failure, as well as considering Pediatric Multisystem Inflammatory Syndrome (MIS-C) as one of the diagnostic possibilities, as it is an entity that, despite a reduction in COVID-19 cases, We do not have complete immunization in our pediatric patients, so they are still at risk. As more is known about its pathophysiology and the spectrum of manifestations that could be found is evidenced, we will achieve early detection, improve treatment options and reduce morbidity and mortality in these cases.

Patient Consent
Yes, I received consent

Disclosure of Interest
None declared

References

1. Radia T, Williams N, Agrawal P, Harman K, Weale J, Cook J, et al. Multi-system inflammatory syndrome in children & adolescents (MIS-C): A systematic review of clinical features and presentation. Paediatr Respir Rev [Internet]. 2021;38:51–7. Available from: https://doi.org/10.1016/j.prrv.2020.08.001

P240

MIS-C and methylmalonic acidaemia. The role of anakinra to abate the cytokine storm

G. Corsello[1,2], C. Castana[2], M. Caserta[2], A. Di Fiore[2], V. Siciliano[3], M. C. Maggio[1,2]
[1]University Department PROMISE "G. D'Alessandro", University of Palermo; [2]ARNAS, Palermo, Paediatric Clinic, Children Hospital "G. Di Cristina"; [3]ARNAS, Palermo, Paediatric Anesthesia and Resuscitation, Children Hospital "G. Di Cristina", Palermo, Italy
Correspondence: M. C. Maggio
Pediatric Rheumatology 2023, **21(Suppl 2):**P240

Introduction: Multisystem Inflammatory Syndrome in Children (MIS-C) is a condition characterized by fever, inflammation, multiorgan impairment that manifests late in the course of SARS-CoV-2 infection.
Methylmalonic acidemia (MMA) secondary to mutase deficiency, *mut0,* is an inborn error of metabolism causing complete enzyme deficiency. The disease is characterized by fever, recurrent ketoacidotic crises or transient vomiting, dehydration, fatigue, vomiting, dehydration, hypotonia, frequent infections, developmental delay, intellectual disability, hepatomegaly, chronic kidney disease, pancreatitis cardiomyopathy, metabolic stroke, coma and death. Despite dietary treatment, patients undergo to life-threatening metabolic imbalance. Other long-term complications include progressive renal failure, metabolic stroke, and other neurological symptoms.
Objectives: We describe the clinical case of a 2-year-old child with MMA secondary to mutase deficiency, *with the documented homozygous mutation* c.2179 C>T of MMUT gene, associated to mut0 phenotype. Although it was treated from the first days of life with the diet, carnitine, hydroxocobalamin, he showed a poor metabolic control, with the program to be subjected to liver transplantation.
Methods: One month after SARS-CoV-2 infection, he presented fever, rash, significant increase of CRP, ferritin, triglycerides, IL-6, PRO-BNP, compatible with the diagnosis of MIS-C. He was treated with intravenous immunoglobulins (2gr/Kg), methylprednisolone (2 mg/Kg/day), with rapid clinical improvement. Ten days later, he showed the worsening of clinical conditions, with the recurrence of fever, vasculitic rash with palmoplantar extension, further increase of ferritin (1033 ug/l), IL-6 (146 pg/ml), PRO-BNP (5117 pg/ml), triglycerides, anemia, thrombocytopenia, metabolic acidosis with hyperlactatemia (180 mg/dl), increased urinary methylmalonic acid (200 mmol/mCreat), multiorgan failure. He was treated with sodium bicarbonate, thiamine, coenzyme Q, vitamin C, methylprednisolone and anakinra (2 mg/Kg/day).
Results: Three days after the start of anakinra, he showed a significant improvement of clinical and biochemical parameters, with the resolution of fever, vasculitis, rash, and the reduction of CRP, triglycerides, PRO-BNP (50 pg/ml), IL-6 (8 pg/ml), ferritin (579 ug/l). Unfortunately, 20 days later, a sepsis from Staphylococcus Aureus and Candida Albicans required the interruption of anakinra, with the worsening of the clinical and haematological parameters and the exitus.
Conclusion: We describe this case, to highlight the role of anakinra in a child with a severe form of MMA and MIS-C, with the significant clinical and biochemical improvement and the resolution of MOF, secondary to with MIS-C and metabolic imbalance. At our knowledge, this is the first case of MIS-C in MMA, treated with anakinra, described in the literature.
Further studies are needed to define the appropriateness and safety of therapy with anakinra in sepsis from Candida Albicans, a lethal complication to which the child went through prolonged venous catheterization and the high risk of infections, typical of MMA.

Patient Consent
Yes, I received consent

Disclosure of Interest
None declared

References

1. Maggio MC, Giordano S, Failla MC, Campione MG, Alaimo A, Corsello G. Ten-month follow-up of patients with covid-19 temporally related multi-system inflammatory syndrome in children: the experience of the children hospital of Palermo. Ital J Pediatr. 2023;49(1):37.

P241

A case report of post-COVID polymyositis in a child

J. Marčiulynaitė[1,2], A. Šnipaitienė[1,2], R. Šileikienė[1,2]
[1]Department of Pediatrics, Medical Academy, Lithuanian University of Health Sciences; [2]Pediatric Clinic, Hospital of Lithuanian University of Health Sciences Kaunas Clinics, Kaunas, Lithuania
Correspondence: J. Marčiulynaitė
Pediatric Rheumatology 2023, **21(Suppl 2):**P241

Introduction: Over the past few years, several post-COVID-19 autoinflammatory conditions have been reported. One of them, polymyositis has been described in adults [1,2], but there are no cases described in children so far. We present a severe pediatric polymyositis with possible relation to previous SARS-CoV-2 infection.
Results: Following unspecified respiratory viral infection, 7-year-old boy complained of increasing leg muscle pain for about a month and was worsening: limping, inable to climb stairs, NSAIDs were ineffective. Signs of tonsillitis, leg and face edema, painful leg muscles on palpation and pain during passive movements were seen on admission. A patient had no fever and no rashes. Laboratory studies showed thrombocytopenia ($60x10^9$/l), normal CRP, high ESR (34mm/h) and liver enzymes (ALT 2059IU/l, AST 674IU/l, GGT 220IU/l), low albumin (30.1g/l) and increased levels of creatin-kinase (CK) (1148IU/l). The patient progressed to acute renal failure, anemia (Hgb 97g/l) and polymyositis in a few days, with increasing levels of CK to 28045IU/l and myoglobin (1297.6→8987.9µg/l). Infectious agents such as EBV, CMV, atypical bacteria were disproved. From anamnesis, the boy was not vaccinated against SARS-CoV-2 and had no PCR proven COVID-19 infection during all pandemic years. However, IgG antibodies against SARS-CoV-2 were positive (140.3BAU/ml, positive >31.5BAU/ml). Immunodeficiency, hemophagocytic lymphohistiocytosis and neurological disorders were excluded. Immunological tests (muscle-specific tyrosine kinase, ANA, anti-DNR, ANCA and myositis related autoantibodies) were negative as well as genetic acylcarnitine profile analysis. Taking into account all the exams performed, previous SARS-CoV-2 infection was the most likely cause of polymyositis. Methylprednisolone 20mg/kg pulses were given for 5 days, then switched to prednisolone 2mg/kg/d. Despite of the initiation of glucocorticoids, the condition progressed to dysphagia, leading to intubation and mechanical ventilation. Head MRI and chest CT scan showed edema of neck and chest muscles. Intravenous cyclophosphamide 750mg/