Heart failure is a chronic illness that is increasing in frequency as more infants and children with complex cardiac malformation survived following corrective or palliative surgery. It is primarily defined as a syndrome caused by the overall inadequate performance of the heart, leading to a constellation of signs and symptoms.

The pathophysiology of heart failure involves a complex series of structural, functional and biochemical events that are responsible for the overall progressive nature of the disease.

The term intractable heart failure is used to describe symptomatic patients who have progressed to NYHA Class IV or American Heart Association (AHA) Stage D.

Normal ventricular function requires structural integrity, optimal interaction between the heart cardiac matrix and myocytes, optimal interaction between the heart and peripheral organs, optimal filling of the heart and a gradual onset of haemodynamic changes allowing the heart to adapt correspondingly.

Several mechanisms interfere with the normal cardiac function causing disturbances that lead to heart failure. These mechanisms include structural lesions, suboptimal interaction between the atria and ventricles, suboptimal interaction with the peripheral organs and abnormal filling.

Regardless of whether the failing systemic ventricle is morphologically left, right, or single, the cascade of events that leads to the symptoms of CHF is the same. An underlying myocardial insult of whatever aetiology results in ventricular systolic dysfunction, leading to activation of the renin-angiotensin-aldosterone pathway and sympathetic over-stimulation. This results in peripheral vasoconstriction, water retention, pulmonary oedema, and tachycardia. All of these impose an even greater load on the already failing myocardium. Medical and mechanical support for the failing ventricle tends to address these pathophysiologic issues.

Medical treatment of CHF targets not only the augmentation of ventricular contractility (positive inotropy) but also addresses the neuro-humoral derangements associated with it. Positive inotropic intravenous agents that have been mainstays in the acute setting include dopamine, dobutamine, and the phosphodiesterase inhibitors, amrinone and milrinone. Agents that have a marked chronotropic (increased heart rate) or vasoconstrictive effect, such as epinephrine and noradrenaline tend to be avoided in the presence of ventricular dysfunction, as they aggravate the sympathetic over stimulation seen intractable CHF.

Conventional wisdom suggests that diuretics are useful to counteract the fluid retention and pulmonary oedema seen in ventricular dysfunction. The loop diuretic frusemide and potassium-sparing aldactone are still the most commonly used agents nowadays. After-load reduction to counter-act the vaso-constrictive effects of CHF has gained popularity over the last few years. Angiotensin converting enzyme inhibitors such as Captopril and Enalapril are used for this purpose. These are preferred to direct vasodilating agents because of their additional ventricular remodeling effect. It has been our practice to start at low doses initially, gradually increasing as tolerated.

Still somewhat controversial at present is the use of beta-blockers. Current research suggests that their benefit stems from reversal of sympathetic over stimulation, myocardial remodeling, prevention of arrhythmias and up-regulation of beta adrenergic receptors, which were found to be decreased in chronic CHF. A recent study suggests that the 3 rd
generation b-blocker (Carvedilol) in addition to the standard drug regimen outlined above improves ventricular function and clinical symptomatology in children with intractable CHF.

Despite aggressive medical therapy, some patients with intractable heart failure continue to deteriorate. They may then require mechanical support with left ventricular assist devices (LVADs), intra-aortic balloon counter pulsation pumps (IABP) or extra-corporeal membrane oxygenation (ECMO), either to gain time while awaiting myocardial recovery or as a bridge to transplantation, depending on the underlying etiology of CHF. Non-recovery of ventricular function within 72 hours of institution of mechanical support is a poor prognostic indicator.

When medical therapy and mechanical support have failed, cardiac transplantation is a possible option.