arthralgia was present in 5/8, parotitis in 4/8, oral symptoms in 4/8, rash in 4/8, fatigue in 3/8 and ocular symptoms in 2/8 patients. One patient developed calcinations on fingers. All patients had high titer of antinuclear antibodies, 7/8 were positive for anti-Ro and 5/8 for anti-La antibodies. Three patients were tested for rheumatoid factor and all were positive. 6/8 patients had elevated ESR and hypergammaglobulinemia. 4/8 patients had elevated serum amylase.

Biopsy of salivary glands was performed in 5 patients and foci of lymphocytic infiltration were shown in all of them. Further two patients had salivary gland changes on MRI and/or US. On ophthalmologic evaluation 3 patients had positive Schirmer test and one had unstable tear film. One patient had CNS vasculitis and two decreased diffusing capacity of the lungs. Other patients showed no signs of internal organ involvement. 4/8 patients were treated with NSAIDs, 7/8 patients with hydroxychloroquine, 2/8 also received corticosteroids and one patient MMF. At this point the patient with CNS vasculitis has stable changes on head MRI without clinical symptoms. Other patients have no signs of internal organ damage. Calcinations that appeared on fingers in one patient are not progressing.

Conclusion: Most common symptoms in our cohort were arthritis and/or arthralgia, parotitis, oral symptoms and rash. Half of the children presented with recurrent parotitis. One child developed calcinations on fingers, which have not yet been described in patients with SS. Pediatric SS differs from adult SS and specific pediatric diagnostic criteria would improve management of these patients.

REFERENCES

- [1] Yokogawa N, Lieberman SM, Sherry DD, Vivino FB. Features of childhood Sjogren's syndrome in comparison to adult Sjogren's syndrome: considerations in establishing child-specific diagnostic criteria. *Clin Exp Rheumatol* 2017; 34(2): 343–351.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.3817

AB0945 CO-MORBID AFFECTATIONS FORMATION IN CHILDREN WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: In children with systemic lupus erythematosus (SLE) due to the cascade of immune-inflammatory reactions there is a development of systemic vascular endothelium lesion, which causes not only clinical manifestations of the main process, but also leads to the damage of vital organs and systems, the development of metabolic disorders. Extensive damage aggravates more the course of the disease, worsens its prognosis, complicates the response to therapy and reduces the quality of life of patients.

Objectives: The aim of study was to determine the frequency and variants of comorbid conditions in children depending on the duration and activity of the disease and features of the therapeutic complex.

Methods: A survey was conducted on 41 patients with SLE: 37 girls (90.2%), 4 boys (9.8%); the duration of the disease is up to 3 years in 20 patients (48.8%) and more than 3 years in 21 (51.2%). Presence of comorbid conditions determinates as the cardiovascular system, kidney, liver and lungs functions changes. General clinical trials included autoantibodies, disease activity and drugs assessment. Total cholesterol, triglycerides, high density lipoprotein cholesterol, low-density lipoprotein cholesterol, apolipoprotein B, ApoA-I and lipoprotein- α were evaluated. The state of the blood coagulation system was also studied: fibrinogen of the blood, prothrombin index, thrombin time, active partial thrombin time, D-dimer, international normalized ratio. Bone mineral density was determined.

Results: In 74.3% of children with SLE, the presence of comorbid conditions characterized by pathology of the cardiovascular system, kidneys, liver, the organ of vision, pulmonary system is established. Besides, atherogenic dislipoproteinemia has been detected in 60.0%, disorders in the hemostasis system were in 25.0% of patients and 43.0% persons have osteopenia. Children with SLE and the presence of comorbid conditions, especially with the involvement of the liver and kidneys and the long-term preservation of the disease activity (SRP, ANA, anti-DNA), are more likely to develop atherogenic dislipoproteinemia and hypercoagulation (increase in the prothrombin index, fibrinogen and D- dimer). The values

of the comorbidity index in patients with SLE increased while maintaining the activity of the process and using high doses of glucocorticosteroids (GCS) including pulse therapy. Comorbidity index was 2.9 ± 0.5 points in the patients who received the GCS pulse therapy; 2.5 ± 0.3 in the cases of treatment by combination GCS + azathioprine; 2.0 ± 0.6 in the children without GCS; $p < 0.05$). ANA-positivity was also accompanied by higher values of the comorbidity index (2.8 ± 0.3 vs. 1.4 ± 0.5 , $p < 0.05$).

Conclusion: Children with SLE in the long-term course of the disease and while maintaining the activity of the pathologic process (ANA, DNA antibodies high level) have a formation of lesions of systems and organs, the lipids metabolism disorders, prolonged hypercoagulation and the development of osteopenia. Aggressive therapy for reducing the activity of the autoimmune process helps to prevent the formation of comorbid states and persistent metabolic disorders.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.3858

AB0946 QUALITY IMPROVEMENT IN PEDIATRIC RHEUMATOLOGY: A NEW APPROACH TO CARE IN QATAR

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Background: Quality improvement (QI) projects have been established in many pediatric and adult rheumatology centers around the world to enable real time improvement of health delivery and outcomes. QI has been driven by the Institute of Medicine, patients, professional organizations and the providers. Notably, another driver is the discrepancy between the availability of better and targeted therapies and poor disease outcomes.¹ QI allows for healthcare teams to decide on improvement goals to address gaps in care and implement these goals in sustainable ways, evaluate the impact and start a new cycle of improvement. In so doing, teams collaborate, learn from each other, and improve variability in a short time frame. In small practices with limited resources, effective QI projects allows for understanding the gaps in the processes, data collection and several process improvements to be done by different team members, in short cycles that allow for real time data evaluation and fine tuning processes.

Objectives: We describe the chosen quality measures, implementation of processes and first 6 months of data at a newly opened Children's Hospital in Doha, Qatar.

Methods: We chose quality measures that have been proposed in the literature and have been implemented in large learning collaboratives.¹ We worked with the informatics team to build measures that are accurately captured and retrievable from the electronic medical record. Additionally we worked with our Ophthalmology clinic to ensure a smooth referral pathway that is coordinated by nursing staff. Our EMR measures were 1) the screening for drug toxicity among patients receiving methotrexate or leflunomide within 3-4 month of receiving Methotrexate 2) the time between referral to and visit in Ophthalmology, to screen for toxicity among patients receiving hydroxychloroquine 3) time between referral to and visit in Ophthalmology, to screen for uveitis in children diagnosed with juvenile idiopathic arthritis.

Our nursing teams developed a manual joint injection log 1) to capture whether the time between referral for procedure and procedure date occurred within 2 weeks, and monitor side effects.

We examined data between June to December 2018 from the pediatric rheumatology patient population of 420 unique patients.

Results: 96% of children receiving methotrexate or leflunomide were screened for methotrexate toxicity within 3-4 months of the medicine being dispensed.

50% of children receiving hydroxychloroquine were seen by Ophthalmology within 4 months of the referral.

71% of children with JIA were seen by Ophthalmology within 1 month of the referral.

100% of children had joint injections performed within 14 days of the referral.

Conclusion: We described the development, implementation and results of our pediatric rheumatology QI projects. To our knowledge it is the first of a kind in Qatar and in the area. For our growing practice of 600 patient visits our data shows areas of great performance and poor performance, specifically in eye care for our patients. Barriers included education of teams about QI measures and processes, collaborating with other services on negotiating best patient practices.

Successful implementation in our processes underlines the need for further data collection and additional improvement cycles. Additionally, in resource poor areas, it is essential to make good use of the EMR and

work in collaborative teams to ensure participation and sustainability of processes. Next steps would be to add additional quality and disease outcome measures.

REFERENCES

- [1] Harris, J. et al. Improving Care Delivery Outcomes in Pediatric Rheumatic Diseases. *Curr Opin Rheumatol.* 2016 March; 28(2): 110–116.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6188

AB0947

TREATMENT WITH SIMULTANEOUS BIOLOGICAL AGENTS IN JUVENILE IDIOPATHIC ARTHRITIS: SINGLE-CENTER CASE SERIES AND REVIEW OF LITERATURE

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Background: Several therapeutic choices are currently available for juvenile idiopathic arthritis (JIA) patients. The classical first line therapies are NSAIDs and corticosteroids (CTCs) and the second line encompasses syntheical and biological DMARDs. The clinical efficacy of DMARDs (alone or in combination) has been well demonstrated in RCTs and the treatment is often tailored on an individual basis. Combination of bDMARDs has not been formally evaluated in rheumatology and few publications are available concerning the combination of bDMARDs in patients with severe active forms of JIA, mainly systemic [SoJIA] and polyarticular [PA] categories. (1)

Objectives: To describe efficacy and safety of JIA patients resistant to MTX and bDMARDs requiring a simultaneous bDMARDs treatment.

Methods: Retrospective analysis of 7 (4.7 ± 2.1 years, male sex 75%) patients with JIA of a single tertiary center. A systematic review of literature was made to search observational studies from MEDLINE database (January 1946 to January 2019).

Results: All patients in the Paediatric Rheumatology Department (Lyon University Hospital) receiving combination therapy with bDMARDs were eligible. Seven patients with JIA diagnosis (4 SoJIA and 3 PA) were included. Genetic studies were performed in three patients, with positive LACC1 mutation in one. The exposure to the bDMARDs of the whole cohort was 79 patient-years (PY), including 16.5 PY of combination with simultaneous bDMARDs. The delay between the date of diagnosis and the first prescription of a combination of simultaneous bDMARDs treatment was 8.3 ± 4.8 years. Nine bDMARDs drugs were prescribed: anti-TNFs, tocilizumab and rituximab (nine times each). Ten co-prescriptions of the bDMARDs were administrated with 5 possible types of combinations between them, the most frequent associating tocilizumab/rituximab. Rituximab was used most frequently in combination with another bDMARD. The rituximab schedule was different between patients. Disease activity decreased for all patients, primarily at the biological level without reaching complete clinical remission.

In general, the clinical tolerance to the co-prescription of bDMARDs was acceptable. One patient (P2) experienced a severe hepatitis event in adulthood attributed to the treatment (tocilizumab), dropping the bDMARDs combination. During the exposure time, the most frequent adverse events were infections: repeated otitis media (P1), persisting vaginitis and fungal urinary infection (P3) not requiring hospitalization. One patient (P6) had 2 episodes of purpuric rash on the days of the canakinumab injection requiring oral CTC treatment. No severe acute reactions were found and no patients died.

The systematic review found a publication: retrospective series of 4 patients (SoJIA) combining abatacept and canakinumab (2).

Conclusion: Severe forms of JIA that are refractory to the current therapeutic options available could benefit from the combination of bDMARDs. The combination of bDMARDs may be an alternative therapy to explore in this group of patients.

REFERENCES

- [1] Cimaz R, Marino A, Martini A. How I treat juvenile idiopathic arthritis: A state of the art review. *Autoimmun Rev.* 2017 Oct;16(10):1008–15.
- [2] Record JL, Beukelman T, Cron RQ. Combination Therapy of Abatacept and Anakinra in Children with Refractory Systemic Juvenile Idiopathic Arthritis: A Retrospective Case Series. *J Rheumatol.* 2011 Jan;38(1):180–1.

Acknowledgement: Sincere thanks to Dr. Maria-Luisa Cabañas (INCAN, Ministry of Health of Paraguay)

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5712

AB0948

PARADOXICAL TINEA AMIANTACEA IN A PATIENT WITH JUVENILE IDIOPATHIC ARTHRITIS RECEIVING ADALIMUMAB

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Background: Tinea amiantacea is a papulo-squamous condition of the scalp that can lead to scalp fibrosis and subsequent permanent hair loss. It is thought to represent a reaction pattern to inflammatory skin disease as psoriasis or seborrheic dermatitis (1).

Objectives: To highlight an adverse reaction which involved the skin in the disease course of a young JIA (juvenile idiopathic arthritis) patient, during treatment with adalimumab.

Methods: A 16-month-old female patient presented to our clinic with a 4-week history of knee swelling, associated with functional limitation and morning stiffness. Family history was unremarkable, while past medical history revealed atopic dermatitis in the first year of life. The baby was initially treated with NSAIDs, but one month later, due the persistence of arthritis and the appearance of uveitis, subcutaneous methotrexate was started (15 mg/m²/weekly). However 5 months later, given the persistence of uveitis and the onset of a severe hypertransaminasemia, methotrexate was interrupted and adalimumab (24 mg/m² every 2 weeks) was introduced with a prompt and stable control of ocular and articular disease and a gradual normalization of transaminases. One year later the patient developed dry, itchy, red and cracked skin behind her ears, with fissuring in the lower attachment of the ear lobe, and presented right parietal yellowish scalp lesions which were pruriginous, thick, and scaly, attached both to the scalp and to the proximal hair shafts. A first diagnosis of pityriasis amiantacea secondary to atopic dermatitis was made. A paradoxical cutaneous reaction to the anti-TNF therapy was later hypothesized (2), and 7 months later adalimumab was interrupted with quick resolution of the dermatologic lesions. However, both arthritis and uveitis rapidly recurred, showing an inadequate response to a six month cycle of abatacept treatment (10 mg/kg/month). Adalimumab was than reintroduced with a rapid improvement.

Results: Currently, after 16 months of adalimumab treatment, the patient still shows complete disease control, without any new dermatologic lesions up to now.

Conclusion: TNF antagonist-induced tinea amiantacea is a rare adverse reaction that may require the drug discontinuation. Although the exact pathogenetic mechanism is unclear, an imbalance in the cytokine milieu with a selective overexpression of type I interferon has been hypothesized.

REFERENCES

- [1] F. Osório, F. Magro, C. Lisboa, S. Lopes, G. Macedo, H. Bettencourt, F. Azevedo, S. Magina, Anti-TNF-Alpha induced psoriasiform eruptions with severe scalp involvement and alopecia: report of five cases and review of the literature. *Dermatology.* 2012;225(2):163-7. doi: 10.1159/000342503
- [2] J. Ettler, D.A. Wetter, M.R. Pittelkow. Pityriasis amiantacea: a distinctive presentation of psoriasis associated with tumour necrosis factor- α inhibitor therapy. *Clin Exp Dermatol.* 2012 Aug;37(6):639-41. doi: 10.1111/j.1365-2230.2011.04286.x.

Disclosure of Interests: Maria Costanza Caparello: None declared, Francesca Tirelli: None declared, Gabriele Simonini Grant/research support from: Abbvie, Speakers bureau: Abbvie, Rolando Cimaz: None declared, Teresa Giani: None declared

DOI: 10.1136/annrheumdis-2019-eular.4311

AB0949

IS PEDIATRIC ONSET LUPUS MORE SEVERE IN BOYS? OUR EXPERIENCE AT A TERTIARY CARE CENTER IN NORTH-WEST INDIA

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Background: There is diversity in clinical presentation of pediatric onset SLE (pSLE) and the manifestations are more severe as compared to