Learning objectives

- To demonstrate the anatomical variants found in the spectrum of univentricular or "single" ventricle hearts.
- To describe the imaging modalities needed to accurately delineate the features of these complex cardiac defects to ensure the best outcome for these patients.
Background

The term "single ventricle heart" includes a heterogeneous group of complex congenital cardiac malformations characterized by a univentricular atrioventricular connection; i.e. both atria are completely or predominantly connected to a single ventricle. This ventricle has to maintain both the pulmonary and systemic circulations and inevitably its function will deteriorate. Mixing of arterial and venous blood occurs at the atrial or ventricular level. In severe pulmonary or systemic outflow obstruction, the patient is dependent on shunting via a patent ductus arteriosus [1].

Patients with single ventricle hearts present in infancy with the majority requiring a staged surgical approach culminating in the Fontan procedure. Surgical cavopulmonary shunts reduce the workload of this sole ventricle, increasing its longevity. Adult survival has improved with surgical and medical advancements: current literature reports 91% survival rate at 10 years [2]. Nevertheless, most patients develop abnormal and complicated physiology such as protein losing enteropathy, thromboembolism and liver dysfunction. Death is usually due to arrhythmias or congestive heart failure [2] and many patients will thus need cardiac transplantation as their cardiac function deteriorates. Timeous assessment of morphology on echocardiography and physiology at cardiac catheterization is essential in planning the surgical management of these patients to optimize their life expectancy [1]. This is especially true of patients in South Africa where end-stage cardiac transplantation is not readily available.

The incidence of single ventricle heart is less than 1% of all congenital cardiac defects [3]. The etiology of single ventricle heart has not yet been established but it has been suggested to be polygenic. Hypoplastic left heart syndrome (HLHS) (fig. 1) is the most common type of single ventricle with an incidence of 2.3:10 000 live births. It is characterized by under-developed left ventricle with mitral and aortic valve atresia, hypoplastic aortic arch and coarctation [2]. AV valve atresia is the second most common type occurring in 1: 10 000 live births. The tricuspid valve is replaced by fibrous or muscular tissue with the right ventricle size varying according to the ventricular septum defect (VSD) size. 30% of these patients have transposition of the great vessels (fig. 2) [2]. The third most common type is the double-inlet left ventricle (DILV) (fig. 3) where both atria are connected to the left ventricle with L-transposition of the great arteries [2].

Other rare types of single ventricle hearts include unbalanced atrial ventricular septum defect (AVSD) and double outlet right ventricle (DORV). Unbalanced AVSD (fig. 4) is characterized by a common AV valve and one well-developed ventricle. In the right-dominant AVSD, the common AV valve is directed more towards the right ventricle, which is then better developed, whereas in the left-dominant AVSD, flow through the AV valve is directed preferentially towards the left ventricle. The right dominance variety is more
common than the left[3]. DORV (fig. 5) is characterized by both the aorta and pulmonary artery arising from the right ventricle with outflow from the LV through the VSD [2].

Surgical management of the single ventricle or "single ventricle repair" is achieved by two cavopulmonary shunts which offload the systemic venous return's load to the heart by directing it straight to the pulmonary arteries. The bidirectional Glenn shunt (fig. 6) is usually performed after 2 months of age, once PA pressures have decreased after birth. The SVC is divided and anastomosed end-to-side to the right pulmonary artery [4]. The Fontan completion (fig. 7) is then typically carried out between 18 months to 4 years of age. The extra-cardiac conduit Fontan completion is the preferred practice at our institution: IVC flow is directed via an extra-cardiac connection to the right pulmonary artery, increasing the level of oxygenation [4]. In an HLHS, a Norwood procedure precedes the Glenn and the Fontan cavopulmonary shunts. The Norwood (fig. 8) consists of performing an atrial septectomy to provide unrestricted blood flow from the LA to the RA across the atrial septum, ligating the ductus arteriosus and augmenting the hypoplastic aortic arch by anastomosing it to the main pulmonary artery and removing any coarctation of the aorta. Pulmonary bloodflow is supplied via either a modified Blalock-Taussig (aorta-to-pulmonary) shunt or Sano operation where a conduit is placed from the right ventricular outflow tract to the pulmonary artery [4].
**Fig. 1:** Illustration of hypoplastic left heart syndrome.

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Fig. 2: Illustration of tricuspid valve atresia.

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Fig. 3: Illustration of double-inlet left ventricle.

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Fig. 4: Illustration of balanced and unbalanced AVSD.

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Fig. 5: Illustration of double-outlet right ventricle.

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Fig. 6: Illustration of Glenn procedure.

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**Fig. 7:** Illustration of extra cardiac Fontan.

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**Fig. 8:** Illustration of Norwood.

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Imaging findings OR Procedure details

There are no definitive findings on chest radiograph in single ventricle hearts, however cardiac silhouette and pulmonary vasculature can be assessed. With associated severe pulmonary stenosis (PS) or atresia, the pulmonary vasculature may be oligaemic or normal with normal or dilated cardiac shadow. In the absence of severe PS, plethoric lung fields and increased cardiac shadow are seen [3].

Echocardiography using 2D imaging and colour-flow doppler provides comprehensive assessment of the anatomy of single ventricle hearts. The diagnosis and morphological subtypes can be appraised. AV valve morphology, atresia, competence and straddling can be assessed as well as relationship and over-riding of great vessels [1].

HLHS typically present with increased cardiac thoracic ratio with plethoric lung fields on chest radiograph (fig. 9), however, other findings are seen. Echocardiography demonstrates underdeveloped left ventricle, with mitral and aortic valve atresia, hypoplastic ascending aorta with coarctation (fig. 10) [5].

In tricuspid atresia, chest radiograph shows increased cardiac silhouette with prominent and rounded LV border and oligaemic pulmonary vasculature (fig. 11). Echocardiography can demonstrate the atretic tricuspid valve with hypoplastic RV as well as the size of the VSD and severity of PS (fig. 12) [3].

In DILV chest radiograph, the aortic shadow can be altered by the position of the rudimentary right ventricle: in rudimentary left-sided right ventricle, the left heart border is prominent; in rudimentary right-sided right ventricle, the aortic silhouette is convex to the right (fig. 13). In echocardiography, the more common rudimentary right ventricle can be identified and localized in the parasternal short-axis and 4-chamber view. The rudimentary RV is attached as an appendage to the well-developed left ventricle and gives rise to the aorta, and the pulmonary artery (fig. 14). Rudimentary left-sided right ventricle is inferred when the aorta arises from the anterior rudimentary chamber [1].

Chest radiographs in patients with unbalanced AVSD demonstrate cardiomegaly, increased pulmonary vascular markings and prominent PA secondary to pulmonary hypertension (fig. 15) [3]. Echocardiography can assess the alignment of the AV valve and any regurgitation as well as the size of the dominant ventricle and any associated abnormalities (fig. 16) [6].
In the DORV chest radiograph shows cardiomegaly with pulmonary plethora and prominent PA (fig. 17). Diagnostic features at echocardiography include discontinuity of the mitral and semilunar valves, the absence of LV outflow other than through the VSD and the origin of both aorta and pulmonary artery from the anterior RV. The size and position of the VSD can further be assessed as well as the presence of associated abnormalities such as coarctation of the aorta (fig. 18) [3].

Cardiac catheterization offers detailed analysis of the anatomic and functional status of the patient. Suitability for a single ventricle repair is based on Choussat's criteria, first published in 1977. These criteria include a mean PA pressure <15 mmHg and a pulmonary vascular resistance of <4 Wood units [7]. Cardiac catheterization is the gold standard for measuring pulmonary vascular resistance and it can also identify abnormal venous return, pulmonary artery distortions and the presence of aorto-pulmonary collateral vessels that may need coil embolization. Patients not suitable for Fontan completion can benefit from repeated catheterization to serially monitor pulmonary artery pressure and to address complications such as shunt stenosis, pulmonary artery stenosis and pulmonary arteriovenous fistula [1].

Cardiac MRI is superior in demonstrating detailed anatomical anomalies and is used for pre and post-op assessment of patients [1]. In some institutions this has replaced cardiac catheterizations to a large extent.
Fig. 9: Frontal chest radiograph demonstrating abnormal cardiac silhouette and plethoric lung fields.

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Fig. 10: 2D 4 chamber echocardiograph demonstrates mitral valve atresia with hypoplastic left ventricle, and small VSD.

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**Fig. 11:** Frontal chest radiograph demonstrating mild cardiomegaly and rounded LV border with diminished peripheral broncho-vascular markings.

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**Fig. 12:** 2D 4 chamber echocardiograph showing tricuspid valve atresia with hypoplastic rudimentary RV with a small VSD and large ASD.

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**Fig. 13:** Frontal chest radiograph demonstrating dextrocardia with situs solitus and a left-sided aorta.

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Fig. 14: 2D 4 chamber echocardiograph demonstrates both the tricuspid and mitral valves are committed to the LV. The RV is rudimentary and connected to the LV by a large VSD.

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Fig. 15: Frontal chest radiograph demonstrating gross cardiomegaly and plethora.

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Fig. 16: 2D 4 chamber echocardiograph showing large, complete Rastelli B AVSD; unbalanced ventricles with single AV valve committed to larger LV and hypoplastic RV and additional atrial septum defect (ASD) secundum.

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**Fig. 17:** Frontal chest radiograph demonstrating dextrocardia with widened mediastinum and mid-thoracic hemi-vertebrae and 13 ribs on the right side.

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**Fig. 18:** Colour Doppler 2D 5 chamber echocardiograph showing both great arteries coming off the RV side-by-side. Smaller PA with persistent sub-pulmonary conal tissue.

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Conclusion

Single ventricle hearts are complex defects which need systematic radiological assessment to guide prompt surgical intervention and thus optimize the patient's outcome. Timeous assessment of morphology on echocardiography and physiology at cardiac catheterization is essential in planning the surgical management of these patients to optimize their life expectancy [1]. This is especially true of patients in South Africa where end-stage cardiac transplantation is not readily available.
References