Multisystem inflammatory syndrome in children

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1 Multisystem inflammatory syndrome in children (MIS-C) is a hyperinflammatory condition associated with a history of SARS-CoV-2 infection

From March 2020 to May 2021, 406 cases were reported in Canada.[1] As of January 2022, the United States reported 1 MIS-C case per 3200 SARS-CoV-2 infections, and 59 deaths.[2] Patients usually present within 6 weeks after SARS-CoV-2 infection, and most are positive for SARS-CoV-2 antibodies.[3–5]

2 Diagnostic criteria include fever and multiorgan involvement

Most patients are school-aged children (median age 8 yr). Risk factors include male sex; obesity; and Black, Hispanic or South Asian ethnicity.[2,3,5] Although case definitions vary worldwide, diagnostic criteria include fever for at least 24 hours, multisystem involvement and evidence of inflammation.[2–4] Cardiovascular shock is common. Older children usually present with gastrointestinal involvement (abdominal pain, diarrhea and vomiting), whereas younger children present with rash, conjunctivitis, strawberry tongue or swollen extremities, similar to Kawasaki disease.[3–5] Differential diagnoses include bacterial sepsis, appendicitis and toxic shock syndrome.

3 Suspected or confirmed MIS-C should prompt hospitalization

Initial symptoms may be mild; however, children can deteriorate quickly and may require intensive care support. Most patients have elevated inflammatory markers (C-reactive protein, ferritin), lymphopenia, elevated cardiac enzymes (troponin, B-type natriuretic peptide) and evidence of coagulopathy, including elevated D-dimer.[3–5] Initial treatment with broad-spectrum, intravenous (IV) antibiotics is essential as clinical features overlap with sepsis.[1,4,5] Immunomodulation with IV immunoglobulin, with or without methylprednisolone, is the first-line treatment, and should be guided by specialists.[2,4,5]

4 Two-thirds of patients will have cardiac involvement

Electrocardiography and echocardiography should be performed in all patients, and may identify features of myocarditis, pericarditis, valvulitis and pathological changes in coronary arteries.[2–5] Treatment includes fluid resuscitation, inotropic support and antiplatelet therapy.

5 Most patients recover fully from the hyperinflammatory state

Response to immunomodulation is usually excellent. Patients should be reviewed by a cardiologist to check resolution of cardiac dysfunction and development of aneurysms of the coronary arteries. Follow-up data are limited; however, in a cohort of 46 children, most (98%) were able to resume full-time education by 6 months, although some continued to report reduced exercise capacity and emotional dysregulation.[6]

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