INTRODUCTION

The concept of univentricular heart moved from hearts with only one ventricle to hearts with two ventricles unable to sustain separately pulmonary and systemic circulations in sequence ***(Frescura and Thiene, 2014).***

Surgical correction of the univentrivular heart is an achievement of the last 4-5 decades. A Fontan-type operation, involving connection of the systemic veins and pulmonary arteries without subpulmonary ventricle, is nowadays the only treatment option for patients with a functionally univentricular heart (UVH) ***(Andrzej et al., 2013).***

Since the first report in 1971 by Fontan and Baudet ***(Fontan and Baudet, 1971)***, the results of Fontan-type operations have improved significantly due to better understanding of the Fontan circulation, better patient selection, and appropriat pre-Fontan palliative procedures. The multi-stage approach most often performed by means of pulmonary arterial blood flow control procedures (Blalock-Taussig shunt or pulmonary artery banding) followed by the bidirectional Glenn (BDG) (superior vena cava with pulmonary artery anastomosis) is considered one of the most important factors behind the good results of Fontan-type operations ***(Masuda et al., 1998; Uemura et al., 2000)***

In 1958 William Glenn, a professor of surgery at Yale University, first reported the clinical application of this concept

(Bonita et al., 2007). The bi-directional cavo-pulmonary shunt which was first performed in 1966 is currently most commonly employed as a first stage in a staged Fontan procedure ***(Moller, 1998).***

The ultimate goal of staged univentricular palliation is to normalize the volume and pressure work of the functional ventricle while pumping blood fully saturated with oxygen, regardless of the underlying cardiac anatomy. However, the elevated pulmonary vascular resistance (PVR) present in the early post-natal period means that attainment of this long-term goal must be delayed, resulting in the need for a staged management strategy in most cases ***(Sglimbea et al., 2013).***

The Glenn shunt is palliative, not corrective. Depending on the diagnosis and the surgical era, the Glenn shunt may be the only palliation for the cyanotic patient, one of several palliative surgeries, or a step prior to corrective surgery or the Fontan form of total right heart bypass ***(Lamberti et al., 1990; Lal and Mahant, 1996)***

The term "single ventricle anomaly" is purposely non-specific. It is used to describe a group of cardiac defects that may differ quite dramatically from each other but share the common feature that only one of the two ventricles is of adequate functional size ***(Goor and Lillehei. 1975)***. Some of the anomalies described as single ventricle defects are tricuspid atresia, hypoplastic left heart syndrome, double inlet left ventricle, many of the heterotaxy defects and some variations

of double outlet right ventricle ***(Perloff and Child, 1998; Azzolina et al., 1972).***

Aortopulmonary collaterals may occur after a bi-directional cavo-pulmonary anastomosis, providing competitive pulsatile pulmonary blood flow. The prevalence may be as high as 36% in patients after a bi-directional cavo-pulmonary anastomosis. The significance of these vessels and indications for closure are not yet clear ***(McElhinney et al., 2000).***

The Glenn operation has two major advantages in most children. First, because the connection is a direct one between two blood vessels, rather than made of artificial matter, it has the ability to grow with the child. Second, it removes some of the work from the single ventricle so that the ventricle will no longer have to pump all of the blood to the lungs in addition to all of the blood to the body. This replaces the risk for early heart failure. In most cases this stage is tolerated the best of all the stages with a survival rate of 95 percent or better ***(Tireli et al., 2003; Fontan and Baudet, 1971).***

After the Glenn operation most children will have oxygen saturation levels of 75 percent to 85 percent. The third and final stage in the reconstruction of a single ventricle heart defect is the Fontan completion operation. This operation is usually performed at 2 or 3 years of age, based on the child's size and clinical status. After a Fontan operation, oxygen levels will be nearly normal (90s) ***(Behrman et al., 2004).***

AIM OF THE WORK

Prospective study to evaluate all patients post bi directional glennn shunts presenting to Ain Shams university Hospital Cath lab for hemodynamic study. To assess the status of patients post bi-directional Glenn shunt as regard pulmonary pressure, aortopulmonary collaterals, veno venous collaterals, oxygen saturation and cardiac function of single functioning ventricle as indicator of Glenn shunt success and need of Fontan completion.

Chapter 1

ANATOMY OF SINGLE VENTRICLE

Heart is developed during the first 8 weeks of fetal development. Normally, the heart has two chambers (ventricles) that serve as the heart's pumping chambers. The right ventricle normally pumps deoxygenated blood out of the heart through the pulmonary artery to the lungs for oxygen, and the left ventricle normally pumps oxygenated blood through the aorta out of the heart to the body ***(Anderson et al.,2003).***

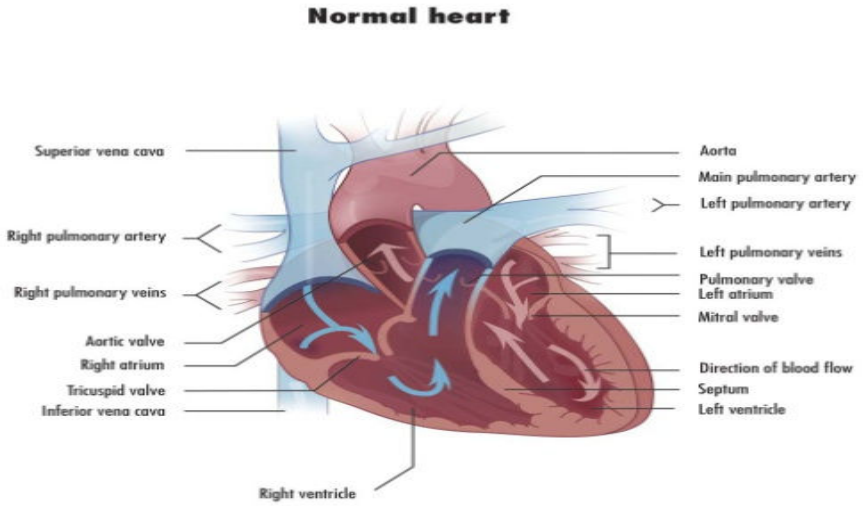

Figure (1): Normal heart.

Single ventricle accounts for about 1% of all cardiac anomalies with an incidence of about 0.05 to 0.1 per 10,000 live births ***(Samanek and Voriskova, 1999).*** The cause is unknown, but it is most likely multifactorial with a genetic predisposition.

Hearts that have previously been called univentricular hearts, or single ventricles, can be described as having a univentricular atrioventricular connection. Most such hearts have two ventricular chambers, albeit one is small and incomplete lacking an inlet component. The atria of these hearts connect only to one of these ventricular chambers, which is usually the larger and dominant ventricle. Other hearts, with biventricular atrioventricular connections, may have hypoplasia of one ventricle, making it impossible to restore a biventricular circulation and such hearts are functionally univentricular. The term "functionally single ventricle" (or functionally univentricular heart) encompasses both these categories of malformation ***(Wilkinson and Anderson, 2012).***

Single ventricles may be classified based on the location of the great arteries ***(Fulton and Freed, 2004).*** There may be normally related great arteries (typeI), D-transposition of the great arteries (type II), or L-transposition (type III). The existence of pulmonic stenosis or pulmonary atresia further subdivides the types of single ventricle. A single ventricle may be accompanied by pulmonary atresia (type A), pulmonic stenosis (type B), or absence of pulmonic stenosis (type C). Depending on the ventricular morphology, the single ventricle can be subdivided as left ventricular type (65% to 70%), right ventricular type (20%), or indeterminate type (10% to 14%) ***(Fulton and Freed, 2004)***. All patients with a single ventricle have some degree of hypoxemia caused by intracardiac

shunting. Clinical manifestations are usually apparent shortly after birth. The most common findings are dyspnea, tachycardia, cyanosis, and progressive heart failure. Later, secondary erythrocytosis and clubbing are usually present. They virtually always require one or more surgeries.

Examples include:

- Tricuspid atresia (TA).
- Hypoplastic left heart syndrome (HLHS).
- Mitral valve atresia (usually associated with HLHS).
- Single left ventricle.
- Double outlet right ventricle (DORV).
- Pulmonary atresia with intact ventricular septum (PA/IVS).

Single ventricle physiology

This term is used to describe the situation where in complete mixing of pulmonary venous and systemic venous blood occurs at the atrial or ventricular level and the ventricle(s) then distribute output to both the systemic and pulmonary beds.

As a result of this physiology the:

- Ventricular output is the sum of pulmonary blood flow (Qp) and systemic blood flow (Qs)

- Distribution of systemic and pulmonary blood flow is dependent on the relative resistances to flow (both intra and extra-cardiac) into the two parallel circuits
- Oxygen saturations are the same in the aorta and the pulmonary artery

This physiology can exist in patients with one well-developed ventricle and one hypoplastic ventricle as well as in patients with two well-formed ventricles ***(Furey et al., 1984).***

In the case of a single anatomic ventricle there is always obstruction to either pulmonary or systemic blood flow as the result of complete or near complete obstruction to inflow and/ or outflow from the hypoplastic ventricle. In this circumstance there must be a source of both systemic and pulmonary blood flow to assure post-natal survival ***(Khambadkone et al., 2003).***

In some instances of a single anatomic ventricle a direct connection between the aorta and the pulmonary artery via a patent ductus arteriosus (PDA) is the sole source of systemic blood flow (hypoplastic left heart syndrome) or of pulmonary blood flow (pulmonary atresia with intact ventricular septum). This is known as ductal dependent circulation. In other instances of a single anatomic ventricle intra-cardiac pathways provide both systemic and pulmonary blood flow without the necessity of a PDA. This is the case in tricuspid atresia with normally related great vessels, a nonrestrictive VSD and minimal or absent pulmonary stenosis.

In certain circumstances single ventricle physiology can exist in the presence of two well-formed anatomic ventricles:

- Tetralogy of Fallot (TOF) with pulmonary atresia (PA) where pulmonary blood flow is supplied via a PDA or multiple aortopulmonary collateral arteries (MAPCAs)
- Truncus arteriosus
- Severe neonatal aortic stenosis and interrupted aortic arch; in both lesions a substantial portion of systemic blood flow is supplied via a PDA
- Heterotaxy syndrome

With single ventricle physiology the arterial saturation (SaO2) will be determined by the relative volumes and saturations of pulmonary venous and systemic venous blood flows that have mixed and reached the aorta.

This is summarized in the following equation:

Aortic saturation = [(systemic venous saturation) (total systemic venous blood flow) + (pulmonary venous saturation) (total pulmonary venous blood flow)] / [total systemic venous blood flow + total pulmonary venous blood flow].

From this equation, it is apparent that with single ventricle physiology, three variables will determine arterial saturation:

1. The ratio of total pulmonary to total systemic blood flow (Qp:Qs). A greater proportion of the mixed blood will

consist of saturated blood (pulmonary venous blood) than of desaturated blood (systemic venous blood) when Qp:Qs is high. An arterial saturation approaching 100% is possible only with an extremely large Qp:Qs.

2. Systemic venous saturation. For a given QP:QS and pulmonary venous saturation, a decrease in systemic venous saturation will result in a decreased arterial saturation. Decreases in systemic venous saturation occur as the result of decreases in systemic oxygen delivery or increases in systemic oxygen consumption. Systemic oxygen delivery is the product of systemic blood flow and arterial oxygen content. Arterial oxygen content, in turn, is dependent on the hemoglobin concentration and the arterial saturation.
3. Pulmonary venous saturation. In the absence of large intrapulmonary shunts and/or V/Q mismatch pulmonary venous saturation should be close to 100% breathing room air. In the presence of pulmonary parenchymal disease, pulmonary venous saturation may be reduced. The V/Q mismatch component of pulmonary venous desaturation will be largely eliminated with a FiO2 of 1.0 while the intrapulmonary shunt contribution will not be eliminated.

For any given systemic venous saturation and QP:QS a reduction in pulmonary venous saturation will result in a decreased arterial saturation ***(Mace et al., 2000; Senzaki et al., 2002; Tanoue et al., 2003; Senzaki et al., 2006; Davos et al., 2003).***

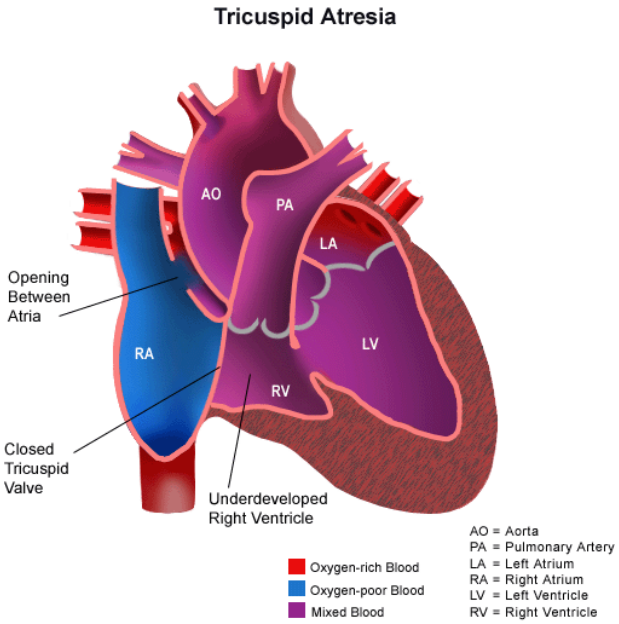

Figure (2): Tricuspid atresia.

- ***Tricuspid Atresia:***

Tricuspid atresia is the third most common form of cyanotic congenital heart disease, with a prevalence of 0.3-3.7% in patients with congenital heart disease ***(Rao, 1997).***

The deformity consists of a complete lack of formation of the tricuspid valve with absence of direct connection between the right atrium and right ventricle. It is the most common cause of cyanosis with left ventricular hypertrophy ***(Rao, 2000).***

Although some authors state that Holmes (1824) or Kuhne (1906) first described tricuspid atresia ***(Kuhne, 1906).*** Rashkind's methodical and thorough historical review indicates that Kreysig (1817) reported the first case in 1817. An 1812

report by the editors of the London Medical Review (1812) appears to fit the description of tricuspid atresia, but they did not use this specific term ***(Rashkind, 1989).***

The cause is unknown. Although specific genetic causes of the malformation remain to be determined in humans, the ZFPM2/FOG2 and HEY2 genes may be involved in the process. Mice in which the FOG2 gene is knocked out are born with tricuspid atresia ***(Sarkozy et al., 2005).***

Anatomy

The most common type of tricuspid atresia is muscular ***(Van Praagh et al., 1971; Rao, 1992).*** It is characterized by a dimple or a localized fibrous thickening in the floor of the right atrium at the expected site of the tricuspid valve. The muscular variety accounts for 89% of cases ***(Anderson et al.,1977).*** In the membranous type (6.6%), the atrioventricular portion of the membranous septum forms the floor of the right atrium at the expected location of the tricuspid valve. This particular type appears to be associated with absent pulmonary valve leaflets.

Minute valvar cusps are fused together in the valvar type (1%). In the Ebstein type (2.6%), fusion of the tricuspid valve leaflets occurs; attachment is displaced downward, and plastering of the leaflets to the right ventricular wall occurs ***(Rao et al., 1973).*** This variant is rare but well documented. The atrioventricular canal type is extremely rare (0.2%). In this type, a leaflet of the common atrioventricular valve seals off the only entrance into the right ventricle ***(Wenink and Ottenkam, 1987).***

In the final type, unguarded with a muscular shelf (0.6%), the atrioventricular junction is unguarded, but the inlet component of the morphologic right ventricle is separated from its outlet by a muscular shelf ***(Scalia et al.,1984).***

The right atrium is enlarged and hypertrophied. An interatrial communication is necessary for survival. This communication most commonly is a stretched patent foramen ovale. Sometimes, an ostium secundum or an ostium primum atrial septal defect (ASD) is present. In rare cases, the patent foramen ovale is obstructive and may form an aneurysm of the fossa ovalis, which is sometimes large enough to produce mitral inflow obstruction. The left atrium may be enlarged, especially when the pulmonary blood flow is increased. The mitral valve is morphologically normal; it is rarely incompetent and has a large orifice. The left ventricle is enlarged and hypertrophied but often morphologically normal. The ventricular septal defect (VSD) is usually small; however, it can be large, or several VSDs may be present ***(Rao, 1992).*** The ventricular septum is rarely intact. When present, the VSD may be conoventricular or perimembranous in type (inferior to the septal band), it may be of conal septal malalignment type (between the limbs of the septal band), or it may be of the muscular or atrioventricular canal type ***(Weinberg, 1980; Weinberg, 1992).***

Muscular VSDs are the most common defects and are usually restrictive; they produce subpulmonary stenosis in patients with normally related great arteries and simulate

subaortic obstruction in patients with transposition of the great arteries ***(Rao, 1977; Rao, 1983).***

The right ventricle is small and hypoplastic, and its size largely depends on the anatomic type ***(Ottenkamp et al., 1985).*** In patients with a large VSD or transposition of the great arteries, the size of the right ventricle may be larger, but, even in these patients; the right ventricle is smaller than normal. In patients with pulmonary atresia and normally related great arteries, the right ventricle is small and may escape detection. However, it is a true right ventricle in most patients; it is composed of a sharply demarcated infundibulum with septal and parietal bands and a sinus with trabeculae, which may communicate with the left ventricle by means of a VSD. By definition, the inflow region is absent, although papillary muscles may occasionally be present.

The great artery relationship is variable and forms the basis of a major classification. Obstruction to the pulmonary outflow tract is present in most cases of tricuspid atresia and is used in the scheme of classification. The aorta is either normal or slightly larger than normal. In 30% of patients, various associated cardiac defects are present; aortic coarctation and persistent left superior vena cava are particularly notable ***(Rao et al., 1991; Rao et al., 1992).***

Classification:

Tricuspid atresia is classified according to the morphology of the valve, ***(Van Praagh, 1992)*** the radiographic

appearance of pulmonary vascular markings, and the associated cardiac defects ***(Rao, 1980).***

Van Praagh and associates (1971) initially proposed a classification based on the morphology of the atretic tricuspid valve ***(Van Praagh et al., 1971).***

Astley and associates (1953) proposed a classification based on pulmonary vascular markings on a chest radiograph: Group A are cases with decreased pulmonary vascular markings, and group B are those with increased pulmonary vascular markings ***(Astley et al., 1953; Dick et al., 1975)*** added a third group, group C, to describe cases with a transition from increased to decreased pulmonary vascular markings ***(Dick et al., 1975)***. This type of classification has some clinical value, although a more precise definition than these can often be made by using noninvasive 2-dimensional (2D) and Doppler echocardiography.

In 1906, Kuhne first proposed a classification based on great-artery relationships ***(Kuhne, 1906)*** which Edwards and Burchell expanded in 1949 ***(Edwards and Burchell, 1949)***. Keith, Rowe, and Vladpopularized this classification in 1967 ***(Keith et al., 1967).*** Other investigators have offered various other classifications. Although these classifications are generally good, their exclusion of some variations in great-artery relationships and the lack of consistency in subgroups are problematic.

Therefore, the authors proposed the following, comprehensive-yet-unified classification ***(Rao, 1980):***

- The principle grouping continues to be based on the following interrelationships of the great arteries: