

Paediatric heart failure from the eyes of the cardiologist

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Proceedings

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Abstract

The causes of heart failure (HF) in children differ substantially from those found in the adult population. They comprise cardiac and noncardiac causes. Today, with the near-disappearance of rheumatic fever, the most common cause of pediatric HF are congenital heart diseases, whilst cardiomyopathies are the most common cause of HF in children with a structurally normal heart. Signs and symptoms of pediatric HF are often not specific, most of all in newborns (difficulties in breastfeeding, excessive sweating, growth retardation and poor weight gain). The aim of the therapy is to clinically control HF through general measures as well as drugs administration. In this respect, due to the lack of large interventional trials, the HF in newborns and children is often extrapolated from adult large trials. Cardiac transplantation is currently the treatment option with good outcome and long-term survival in pediatric patients with end-stage or refractory HF.

Keywords

Heart failure, newborns, children, congenital heart disease, therapy.

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Background

Heart failure (HF), also called cardiac insufficiency, is defined as a pathological condition in which the heart is unable to keep up with the demands and, specifically, to pump blood with normal efficiency.

HF presents similarities and differences between children and adults; in pediatric age the etiology of HF is wide and congenital heart defects (CHD) are the most common cause. However there are some peculiarities in clinical presentation of HF in infants and small children [1].

Physiopatology

A number of conditions, not always of cardiac origin, can damage or weaken the heart, thus causing HF [2].

Five possible HF underlying pathophysiological mechanisms have been identified:

1. volume overload;
2. pressure overload;
3. impaired myocardial contractility;
4. arrhythmias;
5. combination of the above stated mechanisms.

Volume and pressure overload are the most frequent causes of HF in paediatric age. Volume overload may be caused by CHD with left to right shunts, arteriovenous fistulas, valvular insufficiency. Pressure overload may be caused by a reduced ventricular filling (mitral stenosis) or output (pulmonary valve stenosis, aortic valve stenosis, aortic coarctation).

Impaired myocardial contractility as a cause of HF in neonates and children is rare, being mainly due to cardiomyopathies, myocarditis, metabolic disorders (hypoxia, hypoglycemia, hypocalcemia, acidosis), endocrine disorders, and coronary anomalies (anomalous left coronary artery from pulmonary artery).

Arrhythmias include tachycardias (such as paroxysmal supraventricular tachycardia) and bradycardias (too slow heart rate and atrio-ventricular blocks).

All these mechanisms lead to a reduced stroke volume, which is the volume of blood pumped from the left ventricle of the heart with each beat.

Cardiac dysfunction induces several changes in vascular function, blood volume, and neurohumoral status. All these changes serve as compensatory mechanisms to help maintain stroke volume (primarily by the Frank-Starling mechanism) and arterial blood pressure (by systemic vasoconstriction). However, these compensatory changes can worsen the cardiac function as time passes. They are:

- ventricular myocardial hypertrophy;
- sympathetic nervous system activation;
- renin-angiotensin-aldosterone system activation;
- ventricular end-diastolic volume enlargement.

Myocardial hypertrophy leads to increased ventricular contractility, whilst the activation of the sympathetic nervous system (through the increase in catecholamines levels) improves the ventricular contractility and heart frequency. Furthermore, a beneficial blood flow redistribution to the main organs – rather than to skin, kidney, and spleen – is registered. Conversely, sympathetic nervous system activation increases the peripheral vascular resistances (vasoconstriction) and the ventricular overload as well. The latter counteracts the cardiac output.

Chronic renal hypoperfusion triggers a vasoconstriction (increased overload), as well as a hypervolemia induced by salt and water retention. Hypervolemia implies an increase in venous return to the heart (preload) and therefore in the filling pressure of the ventricle, thus leading to increased stroke volume.

The ability of the heart to change its force of contraction and therefore stroke volume in response to changes in venous return is called the “Frank-Starling mechanism”. In fact, cardiac myocytes initial stretching increases the sarcomere length, which causes an increase in force generation. This mechanism enables the heart to eject the additional venous return, thereby increasing the stroke volume.

However the Frank-Starling compensatory mechanism induces a ventricular end-diastolic volume enlargement throughout time, with a consequent raise in the systemic and pulmonary venous pressures.

Systemic and pulmonary venous congestion are just responsible of several HF signs and symptoms.

Signs and symptoms

Signs and symptoms of HF are frequently not specific. Difficulties in breastfeeding is a common feature in newborns and correspond to reduced

exercise tolerance and dyspnea in children. In the latter, oedematous legs are rare [3].

The other signs and symptoms of HF may be classified on the basis of their pathophysiological mechanism:

1. reduced stroke volume: excessive, unexplained irritability and tachycardia (due to the increment in catecholamines blood levels in order to increase the stroke volume); cold skin and excessive sweating that is increased with feeding and may occur during sleeping (due to the increment in catecholamines blood levels); growth retardation and poor weight gain (due to the reduction in calories intake because of the difficult feeding as well as to the increase in the calories need because of the raise in cardiopulmonary work);
2. pulmonary venous congestion: pressure increases inside the left atrium and then in the veins and capillaries in the lungs, causing fluid to be pushed through the capillary walls into the air sacs. This condition results in tachypnea and increased work of breathing. Unusual breathing movements (such as drawing back of the chest muscles at epigastrium, intercostals spaces, and jugular notch) as well as nasal flaring are common features in newborns suffering from HF. Orthopnea is rare, being relatively more frequent in children. The most severe consequence of HF is pulmonary edema, with the progressive development of wheezing in the lungs from bases to apices;
3. systemic venous congestion: hepatomegaly (which is the most common sign of HF in this group of patients. It is not pathognomonic of HF, being present in case of acute respiratory distress as well, due to the downwards thrust of the lungs. Absence of hepatomegaly does not exclude the little patient is suffering from HF, because it may appear later. Peripheral edema (legs, eye lids) may be a late sign of HF in children, not in newborns. The same for jugular veins distention and ascites. Excessive weight gain owing to idrosaline retention is another sign which may involve newborns as well as children.

Diagnostic tools

Chest X-ray may show a heart enlargement, which is always present in case of HF, with the exception of the cardiac insufficiency due to reduced preload, such as in CHD like total pulmonary venous return, mitral valve stenosis, cor triatriatum.

The heart is enlarged if the cardiothoracic ratio is greater than 50% on a posterior-anterior view.

However, cardiomegaly is not pathognomonic of HF. For example, children suffering from CHD with left-to-right shunt may show heart enlargement at radiograph, without HF. Chest X-ray is a useful tool to evaluate the pulmonary venous congestion as well as to identify the underlying cause of HF (for example “snowman image” in case of total pulmonary venous return; “coeur en sabot” in case of tetralogy of Fallot).

ECG may be a diagnostic tool if the cause of HF are arrhythmias.

Echocardiography is able to detect the dilation of the cardiac chambers, the ventricular ejection fraction, and the underlying cause of HF [4].

Differential diagnosis

HF signs and symptoms are similar to those in patients affected by respiratory infections. However anamnesis, clinics, inflammatory markers, and chest X-ray are useful tools to distinguish HF from pulmonary disease patients [4].

Therapy

The aim of the therapy is to clinically control HF through general measures and drugs administration.

Specifically, if the underlying cause of HF is a CHD, the first step is to prevent or control acute HF through drugs such as prostaglandins and inotropic agents i.v. The second step is the surgical correction of the CHD.

If HF is due to severe anemia, hypervolemia, hypoglycemia, hypocalcemia or hypertyroidism, the goal of the HF therapy is to treat the above stated conditions.

If HF is due to arrhythmias, the aim of the therapy is to restore a normal cardiac frequency, by means of antiarrhythmic agents in case of tachycardias or isoproterenol and/or external pacing in case of severe bradycardias and atrio-ventricular blocks [5].

General measures

- Thirty degrees sitting position, in order to improve pulmonary venous congestion and dyspnea.
- Increased oxygen administration (concentration 30-50%), even in case of no cyanosis. The only contraindication to this therapeutic measure are the ductus-dependent systemic CHDs, because

an elevated oxygen concentration in the blood may be responsible of the ductal closure.

- Decrease in oxygen consumption, through morphine-induced sedation (0.1 mg/kg i.v.), most of all in case of pulmonary edema.
- Forced feeding (through a tube passing through the nose and into the stomach) or alimentionation subdivided into most frequent hypercaloric feedings. In both cases the aim is to reduce the patient's fatigue.
- Cardio-respiratory monitoring (weight, cardiac and respiratory frequencies, blood pressure, diuresis).
- Continuous positive airway pressure (CPAP) with oxygen 100%, with the aim to improve oxygen blood saturation and reduce the amount of fluid which passes through the capillary walls into the air sacs.

Drug therapy

In paediatric age there are few prospective studies about pharmacology of HF. Due to the lack of large interventional trials, the HF therapy in newborns and children is often extrapolated from adult large trials.

Therapeutic interventions to improve cardiac function in HF include the use of cardiostimulatory drugs (i.e. beta-agonists and digitalis) in order to stimulate heart rate and contractility, and vasodilator drugs with the aim to reduce ventricular afterload and thereby enhance the stroke volume (angiotensin-converting enzyme inhibitors).

Treatment efforts with loops diuretics are directed towards the lung congestion reduction.

The use of beta-blockers, which is strongly recommended in adult HF guidelines, is still in its pioneering phase in newborns and children. In this respect, in literature only few studies performed in single Centers with a small sample size have been reported.

There are also some reported experiences with new inotropics drugs in acute HF.

Non pharmacological treatment with autonomic implantable cardioverter defibrillators and resynchronization therapy as well as the surgically

implant of ventricular assist devices are increasingly employed in children over the last years. Cardiac transplantation is currently the treatment option with good outcome and long-term survival in pediatric patients with end-stage or refractory HF.

Declaration of interest

The Authors declare that there is no conflict of interest.

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