

Canadian Cardiovascular Society/Canadian Pediatric Cardiology Association Focused Guidelines on Acute Heart Failure in Children.

Canadian Cardiovascular Society/Canadian Pediatric Cardiology Association · 2026

Pediatric Acute Heart Failure: CCS/CPCA Focused Guidelines Study Guide

This study guide provides a comprehensive synthesis of the Canadian Cardiovascular Society (CCS) and Canadian Pediatric Cardiology Association (CPCA) focused guidelines for the management of acute heart failure (AHF) in children. It covers diagnostic evaluation, pharmacological interventions, monitoring strategies, and advanced mechanical support.

Core Guidelines and Recommendations

The guidelines address ten prioritized clinical questions regarding the diagnosis and management of pediatric AHF. The following table summarizes the strength of each recommendation and the certainty of the underlying evidence.

Recommendation Number	Topic	Summary of Recommendation	Strength; Certainty
1	CMR	Perform Cardiac Magnetic Resonance (CMR) in children with suspected myocarditis to confirm diagnosis and risk stratify.	Conditional; Very Low
2	Biomarkers	Use natriuretic peptides (BNP or NT-proBNP) as prognostic factors.	Conditional; Very Low
3	Genetics	Perform Whole Exome Sequencing (WES) rather than targeted panels.	Conditional; Very Low
4	Diuretics	Use early diuretic therapy (within 6 hours of presentation) for new-onset symptomatic HF.	Conditional; Low
5	GDMT	Alternating (simultaneous) initiation of medications can be used instead of sequential titration.	Conditional; Low
6	Inotropes	Milrinone is the preferred inotrope over dobutamine or levosimendan.	Conditional; Very Low
7	Monitoring	Routine invasive monitoring with a pulmonary artery catheter (PAC) is suggested against.	Conditional Against; Very Low
8	VAD Timing	Implant Ventricular Assist Devices (VAD) before the development of cardiogenic shock (INTERMACS/PEDIMACS profile ≥ 2).	Strong; Very Low
9	Temporary MCS	Use temporary mechanical circulatory support (tMCS) over VA-ECMO when feasible.	Conditional; Very Low
10	ECMO Decompression	Perform left heart decompression within the first 24 hours of VA-ECMO support.	Conditional; Low

Key Concepts and Clinical Workflows

1. Diagnostic Evaluation and Risk Stratification

The diagnostic pathway for suspected AHF begins with clinical evaluation (history, physical examination, ECG, and chest X-ray), followed by biomarker testing and echocardiography to confirm pump failure.

- **Cardiac Magnetic Resonance Imaging (CMR):** Used to differentiate myocarditis from noninflammatory conditions like dilated cardiomyopathy. Tissue characteristics in myocarditis include

myocardial edema (bright on T2-weighted images), prolonged T1 and T2 mapping times, and positive late gadolinium enhancement.

- **Biomarkers:** Universal measurement of BNP or NT-proBNP is endorsed. Peak levels should be established within 12–24 hours of admission. PredischARGE measurements are recommended to establish a patient-specific baseline.
- **Genetic Testing:** WES is preferred as a first-tier test, particularly for patients under two years of age or those with multisystem disease.

2. Pharmacological Management

Management focuses on rapid stabilization and the initiation of Guideline-Directed Medical Therapy (GDMT).

- **Diuretic Strategy:** Early initiation (within 6 hours) via intravenous bolus is preferred for congested patients. Monitoring must include urine output (mL/kg/h), renal function, and electrolytes.
- **Inotropic Support:** Milrinone is the first-choice agent for AHF. It should be used for the shortest duration possible, with clinicians weaning the patient as they transition to enteral (oral) therapies.
- **GDMT Initiation:**
 - **Sequential:** One medication is started and titrated to the target dose before the next is introduced.
 - **Alternating:** Multiple medications (e.g., RAAS inhibitors and Beta-blockers) are started simultaneously at low doses and titrated alternately. This aim is to reach therapeutic dosing within approximately 14 days.

3. Advanced Monitoring and Mechanical Support

The guidelines prioritize early intervention to prevent the "crash and burn" scenario of established cardiogenic shock.

- **INTERMACS/PEDIMACS Profiles:** A scale used to determine the urgency of support. Profile 1 represents critical cardiogenic shock ("crash and burn"). The guidelines strongly recommend VAD implantation at Profile 2 or higher (stable but inotrope-dependent) to improve survival outcomes.
 - **VA-ECMO and Decompression:** In patients supported by Veno-Arterial Extracorporeal Membrane Oxygenation (VA-ECMO), early left heart decompression (within 24 hours) is critical to prevent the clinical sequelae of left atrial (LA) hypertension, such as loss of arterial pulsatility and increasing pulmonary congestion.
 - **tMCS vs. VA-ECMO:** Temporary mechanical circulatory support (e.g., centrifugal flow pumps or percutaneous axial flow pumps) is suggested over VA-ECMO for pediatric AHF, though implementation may depend on institutional expertise.
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Short-Answer Quiz

Q1: What is the recommended timeframe for initiating diuretic therapy in a child with new-onset symptomatic heart failure? *Answer:* Within 6 hours of presentation.

Q2: Why is Whole Exome Sequencing (WES) preferred over targeted cardiomyopathy panels in young children with AHF? *Answer:* WES is more likely to identify underlying multisystem disorders that may not be represented on a standard cardiomyopathy panel, particularly in patients younger than 2 years old.

Q3: According to the guidelines, which inotropic agent is preferred for children with AHF? *Answer:* Milrinone.

Q4: What are the three primary tissue characteristics of myocarditis visualized via CMR? *Answer:* Myocardial injury/edema (T2-weighted), prolonged T1/T2 mapping times, and late gadolinium enhancement.

Q5: At what INTERMACS/PEDIMACS profile level does the guideline recommend VAD implantation? *Answer:* Profile ≥ 2 (before the development of profile 1/cardiogenic shock).

Q6: What is the clinical rationale for left heart decompression within 24 hours for a patient on VA-ECMO? *Answer:* To avoid the morbidity associated with left atrial (LA) hypertension and pulmonary congestion.

Essay Questions for Deeper Exploration

1. Compare and Contrast: Alternating vs. Sequential GDMT Initiation. Discuss the traditional sequential approach to Guideline-Directed Medical Therapy versus the newly suggested alternating approach. In your response, address the rationale behind rapid up-titration, the specific drug classes involved (RAAS inhibitors, BBs, MRAs), and the necessary safety monitoring required for hospitalized pediatric patients.

2. The Evolution of Mechanical Circulatory Support (MCS) in Pediatric AHF. Explain the transition from using VA-ECMO as a primary support tool to the preference for temporary mechanical circulatory support (tMCS) and durable VADs. Analyze how INTERMACS/PEDIMACS profiling influences the timing of these interventions and the impact of "early" vs. "late" implantation on pediatric survival rates.

3. The Role of Advanced Diagnostics in Risk Stratification. Evaluate the importance of CMR and natriuretic peptides in the management of pediatric AHF. How do these modalities assist clinicians in differentiating etiologies (e.g., myocarditis vs. dilated cardiomyopathy) and establishing long-term prognostic baselines for patients?

Glossary of Important Terms

- **AHF:** Acute Heart Failure; a complex clinical syndrome with high morbidity/mortality requiring intensive management.
- **BNP / NT-proBNP:** B-type Natriuretic Peptide and N-terminal pro-hormone B-type natriuretic peptide; biomarkers used for diagnosis and prognosis.
- **CMR:** Cardiac Magnetic Resonance; an imaging modality used to define myocardial injury, edema, and inflammation.
- **GDMT:** Guideline-Directed Medical Therapy; the mainstay of medical management involving RAAS inhibitors, Beta-blockers, and Mineralocorticoid Receptor Antagonists (MRA).
- **INTERMACS / PEDIMACS:** A classification system for the severity of heart failure and the urgency of mechanical support; Profile 1 is the most critical.
- **Left Atrial Decompression:** A procedure (e.g., balloon atrial septostomy or venting) to reduce pressure in the left atrium, often necessary during VA-ECMO support.
- **RAAS Inhibitors:** Renin-angiotensin-aldosterone system inhibitors, including ACE inhibitors (ACEi) and Angiotensin-receptor/neprilysin inhibitors (ARNI).
- **tMCS:** Temporary Mechanical Circulatory Support; devices such as centrifugal flow pumps or percutaneous axial flow pumps used for short-term cardiac support.
- **VA-ECMO:** Veno-Arterial Extracorporeal Membrane Oxygenation; a form of life support that provides both cardiac and respiratory gas exchange.
- **WES:** Whole Exome Sequencing; a comprehensive genetic test that sequences all protein-coding regions of the genome.