WC is a rare task-specific limb dystonia reported in fourth fifth decade of life triggered by writing/typing/playing musical instruments and characterized by wrist flexion/extension, dystonic hand posturing, contralateral limb mirror dystonia and sensory trick phenomena [2]. The entity is extremely uncommon in childhood. The prominent differentials include essential tremor, writer’s cramp, neurodegenerative disorders (like Wilson disease), stroke, focal nerve entrapment (like cervical rib syndrome), dystonic tremor, writer’s cramp, and dystonia myoclonia syndrome [3]. Its patho-physiology is attributed to parietal-premotor pathway dysfunction secondary to etiologies such as genetic factors (e.g., DYT-1 mutation) and hand trauma [2,4]. It is not reported with SLE. Antibody-mediated phenomena against basal ganglia and frontal lobe probably led to the index child’s symptomatology. Treatments include pharmacological therapy, botulinum-toxin, physiotherapy, neurosurgery and assistive devices [3].

In view of features suggestive of autoimmune inflammatory disorder, transfusion dependent anemia, and auto-splenectomy, clinical exome sequencing was done, which revealed homozygous nonsense variant in exon 3 of HMOX1 gene (OMIM*141250) which causes human heme hemoxygenase-1 deficiency, confirmed by Sanger sequencing. Both parents were asymptomatic heterozygous carriers of the pathogenic variation detected in our patient.

When comparing our case with previous cases published in literature, we found our child had delayed development, growth retardation and dysmorphic features as reported earlier. He presented with fever but did not have lymphadenopathy or rash as seen in earlier cases. Also, he did not have asplenia from the beginning but had autosplenectomy during the course of treatment. Laboratory features similar in our case to the previous cases were, features of hemolysis (raised LDH and SGOT), raised inflammatory markers (raised CRP, ferritin). Our case is different from previously reported cases as he did not have coagulation abnormalities, features of nephritis or abnormal lipid profile [1-3]. Subsequently our patient developed acute arterial stroke confirmed on MRI and later was transfusion dependent. He succumbed to his illness at the age of 3½ years at a peripheral hospital.

Human heme oxygenase-1 deficiency is a disease which is known to be associated with impaired stress hematopoiesis. This results in marked red blood cell fragmentation, intravascular hemolysis, coagulation abnormalities and endothelial damage. This leads to deposits in the kidney and liver. Clinical features include persistent hemolytic anemia, asplenia, nephritis, generalised erythematous rash, growth retardation and hepatomegaly. There is one case report of successful HLA matched stem cell transplantation in the literature.

Though a rare disorder if a patient presents with features of hemolysis, generalised 3 inflammation, bleeding diathesis, nephropathy and asplenia diagnosis of human heme oxygenase

Heme Oxygenase-1 Deficiency

The oxidation of heme to biliverdin is facilitated by a stress-induced enzyme, heme oxygenase-1. This enzyme has antioxidant properties and plays an important role against inflammation. First case of heme oxygenase-1 deficiency was reported in 1999 [1]. The key features of heme oxygenase-1 deficiency, a recently described disorder, are hemolysis, generalized inflammation, bleeding diathesis, nephropathy and asplenia [1,2].

An 8-month old child presented with fever for 1-month. He was first of nonconcordant twins and the sibling was healthy. On examination, he had dysmorphic facies, frontal bossing, depressed nasal bridge and large ears. General examination revealed pallor and on systemic examination he had hepatomegaly. Investigation revealed increased inflammatory markers (raised CRP 84 mg/L and ferritin 3503 ng/mL), features of hemolysis (raised LDH 5253U/L and SGOT 210 units/L and SGPT 114 units/L), anemia and thrombocytopenia. He was worked up for bicytopenia (EBV, Parvo virus and CMV work up was negative). Other infections like tuberculosis and HIV were also ruled out. Bone marrow examination revealed few hemophagocytes. His initial USG abdomen revealed a normal spleen but on repeat CT abdomen after 6 months, spleen was not seen and only a small focal nodular calcified area within splenic fossa was seen. In view of the unclear primary diagnosis, hemophagocytes on marrow, increased liver enzymes and bicytopenia, he was started on oral steroids, pending further investigations. He improved, fever disappeared and liver enzymes returned to normal but he needed transfusion once in 2-3 months. However, on tapering steroids, fever reappeared and he became pale again. He was readmitted for investigations. USG showed absent spleen. Hemoglobin electrophoresis showed sickle cell trait.

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Providing Medical Services Online to Children With Chronic Kidney Disease During the COVID-19 Pandemic

The coronavirus disease 19 (COVID-19) pandemic has thrown up unprecedented challenges for child care the world over [1-3]. In Kerala, government policy entails that children below 10 years are not allowed in public spaces. As government hospitals are handling the majority of COVID-19 cases, child-care areas and child-care professionals in these hospitals have been diverted to adult healthcare.

Thus, children with chronic kidney disease, who require regular follow-up, have been badly impacted. Being immunosuppressed, both due to the inherent nature of their disease, as well as their medications, they cannot attend regular outpatient services at the hospital. The pandemic has forced health professionals and patient-caregivers to find new ways to cope. Guidelines for the same have been published recently [4].

At our center, the follow-up clinic for pediatric renal disease was modified to adapt to the situation. Whatsapp was used to keep in touch with patients and caregivers. The social worker acted as liaison between caregivers and clinicians. Follow-up appointments were given as was usual in non-COVID times, acute problems were assessed via text- and voice-messages, and images, when necessary. Prescriptions were photographed and sent on Whatsapp, as were reports of laboratory investigations and recordings of weight, height and blood pressure. If face-to-face consultation was deemed necessary, it was fixed in the ward or the casualty, and duty worker acted as liaison between caregivers and clinicians. Follow-up appointments were given as usual in non-COVID times. We handled 633 visits during March-November, 2020 in this manner, as compared to 391 physical visits in whole of 2019.

Expensive or less easily procured drugs were made available using government schemes. This was not easy for children staying in other districts who needed to travel long distances to reach the hospital. In such cases, liaison was established with the Reproductive and Child Health (RCH) officer of those districts, or the doctors in peripheral rural hospitals, who went out of their way to make the drugs available locally.

Patient information material, as documents, pictures and videos were circulated, such as procedure for testing urine, balanced diet and exercise routines. A Google form was used to check compliance with drugs, immunization and lifestyle modifications.

Psychiatric Problems Among Adolescents With HIV

We read the recently published article by Pilania, et al. [1] with interest. Authors deserve appreciation for conducting a study on this novel and sensitive topic. However, we would like to draw attention towards following observations. Firstly, authors try to associate the CD4 count with depression, which is not part of the design and methodology of the study. Moreover, there are studies which paradoxically state better psychological status leads to high CD4 count [2,3]. Additionally, low prevalence of psychiatric illness in this study (12%) in comparison with most of the previous studies (up to 50%) [2,4,5], as described to be due to...