is effective in controlling inflammation, fever and cardiac dysfunction. Side effects are transient and usually mild. Overall the reported incidence of CAA in MIS-C cohorts is 10%, interestingly in our cohort no patient has developed CAA after beginning ANK, possibly suggesting a protective role of IL1 inhibition in aneurysm formation. Further studies in bigger cohorts are needed to define the most effective timing and dose of ANK in MIS-C.

Disclosure of Interest
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P372

COVID-19 temporally related multisystem inflammatory syndrome (MIS-C): an early window of opportunity is a good treatment strategy? The experience of the paediatric covid center in Palermo

S. Giordano[1], M. C. Failla[2], M. G. Campione[3], L. Siracusa[2], L. Messina[2], L. Alessi[2], M. C. Maggio[4]
[1]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS, Palermo, Via dei Benedettini 1, 90100 Palermo; [2]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS, Palermo, Via dei Benedettini 1, 90100 Palermo; [3]University Department PROMISE "G. D'Alessandro", University of Palermo, Via del Vespro 129, 90100 Palermo; [4]University Department PROMISE "G. D'Alessandro" - Children Hospital "G. Di Cristina", University of Palermo, via dei benedettini n. 1, Palermo, Italy
Correspondence: M. C. Maggio
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Introduction: Multi-system inflammatory syndrome in children (MIS-C) shows a presentation mimicking Kawasaki Disease (KD), Toxic Shock Syndrome (TSS), Macrophage Activation Syndrome (MAS). Furthermore, many children show respiratory or abdominal symptoms.
Objectives: Intravenous immunoglobulin (IVIG) is recommended as first line treatment as in KD, followed by aspirin, steroids and, in IVIG-resistant patients, IL-1 or IL-6 blocking agents.
Methods: We describe a cohort of 16 Sicilian children (6M;10F; age:1.4-14 years), with MIS-C, with clinical features compatible with classical or incomplete KD, in some cases with MAS and/or TSS. Demographic, clinical, laboratory, echocardiographic and imaging findings, treatment strategy and outcome were collected.
Results: Common presenting symptoms included: fever (94%), abdominal pain or vomiting (50%), mucocutaneous rash (50%), conjunctivitis (44%), latero-cervical lymphadenitis (63%), cheilitis/pharyngeal hyperaemia (81%), hands and feet oedema (13%). Symptoms started 1-8 days before the hospitalization. Nasopharyngeal swab for SARS-CoV-19 was positive in 12/16 patients, with positive serological IgG, negative or grey zone IgM-type antibodies. 2 patients with negative swab had a history of recent infection and positive IgG-type antibodies; 2 patients had parents with positive swab.
All the patients showed significant increase of C-reactive protein (CRP). AST, ALT, gamma-GT were increased in 25%. Pancreatic amylase and lipase were increased in 13%, 19% showed lymphocytopenia.
Pro-BNP was increased (129-3980pg/ml) in 44% and troponin was increased (27.3-246ng/ml) in 31%. In addition, hyponatraemia was found in 100% of cases. Furthermore, 31 % had proteinuria. 50% showed cardiac involvement (3 pericardial effusion; 5 mitral insufficiency; 2 mitral and aortic insufficiency; 1 coronaritis). Pleural, ascitic, pericardial effusion and abdominal adenitis were found in 19%, 25%, 19% and 31% of cases, respectively.
IL-6 levels were evaluated in 9/16 patients and 8/9 showed a significant increase (30.2-285pg/ml) with a rapid normalization after steroids and IVIG treatment. Pro-BNP persisted increased for 7-10 days after IVG and steroids treatment. 25% of patients dramatically and rapidly evolved in a MAS-like form, fulfilling the classification criteria for the diagnosis of MAS (ACR/EULAR 2016). High doses of steroids and IVIG were promptly started with a significant improvement of the clinical course. In all the patients, treatment was started within 72 hours of admission, with IVIG (2 g/Kg/dose), methylprednisolone (2mg/Kg/day in 56% of patients; 30 mg/Kg/day for 3 days, followed by 2 mg/Kg/day in 38% of patients). 2 patients were treated with enoxaparin. TSS was described in 2 patients, who received additionally vasoactive drugs, albumin and diuretics.
Conclusion: In our series, most of patients received a prompt treatment with IVIG and steroids. This approach could explain the good outcome in all the cases and the rapid restoring of cardiac function also in patients with MAS or TSS. Patients showed a wide spectrum of presenting signs and symptoms; evidence of inflammation with pathological values of CRP, ESR, D-dimer, ferritin, pro-BNP, troponin, transaminase, pancreatic amylase and albumin; a multi-organ involvement was documented in a high percentage of cases, inducing the clinician to perform a multi-specialistic approach.
Patient Consent Received
Yes

Disclosure of Interest
None declared

P373

COVID-19 temporally related multisystem inflammatory syndrome (MIS-C) and cardiovascular involvement assessed with cardiac magnetic resonance (CMR). Experience of the children hospital of Palermo

F. Finazzo[1], A. Alaimo[2], S. Giordano[3], M. C. Failla[3], V. Vanella[3], L. A. Canduscio[3], A. Lembo[4], M. C. Maggio[4]
[1]UOC of Paediatric Radiology, Children Hospital "G. Di Cristina"; [2]U.O.C. of Paediatric Cardiologic Division, Children Hospital "G. Di Cristina"; [3]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS; [4]University Department PROMISE "G. D'Alessandro" – Children Hospital "G. Di Cristina", ARNAS Palermo, University of Palermo, Palermo, Italy
Correspondence: M. C. Maggio
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Introduction: MIS-C is a hyperinflammatory syndrome that follows exposure to SARS-CoV-2 by 2-6 weeks. However, some aspects remain unclear, such as cardiac involvement.
Objectives: to evaluate the role and effectiveness of cardiac magnetic resonance (CMR) in heart involvement in children affected by MIS-C; to review the expert groups' clinical experience in the field.
Methods: we describe a case series of 7 children (age: 2-11 years), admitted to the tertiary care Children Hospital "G. Di Cristina", Palermo, between December 2020 and May 2021 with clinical symptoms meeting the criteria for the diagnosis of MISC-C. All the patients showed findings of cardiac involvement without coronary artery lesions. Transthoracic echocardiography demonstrated temporary systolic dysfunction that lasted for 2-5 days. CMR was performed during the recovery phase or after the discharge (the median time to CMR was 10-30 days after the onset of illness). CMR was performed with a 1,5 Tesla scanner (GE Signa Explorer). 5/7 didn't undergo CMR study during the acute phase because they were clinically unstable and needed general anesthesia or sedation.
The protocol included, before intravenous contrast media injection, retrospective ECG-Gated fiesta cine sequences (short axis, 4, 3 and 2 chamber views), sequences for edema, and hyperemia T2 -short tau inversion recovery (Stir) (repetition time =1689ms, echo time55.10 ms).
Myocardial edema was evaluated by following the Lake Louise criteria. Because normal value in native T1 mapping and T2 relaxation time in children have poor reference, myocardial edema was characterized by increased signal intensity on T2-weighted imaging and myocardial damage by non-ischemic patterns late gadolinium enhancement.

Study for evaluating myocyte necrosis and fibrosis: Late gadolinium-enhanced 2D inversion recovery sequences performed at 6 min following intravenous contrast medium administration (0,2 mmol/kg).
Results: In 5/7 patients, T2-Stir sequences didn't show myocardial edema and hyperemia. Mean indexed left ventricular end-diastolic volume (iLVEDV), indexed left ventricular end-systolic volume (iLVESV), and indexed left ventricular stroke volume (iLVSV) were within normal range corrected for BSA. In 2 patients CMR showed late gadolinium enhancement in non-ischemic pattern. 1 patient, studied in subacute phase, after steroids and IVIG treatment, showed ventricular apical septum and lateral wall myocardial oedema, without fibrosis and an imaging compatible with focal acute myocarditis. Ventricular systolic function was normal. 1 patient, studied 1 month after the acute phase, and showed myocardial fibrosis.
Conclusion: international literature reports that children with MIS-C develop a transitory myocardial impairment, resembling myocarditis, with full recovery in most of them. Until now, the pathophysiology of the event is still object of debate.
CMR is an excellent noninvasive diagnostic tool for the diagnosis and follow-up of myocarditis. Furthermore, CMR can predict prognosis and recognize children at high risk to develop arrhythmias and unfavorable events. CMR is a codified method highlighting specific features of myocardial damage: inflammation, edema, necrosis, contractile scar impairment, and pericardial effusion. 6/7 didn't demonstrate myocardial oedema, probably because the CMR was performed during the recovery.

Disclosure of Interest
None declared

P374

Livedo reticularis as a late sign of COVID-19 temporally related multisystem inflammatory syndrome (MIS-C): a case series

S. Giordano[1], M. C. Failla[1], V. Vanella[1], L. A. Canduscio[1], M. Scalisi[2], M. C. Maggio[3,4]
[1]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina"; [2]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS, ARNA S; [3]University Department PROMISE "G. D'Alessandro", Children Hospital "G. Di Cristina", ARNAS; [4]University Department PROMISE "G. D'Alessandro" – Children Hospital "G. Di Cristina", ARNAS Palermo, University of Palermo, Palermo, Italy
Correspondence: M. C. Maggio
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Introduction: In children, the dermatologic features appear to occur early with other COVID-19 manifestations. Dermatologists play a key role in the early diagnosis of COVID-19. Multi-system inflammatory syndrome in children (MIS-C) shows a presentation mimicking Kawasaki Disease (KD), with mucocutaneous signs. However, late onset dermatological signs are poorly described.
Objectives: to evaluate children with MIS-C during the follow-up and to describe late dermatological signs in these patients.
Methods: We followed 14 children (3M; 11 F) with MIS-C, with clinical, biochemical, imaging data. Autoantibodies, D-Dimer, CRP, ESR, C3, C4, ferritin, serum amyloid, IgA, IgM, IgG were detected 1-2 months after the resolution of the clinical manifestations of MIS-C.
Results: 8/14 children (58%) showed livedo reticularis at the legs, arms, trunk. The livedo was more evident at the legs in all the patients. The livedo started at the remission, after normalization of CRP, ESR, D-Dimer; the sign lasted also for 1-2 months after the discontinuation of steroids and the normalization of haematochemical parameters. 4/8 showed low-title positive autoimmune tests (ANA in 2; ENA anti-Sm in 2; anti-cardiolipin IgG in 1; ASCA in 2).
Conclusion: In our series, 8/14 patients showed a livedo reticularis, more marked in the legs, however in some cases with a wide distribution to arms and the trunk. Low-title autoantibodies were transiently positive in 50% of these cases, negative in later detections.
Livedo reticularis was a late sign, linked to MIS-C related vasculitis, persisting 1-2 months after the resolution of MIS-C. A different treatment regimen (IVIG plus steroids at 1-2 or 30 mg/Kg/day) did not influence the progress of this clinical manifestation. In 50% of children we documented a transient autoimmune response.
Patient Consent Received
Yes

Disclosure of Interest
None declared

P375

Acute cardiovascular manifestations in children with multisystem inflammatory syndrome associated with COVID-19 infection in a Sicilian case series

A. Alaimo[1], S. Giordano[2], M. C. Failla[3], F. Finazzo[4], L. Siracusa[3], L. Messina[3], L. Alessi[3], M. Scalisi[3], M. C. Maggio[5]
[1]U.O.C. of Paediatric Cardiologic Division, Children Hospital "G. Di Cristina", ARNAS; [2]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS; [3]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina"; [4]UOC of Paediatric Radiology, Children Hospital "G. Di Cristina", ARNAS; [5]University department PROMISE "G. D'Alessandro" - Children Hospital "G. Di Cristina", ARNAS, Palermo, University of Palermo, Palermo, Italy
Correspondence: M. C. Maggio
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Introduction: Multisystem inflammatory syndrome in children (MIS-C) is a severe complication of COVID-19 infection, typically evidenced 4-6 weeks after the infection. The debated pathogenesis is a dysregulation of inflammatory response to SARS-CoV-2 infection ad a cytokine hyperexpression. Persistent fever, respiratory and gastrointestinal symptoms are the most common manifestations, associated with typical clinical signs described in Kawasaki Disease (KD). Furthermore, pleiomorphic cardiac manifestations are described, including ventricular dysfunction, coronary artery dilation and aneurysms, arrhythmia, conduction abnormalities and pericardial effusion. These manifestations are a strong link with KD, even if in MIS-C they are more frequently documented. Severe cases can present as Toxyc Shock Syndrome (TSS) with vasodilatory or cardiogenic shock, requiring treatment with plasma expanders, inotropic drugs, diuretics, albumin and -in the more severe patients- extracorporeal membrane oxygenation and mechanical ventilation. KD experience guided the clinicians to treat these children with intravenous immunoglobulin (IVIG), steroids, aspirin (ASA) and, in refractory cases, anti-IL-1 monoclonal antibodies.
Objectives: Most patients recover within days to a couple of weeks and mortality is rare, although the medium- and long-term sequelae, particularly cardiovascular complications, are not yet known.
Methods: We describe the short-term outcome in a case series of 12 Sicilian children (4M; 8F; age: 1.4-14 years) with MIS-C and a documented recent or actual infection by SARS-CoV-2 who showed cardiac involvement.

Results: The cardiac features were: 3 patients showed pericardial effusion; 1 coronaritis; 6 transient mitral valve regurgitation; 1 Brugada pattern, evidenced when he was febrile; 2 showed associated mitral and aortic valve regurgitation). 7/8 patients with valve regurgitation showed a significant increase of pro-BNP, normalized during the follow-up.
TSS was described in 2 patients, showing a significant increase of troponin, promptly treated with high dose of methylprednisolone, IVIG, vasoactive drugs, albumin and diuretics.
3 patients (21%), after the resolution of the acute phase, showed bradycardia (heart rate < 50/min), persisting for 7-10 days. The bradycardia was not associated with first-degree AVB, or a pathological PR.
6 patients (42%) showed an altered ventricular repolarization phase, in association with an increase of pro-BNP (129-3980 pg/ml). 4/12 (33%) had increased troponin levels (27.3-246 ng/ml) in the acute phase, with the normalization of troponin after IVIG and steroids treatment. Pro-BNP persisted increased for a longer time, besides the clinical improvement and the normalization of blood chemistry parameters.
Conclusion: Generally, pro-BNP and troponin levels in MIS-C are higher than in KD, reflecting vasculopathy and cardiomyocytes damage extent. Persistence of increased levels of pro-BNP, in patients with a normalization of inflammatory parameters, suggests a mechanism of myocardial oedema, persisting besides the intensive care approach useful, however, to limit effects on cardiac function and normalize inflammatory parameters. Patients admitted with MIS-C require close electrocardiogram monitoring during the acute phase and the recovery, even if they do not manifest dyselectroliteemia, coronary lesions, pericardial effusion, myocarditis, shock. This approach can avoid severe arrythmia.
Patient Consent Received
Yes

Disclosure of Interest
None declared

P376

Step-by-step surveillance in children with multisystem inflammatory syndrome associated with COVID-19 infection: proposal of a cardiologic follow-up

F. Finazzo[1], A. Alaimo[2], S. Giordano[3], M. C. Failla[3], M. C. Maggio[4]
[1]UOC of Paediatric Radiology, Children Hospital "G. Di Cristina"; [2]U.O.C. of Paediatric Cardiologic Division, Children Hospital "G. Di Cristina"; [3]U.O.C. of Paediatric Infectious Diseases, Paediatric COVID Center, Children Hospital "G. Di Cristina", ARNAS; [4]University Department PROMISE "G. D'Alessandro" – Children Hospital "G. Di Cristina", ARNAS Palermo, University of Palermo, Palermo, Italy
Correspondence: M. C. Maggio
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Introduction: Multisystem Inflammatory Syndrome Associated with COVID-19 Infection (MIS-C) shows many matches with children with Kawasaki disease (KD) and most children with MIS-C have incomplete or complete KD-like phenotype. In these patients cardiologic involvement mimics KD, showing, however, a higher incidence of severe acute manifestations.
Objectives: In children with MIS-C and clinical findings related to heart disease, ECG and echocardiography are the first-line imaging. Until now, there are no international guidelines on the management of these patients. We suggest to take inspiration from the recommendations for KD for the significant phenotypic overlap between MIS-C and KD.
Methods: We propose a step-by-step cardiologic imaging follow-up:
- in all the patients, we recommend ECG and echocardiography at the diagnosis, at the worsening of the clinical and/or blood chemist parameters (CRP, ESR, ferritin; proBNP, troponin, D-Dimer), at any change of treatment supported by clinical worsening, at 8, 30, 45, 60, 90, 180 days since the diagnosis. The time-table may be changed in consideration of the outcome of the patient.
-In patients with coronary artery dilatation (CAL), documented by echocardiography, it is advisable to follow-up them, since the diagnosis, with ECG, echocardiography, D-dimer, pro-BNP, troponin.
-If CAL are oversized with z-score >- 2,5, according to age and body surface or increase during the follow-up:
-it is recommended to perform Coronary Computed Tomography (CT) (CCA) or Cardiac Magnetic Resonance Angiography (CMRA). In fact, echocardiography cannot visualize the whole coronary artery vessels.
Results: Both allow visualization of coronary artery aneurysms, vessels thickening, myocardial perfusion defects, permitting risk stratification and handing treatment decisions.
CMRA is the first choice, because it is a radiation-free imaging method. It can evaluate the entire coronary artery system and provides details on myocardial function ischemia (detecting areas of inducible myocardial ischemia with pharmacological stress), infarction, inflammation, fibrosis.
However, the new generation Multidetector Single –Source CT scanners and Dual Source Ct scanner allow a fast heart CT study with low radiation dose and reduce the need for sedation.
CMR is less suitable because it is a lengthy examination and very often requires general anesthesia.
If echocardiography demonstrates myocardial dysfunction or valve regurgitation at admission or during hospitalization, we suggest performing CMR. There is no consensus on the right timing.
Conclusion: We suggest performing CMR during the acute or subacute phase: it is a step of the relieve in the cardiologic diagnosis to assess ventricular function and myocardial active injuries (oedema, hyperemia, ischemia, necrosis) and to repeat the imaging at the discharge in patients with the first pathological CMR, to evaluate fibrosis by myocardial delayed enhancement. CMR at the discharge is suggested also in cases with the first CMR normal, who showed a worsening of the echocardiographic parameters, the relieve of new-insurance valvulitis, persistent arrhythmia. At 3-6 months since the patient showed the remission, the CMR must be repeated to avoid any fibrotic lesions.

Disclosure of Interest
None declared

P377

Safety and tolerability of the biontech COVID-19 vaccine in adolescent patients with JIA on TNFi

D. Maritsi, F. Dasoula, G. Vartzelis, D. Dimopoulou, C. Papadaki, M. Tsolia
Infectious Diseases, Immunology and Rheumatology Unit, Second Department of Pediatrics, Athens Medical School, Athens, Greece
Correspondence: D. Dimopoulou
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Introduction: The post-authorization safety reports of the novel mRNA vaccines against COVID-19 are generally reassuring; nonetheless their safely profile has not been evaluated in adolescents with MRDs on immune-modulating treatment.
Objectives: To evaluate the safety and tolerability of the BNT162b2(Pfizer-BioNTech) COVID-19 vaccine in adolescent and young adult patients with juvenile idiopathic arthritis (JIA) on TNFi treatment.
Methods: Study population: The study involved 21 subjects aged 16-21 years (median 17 years) with stable JIA who have been diagnosed and treated for at least 1 year with TNFi. In particular, 10 were receiving adalimumab at two weekly intervals, eleven were given etanercept once a week, whereas 15 patients were on concomitant weekly subcutaneous methotrexate. Eight patients had poly-articular JIA, 7 psoriatic JIA and 6 ERA. Written informed consent was obtained at enrolment. Study procedures: The patients received two doses of the COVID-19 vaccine (Pfizer-BioNTech) intramuscularly at 0 and 3 weeks. In addition to the visits for vaccine administration, further visits were planned at 2, 6 and 12 months after enrolment. A blood sample for the evaluation of vaccine immunogenicity is planned to be taken from all of the subjects at the time of enrolment, after 2, 6 and 12 months after the last vaccine dose. All participants were observed for 30 min after the injection in order to assess vaccine safety and