were statistically significant differences in 26 conditions (Figure), with the most striking differences comparing the GP cohort with the JIA cohort in ankylosing spondylitis (0.001% vs 0.98%), psoriatic arthritis (0.01% vs 4.44%), systemic sclerosis/scleroderma (0.004% vs 0.6%), uveitis (0.07% vs 7.82%) and sarcoidosis (0.003% vs 0.3%). Only alopecia areata was more frequent in the GP cohort than in patients with JIA (4.5% vs 0.2%).

Conclusion: Patients with JIA have more pronounced autoimmunity prevalence than the general paediatric population, especially for ankylosing spondylitis, psoriatic arthritis, uveitis, systemic sclerosis/scleroderma and sarcoidosis.

REFERENCES

- [1] Eaton WW, et al. *J Autoimmun* 2007;29:1–9.
- [2] Jacobson DL, et al. *Clin Immunol Immunopathol*
- [3] American Autoimmune Related Disease Association. Autoimmune statistics. 2016. <https://www.aarda.org/news-information/statistics/>. Accessed 7 July 2016.

Disclosure of Interests: Daniel J Lovell Consultant for: Consulting fees and/or honoraria from Astra Zeneca, Wyeth Pharma, Amgen, Abbott, Pfizer, F. Hoffmann-La Roche, Novartis, UBC, Takeda, GSK, Boehringer, and Celgene, Bin Huang: None declared, Chen Chen: None declared, Sheila Angeles-Han: None declared, Teresa Simon Employee of: Bristol-Myers Squibb, Hermine Brunner Grant/research support from: Bristol-Myers Squibb, Pfizer, Consultant for: Pfizer, Bristol-Myers Squibb, Janssen, Novartis, Lilly, Roche, GlaxoSmithKline, Sanofi, Speakers bureau: Novartis, Roche

DOI: 10.1136/annrheumdis-2019-eular.1844

SAT0490

IL-1 BLOCKADE IN PEDIATRIC RECURRENT PERICARDITIS: A MULTICENTRIC RETROSPECTIVE STUDY ON THE ITALIAN COHORT

Roberta Caorsi¹, Antonella Insalaco², Chiara Longo³, Giorgia Martini⁴, Marco Cattalini⁵, Rita Consolini⁶, Giovanni Filocamo⁷, Alessandro Rimini⁸, Silvia Federici⁹, Camilla Celani¹⁰, Maria Cristina Maggio⁹, Micol Romano¹⁰, Barbara Lia Teruzzi¹¹, Andrea Taddio¹², Angela Minaichi¹³, Silvana Martino¹⁴, Francesco La Torre¹⁵, Alessandro De Fanti¹⁶, Lorenzo Dagna¹⁷, Barbara Bigucci¹⁸, Antonio Brucato¹⁹, Fabrizio De Benedetti², Marco Gattomo¹. ¹I.R.C.C.S. Giannina Gaslini, Center for autoinflammatory diseases and immunodeficiencies, department of Pediatrics and Rheumatology, Genova, Italy; ²Bambino Gesù, Department of Rheumatology, Fiumicino, Italy; ³University of Genoa, Department of Pediatrics, Genova, Italy; ⁴University of Padua, Department of Pediatric rheumatology, Padova, Italy; ⁵ASST Civil Hospital of Brescia, Department of Pediatric rheumatology, Brescia, Italy; ⁶Università di Pisa, Department of Pediatrics, Pisa, Italy; ⁷Ospedale Maggiore, Department of Pediatric rheumatology, Milano, Italy; ⁸I.R.C.C.S. Giannina Gaslini, Department of cardiology, Genova, Italy; ⁹Children's Hospital G. Di Cristina, Department of Pediatrics, Palermo, Italy; ¹⁰ASST Gaetano Pini-CTO Polo Isocrate, Department of Rheumatology, Milano, Italy; ¹¹ASST Great Metropolitan Niguarda, Department of Pediatric rheumatology, Milano, Italy; ¹²Hospital Burlo Garofolo, Department of Rheumatology and Immunology, Trieste, Italy; ¹³Policlinico S. Orsola – Malpighi, Department of Pediatrics, Bologna, Italy; ¹⁴Ospedale Regina Margherita, Department of Rheumatology and Immunology, Torino, Italy; ¹⁵Oncology Hospital Vito Fazzi, Department of Pediatrics, Lecce, Italy; ¹⁶Azienda USL IRCCS Reggio Emilia, Pediatrics, Reggio Emilia, Italy; ¹⁷San Raffaele Hospital, Department of Rheumatology, Segrate, Italy; ¹⁸Ospedale "Infermi" di Rimini, Department of Pediatrics, Rimini, Italy; ¹⁹OSPEDALE PAPA GIOVANNI XXIII. BERGAMO, Department of internal medicine, Bergamo, Italy

Background: Acute pericarditis is an inflammatory condition causing the occurrence of pericardial effusion. In a third of patients, the disease is recurrent. Most of the cases are idiopathic or occur after a pericardial procedure. First line treatment of idiopathic pericarditis consists in NSAIDs and colchicine; glucocorticoids represent the second line treatment in resistant or intolerant cases. The use of different biologics and immunosuppressant has been reported, with variable responses. A recent clinical trial has enlightened the effectiveness of anakinra in patients with colchicine-resistant recurrent pericarditis.

Objectives: To describe the clinical characteristics and response to treatment in a cohort of paediatric patients with recurrent pericarditis treated with IL inhibitors.

Methods: Pediatric patients with recurrent pericarditis followed at 19 Italian centers of paediatric rheumatology or cardiology and treated with IL1 inhibitors were included in the study. Demographic, clinical and response to treatment data were retrospectively collected.

Results: 55 patients were included in the study. The mean age at onset of the first episode of pericarditis was 12.53 years. The mean number of relapses of pericarditis before the beginning of treatment with IL1 inhibitors was of 3.4. 53 out of 55 patients had previously received treatment with NSAIDs and 44 colchicine. 44 patients received steroidal treatment: 2 of them displayed a steroidal-resistance and 39 steroidal-dependence with recurrence of the symptoms following any attempt to reduce or withdraw this treatment.

Anakinra (mean dosage of 1.67 mg/kg/day) was used as first IL-1 inhibitor in 54 of the 55 patients, Canakinumab (150 mg every 4 weeks) in one. 53 out of 54 patients treated with anakinra displayed a complete clinical response to treatment within a mean of 2 days: NSAIDs, glucocorticoids and colchicine were withdrawn in 25 out of 26, 31 out of 35 and 15 out of 32 patients respectively. 50 of 54 patients displayed a complete response; among these, three were switched to Canakinumab, 17 patients continued treatment at the same dosage while in 30 patients a reduction of treatment was attempted. 12 patients presented, during anakinra tapering, a disease flare, promptly resolved after an increasing of the dosage. The remaining 18 patients did not present any flare despite the reduction of the drug. Anakinra was withdrawn in 16 patients, with recurrence of the symptoms in 11 (9 restarted anakinra, 2 were treated with glucocorticoids and colchicine, with good response).

5 patients were treated with Canakinumab: 1 as first anti-IL1 drug, 4 were switched from anakinra (two for poor compliance, one for side effects and one for incomplete control of the disease). 2 out of five patients had a complete control of the diseases, 2 patients discontinued the treatment because of inefficacy and 1 patient required low dose of glucocorticoids to control the disease.

At last follow-up 34 patients were on anakinra, 7 on anakinra and colchicine, 2 on canakinumab, 1 on canakinumab plus colchicine and NSAIDs. In 9 patients all treatments were withdrawn for complete control of the disease.

Conclusion: This study confirm the effectiveness of IL-1 blockade in paediatric patients with recurrent pericarditis. However, most of the patients require prolonged treatment to maintain clinical remission. Moreover, in our cohort of patients the rate of response was higher for anakinra than for canakinumab, suggesting a possible role of IL1 α in the pathogenesis of this condition.

Disclosure of Interests: Roberta Caorsi Consultant for: Novartis, Eli Lilly, Antonella Insalaco: None declared, Chiara Longo: None declared, Giorgia Martini: None declared, Marco Cattalini: None declared, Rita Consolini: None declared, Giovanni Filocamo: None declared, Alessandro Rimini: None declared, Silvia Federici: None declared, Camilla Celani: None declared, Maria Cristina M

DOI: 10.1136/annrheumdis-2019-eular.5854

SAT0491

METABOLIC DISORDERS IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS

Iryna Chyzeuskaya¹, Ludmila Byelyaeva¹, Tatyana Matsushko², Tamara Yuraga¹. ¹Belarusian Medical Academy of Postgraduate Education, Minsk, Belarus; ²4th Minsk City Children's Clinical Hospital, Minsk, Belarus

Background: The urgency of identifying metabolic disorders causing the formation and complication of rheumatic diseases is associated with the difficulties of early and differential diagnosis, and often unfavorable outcomes with a high incidence of patient disability.

Objectives: The purpose of the study is to study the lipid, lipoprotein spectrum of blood serum, lipid peroxidation and antioxidant protection in children with juvenile idiopathic arthritis (JIA).

Methods: On the state of lipid metabolism was evaluated by the study content of total lipids, total cholesterol, triglycerides, lipoprotein spectrum of blood serum apolipoprotein atherosclerotic and antiatherogenic lipoproteins (apoB, apoA, apoE), activity of lipoprotein lipase, primary and secondary products of lipid peroxidation (LPO) and the content of antioxidant ability of water - and fat-soluble antioxidants.

Results: 147 children with juvenile idiopathic arthritis (JIA) aged from 2 to 17 years were examined. The control group consisted of 50 healthy children. In children with JIA, a significant increase in the content of triglycerides, total cholesterol ($p < 0.001$) and a decrease in cholesterol-HDL levels ($p < 0.05$) in comparison with the same indicators in the control group was established. The mean concentrations of cholesterol-HDL significantly decreased with an increase in the degree of disease activity: 1.15 [0.98; 1.3] mmol/l with minimal activity, 1.02 [0.87; 1.17] mmol/l with moderate activity and 0.94 [0.78; 1.01] mmol/l with a high activity of the disease.

Lipoprotein lipase is a key enzyme in lipid metabolism, ensuring the hydrolysis of plasma triglycerides found in chylomicrons and very low density lipoproteins. An increase in the catalytic activity of lipoprotein lipase leads to a decrease in the level of TG and an increase in the level of HDL in the blood. According to the results of the survey in children with JIA, the concentration of lipoprotein lipase was 13.9 [7.1; 25.3] µg/l, with obesity 27.5 [9.9; 47.8] µg/l. A direct relationship was established between its content in blood serum and the concentration of LDL ($r_s=0.37$; $p<0.05$) and the inverse with the level of LDL ($r_s = -0.4$; $p<0.05$). There was a significant difference ($p<0.05$) by gender: the level of lipoprotein lipase in the blood serum in girls was 25.8 [16.7 - 54.4] µg/l, in boys - 15.6 [7.8 - 29.0] µg/l. A correlation was also established between the level of lipoprotein lipase and body mass index ($r_s = 0.35$; $p<0.05$).

According to the results of the study, a significant ($p<0.05$) increase in the level of primary (DK233) and secondary (DK278 and MDA) LPO products in the blood serum of children with JIA was found in comparison with the control group. During the correlation analysis, a positive correlation was established between the levels of DK233, DK278 in serum and ESR ($r_s = 0.587$, $p<0.01$). The results obtained indicate the intensification of lipid peroxidation processes. A significant ($p<0.05-0.01$) decrease in the content of water-soluble (ACW) and fat-soluble (ACL) antioxidant capacity of substances in the blood serum of children with JIA was found when compared with similar indicators in the control group. During the correlation analysis, a negative correlation was established between the level of CRP and the content of ACL in serum ($r_s = -0.346$, $p<0.05$), between the content of ACW and CRP in serum ($r_s = -0.54$, $p<0.01$).

Conclusion: It can be concluded that the activation of lipid peroxidation and the failure of the antioxidant system play a significant role in the development and progression of JIA in the examined children, as well as in the formation of atherogenic disorders by the lipid peroxidation mechanism. In addition, children with JIA have abnormalities in the lipid spectrum, acquiring an atherogenic orientation, which, together with hemostasiological changes, can be regarded as a risk factor for atherothrombosis.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7633

SAT0492 GASTROINTESTINAL MANIFESTATIONS IN CHILDHOOD BEHÇET'S DISEASE

Ferhat Demir¹, Nelgin Gerenli², Seval Simsek¹, Betül Sözeri¹. ¹University of Health Sciences, Umraniye Training and Research Hospital, Pediatric Rheumatology, Istanbul, Turkey; ²University of Health Sciences, Umraniye Training and Research Hospital, Pediatric Gastroenterology, Istanbul, Turkey

Background: Behçet's disease (BD) is an idiopathic, chronic, inflammatory systemic vasculitis characterized by recurrent oral and genital aphthous ulcers, skin lesions, and ocular involvement. It can also affect multiple visceral organ and system such as heart, lung, blood vessels, gastrointestinal (GI) system, and central nervous system (1). The etiology of the disease is still unknown. New diagnostic criteria for pediatric Behçet's patients have been established in 2015, and its largely based on clinical findings (2). Although the GI manifestations are not take part in to the diagnostic criteria, it can be emerge in great deal of patients and associated with significant morbidity and mortality. The most common GI symptoms in BD are abdominal pain, nausea, diarrhea and GI bleeding in, respectively. Also, the most common reported site of intestinal involvement was ileocecal. The literature data related to GI involvement of BD are frequently concern adult patients and pediatric studies are limited.

Objectives: We analyzed the GI manifestations and its endoscopic findings in children diagnosed with BD.

Methods: We retrospectively reviewed the clinical records of 64 patients who fulfilled the international pediatric classification criteria for BD from December 2015 to December 2018. Endoscopic examination results of 13 patients were also collected and analyzed.

Results: Of the 64 children with BD, 37 were male and 27 were female, and sex ratio (male/female) was 1.37/1. The mean age of our patients was 9.49±4.10 and 11.52±3.50; at onset of symptoms and at the time of diagnosis, respectively. Assessment of our 64 patients revealed that the prevalence of GI symptoms is 60% (39/64). The most frequent GI symptoms were abdominal pain in 56% (36/64), chronic diarrhea in 18% (12/64), nausea/vomiting in 11% (7/64), chronic constipation in 12% (8/64), and regurgitation or heartburn in 7% (5/64). Upper GI endoscopy was performed in 7 patients and colonoscopy in 6. All of the patients who underwent the endoscopy or colonoscopy, had findings support the GI

inflammation. The endoscopic findings shown in the patients are; gastric inflammation in 100% (7/7), oesophageal inflammation in 57% (4/7), duodenal inflammation in 42% (3/7), colonic erythema and inflammation in 83% (5/6), polyps in 50% (3/6), ulceration in 66% (4/6), and hemorrhoids in 16% (1/6). Upper GI ulceration was not observed. Pathology specimens were taken in 7 upper endoscopies and 6 colonoscopies within our patients. The findings of pathology specimens were vascular congestion and inflammation in 100% (7/7) of upper GI biopsies and 83% (5/6) of colonic biopsies, cryptitis in 66% (4/6) of colonic biopsies. Acute colitis in pathology specimens was shown in 83% (5/6) of the patients. Three of the patients with colitis had punch ulcers, and these patients were evaluated as BD-associated colitis. The other two patients were diagnosed and treated as Crohn's colitis accompanying BD.

Conclusion: The GI system is one of the most frequently affected organ by the BD. It may cause the inflammation and ulceration in the GI tract. Inflammatory bowel diseases can also cause GI inflammation similar to BD. In patients with symptoms of GI inflammation, upper and lower GI endoscopic examination and pathological analysis may be helpful in determine the underlying etiology.

REFERENCES

- [1] - Sakane T, Takeno M, Suzuki N, Inaba G. Behçet's disease. *N Engl J Med* 1999;341:1284-91
- [2] - Koné-Paut I, Shahram F, Darce-Bello M, Cantarini L, Cimaz R, Gattorno M et al, PEDBD group. Consensus classification criteria for paediatric Behçet's disease from a prospective observational cohort: PEDBD. *Ann Rheum Dis*. 2016;75:958-64.

Acknowledgement: We are grateful to all participating children and their families.

Disclosure of Interest: None Declared

DOI: 10.1136/annrheumdis-2019-eular.4053

SAT0493 THE CHALLENGE OF TREATING PULMONARY VASCULITIS IN BEHÇET'S DISEASE: TWO PEDIATRIC CASES

Selcan Demir¹, Erdal Sag¹, Ummusen Kaya Akca¹, Tuncay Hazirolan², Yelda Bilginer¹, Seza Özen¹. ¹Hacettepe University Faculty of Medicine, Pediatric Rheumatology, Ankara, Turkey; ²Hacettepe University Faculty of Medicine, Radiology, Ankara, Turkey

Background: Behçet's Disease (BD) is a multisystemic autoinflammatory disease and the most severe complication of BD is pulmonary artery involvement (PAI). Data regarding treatment and outcomes of pediatric patients with PAI is very limited.

Objectives: Herein, we report two pediatric patients with BD presenting with PAI and treated successfully with aggressive immunosuppressive treatment.

Methods: Demographic data, clinical manifestations, laboratory and radiological findings, and treatments of patients were documented from patient charts retrospectively.

Results:

Case 1: A 15-year-old boy was admitted with abdominal pain and fever. An abdominal Doppler ultrasonography (USG) showed stenosis of vena cava inferior (VCI) with a thrombus. Transthoracic echocardiography (TTE) detected that the thrombus extended from VCI to the right atrium. When he started to have hemoptysis, he was referred to our hospital. TTE showed a mass in RV. The computed chest and abdominal tomography angiography (CTA) showed bilateral aneurysmatic dilatation with thrombi in the pulmonary arteries and thrombosis in vena hepatica. The pathergy test was negative and the HLA B5 was negative. According to the ICBD, the patient had been diagnosed as BD due to genital ulcers and vascular involvement. He was given pulse methylprednisolone (MP) 500 mg 3 days along and followed by oral prednisolone 1 mg/kg/day, intravenous cyclophosphamide at a dose of 15 mg/kg every 3 weeks for a total of 6 cycles, and Interferon-α2a (IFN-α2a) 3 times a week. Within one month, hemoptysis and fever disappeared, and CRP values normalized. After a three-month treatment, TTE and CTA revealed that thrombi shrank significantly. The dosage of prednisolone was tapered gradually and stopped 2 years later. Immunosuppressive treatment was continued with adalimumab. The patient has been followed in remission for nearly 6 years.

Case 2: A fifteen-year-old boy was referred to our hospital for the evaluation of fever for over 4 months and a thrombus in his right ventricle. He had a medical history with cough, fever, intermittent hemoptysis and weight loss for the past 3 months. Physical examination revealed acne-like rashes over face and back, oral ulcers. A CTA confirmed the