Heart Failure: Signs and symptoms, pathophysiology, how does it present in children?

General Presentation

Background

Heart Failure (HF) is the clinical condition in which the heart fails to meet the metabolic and circulatory demands of the body. It presents as the final common pathway of a combination of structural, functional and biologic mechanisms. Pulmonary and/or systemic congestion may develop as a consequence of heart failure, resulting in Congestive Heart Failure (CHF). Although the incidence of Heart Failure in children is difficult to estimate, its importance lies in its many causes and etiological factors.

Presentation

Fundamentally, differences exist depending on whether or not HF presents in patients with a structurally normal heart, or patients with congenital heart disease. Virtually all patients who develop HF from congenital heart lesions do so by 6 months of age. Patients who acquire HF from acquired conditions may do so at any age.

Infants with heart failure often present with non-specific signs, including irritability, diaphoresis with feeds, failure to thrive. Older Children with HF may present with more classic features such as fatigue, exercise intolerance, breathlessness, and/or evidence of pulmonary congestion.

Pathophysiology

CHF occurs due to the heart's inability to keep up with hemodynamic needs, and can result from worsening systolic and/or diastolic dysfunction. In systolic dysfunction, the stroke volume decreases, thereby reducing cardiac output. Subsequently, the heart responds with 3 compensatory mechanisms: a) increasing left ventricular volume or elasticity, b) increasing contractile state by activation of circulating catecholamines, or c) increasing filling or preload. Each compensatory mechanism is limited, so in an untreated patient, the heart fails, leading to HF.

On the other hand, in diastolic dysfunction, stroke volume is decreased from decreased ventricular filling. To compensate, left ventricular end-diastolic pressure is increased. Diastolic dysfunction may be caused by conditions such as hypertension (causing ventricular hypertrophy).

Particularly in children, one of many congenital abnormalities may lead to dysfunction of the heart, causing increased compensatory mechanisms and the
HF. The mechanism for heart failure include volume overloading (from right to left sided shunting, for example), valvular incompetence, increased afterload (such as valvular stenosis or coarctation), and others.

Questions to Ask

- Infants: (asking the parents)
  - How are they feeding? Does the baby “tire out”, or have to rest in the middle of feeding? Does the baby change colour during feeds?
  - Is the baby growing?
  - Any episodes of blueness around the lips or face?

- Children
  - Do you feel short of breath when exercising? Can you keep up with other children? Can you run or play as much as before?
  - Do you feel short of breath when lying down?
  - Do your hands and/or feet feel constantly cold?
  - Do you often feel sweaty?
  - To parents: Have you noticed a change in their activity level? Are they keeping up with other kids? Any episodes of blueness? Noticed any facial puffiness? (facial edema) Do they seem tired?

Differential Diagnosis

The etiology of HF varies depending on whether or not the congenital abnormalities exist. Tables 1 and 2 list common causes of HF in patients with a structurally normal heart, and with congenital heart disease, respectively.

Table 1: Common causes of heart failure in the structurally normal heart

<table>
<thead>
<tr>
<th>Prenatal</th>
<th>Neonates and infants</th>
<th>Childhood</th>
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</thead>
<tbody>
<tr>
<td>Anemia</td>
<td>Anemia</td>
<td>Acquired valve disorders</td>
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<tr>
<td>Arrhythmia</td>
<td>Arrhythmia</td>
<td>Anemia</td>
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<tr>
<td>Arteriovenous fistula</td>
<td>Arteriovenous fistula</td>
<td>Arrhythmia</td>
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<tr>
<td>Cardiomyopathy</td>
<td>Dilated cardiomyopathy</td>
<td>Dilated cardiomyopathy</td>
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<tr>
<td>Twin-twin transfusion</td>
<td>Endocrinopathies</td>
<td>Hypertension</td>
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<tr>
<td></td>
<td>Hypoglycemia</td>
<td>Renal failure</td>
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</tbody>
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Hypothyroidism  
Restrictive cardiomyopathy  
Hypoxic ischemic injury  
Hypertension  
Infection/sepsis  
Kawasaki syndrome

Table 2: Common causes of heart failure in patients with congenital heart disease.

<table>
<thead>
<tr>
<th>Prenatal</th>
<th>Neonates and Infants</th>
<th>Children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrioventricular valve regurgitation</td>
<td>Systemic outflow obstruction</td>
<td>Aortic regurgitation</td>
</tr>
<tr>
<td>Mitral stenosis with intact atrial septum</td>
<td>Aortic valve stenosis</td>
<td>Mitral regurgitation</td>
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<tr>
<td>Coarctation of the aorta</td>
<td></td>
<td>Mitral stenosis</td>
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<tr>
<td>Subaortic stenosis</td>
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<td>Pulmonary vein stenosis</td>
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<tr>
<td>Systemic inflow obstruction</td>
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<td></td>
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<tr>
<td>Pulmonary venous stenosis</td>
<td></td>
<td></td>
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<tr>
<td>Systemic ventricular volume overload</td>
<td></td>
<td></td>
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<tr>
<td>Aortic or mitral regurgitation</td>
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<td></td>
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<tr>
<td>Atrial/Ventricular septal defect</td>
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<tr>
<td>Mitral stenosis</td>
<td></td>
<td></td>
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<tr>
<td>Patent ductus arteriosus</td>
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</tbody>
</table>

Physical Examination: a complete general physical exam with vitals
- **Vital signs:**
  - Tachycardia (>160 beats per minute in the neonate; >120 beats per minute in the older infant)
  - Tachypnea (>60 breaths per minute in the neonate; >40 breaths per minute in the older infant)
  - Blood pressure. Do 4 limb blood pressures if aortic coarctation is suspected.
  - Oxygen saturation is present in cyanotic congenital heart diseases.
- **Growth parameters, especially weight** – poor weight gain is a key indication of poorly compensated heart failure.
- **General appearance:**
  - Perspiration, Dysmorphic features (often associated with syndromes), cyanosis, increased work of breathing
- **Cardiovascular Exam:**
  - Pulses – feel for brachial, femoral, and pedal pulses. Pulses may be bounding or weak, depending on the underlying cause and the significance of the heart failure. There may also be a delay between the brachial and femoral pulses, in the case of coarctation
  - Capillary refill time
  - JVP – Useful in children older than 5-6 years old, although it may be difficult to obtain. In infants and younger children, right sided congestion tend to present as hepatomegaly and facial edema.
  - Precordial exam:
    - Palpate for thrills and right and left sided heaves
    - Listen for S1, S2. Abnormal S1 S2 may be a clue to valvular disease. A loud P2 is in strong indication of pulmonary overload.
    - Listen for gallop rhythms (S3, S4) and murmurs
    - Infants with cardiomyopathy often present with a quiet precordium
- **Respiratory Exam:**
  - Signs of increased work of breathing, including tachypnea, indrawing, tracheal tugging.
  - Auscultation, listening for signs of pulmonary edema
Laboratory Investigations:

- **Chest X-ray**: very commonly demonstrates cardiomegaly. The shape of the cardiac silhouette may give clues to certain structural heart diseases, and also to right versus left sided heart involvement. Pulmonary markings are often increased, showing pulmonary congestion. However, right sided heart failure may result in decreased pulmonary perfusion.

- **Electrocardiogram**: usually abnormal, and although not useful in assessing HF, may give diagnostic hints for the underlying disorder through demonstrating ventricular enlargement, atrial enlargement, ST changes associated with myocarditis / pericarditis, and arrhythmias.

- **Urine test**: In chronic heart failure, proteinuria and high specific gravity of urine are common.

- **Blood test**: An increase in blood urea nitrogen and creatinine levels may be present, as renal function decreased due to decreased perfusion. CBC, differential may give clues to anemia and infection causing or complicating HF. Brain natriuretic peptide (BNP) may be used in some cases to track heart failure.

- **Echocardiogram**: Invaluable in ruling out structural heart disease and evaluating cardiac function, including atrial and ventricular size, systolic and diastolic function, valve anatomy and function, and the presence and hemodynamic significance of intra-cardiac shunts.

- **Endomyocardial biopsy**: for evaluation of myocarditis

- **Thyroid, Renal and Hepatic function tests**

References

