juvenile idiopathic arthritis (JIA), although its occurrence in patients with other autoimmune or autoinflammatory conditions, i.e., adult- and childhood onset systemic lupus erythematosus, Kawasaki disease, and periodic fever syndromes, is being reported with increased frequency. Virus-associated MAS is a well-recognized entity. Most cases are related to Epstein–Barr virus (EBV), cytomegalovirus (CMV), and herpesvirus, while streptococcus and parvovirus B19-induced MAS is very rare. Complete recovery can be provided with early and efficient treatment in macrophage activation syndrome.

Disclosure of Interest
None Declared

Vasculitides II

P221
PARVOVIRUS INFECTION AND KAWASAKI DISEASE: ONE DISEASE FOR TWO SIBLINGS

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Introduction: Kawasaki disease (KD) is rarely described in siblings in the same time. In these cases, an infectious trigger must be excluded.
Objectives: We describe the clinical course of two brothers who showed severe KD all at once, secondary to Parvovirus infection.
Methods: A 9-month-old female showed fever, pallor, vomiting, bilateral non-secreting conjunctivitis, rash. Anamnesis revealed that 12 days before, she had fever, spontaneously resolved. At admission, 9 days after fever onset, she showed fever, conjunctivitis, pharyngitis, rash, and cervical adenopathy. Haematological parameters showed: leukocytosis, neutrophilia; anaemia; CRP: 2.31; ESR: 120. ECG and echocardiography were normal, including coronary Z-scores. She showed positive Parvovirus IgM. Spontaneous defervescence occurred. Further cardiological evaluation was performed to exclude a pericarditis secondary to Parvovirus, and at day 26 after fever onset, coronary artery lesions (CAL) were documented: proximal right coronary artery Z-score of 6.02; left main coronary Z-score: 5.72; left anterior descending Z-score: 5.78. The child was promptly treated with IVIG plus ASA.
A further echocardiographic evaluation showed worsening of CAL, with a sacciform aneurysm in the left anterior descending artery (Z-score:5.08). Laboratory test did not show inflammation; however, the girl was treated with 3 bolus doses of intravenous methylprednisolone (30 mg/kg/dose). The Z-score of CAL did not change and the patient was treated with anakinra (4 mg/kg/day), with a progressive improvement of CAL, and after 2 months, Z-scores normalized.
The 7-year-old brother presented fever, vomiting at the same time of the sister, with spontaneous resolution after 4 days. Four days later, he presented again fever with abdominal pain, tachypnoea and tachycardia, secondary anuria. He had: leukocytosis, neutrophilia, anemia; CRP: 0.24; CPK: 773; creatinine: 0.77; BUN: 111; elevated myocardial necrotic enzymes (c-Troponin T: 91.4; Pro-BNP: > 70.000).
Echocardiogram showed generalized hypokinesia, a severe reduction of the ejection fraction (EF) (20-25%); increased left atrium (Z-score: 3.3) and mitral valve with moderate insufficiency. He received dopamine, dobutamine, furosemide plus steroids. He showed a constant improvement of echocardiographic parameters, plasmatic enzymes and clinical signs. In 16th day he was discharged with an EF of 45% and persistent septal hypokinesia. However, specific serology anti-Parvovirus was tested and showed increased IgM, with negative IgG. The cardiological outcome revealed a progressive improvement of EF, which reached the 50%.
Results: CAL significantly improved after anakinra, at the contrary, the clinical evolution in the brother was different.
Conclusion: We describe familial KD in two siblings which had the same infectious trigger (Parvovirus). The brother was diagnosed as a post-viral myocarditis. However, considering the two parallel and different evolution, the girl showed late CAL with aneurisms, and the brother a Kawasaki shock syndrome picture with myocardial dysfunction. Viral illnesses are recognised trigger of KD, and in these cases the rareness is the coincident KD in two siblings, with different and severe clinical course. Noteworthy, the girl had aneurisms which resolved with anakinra, a therapy which has been recently shown to be promising for this disease. Informed consent to publish had been obtained from the parents.

Disclosure of Interest
None Declared

P222
PANCREATITIS IN HENOCH-SCHONLEIN PURPURA. A SINGLE-CENTRE OBSERVATIONAL STUDY

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Introduction: Henoch-Schönlein purpura (HSP) is the most frequent vasculitis in children. Typically, it is characterized by palpable purpura, joints swelling, arthralgia, abdominal pain with possible intestinal bleeding. In more severe cases, the patients show acute abdomen.
Acute pancreatitis is a rare dramatically evolutive, life-treating manifestation of SHS and it can be associated with a fulminant course. Persistent abdominal pain, need to be investigated by the dosage of serum pancreatic amylase, lipase and by abdominal MRI. In these patients, corticosteroid treatment is recommended and must be associated with parenteral feeding.
Objectives: We analysed the full series of children with HSP admitted to our paediatric unit in the period 2011-2018.
Methods: We retrospectively collected data of 50 children (age: 4-14 years), with HSP who needed hospitalization.
4/45 patients (9%) developed an acute pancreatitis.
All the patients were males, age:3-10 years. All the patients did not show other risk factors of pancreatitis.
Results: The treatment was parenteral feeding in 100% of the patients. One patient with pancreatitis and nephrotic syndrome received 3 bolus doses of methylprednisolone (30 mg/kg/dose) followed by prednisolone (2 mg/kg/day) and mycophenolate; one patient with pancreatitis and acute renal failure, received 3 bolus doses of methylprednisolone (30 mg/kg/dose) followed by a single-dose of cyclophosphamide (750 mg/m2), followed by azathioprine (50 mg/day). One patient showed a mild pancreatitis and healed with prednisolone (2 mg/kg/day) and parenteral feeding. One patient showed acute pancreatitis, associated with acute intestinal bleeding, orchitis, was treated with 3 bolus doses of methylprednisolone (30 mg/kg/dose) followed by prednisolone (2 mg/kg/day). Steroids treatment was progressively tapered until the complete resolution of the pancreatic involvement.
Conclusion: Acute pancreatitis is a rare and life-treating manifestation of HSP. High-doses steroids are a recognised and useful treatment, associated with parenteral feeding. In non-responders to steroids, immune suppression treatment is the second-line treatment to induce remission. The choice depends on associated manifestations of HSP and on associated failure of other organs.

Disclosure of Interest
None Declared

P223

A CASE OF KAWASAKI DISEASE SUCCESSFULLY TREATED WITH ANAKINRA

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Introduction: Kawasaki disease (KD) is a systemic vasculitis that affects the medium-and small-size arteries.Some children do not respond to the first line of therapy.The treatment for IVIG non-responsive patients is controversial,but adding steroids to the second IVIG dose can be effective at reducing the incidence of CAA. For those non-responders to this second step of therapy,other approaches have been described.In recent years,some reports have suggested the role that Anakinra can play in the treatment of severe or resistant cases of KD.
Objectives: We present a case of an IVIG and steroids-resistant KD treated with Anakinra.
Methods: We present the case of a one-year-old with persistent fever for six days,a generalized rash,lymphadenopathy,conjunctivitis,cheilitis,swollen hands and feet,diarrhoea and irritability.Blood tests revealed increased C-reactive protein (100.80 mg/L),D-Dimer (532.00 ng/mL) and Fibrinogen (810.00 mg/dL).There was increased platelet count (532,000/mm3) and hyperneutrophilia (neutrophils 79.80 %);He showed low levels of serum albumin (2.9 g/dl),serum sodium (130 mEq/L) and serum chloride (90 mEq/L).Blood cultures and serological tests for infection were negative.He underwent an echocardiography that showed an enlarged coronary diameter,(right 3.9 mm, left 3.6 mm; circumflex 3.1 mm, proximal anterior inter-ventricular 3.1 mm; worse Z score +5).
Results: From the presence of fever that had lasted for more than five days,the lymphadenopathy,conjunctivitis,the cutaneous rash and the oedema of the hands and feet with cardiac involvement,we diagnosed complete Kawasaki disease and started IVIG (2 g/Kg) and Aspirin (80 mg/Kg).Despite the initial IVIG treatment, the fever,rash,conjunctivitis,cheilitis and mild oedema of hands and feet remained,and additional IVIG doses and three metilprednisolone (30 mg/kg) pulses were administered in the following days.The patient underwent another echocardiography that did not show any changes.Maintenance treatment with oral prednisone (0.5 mg/kg/day) was initiated.The patient became afebrile, and the rash, cheilitis and conjunctivitis gradually disappeared.Serological markers of inflammation returned to the normal range.After ten days, the patient's fever recurred with irritability, exanthema,conjunctivitis, and hand and feet desquamation.Blood tests showed a significant increase of CRP (65.30 mg/L),ESR (50 mm/h);anaemia (Hb 10.4 g/dL) and more platelets (566,000) were also registered.Echocardiography did not show changes.Further Metilprednisolone pulses were administered over three days.After one day the patient repeated the Echocardiography, which showed a worsening of the enlargement of the coronary diameter,(right 6 mm (Z score 13),left 6 mm (Z score 9.9),proximal anterior interventricular 5.7 mm (Z score 15).There was an aneurism in the diagonal branch of left anterior descending coronary (7.2 mm; Z Score >10) with a thrombus inside.After three days, the patient's fever continued.Due to the persistence of the fever,the worsening of the echocardiography features and blood tests,with the agreement of his parents,other intravenous treatments with IL-1RA were initiated (Anakinra 2 mg/kg subcutaneously once a day for two weeks).One day later, the fever disappeared and CRP, ESR, platelet and haemoglobin levels returned to normal in the subsequent blood tests.The Aspirin dose was reduced to the antiplatelet dose and Clopidogrel (5 mg) was added.
Conclusion: Treatment with Anakinra was maintained for two weeks, and Prednisone (0.5 mg/kg/dy), Aspirin and Clopidogrel.There were no flares of the disease after discontinuing the Anakinra and blood tests were within the normal range. Echocardiography tests showed an improvement in his coronary enlargement. Written informed consent was obtained from the patients for publication of Case Report.

Disclosure of Interest
None Declared

P224

SUCCESSFUL TREATMENT OF HAEMORRHAGIC BULLOUS HENOCH-SCHONLEIN PURPURA WITH INTRAVENOUS IMMUNOGLOBULINS

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Introduction: Henoch-Schonlein purpura(HSP) is the most common childhood systemic vasculitis.The disease affects the skin,the gastrointestinal tract,the joints and kidneys.In rare cases the skin can present as haemorrhagic,bullous or necrotic lesions.
Objectives: We describe a case of HSP presenting with severe skin lesions that did not respond to standard therapy with corticosteroids,and was therefore treated with intravenous immunoglobulins(IVIG).
Methods: An 11-year-old girl was admitted to our department with fever,abdominal pain,arthralgia,severe purpuric palpable rash and haemorrhagic bullae distributed on the buttocks and lowerextremities.Physical examination showed a purpuric palpable rash,multiple haemorrhagic bullae and vesicles ranging in size from five to 30mm on the buttocks and lower limbs.The abdomen was painful but soft.-Blood tests revealed a white blood cell (WBC) count of 20.730/ml with normal differential, platelets (PLT) 336.000/ml,haemoglobin (Hb) 13.3 g/dl,erythrocyte sedimentation rate (ESR) 30 mm/1h (nv 20 mm/1h), C-reactive protein (CRP) 13 mg/L.She tested negative for streptococcal tests showed Antistreptolisine (ASLO) 778 IU/mL (nv 0-200) and anti-DNAse 1170 IU/mL (nv 0-200).Microscopic analysis of the patient's urine showed mild proteinuria (30mg/dl) and an occult blood test on a stool sample was negative.Abdominal ultrasound was unremarkable.A skin biopsy revealed peri-nuclear infiltration of polymorphs,nuclear leukocyte and mononuclear cells with IgA deposition in immunofluorescence studies,typical of leukocytoclastic vasculitis.
Results: Clinical findings were consistent with HSP and she was started on oral prednisone (1mg/kg/daily),as well as antibiotics with teicoplanin for the super-infection of skin lesions (Staphylococcous Aureus).After ten days she was discharged,with an improvement in skin appearance.She was given a schedule to progressively taper and stop steroid therapy within a month.At the 15-day follow-up,she presented new onset of purpuric rash on her lower extremities and scars of previous bullae.We decided to increase the dosage of corticosteroid therapy from 0.5mg/Kg to 1mg/kg/die.At the one-month follow-up she presented a new purpuric rash and haemorrhagic bullae on her lower extremities,associated with abdominal pain.She was therefore readmitted to our department.Blood tests revealed WBC 10.730/ml with normal differential count,PLT 300.000/ml,Hb13.2 g/dl,ESR 40 mm/1h,CRP 15 mg/L;creatinine,were within range.The results of urinalysis,an occult blood test of a stool sample and an abdomen ultrasound were all negative.She started an IVIG infusion (2g/kg),while still on oral prednisone (1mg/kg/daily),progressively tapered.After treatment she showed good improvement in respect of her skin lesions and was discharged.At the 15-30-60 day follow-up visits the patient was in good general condition;she did not have any new lesions but only scars on her lower limbs.
Conclusion: There is no consensus about the management for cutaneous manifestations in HPS.In the case of rare skin complications,-such as haemorrhagic bullae lesions,immunosuppression with corticosteroid therapy is often necessary in order to control the inflammation and limit the expanse of necrosis.Unfortunately,our patient gained only partial benefit from corticosteroid therapy.As a consequence,we decided to treat her with an IVIG (2g/kg as a single infusion) associated with oral prednisone (1mg/kg/daily),progressively tapered,which induced rapid and persistent resolution of symptomatology.Written informed consent was obtained from the patients for publication of Case Report.

Disclosure of Interest
None Declared