

# Management of Heart Failure in Infancy and Childhood

By

DR. KALIMUDDIN AZIZ\*

Heart failure is a complex state of cardiac muscle dysfunction in which ventricular ejection from left ventricle to aorta and right ventricle to pulmonary artery is insufficient to empty the pulmonary and systemic venous reservoirs<sup>1</sup>. The consequence of such a state is an increase in the systemic and pulmonary venous blood volumes and pressures. In-sufficient right ventricular ejection causes an increased systemic venous return with elevation of pressure in the right atrium and peripheral veins. The right ventricular filling volume and pressure are elevated resulting in increased diastolic ventricular volume. Failure of the left ventricle to adequately empty the pulmonary venous reservoir results in elevated left atrial and pulmonary venous blood volumes and pressures. The left ventricular pre load is increased with elevation of diastolic pressure and chamber volume. Cardiac failure can result from increased cardiac work, which is defined as a product of pressure stroke volume, Heart rate<sup>2</sup>. One of the mechanisms that increases cardiac work is an elevation of after load (systemic and pulmonary arterial pressure) which is defined as a force opposing myocardial fibre shortening. The lesions which impose increased right ventricular after load are obstructive lesions of the right ventricle such as pulmonary valve stenosis and obstructive diseases of the pulmonary vascular bed. Some

of the obstructive lesions imposing increased left ventricular after load are aortic valve stenosis, coarctation of aorta and systemic arterial hypertension. The mechanism by which left ventricle deals with increased after load is to develop hypertrophy of its walls. The left ventricular mass/volume ratio is increased and thereby greater left ventricular peak systolic pressure is generated<sup>3-4</sup>. When after load is chronically present over number of years increased fibrosis due to myocardial cell necrosis results and myocardial contractility is reduced<sup>5</sup>. The consequence is inadequate emptying of the pulmonary venous reservoir. The second type of increased work imposed on the heart is ventricular volume overload. This is a very common occurrence in many congenital cardiac malformations such as ventricular septal defect, insufficiency of atrioventricular or semilunar valves. During the initial stages of increased work requirement the ventricular ejection is increased. The diastolic volume is enlarged due to increased stretch imposed by the increased blood volume. The stroke volume is thereby increased in accordance with Starling's law which dictates that the force of contraction is directly related to the (Sarcomere) fibre length. The myocardial hypertrophy occurs, however, the left ventricular mass/volume ratio remains within normal range. The state at which an increased volume

---

\*Pediatric Cardiologist, N.I.C.V.D, Karachi.

overload is adequately cleared from the ventricles is a compensated one and may last for a number of years. Ultimately however, myocardial fiber death and necrosis leads to inadequate emptying of the volume load decompensated state<sup>5</sup>. Stroke volume and cardiac output are reduced due to decreased ejection fraction, which is defined as  $EDV-ESV/EDV$  (End diastolic volume—end systolic volume/End diastolic volume.) There is evidence to suggest that during cardiac decompensation the left ventricular mass/left ventricular volume ratio is reduced and may suggest loss of myocardial cells and replacement by fibrous tissue<sup>5-6</sup>.

The third situation which produces heart failure is reduced contractility<sup>7</sup> due to intrinsic myocardial diseases. This means that for a given preload the rate of shortening of myocardial cell is reduced. Etiologic agents which produce myocardial disease are infections such as bacterial and viral myopathy, alcoholic myopathy and group of diseases classified as idiopathic myopathies.

### Causes of Heart Failure:

1. Pressure overload lesions: aortic valve stenosis, coarctation of aorta and systemic hypertension for the left ventricle. Pulmonary valve stenosis for the right ventricle.
2. Volume overload lesions; aortic and mitral valve insufficiency, ventricular septal defect and patent ductus arteriosus for the LV. Tricuspid and pulmonary valve insufficiency for the RV.
3. *Myocardial diseases: Mainly Affecting The Left Ventricle.*
  - (a) Infections: Bacterial i.e. diphtheria, typhoid, viral infections and parasitic infections.
  - (b) metabolic; hypocalcemia and hypoglycemia

- (c) collagenosis; systemic lupus erythematosus
- (d) allergic; rheumatic fever.
- (e) myopathies; endocardial fibroelastosis, Hypertrophic obstructive cardiomyopathy (I.H.S.S.).

Table I: Lesions Causing Left Heart Failure

---

I.	<i>Pressure Lesions</i> (increased Afterload)
	Aortic atresia
	Aortic stenosis (in infancy and adults)
	valvar
	subvalvar
	Coarctation of aorta
	simple
	complicated (with ventricular septal defect and or patent ductus arteriosus)
	II. Mitral atresia or stenosis
	III. Pulmonary venous obstruction
	IV. Total anomalous pulmonary venous return with obstruction.
	V Endocardial fibroelastosis
	VI Anomalous origin of coronary arteries
II.	<i>Volume Overload</i> (Increased preload)
	Aortic insufficiency
	Patent ductus arteriosus and aorta to right ventricle or left ventricle fistulae
	Truncus arteriosus Communis
	Ventricular septal defect—simple and complicated.
	Common or single ventricle
	AV canal defects

---

**Table II: Onset of Symptoms and Predictability of Lesions:**

*First Week of Life:*

1. Aortic and mitral atresia complex
2. Coarctation of aorta—complicated
3. Transposition of the great vessels

*First Month:*

1. Coarctation with patent ductus arteriosus or ventricular septal defect
2. Total anomalous pulmonary venous drainage below the diaphragm.
3. AV canal defect
4. Paroxysmal Atrial Tachycardia
5. Tricuspid atresia
6. Single ventricle
7. Truncus arteriosus communis

*Second Month:*

1. Transposition of the great vessels with ventricular septal defect.

*Second to Third Month:*

1. Ventricular septal defect—simple
2. Endocardial fibroelastosis
3. Patent ductus arteriosus

*Third to Sixth Month:*

1. Ventricular septal defect
2. Anomalous left coronary
3. Truncus arteriosus communis

*Sixth to Twelfth Month (less common age for the onset of heart failure)*

1. Endocardial fibroelastosis—few cases
2. AV canal defects
3. Total anomalous pulmonary venous drainage; subdiaphragmatic, supradiaphragmatic.

**Clinical Features:**

The clinical features of heart failure are primarily related to the failure of emptying of either the systemic venous or pulmonary venous reservoirs. The effect on the systemic venous circulation of an increased right ventricular end-diastolic pressure is an increase in superior vena caval, inferior vena caval and peripheral venous pressure. The raised level of jugular venous pulse can be directly observed in older children. In infants, due to increased amount of subcutaneous fat and short neck, the elevated jugular venous pressure cannot usually be appreciated. Puffiness of the eyelids due to elevated venous pressure is more often present in infants in severe cases. Hepatomegaly due to passive venous congestion can be easily evaluated even in the newborn. Liver enlargement of greater than two centimeters when measured in the mid-clavicular line in the infant suggests enlargement. Careful assessment of the hepatic size provides not only a diagnostic but also prognostic index to therapy.

Peripheral edema is uncommon in infancy and childhood and when present is most often due to severe left-sided obstructive lesions such as coarctation of aorta or myocardial disease. The signs and symptoms of pulmonary venous congestion due to failure of emptying of the pulmonary venous reservoir results from left ventricular decompensation. Tachypnea results from increased exudation of fluid in the alveolar septae of the lung. Dyspnea is a sensation of awareness of breathing; the exact mechanism as to how this sensation is provoked is unknown. Tachypnea, that is, increased respiratory rate due to decreased lung compliance, is better appreciated in children. A respiratory rate greater

than 40-60/min in infants and greater than 20/min in adults, is abnormal. Short and shallow respiratory efforts involving increased work by the intercostal and subcostal muscles give a good assessment of the severity of reduced compliance of the lung. Increased pulmonary venous pressure in infants is often accompanied by an increased in the pulmonary blood flow. Symptoms of obstructive airways due to engorged intrabronchial mucosa or compression of main stem bronchi by the enlarged pulmonary artery or left atrium clinically can be present. These are manifested as expiratory wheezing and prolongation of the expiratory time and moist crepitations suggesting fluid exudation into the interstitia of the lung may be audible. The exudation of fluid into the alveoli leads to features of gross pulmonary edema. On chest roentgenogram hyperinflation due to air trapping can be observed. If the left atrial pressure continues to rise then reactive pulmonary arterial hypertension develops and signs of right heart failure are superimposed. Decrease in cardiac output due to diminished stroke volume produces symptoms such as fatigue, pallor and cold extremities and constriction of the peripheral vascular bed. Infants show pallor but fatigue is manifested as listlessness and prolonged feeding time, during which the infant pants and becomes "short of breath". Fatigue is caused by diminished perfusion to the muscles. Episodes of cardiac asthma and frank pulmonary edema which are often seen in adults with left ventricular failure are infrequent in children. Reduction in perfusion pressure in the kidney due to renal arteriolar vasoconstriction results in diminished glomerular filtration and reduced urinary output. The blood urea nitrogen and serum creatinine, serum renin and aldosterone levels are increased from sodium retention<sup>8</sup>.

The sympathetic tone is increased in children with congestive cardiac failure and increased sweating is observed in infants. The serum catecholamine levels are elevated<sup>9</sup>. The result of chronic tissue hypoperfusion in a growing child is retarded growth. The weight lags behind the linear growth.

**Table III: Lesions Causing Pure Right Heart Failure**

- 
- I. Tricuspid atresia
  - II. Ebstein's anomaly
  - III. Pulmonary stenosis (critical, in infancy)
  - IV. Cor pulmonale (chronic lung disease, i.e., fibrocystic disease)
  - V. Primary pulmonary arterial hypertension
  - VI. Constrictive pericarditis
- 

**Table IV: Heart Failure due to Arrhythmias**

- 
- I. Bradycardia: 100 in newborn  
60 in older children
    - A. Complete heart block
      - (a) congenital
      - (b) acquired; infection, surgery
  - II. Tachycardia: Paroxysmal supraventricular tachycardia  
Paroxysmal ventricular tachycardia
- 

Cardiac signs of cardiac failure are gallop rhythm, i.e., triple rhythm, which is due to a loud third heart sound indicates elevated volume load and increased filling pressure of the ventricles, while proto-diastolic gallop, i.e., prominent fourth heart sound, suggests reduced myocardial contractility.

Serum electrolyte disturbances may include dilutional hyponatremia (less than 120 mEq/L) and hypochloremia. Hyperkalaemia is present in severe cases and is due to diminished tissue perfusion and hypoxemia<sup>10</sup>. Initially lesion with volume overload produce respiratory alkalosis i.e., diminished Pa CO<sub>2</sub> and elevated pH, however, in later stages respiratory acidosis (i.e., decreased pH and elevated PaCO<sub>2</sub>) results and eventually severe metabolic acidosis supervenes (reduced-HCO<sub>3</sub>) and elevated lactic acid). In infants with obstructive lesions of the right heart, severe hypoxemia and metabolic acidosis (reduced pH and HCO<sub>3</sub> and increased lactic acid) are commonly present<sup>11</sup>. Hypoglycemia and hypocalcemia may complicate the picture in the neonatal period.

#### Management of Heart Failure:

The cause of heart failure must be elucidated and appropriate corrective measures should be undertaken. The congenital heart malformations are now amenable to primary surgical correction, however, palliative procedures such as pulmonary artery Banding which reduces the Pulmonary arterial pressure and blood flow produce dramatic improvement in the symptoms in desperately sick infants in congestive heart failure.

#### Medical Management:

##### *Digitalis:*

Digitalis provides myocardial inotropic support. It improves myocardial contractility, decreases atrioventricular conduction and slows the heart rate. The effect is to increase the stroke volume and cardiac output. The adverse effects of reduced tissue perfusion are thus alleviated with marked improvement in symptoms. The systemic and venous reservoirs are now better

emptied, systemic venous pressure falls with alleviation of hepatomegaly and peripheral edema. Due to reduction in the vasomotor tone, systemic resistance falls. Reduced left atrial pressure results in improved lung compliance and relief of tachypnea. Renal perfusion is increased with correction of renal oligemia and its sequelae.

The mode of action of digitalis is complex. Digitalis seems to interfere with ATPase necessary for the removal of sodium from the cell by an active transport mechanism i.e., inhibits the sodium pump. This action allows accumulation of sodium in the intracellular space and allows influx of calcium into the myofilaments, thereby potentiating contractility. Decreased potassium concentration on the outer surface of myofibril membrane increase the sensitivity of the cell to the glycoside.

Digoxin is the preparation of Digitalis most commonly used in children and infants. Lanoxin is a liquid preparation of digoxin and is available in concentration of 0.05 mg/ml. Orally, 80% of digoxin is absorbed and the peak serum concentration is achieved in 30-60 minute period. Sixty percent of digoxin is excreted in the urine and 17% is lost in the faeces. Therefore, special consideration should be given to the dosages in renal or hepatic failure and monitoring by serum digoxin levels may be necessary.

##### *Dosages:*

For infants under 1 year of age 0.03 mg/lb is utilized as oral digitalizing dose over a 24 hour period. One-half of the dose can be given as a first initial dose and one-half divided into equal portions at 8 hour intervals. After 24 hours of the initial digitalizing dose, maintenance dose

is calculated at 1/4 the digitalizing dose given in two 12-hourly doses. The intravenous dose is 75% of the oral dose. The dose of digoxin is reduced to 2/3 to 1/2 the recommended dose in cases of myocarditis.

For children more than 1 year of age 0.02 mg/lb is used as a digitalizing dose. For premature infants the same dose schedule is advised.

#### *Toxicity.*

Therapeutic serum digoxin levels by radio immune assay are  $2.8 \pm 1.9$  ng/ml for infants and  $1.3 \pm 0.4$  ng/ml for older children. Toxic range in infants ranges from 4-5 ng/ml<sup>12</sup>. General symptoms of digitalis toxicity are vomiting and diarrhea in the infant while older children may complain of weakness, apathy and nausea.

The therapeutic doses of digoxin produce electrocardiographic changes such as sagging of the ST segment, T-wave inversion or prolongation of the P-R interval. These are not indicative of toxicity. Supraventricular arrhythmia, atrial systoles, junctional extra-systoles or tachycardia and ventricular extra-systoles, such as bigeminous rhythm are frequently encountered in childhood. Atrial tachycardia with irregular AV block is characteristic of digitalis over dosage.

Marked bradycardia with complete AV dissociation can occur and may require ventricular pacing to restore circulatory sufficiency.

#### *Treatment of Toxicity:*

In the majority all that is required is to stop the drug. Hypopotassaemia if present should be corrected. Tachyarrhythmias are treated with potassium administration and dilantin (dose

3.5 mg/Kg.) and Inderal 0.01 mg/kg i.v.) can be given. Potassium is contraindicated if bradycardia is present.

#### *Diuretics:*

Oral diuretics used are cholorthiazide (20-40 mg/kg oral daily), and Lasix (2-3 mg/kg daily dose and 1 mg/kg i.v. dose). Additionally aldactone (spironolactone, 1-2 mg/kg/day) can be given to avoid Potassium supplementation. Maintenance diuretics can be given as cholorthiazide 20-40 mg/kg orally on a 5 day a week schedule and do not require potassium supplements. KCl can be given as 1 mEq/kg daily dose.

#### *Position:*

Elevation of the upper body at 45° is helpful in alleviating dyspnea and can be achieved in infants with infant seat.

Children with respiratory distress and hypoxemia should receive adequate oxygen with humidity.

#### *Diet:*

Most children and infants do not require salt restriction. However, in severe failure in older children added salt in the regular diet should be eliminated.

#### *Cardiovascular Support:*

In desperately sick child with acute congestive heart failure and respiratory insufficiency ventilatory support may be required and cardiovascular support can be provided by Isoproterenol and dopamine administration.

Blood gasses, urine output, systemic blood pressure and left atrial pressure should be monitored to maintain fluid balance and evaluate response to the therapy.

## References:

1. Katz, A.M. In *Physiology of the Heart*. Raven press, 1977 p. 397.
2. Yang S.S., Bentivoglio L.G., Marahao V., Goldberg H. In *From cardiac Catheterization data to hemodynamic Parameters* FA Davis CO, 1978, PA, p. 249.
3. Grossman W., Jones D., McLaurin L.P. Wall stress and patterns of hypertrophy in human left ventricle *J. Clin. Invest.* 56-64, 1975.
4. Aziz K.U., Grondelle A. Van, Paul M.H. Muster A.J. Echocardiographic assessment of the relation between left ventricular wall and cavity dimension and peak systolic pressure in children with Aortic stenosis. *A.J. Cardiol* 40, 775:-779, 1977.
5. Meerson F.Z. Myocardium in hyper perfusion hypertrophy and heart failure. A.H.A. monograph No. 26 American Heart Association, New York.
6. Gaasch W.H., Andrias C.W., Levine H.J. Chronic Aortic regurgitation. The effect of aortic valve replacement on left ventricular volume, Mass and Function. *Circulation* 58:825, 1978.
7. Sonnenblick E.H. Determinants of active state in the heart muscle. Force, velocity, instantaneous muscle length, time. *Fed. Proc.* 24:1396-1409, 1965.
8. Davis J.O., Hartroft, P.M., Titus, E.O., Carpenter, C.J., and Spiegel E.H. The role of renin-angiotensin system in control of aldosterone secretion. *J. Clin. Invest.* 41: 378, 1962.
9. Less, M.H. Catecholamine metabolite excretion of infants with heart failure *J. Pediatr* 69-259, 1966.
10. Talner N.S. Pathophysiology of Cardiac failure in the new born. In *Pathophysiology of congenital heart disease* edited by F.H. Adams, H.J. Swan and V.E. Hall University of Cal Press, Berkelay USA. Forum in Medical Sciences No. 10, 1970 p. 119.
11. Talner N.S., Sanyal S.K., Halloran K.H., Gardner T.H., and Ordway N.K. Congestive heart failure in infancy. Abnormalities in blood gases and acid—Base equilibrium. *Pediatrics* 35:20, 1965.
12. Hayes C.J., Butler V.P. Jr. and Gersony W.M. Serum Digoxin studies in infants and children. *Pediatrics* 52:561, 1973.

—:0:—