

Clinical Guidance

Paediatric Critical Care: Myocarditis/Cardiomyopathy

Clinical guideline on the diagnosis and management of paediatric myocarditis and cardiomyopathy, for use on PICU and for hospitals referring to STRS.

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Glossary: ECMO is used to represent mechanical support that would be used acutely. For individual patient, other modalities like ventricular assist device (VAD) may be used. ALCAPA: Anomalous Left Coronary Artery from Pulmonary Artery

MYOCARDITIS is the most common acquired heart disease in children. A wide range of aetiologies cause inflammation of the myocardium, which may resolve, stabilise, or lead to dilated cardiomyopathy (DCM).

CARDIOMYOPATHY refers to disease of the myocardium, which causes it to become dilated, hypertrophic, restrictive or arrhythmogenic.

Suspect myocarditis if viral illness + symptoms of heart failure + persistent tachycardia + abnormal CXR or ECG

Doing both CXR and ECG increases diagnostic sensitivity to 97% from 55% (CXR alone) and 93% (ECG alone)⁶

Clinical Presentation:

- Respiratory: often misdiagnosed as resp illness e.g. bronchiolitis
- Cardiac failure: Symptoms (e.g. fatigue/ poor feeding) plus signs (hepatomegaly, elevated JVP, CXR-cardiomegaly/pulmonary oedema)
- Arrhythmia: palpitations +/- syncope
- Shock presenting with abdominal pain, vomiting, tachycardia, narrow pulse pressure and hypotension

Differential Diagnoses: Coronary anomalies (e.g. ALCAPA), severe sepsis (acute myocardial dysfunction), primary arrhythmia resulting in myocardial dysfunction (e.g. prolonged SVT)

Continual assessment of fluid balance is critical to determine which patients will benefit from fluid

First line investigations

- Blood gas, FBC, CRP, U&E, LFT, Troponin, BNP, CK, vit D, TFTs
- Viral serology (coxsackie, adenovirus, EBV, CMV, Parvovirus B19, SARS-CoV-2), NPA, throat swab, cultures (Blood, ET, urine)
- **ECG:** sinus tachycardia, heart block, ST changes, axis deviation, small voltage (inflammation/effusion), arrhythmias/ectopics
- **CXR:** cardiomegaly, pulmonary plethora, pleural effusion
- **Echo:** Ventricular function, effusion, valve regurgitation; rule out other causes of cardiac failure (e.g. ALCAPA)

Second Line investigations

- Genetic & Metabolic screen often diagnostic
- Ferritin, AutoAbs(ANA, ANCA, ACE, anti-dsDNA)
- ASOT, mycoplasma pneumoniae PCR, viral serology (HIV, HBV, HCV), Borrelia serology
- **Cardiac MRI**
- **FDG-PET:** detects metabolically active disease; inflamed myocardium shows decreased perfusion and increased FDG uptake.

Initial management: Involve paediatric cardiology and STRS early

- Give supplemental oxygen to maintain saturations $\geq 90\%$
- Continuous ECG monitoring, set BP to cycle every 5 min
- Commence CPAP as a temporising measure: positive pressure ventilation aids LV function and reduces metabolic demand
- Consider milrinone 0.5mcg/kg/min- improves cardiac output without O₂ demand (hypotension unlikely as no load)

Assess fluid status:

- Shock and no pulm oedema: trial of *cautious fluid resuscitation* (2-5mL/kg aliquots). Titrate to HR, BP, CRT, lactate, end organ perfusion Reassess++ and stop if worsening respiratory status or hepatomegaly
- Pulmonary oedema: IV furosemide 0.5-1mg/kg (max initial dose 20mg)

Emergency Management and Stabilisation:

- **Intubation:** induction of anaesthesia is \uparrow risk: \downarrow endogenous sympathetic response, sudden \downarrow SVR/ BP, pulmonary oedema risk
- Indications for intubation – persistent desaturation, unresponsive hypotension, reduced consciousness or senior decision
 - Start peripheral low dose adrenaline infusion (0.02-0.1mcg/kg/min) prior to induction. Caution: higher doses of adrenaline may cause tachycardia, arrhythmias, vasoconstriction
 - Prepare resuscitation drugs and dilute adrenaline (0.1mL/kg 1:10,000 adrenaline diluted to 10mL of 0.9% sodium chloride)
 - Use small doses of cardio-stable anaesthetic drugs (fentanyl 1-2mcg/kg, ketamine 1mg/kg and rocuronium 1mg/kg).
The onset of drugs may be slow (longer arm-brain circulation time due to low cardiac output)
 - The most experienced operator should intubate
Have a defibrillator and ECMO team on standby if available
- **Haemodynamic support:** aim to maximise perfusion/ O₂ delivery
 - Milrinone inotrope of choice. Initially 0.5mcg/kg/min but further afterload reduction can be considered by increasing up to 1mcg/kg/min or levosimendan is alternative (senior decision)
 - Consider transfusion if Hb <70 g/L. ⁷ Weigh up benefit of O₂ delivery vs risk of fluid overload (diuresis if necessary)
 - Ideally sedate with morphine/clonidine infusion +/- muscle relaxation if intubated. Caution with midazolam/propofol

Ongoing Management:

- CVC (avoid RIJV anticipating ECMO may be needed)
- Arterial access - titrate MAP to end organ perfusion and monitor NIRS
- Anticoagulation ([Anti-thrombotic therapy guideline](#))
- Severe viral myocarditis: high dose corticosteroids + IVIG improve outcomes^{2,4} and consider anti-virals. Liaise with PIID (IVIG is not commissioned for routine use in myocarditis)⁵
- Immune-mediated disease (e.g. PIMS-TS)- corticosteroids/ immunomodulators
- Rhythm control: Loss of sinus rhythm is associated with 8x increase in need for: ECMO, heart transplant or death¹
 - Maintain normothermia, correct electrolytes, antiarrhythmic drugs (consult cardiology) [PICU arrhythmia guideline](#)
- **Organ support:**
 - GI: caution with enteral feeds in LCOS (risk of NEC),
 - Consider PD/CVVH support if IV furosemide infusion insufficient.
 - CNS: NIRS, consider sedation +/- muscle relaxant to decrease oxygen demand.
- Cardiac transplant: timely discussion

Prognosis: The overall mortality for paediatric myocarditis is 7.3%³. 30% of cases of biopsy-confirmed myocarditis progress to dilated cardiomyopathy. Poor prognosis is associated with age <12 years old, low LV ejection fraction at time of hospital admission, ST changes on ECG.

Post-mortem useful if no definitive diagnosis

References: 1. Lampejo et al. Acute myocarditis. Clin Med. 2021; 21(5):505-510. 2. Yen et al. Role of IVIG in paediatric acute myocarditis: systematic review/meta-analysis. Sci Rep. 2019; 9(10459). 3. Ghelani et al. Paed acute myocarditis in the United States, 2006-2011. Circ Cardiovasc Qual Outcomes. 2012;5(5):622-627. 4. Schauer et al. Pediatric myocarditis-high dose steroids and IVIG. Paed cardio (2023) 44:441-450. 5. Commissioning Criteria Policy IVIG England, 2021 6. Freedman et al. Pediatric Myocarditis: ED Clinical Findings and Diagnostic Evaluation. Ped. 2007;120:1278. 7. Doctor et al. RBC Transfusion in General Critically Ill Children Based on Hb and/or Physiologic Thresholds From the Pediatric Critical Care TAXI. Pediatr Crit Care Med. 2018 Sep;19(9S Suppl 1):S98-S113.