

REFERENCES

1. Bhat CS, Gupta L, Balasubramanian S, Singh S, Ramanan AV. Hyperinflammatory syndrome in children associated with COVID-19: Need for awareness. *Indian Pediatr.* 2020;57:929-35.
2. Fouriki A, Fougère Y, De Camaret C, et al. Case series of children with multisystem inflammatory syndrome following SARS-CoV-2 infection in Switzerland. *Front Pediatr.* 2021;8:594127.
3. Harwood R, Allin B, Jones CE, et al. PIMS-TS National Consensus Management Study Group. A National Consensus Management Pathway For Paediatric Inflammatory Multisystem Syndrome Temporally Associated With COVID-19 (PIMS-TS): Results of a National Delphi Process. *Lancet Child Adolesc Health.* 2021;5:133-41.
4. Henderson LA, Canna SW, Friedman KG, et al. American College of Rheumatology Clinical Guidance for Multisystem Inflammatory Syndrome in Children Associated With SARS-CoV-2 and Hyperinflammation in Pediatric COVID-19: Version 2. *Arthritis Rheumatol.* 2021;73:e13-e29.
5. Nuffield Department of Population Health. Information for site staff 2020. Accessed June 21, 2021. Available from: <https://www.recoverytrial.net/for-site-staff>
6. Shakoory B, Carcillo JA, Chatham WW, et al. Interleukin-1 receptor blockade is associated with reduced mortality in sepsis patients with features of the macrophage activation syndrome: Re-analysis of a prior phase III trial. *Crit Care Med.* 2016;44:275-28.

Acute Severe Heart Failure in a Child With Congenital Heart Defect and Multisystem Inflammatory Syndrome in Children (MIS-C)

Myocardial involvement is a known feature of multisystem inflammatory syndrome in children (MIS-C), with mild ventricular dysfunction being the commonest finding. We describe a child who had congenital heart defect with complete heart block (CHB), and presented to us with MIS-C, but due to a rare complication succumbed to intractable heart failure.

A 13-year-old boy was admitted with a history of high-grade fever, rash, body ache and vomiting for 4 days, and mild swelling over feet for 2 days. There was no history of dyspnea, cyanosis, syncope, loss of consciousness, or neurological symptoms. Hailing from a containment zone area for coronavirus disease 19 (COVID-19) two months back, neither the child, nor his family members had been symptomatic nor tested for severe acute respiratory syndrome coronavirus (SARS-CoV-2) infection. He was a known case of corrected transposition of great arteries (CCTGA), moderate pulmonary stenosis (PS), intact interventricular septum, small ostium secundum atrial septal defect (ASD), and CHB, since infancy without any previous admissions.

On admission, he was conscious, irritable and febrile. Pulse rate was 80/min, respiratory rate 28/min, no distress, blood pressure 100/60 mmHg, and saturation 96% on room air. He was underweight (body mass index 12.4 kg/m²; <-3 SDS) and had a petechial rash over the body with bilateral pedal edema. The first heart sound was normal and pulmonary component of second heart sound was soft. A grade III/VI ejection systolic murmur in the left upper parasternal area was present. Rest of the systemic examination was unremarkable. We considered MIS-C and dengue fever as the possibilities. Investigations revealed hemoglobin of 13.3 g/dL, total leucocyte count of 7.2×10⁹/L, with relative neutrophilia (80%) and lymphocytopenia (14%), and thrombocytopenia (37×10⁹/L). His C-reactive protein (CRP) was elevated (62.9 mg/L), D-dimer 4674 ng/mL was increased, NT-pro

BNP 14352 pg/mL was highly elevated with hypoalbuminemia (2.9 g/dL). Renal function tests were normal. Anti-SARS-CoV-2 IgG antibody was positive, and IgM antibody was negative. Dengue serology was negative and chest X-ray was normal. ECG showed CHB with a ventricular rate of 78/min. Echocardiogram confirmed the anatomy of CCTGA with mild tricuspid regurgitation (TR), trivial mitral regurgitation (MR), mild right ventricular (RV) dysfunction, normal coronaries and no evidence of infective endocarditis.

We made a diagnosis of MIS-C [2]. Increased NT Pro-BNP and ventricular dysfunction on echocardiography reflected the myocardial involvement in MIS-C [2,3].

He was treated with intravenous immunoglobulin (IVIG) infusion (2 g/kg over 48 hours) along with injection methylprednisolone pulse therapy (10 mg/kg/dose), milrinone infusion and low molecular weight heparin. On day 3 of admission, he started complaining of severe abdominal pain, increased irritability, tachypnea but no tachycardia, and started desaturating (SpO₂ 70%), despite being on high flow oxygen. Grade IV/VI pansystolic murmur was now audible over the left lower parasternal area and apex. Repeat chest X-ray showed cardiomegaly and pulmonary venous congestion, but no parenchymal involvement. Despite being electively put-on mechanical ventilation, the child remained hypoxic. Repeat echocardiogram showed severe MR due to ruptured chordae tendinae of the anterior leaflet, with the mobilized chordae giving an impression of a thrombus/vegetation attached to the edges of the flail leaflet. This caused gross coaptation failure and the regurgitant jet was eccentrically directed towards the ASD, through which there was now right to left shunting, causing cyanosis. The right atrium was dilated, right ventricle was dysfunctional and there was no thrombus/vegetation anywhere else. His heart rate did not show much variation, but inotropes doses were escalated in view of persistent hypotension. Extracorporeal membrane oxygenator (ECMO) and/or surgery was contemplated in view of persistent worsening but he went into sudden cardiac arrest and died.

Myocardial involvement is a known feature of MIS-C, and common cardiovascular complications reported are shock, cardiac arrhythmias, pericardial effusion, coronary artery dilatation, and reduced left ventricular ejection fraction [1-4]. Although the exact

mechanism of MIS-C is still unclear, it is treated with IVIG and/or steroids, anticoagulants, and inotropes depending upon the cardiac status. Irrespective of the cardiac involvement, response to treatment and prognosis is generally good in MIS-C. However, in children with comorbidities, course of MIS-C can be complicated [1-4]. Chordae tendinae rupture and severe regurgitation can be either iatrogenic (during intracardiac device implantation or myocardial biopsy), or in conditions like acute rheumatic fever and infective endocarditis [5]. There have been no reports of chordal rupture in MIS-C [1-4]. Whether presence of CHD predisposes to chordal rupture in myocardial involvement due to MIS-C, is open to speculation. However, it can be concluded that the myocardial involvement may be sufficient enough to cause chordal rupture, at least in some individuals.

To conclude, a high index of suspicion for diagnosis of MIS-C, and early proactive intensive and cardiac care management, with preparedness for rare complications, is recommended for children with congenital heart defect and MIS-C.

RAVINDRA S PAWAR,* NITISH S AKHELIKAR, ANIL B KURANE
Department of Pediatrics,
Dr D Y Patil Medical College Hospital

and Research Institute, Kolhapur, Maharashtra.
**dravipawar@gmail.com*

REFERENCES

1. Whittaker E, Bamford A, Kenny J, et al. Clinical characteristics of 58 children with a pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2. *JAMA*. 2020;324:259-69.
2. Dufort EM, Koumans EH, Chow EJ, et al. Multisystem inflammatory syndrome in children in New York State. *N Engl J Med*. 2020;383:347-58.
3. Valverde I, Singh Y, Sanchez-de-Toledo J, et al. Acute cardiovascular manifestations in 286 children with multisystem inflammatory syndrome associated with COVID-19 infection in Europe. *Circulation*. 2021;143:21-32.
4. Belhadjer Z, Meot M, Bajolle F, et al. Acute heart failure in multisystem inflammatory syndrome in children in the context of global SARS-CoV-2 pandemic. *Circulation*. 2020;142:429-36.
5. Otto CM, Nishimura RA, Bonow RO, et al. 2020 ACC/AHA Guideline for the Management of Patients with Valvular Heart Disease: Executive summary: A report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021;143:e35-e71.