INTRODUCTION

Single ventricle is a rare, but complex cardiac disorder. It represents 7.7% of congenital heart disease diagnosed in childhood and has a birth incidence of approximately 4–8 per 10 000. [1]

Functional single ventricle (FSV) is a spectrum of severe congenital heart disease, with multiple anatomic variations but similar surgical treatment strategies.

The cardiac physiology of a fetal SV differs from the fetal normal heart. The fetal SV is volume loaded because it must accommodate both the pulmonary and total systemic venous return, In contrast, the afterload of the fetal SV is not altered (in the absence of outflow obstruction) from the normal fetal heart. Although the ventricle may hypertrophy and improve ventricular function with increased pressure afterload,but the total number of myocytes may be decreased for life.

Studies have demonstrated differences between the ventricles, which would be expected to affect the performance of the right ventricle compared with the left ventricle; there are differences in collagen content and fiber layout between the two ventricles. However, the finding that the fetal SVexhibits normal strain values correlates with the clinical experience that the fetal SV, in the absence of atrioventricular valve

regurgitation, anemia, or arrhythmia, does not experience functional deterioration in utero. Fetal heart failure can occur in conditions of abnormal preload to the fetal heart, such as atrioventricular valve regurgitation, but in the absence of significant valvar regurgitation, the SV appears to be able to adapt to the increased preload in SV physiology with normal ventricular systolic function and normal deformation parameters. [2]

With the advent of advanced palliative and corrective surgical procedures, FSV patients are living longer into adulthood compared to two or three decades ago, and they are more frequently undergoing imaging to assist in clinical and surgical management.

Surgical palliation of FSV began in 1971 with the treatment of tricuspid atresia (TA), described by Fontan and Baudet. These surgical palliative techniques were then further refined in 1980 by Norwood, specifically in patients with hypoplastic left heart. Subsequently, many variations of surgical palliation have been built upon these original techniques enabling neonates, who previously would have suffered significant morbidity and mortality, to have a 70% chance of survival into adulthood.

The goal of surgical palliation is to maximize systemic blood oxygenation by converting blood flow from a parallel to

a serial circuit and to maintain the functional capacity of the systemic ventricle. [3]

And these surgical procedures include (Glenn, Kawashima and Fontan procedures) but Glenn shunt is the most commonly used in case of single ventricle patients in Egypt.

Bidirectional superior cavopulmonary anastomosis Glenn procedure is a palliative surgical procedure for anatomical or physiological univentricular hearts.

The Glenn procedure was introduced in 1958 William Glenn and modifications to the procedure were published by Dr. Azzolina in 1973.

The original description by Dr. Glenn allowed communication only between the right pulmonary artery and the SVC, whereas the modified technique had the SVC connecting at or before the bifurcation between the right and left pulmonary arteries.

This results in deoxygenated blood returning from the head and upper body directly routed to the pulmonary arteries for oxygenation by the lungs, to some extent reducing the ventricular workload. Since the blood passing from the SVC into the pulmonary arterial system flows bidirectionally to both right and left lungs, it is called a bi-directional Glenn procedure. [4]

The bidirectional Glenn cavopulmonary shunt (CPS) represents the standard interim procedure in the palliation of patients with single-ventricle physiology toward eventual Fontan circulation. Its performance provides excellent palliation of single ventricles and has resulted in decreased mortality compared with the Fontan procedure without the intermediate step. (5)

The assumption that, in the past 2 decades, there has been a dramatic improvement in the survival of infants born with a single ventricle is based on a scarcity of data. Numerous reports have demonstrated improvements in outcomes after the Cavopulomanry anastmosis), yet it is unclear whether these improvements can be extrapolated to all patients born with a univentricular heart or not. (6)

Aim of the Work

To determine impact of ventricular morphology on myocardial deformation in patients with single ventricle.

THE FUNCTIONAL SINGLE VENTRICLE

Introduction:

The term "functionally univentricular heart" encompasses a group of cardiac malformations defined by the presence of a single ventricular chamber or a large dominant ventricle associated with a diminutive opposing ventricle. (1)

Definition:

Functionally single ventricle arrangements represent a heterogeneous group of anomalies sharing common feature which is functional single cardiac chamber. In most patients with functionally univentricular hearts (UVH) there are two morphologic ventricles, one of which is too small to sustain one of the circulations.

Incidence:

In different series of congenital heart disease patients, cases of UVH represent about 1–2% of total congenital heart diseases. (2)

Etiology:

Most cases of single ventricle are sporadic. However, several investigators have reported familial cases of tricuspid atresia. Tricuspid atresia and other forms of functionally univentricular hearts have been associated with a variety of genetic syndromes, such as Holt–Oram, Noonan,

velocardiofacial syndrome and, rarely, trisomy 21. Most forms of UVH are considered to be polygenic in nature. [3]

Developmental considerations:

The embryologic cause of the UVH, as for most congenital cardiac defects, is not fully known. Two main theories exist. One states that the bulboventricular septum becomes realigned to form the interventricular septum, and that UVH is a consequence of failure of this realignment. The other states that bulboventricular and interventricular septa are different structures, and that the UVH results from failure of formation of the posterior interventricular septum as showed in figure 1. [3]

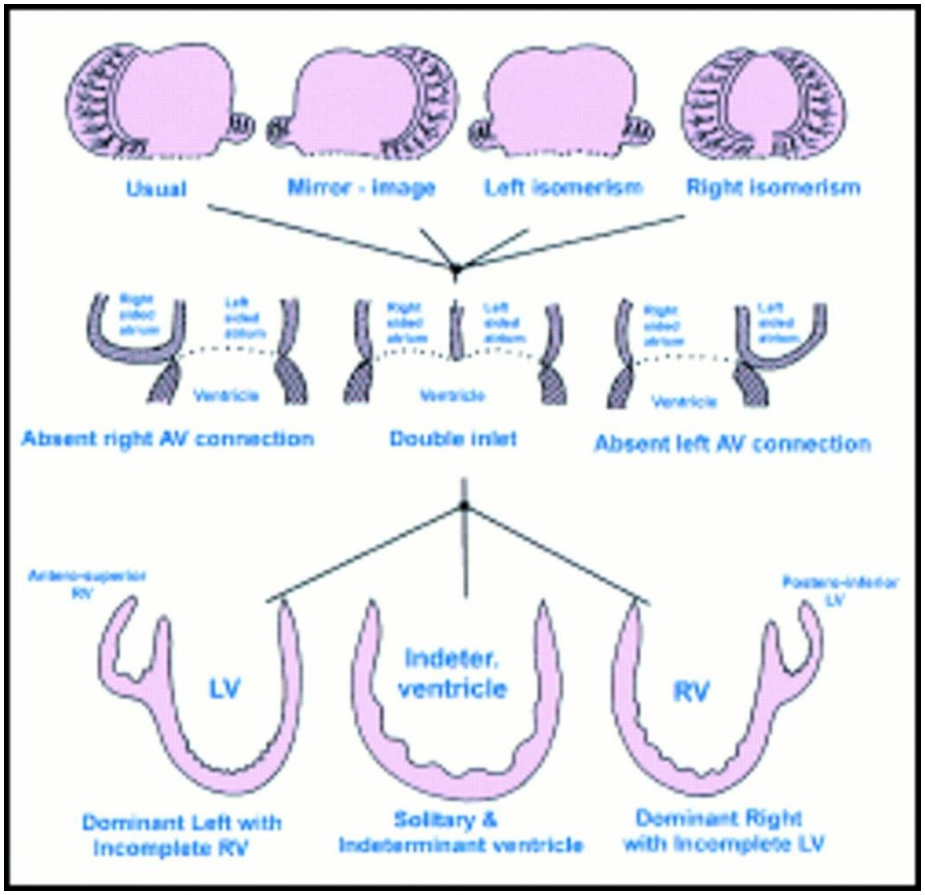

Figure (1): Shows the classification of univentricular heart.

Anatomy and morphology:

The anatomy of complex congenital heart lesions is best approached in a systematic way. The segmental approach has generally been accepted as the standard method for analyzing and categorizing congenital heart malformations.

This method is based on describing the four primary segments of the heart (veins, atria, ventricles and arterial trunks) and the way they are joined together at the venoatrial, atrioventricular and ventriculo-arterial junctions.

Once the morphology of each chamber is defined, the connections can be studied.

The atrial topological arrangement can be solitus (the usual pattern), inversus (mirror-image pattern), or ambiguous in case of left or right isomerism. Either type of atrioventricular connection can be associated with any of the four forms of atrial arrangement. The atrioventricular (AV) connection is, by definition, Univentricular but there may be a double inlet, a common inlet, or an absent right or an absent left AV connection. [4]

Based on these characteristics most ventricles can be recognized as being morphologically right or left. In rare situations there is only one ventricular chamber in the ventricular mass, with a trabecular pattern typical of neither left nor right ventricle (indeterminate or mixed ventricular morphology). The topology (relationship between the two ventricles in space) of the ventricle should also be determined. Chirality can be used to describe ventricular topology. (Fig 2).

A ventricle is called a right-hand ventricle if the palmar surface of the right hand can be placed on the ventricular septum with the thumb pointing to the inlet and the fingers to the outlet, the wrist being in the apical component.

In a normal heart the right ventricle is right-handed, reflecting the normal embryologic development with a D

ventricular looping, whereas in AV and ventricular arterial discordance. [4]

The right ventricle has a left-handed topology consistent with embryologic development with an L ventricular looping. As a final part of the segmental approach, the ventriculoarterial connections should be defined. This connection can be concordant, with the pulmonary artery arising from the right ventricle and the aorta from the left ventricle, or discordant, with transposed great artery relationships. Double-outlet ventricle (with both great arteries arising from one ventricle) and single-outlet ventricle (common arterial trunk or in pulmonary or aortic atresia) have also been described. [5]

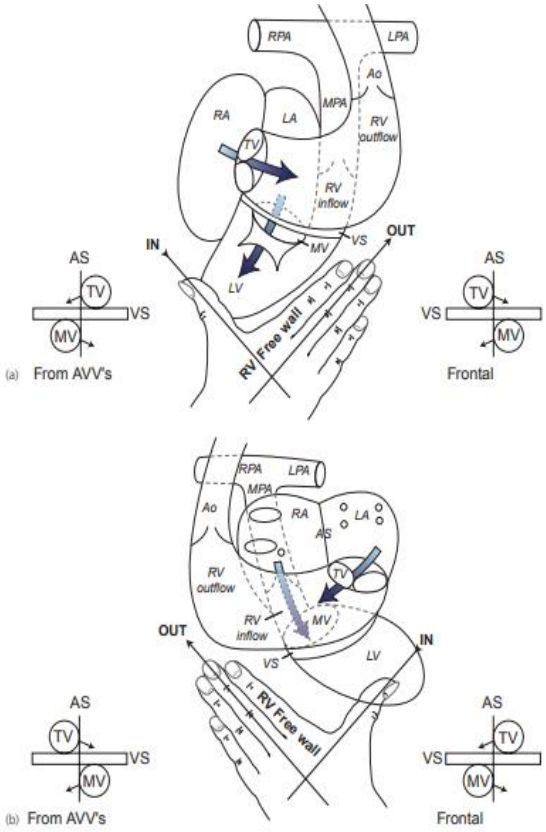

Figure (2): The hand rule a-D-Loop the right ventricle is right handed b-the L-Loop the Right ventricle is left handed.

Specific lesions:

Tricuspid atresia this is sometimes considered to be the "prototype" of hypoplastic right heart disease. Tricuspid atresia has been classified based on the VA alignments and the degree of obstruction.

Type I is "simple" tricuspid atresia with concordant VA connections; type II is "simple" tricuspid atresia with discordant VA connections. Type III is "complex" tricuspid atresia with associated anomalies such as juxtaposition of the

atrial appendages, incomplete AV canal defect, anomalous pulmonary venous connection and other associated anomalies.

In type I patients, a restrictive VSD will result in reduction of pulmonary blood flow, which can result in severe cyanosis and duct-dependent pulmonary blood flow.

In type II patients a restrictive VSD will result in diminished systemic output and can be associated with aortic arch anomalies like aortic coarctation and interrupted aortic arch.(figure 3) [4]

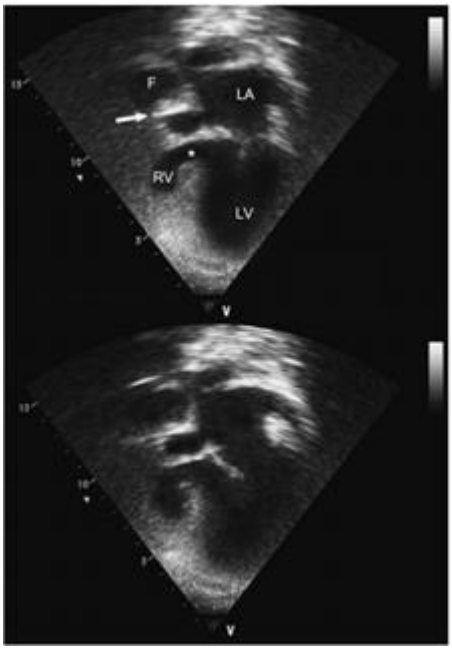

Table 27.1 Tricuspid atresia subtypes

Type I
A No VSD, PA
B Small VSD, PS
C Large VSD, no PS
Type II (D-transposed great arteries)
A VSD, PA
B VSD, PS
C VSD, no PS
Type III: Complex

Figure (3): Apical 4 chamber view shows atretic tricuspid valve.

Double-inlet left ventricle (DILV):

The left ventricle receives inflow from both of the atrioventricular valves (AVVs) and communicates with the right ventricle via a ventricular septal defect (VSD). The right ventricle is usually hypoplastic and DILV is often associated with transposition of the great arteries, in which the aorta arises from the hypoplastic morphologic right ventricle. [6]

The hypoplastic right ventricle (RV) is anterior and leftward to the left ventricle (LV) in 63–74% of cases. The topology of the RV will be left-handed in these cases.

Mostly it is not possible to define clearly the anatomy of the valves and it is better to name them left and right AV valves.

The morphology and function of the AV valves are assessed from the subcostal, apical and parasternal windows (Figs 4).

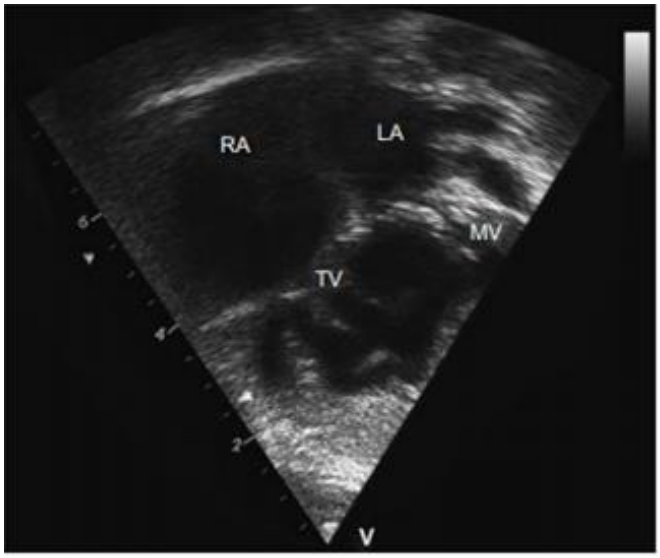

Figure (4): Evaluation of A-V valves in (DILV) patient in apical 4 chamber view.

Double-inlet and common-inlet right ventricles:

Both types of single RV – double-inlet and common-inlet RV – are rare. In double-inlet right ventricle (DIRV), both AV valves are exclusively committed to the RV, and both great arteries originate from it (double-inlet double-outlet RV) [7].

Pathophysiology:

Single ventricle physiology is characterized by obligatory mixing of the systemic and pulmonary venous returns, the aorta and pulmonary artery having mixed oxygen saturation.

The systemic and pulmonary outputs are determined by the presence and degree of hypoplasia or narrowing of one of the great arteries.

Pulmonary blood flow is determined by anatomic obstruction below, at or above the pulmonary valve, pulmonary arteriolar resistance, pulmonary venous obstruction and left atrial pressure.

Surgical palliation of anatomic and functional single ventricle usually requires a series of operative procedures. The final common surgical pathway for all these variants is one of several modifications of the Fontan procedure, in which the systemic venous return is diverted directly to the pulmonary arteries and the pulmonary venous return crosses the AV valve(s) and is pumped by the "single ventricle" to the systemic vascular bed. [8]

3. Clinical presentation

Most children with single ventricle present as neonates. However, an increasing proportion of patients with functional univentricular heart are diagnosed prenatally.

Advances in ultrasound technology and resolution have allowed accurate prenatal diagnoses of all forms of single ventricle. The most frequent postnatal presenting symptom is that of cyanosis.

Heart murmurs, congestive heart failure and neonatal shock can also be seen depending on the associated lesions and degree of outflow obstruction present.. Shock syndromes are