SEGMENTAL OR LOCALIZED OR ADULT-TYPE OF COARCTATION OF THE AORTA**

Here there is maximum point of localized area of narrowing just proximal to or just distal to the ductus arteriosus or ligamentum arteriosus. This is associated with pressure hypertrophy of the left ventricle and in some cases of the left atrium. There is hypertension proximal and hypotension distal to the area of narrowing. Collateral anastomoses may develop, bypassing the area of narrowing between the proximal and distal portions of the aorta by way of the subclavian, dorsal scapular, internal mammary and intercostal arteries.

**HYPOPLASIA OF TRANSVERSE ARCH WITH COARCTATION OF THE AORTA OR FETAL COARCTATION**

There is narrowing of the transverse arch that involves the entire transverse arch or may extend from the left common carotid artery to the left subclavian artery and the left subclavian may emerge at the junction of the transverse arch with the ductus arteriosus. The ascending aorta is smaller than normal. In this entity, there is usually an atrial septal defect of a fossa ovalis type and a widely patent ductus arteriosus that usually forms the descending aorta. There is volume hypertrophy of the right ventricle and volume atrophy of the left side of the heart. It is assumed that in this complex, the smallness of the left side and largeness of the right side are caused by insufficient flow reaching the left side of the heart and more blood reaching the right side of the heart in intrauterine life. The small amount of blood entering the ascending aorta results in a small ascending aorta and an even smaller isthmus. Thus, this type of a coarctation, in many cases, may be considered the result of insufficient flow to the left side of the heart before birth. After birth there may be left to right shunt at the atrial level and right to left shunt at the ductus level.

**SHUNT WITH OBSTRUCTION - TETRALOGY OF FALLOT**

Consists of:

1. Infundibular pulmonary stenosis.
2. Right ventricular hypertrophy.
3. Ventricular septal defect.

4. Overriding aorta.

The outflow tract of the right ventricle is called the infundibulum which consists of septal and parietal groups of muscle bands. The parietal and the septal group of muscles may be abnormally oriented and markedly hypertrophied resulting in infundibular pulmonary obstruction. The ventricular septal defect is a U shaped deformity of the ventricular septum confluent with the aortic valve. Thus, the aorta overrides the defect emerging from both the ventricular chambers.

There are two types of tetralogy:

1. Cyanotic - the common type.

2. Acyanotic.

**CYANOTIC TETRALOGY OF FALLOT – COMMON TYPE**

Here there is pressure hypertrophy of the right atrium and right ventricle with significant infundibular pulmonary obstruction. The left atrium and left ventricle have a tendency to be smaller than normal. The right ventricle is contracting against systemic and infundibular resistance, decreased pulmonary flow, and predominant right to left shunt at the ventricular level.

**ACYANOTIC TETRALOGY OF FALLOT – UNCOMMON TYPE**

In acyanotic, tetralogy there is pressure and volume hypertrophy of the right ventricle, pressure hypertrophy of the right atrium, volume hypertrophy of the left atrium and left ventricle and enlargement of the mitral and aortic orifices. Here we are dealing with a left to right shunt at the ventricular level with increased pulmonary flow and increased volume to the left side of the heart.