

Cardiomyopathy

Left Ventricular Non-Compaction Cardiomyopathy (LVNC)

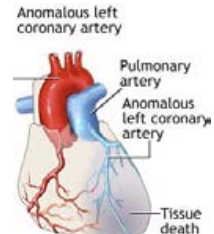
Presentation	Signs and symptoms of heart failure (see CHF section)
Pathophys	During fetal cardiac development, the ventricular myocardium begins as a spongy, highly-trabeculated tissue that should become "compacted" ventricular cavity becomes relatively smooth, especially w/ the LV, which doesn't happen in patients w/ this In patients w/ LVNC
Workup	Echo
Treatment	Heart failure management (see CHF section)

Congestive Heart Failure

Presentation	<ul style="list-style-type: none"> • Infants: Tachycardia, tachypnea, feeding difficulty, diaphoresis (particularly w/ feeding) and poor growth • Children and Adolescents: Shortness of breath, orthopnea, cough, peripheral edema. • PE Finding: Gallops, murmurs (MR/TR), hepatomegaly, edema of ankles or eyelids, tachypnea, tachycardia, crackles, cool extremities, delayed cap refill, weak pulses.
Pathophys	Multiple etiologies-- structural heart disease, arrhythmia, ischemia, cardiomyopathies, myo/ pericarditis, hypertension, and systemic issues including severe anemia, and severe thyroid disease
Workup	<ul style="list-style-type: none"> • CXR: Cardiomegaly and pulmonary edema, Kerley B lines • EKG: Atrial or ventricular enlargement, ischemia, arrhythmia • Echo: Depressed systolic function, +/- ventricular dilation and/or hypertrophy • Labs: If severely depressed cardiac output, may have acidosis, elevated lactate, elevated BNP, abnormal electrolytes and elevated CK and Troponin (if myocardial injury is present). If right sided may have abnormal liver studies.
Treatment	<ul style="list-style-type: none"> • Diuresis: Furosemide or other loop diuretic are first-line. Thiazide diuretics and spironolactone also may be used, usually in chronic CHF. • Inotropes: Digoxin increases contractility. Dopamine, isoproterenol and dobutamine may be used in sicker ICU patients. • Afterload reduction: ACE inhibitors decreased SVR and may positively impact cardiac remodeling. Milrinone infusion has a similar effect and may be used in sicker patients. • Other Measures: O2 and correction of anemia aid O2 delivery. Salt restriction aids diuresis. Treating underlying illness (e.g. infection, arrhythmia, acidosis) can improve contractility. Sedation and mechanical ventilation can decrease demand on the heart.

Coronary Artery Anomalies

Anomalous Left Coronary Artery off the Pulmonary Artery (ALCAPA)

Presentation	Recurrent episodes of irritability and emesis as well as signs of congestive heart failure in infants → diaphoresis, tachycardia, tachypnea, respiratory distress, weak peripheral pulses and cool extremities , +/- gallop or MR murmur
Pathophys	<p>The left coronary artery arises from the pulmonary artery rather than the left coronary cusp of the aortic valve→ can lead to ischemic cardiomyopathy</p> 

Coronary Artery Anomalies continued on next page →