The most important step in situation of massive blood loss through the mouth is to first establish that the child is indeed experiencing hemoptysis. The boy was initially treated as hematemesis, and differentiating hemoptysis from hematemesis is critical, as treatment strategies differ markedly [3]. Definition of massive hemoptysis has not been completely agreed upon and vary between 200 and 600 mL as a cut-off, ideally, a volume >200 mL warrants immediate investigation [4]. Although massive hemoptysis is relatively uncommon, mortality rates as high as 75% been reported. The common cause of death is asphyxia secondary to aspiration rather than blood volume loss. Selective bronchial artery embolization is an effective nonsurgical technique for immediate control of massive hemoptysis with a success rate around of 77 - 94% [5]. Bronchial dieulafoy disease is rare disease but known to cause massive hemoptysis, where bronchial artery embolization should be considered as the first-line treatment.

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The Incompleteness of Incomplete Kawasaki Disease: A Customized Definition Is Needed for Indian Children

We read with interest the Indian Academy of Pediatrics position paper on Kawasaki disease in the journal [1]. It is indeed timely that this statement has come out amidst the coronavirus disease (COVID-19) pandemic and associated multisystem inflammatory syndrome in children. The authors have aimed to present this paper as a practice guideline specific to resource constrained setting like ours. In this context, we have the following comments:

In describing the definition for incomplete KD, the authors have presented the diagnostic approach, which is largely adapted from American Heart Association (AHA) scientific statement on Kawasaki disease [2]. While AHA algorithm considers evaluation for incomplete KD in children with fever ≥ 5 days and 2 or 3 compatible clinical criteria, the algorithm by Shenoy, et al. [1] triggers KD evaluation if fever ≥ 5 days is accompanied by less than four compatible clinical features. Although these two statements appear similar, this approach loses specificity by including children who present with fever and just one compatible clinical feature. Individually, the clinical features like rash, lymphadenopathy, conjunctival injection, oral or extremity changes are nonspecific and may occur with various childhood infections in India. This approach risks huge number of children with underlying infections being referred for echocardiographic evaluation.

Treatment with intravenous immunoglobulin is recommended if 3 of the 5 laboratory features (anemia for age, platelet $\geq 450 \times 10^9/L$, albumin < 3 g/dL, elevated alanine

aminotransferase, leucocyte count $\geq 15 \times 10^9/L$, urine > 10 WBC/hpf) are met in a child lacking echocardiographic abnormalities. Compared to Western cohorts, these criteria should be carefully defined in a low- and middle-income setting like India, with a high prevalence of iron deficiency anemia [3] and associated thrombocytosis (present in up to a quarter of those with iron deficiency) [4]. Iron deficiency when associated with infection accounted for more than half of all cases of reactive thrombocytosis in Indian children [5]. Given these findings, the current definition is likely to overestimate the burden of incomplete KD in Indian children, risking increased cost and potentially delaying the diagnosis of underlying infections. For example, as per the algorithm, a child with undifferentiated fever ≥ 5 days due to measles or a rickettsial infection that has a rash, iron deficiency anemia (and associated thrombocytosis) and hypoalbuminemia (negative acute phase reactant) would be treated for Kawasaki disease even if the echocardiogram is normal. In the absence of a 'gold standard' for diagnosis, we believe that grading recommendations based on available quality of evidence may be more useful for the readers to make informed decisions [6].

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AUTHORS' REPLY

We thank the readers for their query on the proposed algorithm for Incomplete Kawasaki disease in children [1]. This algorithm is indeed adapted from the American Heart Association [2], and we agree about the possible differential diagnoses that can be entertained in these children.

We wish to underscore that incomplete Kawasaki disease (KD) is considered in a child who is having ongoing high grade fever with less than four clinical criteria to satisfy a complete diagnosis. The algorithm raises the need for consideration of this diagnosis in a febrile child, particularly a young infant, with an unexplained persistent high grade fever for >5 days with systemic evidence for inflammation and with no other reasonable explanation for the same. The consequences of a delayed diagnosis of KD, especially the atypical presentation, can be devastating, as the risk of coronary aneurysms is higher

in these patients. It bodes well to have a high index of suspicion for the same, and encourages the consideration for an echocardiogram in these children which can further aid the diagnosis and the decision to treat. A rickettsial illness can be a differential diagnosis; however, it would normally respond to appropriate therapy and is usually recognized in the endemic areas by the pediatricians. One must appreciate the fact that KD is a diagnosis of exclusion, and this is highlighted in Box 1 as 'exclusion of other diseases with similar findings' [1].

Children with incomplete KD have increased risk for coronary artery aneurysms and the current recommendations are meant to ensure a timely diagnosis in these children. Hence, referring children, especially young infants with fever and elevated inflammatory parameters for echocardiography seems prudent, all the more, when they do not respond to the first-line antibiotics. Anemia and thrombocytosis can be noted in iron deficiency and can act as confounders in the diagnosis of incomplete KD. At the moment, we do not have nationwide data to provide new criteria for diagnosis. Studies that collate nationwide data on KD are the need of the hour – these recommendations infact may pave the path for such studies, which shall in turn help us formulate revised definitions for incomplete KD in our setting.

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Writer's Cramp and Psychosis: An Atypical Presentation of Systemic Lupus Erythematosus

A 9-year-old right-handed girl presented with handwriting deterioration and behavioral issues for five months. Initially, she had painful grip tightening and writing difficulty after writing about ten lines, which resolved on resting. After a month, symptoms emerged immediately on writing few words. Symptom fluctuation, weakness, sensory phenomena, ptosis, unresponsiveness, feeding /bladder/bowel/gait abnormalities were absent. Her behavioural problems included unprovoked crying/laughter, auditory hallucinations and tantrums. There was no history of fever, rash, jaundice, head-injury, toxin-exposure, abuse, family history of neurological illnesses. On examination, child's anthropometry, vital parameters, and systemic examination were normal. After writing 3-4 words, her grip tightened, wrist and interphalangeal joints got flexed,

strokes coarsened and slowed, with inability to trace curves. Her left (non-writing) hand had mirror-posturing. Posturing resolved two minutes after discontinuing writing. Percussion myotonia, Trousseau sign and Chvostek's signs were absent. Child was diagnosed with Writer's cramp (WC) and psychosis.

Her hemogram, anti-Streptolysin O titer, electroencephalogram, MRI brain and electromyography were normal. Workup for Wilson disease and autoimmune encephalitis was normal. ESR: 60 mm; Serum antinuclear antibody: strongly positive (speckled); anti double-stranded DNA (dsDNA) titer: 12 IU/mL (reference <5IU/mL); C3 and C4 levels: 30 mg/dL (reference: 80-200 mg/dL) and 12 mg/dL (reference: 15-40 mg/dL), respectively; anti-phospholipid antibodies: absent. She was diagnosed as SLE (European League Against Rheumatism/American College of Rheumatology criteria 2019 score:15) and administered intravenous methyl-prednisolone (30 mg/kg/day × 5 days) followed by maintenance oral prednisolone, hydroxychloroquine, trihexyphenidyl and physiotherapy. Dystonia and behavioural problems reduced significantly in three months.