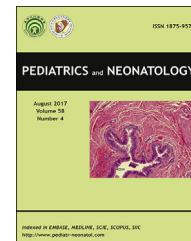




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REVIEW ARTICLE

# Pediatric Heart Failure: A Practical Guide to Diagnosis and Management



Daniele Masarone\*, Fabio Valente, Marta Rubino, Rossella Vastarella, Rita Gravino, Alessandra Rea, Maria Giovanna Russo, Giuseppe Pacileo, Giuseppe Limongelli

*Cardiologia SUN – Heart Failure Unit, Department of Cardiothoracic Sciences, Second University of Naples, Naples, Italy*

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Pediatric heart failure represents an important cause of morbidity and mortality in childhood. Currently, there are well-established guidelines for the management of heart failure in the adult population, but an equivalent consensus in children is lacking. In the clinical setting, ensuring an accurate diagnosis and defining etiology is essential to optimal treatment. Diuretics and angiotensin-converting enzyme inhibition are the first-line therapies, whereas beta-blockers and devices for electric therapy are less used in children than in adults. In the end-stage disease, heart transplantation is the best choice of treatment, while a left ventricular assist device can be used as a bridge to transplantation (due to the difficulties in finding organ donors), recovery (in the case of myocarditis), or destination therapy (for patients with systemic disease).

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## 1. Introduction

Pediatric heart failure (PHF) represents an important cause of morbidity and mortality in childhood.<sup>1</sup> Etiology and pathogenesis are different between adults and children:

the first mainly relates to ischemia (60–70% of cases), the latter as a consequence of congenital heart diseases (CHDs) or cardiomyopathies in most of the cases.<sup>2</sup> Hence, managing PHF requires specific knowledge and skills.<sup>3</sup> Presently, there are well-established guidelines for the management of heart failure (HF) in the adult population,<sup>4</sup> but the equivalent consensus for PHF is lacking. This article offers an overview on the etiology, diagnosis, and therapy of PHF, with a specific focus on practical issues required for management.

\* Corresponding author. Cardiologia SUN – Heart Failure Unit, Department of Cardiothoracic Sciences, Second University of Naples, Via Leonardo Bianchi n°1, Naples 80100, Italy.  
E-mail address: [danielemasarone@libero.it](mailto:danielemasarone@libero.it) (D. Masarone).

## 2. Definition

In the 1950s, HF was described as a clinical syndrome caused by low cardiac output.<sup>5</sup> In recent years, knowledge of the pathophysiology has been expanded and neurohormonal and molecular pathways that modulate cardiac performance in failing hearts have been discovered.<sup>6</sup> The contemporary vision describes HF as a clinical syndrome characterized by typical symptoms and signs associated with specific circulatory, neurohormonal, and molecular abnormalities.<sup>7</sup>

## 3. Etiology

In children, cardiac failure is most often due to CHDs and cardiomyopathies. The cardiac and noncardiac causes of PHF are summarized in Table 1. At birth, HF is caused by fetal cardiomyopathies or extracardiac conditions (such as sepsis, hypoglycaemia, and hypocalcaemia). In the 1<sup>st</sup> week after birth, CHDs with ductus-dependent systemic circulation (such as severe aortic stenosis/aortic coarctation and hypoplastic left heart syndrome), in which the closure of the ductus arteriosus causes severe reduction of end-organ perfusion,<sup>8</sup>

**Table 1** Etiology of pediatric heart failure.

Type of diseases	Pathophysiology	Examples
Congenital heart diseases	Left to right shunt (volume overload)	Ventricular septal defects Complete atrioventricular canal defects Patent ductus arteriosus Aorto–pulmonary windows
	Valvular regurgitation (volume overload)	Mitral regurgitation Aortic regurgitation
	Outflow tract obstruction (pressure overload)	Aortic stenosis  Tunnel type subaortic stenosis Supravalvular aortic stenosis Pulmonary stenosis Pulmonary vein stenosis
	Coronary insufficiency (decreased O <sub>2</sub> supply to cardiomyocyte)	Coronary artery anomalies
Cardiomyopathies (inherited or acquired)	Systolic dysfunction (low cardiac output)	Dilated cardiomyopathy
	Diastolic dysfunction (elevated pulmonary capillary pressure)	- Myocarditis - Barth syndrome - Carnitine deficiency - Familial dilated cardiomyopathy - Neuromuscular disorder (i.e., Becker dystrophy/ Duchenne dystrophy)
		Hypertrophic cardiomyopathy  - Pompe diseases - Noonan syndrome - Maternal diabetes - Mitochondrial diseases - Familial hypertrophic cardiomyopathy
Arrhythmias	Systolic dysfunction (low cardiac output)	Idiopathic restrictive cardiomyopathy
		Tachycardia induced cardiomyopathy  - Atrio–ventricular node reentry tachycardia - Atrio–ventricular reentry tachycardia - Ectopic atrial tachycardia
Infection	Systolic dysfunction	Congenital third degree atrio–ventricular block Sepsis induced myocardial dysfunction
High output state	Volume overload	Thyrotoxicosis Systemic arteriovenous fistula Severe anemia

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