REFERENCES

- [1] Frid P, Nortal E, Bovis F, Giancane G, Larheim TA, Rygg M, et al. Temporomandibular Joint Involvement in Association With Quality of Life, Disability, and High Disease Activity in Juvenile Idiopathic Arthritis. *Arthritis Care Res (Hoboken)*. 2017;69:677-86.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7433

SAT0500

JUVENILE RECURRENT PAROTITIS: A RARE MANIFESTATION OF RHEUMATIC IMMUNE-MEDIATED DISEASES

Sara Ganhaõ¹, Francisca Aguiar², Mariana Rodrigues², Ana Maia³, Iva Brito^{2,4}.

¹Centro Hospitalar e Universitário de São João, Department of Rheumatology, Oporto, Portugal; ²Centro Hospitalar e Universitário de São João, Pediatric Rheumatology Unit, Oporto, Portugal; ³Centro Hospitalar e Universitário de São João, Pediatrics Department, Oporto, Portugal; ⁴Faculty of Medicine of Oporto University, Oporto, Portugal

Background: Juvenile recurrent parotitis (JRP) is the second most common childhood disease of the salivary glands after mumps and mainly affects children between the ages of 3 and 6. There is a male predilection, often with spontaneous resolution at puberty. However, in some cases, it can be the first manifestation of a rheumatic immune-mediated disease.

Objectives: To analyze the clinical, laboratory and imaging profile of children with JRP and investigate the prevalence of rheumatic immune-mediated diseases in these patients.

Methods: Retrospective study from 2008 to 2018 including all cases of recurrent parotitis with juvenile onset at our center. Parameters evaluated included gender, age, laterality, number of recurrences, symptoms of presentation, associated conditions, imaging details, blood tests, treatment, outcome and follow-up.

Results: 40 patients were identified over a 10-year period, with a female to male ratio of 5:3; 62.5% females. The youngest child was nine months old and the eldest was 16 years old with median age at presentation of 5 years. The median (min-max) follow-up period was 3 (0.04-33) years. 29 children (72.5%) only ever reported unilateral symptoms, while the rest had both glands affected, although not usually at the same time. Pain and swelling were the most common presenting symptoms, seen in all cases. Fever in 15 (37.5%) and whitish discharge from Stenson's duct in one (2.5%). 14 (35%) had palpable cervical lymphadenopathy and 4 (10%) had dental caries. The average reported frequency was 1.25 episodes per year, with a range of 0.43-5 episodes. Numbers of recurrences were in range of one to 15 with average of 3 times. Younger age at first episode didn't increase the likelihood of recurrences in our sample ($p=0.123$). Sonography was performed in 34 cases as primary investigation. Reports showed multiple hypoechoic lesions with enlarged intraglandular lymph nodes and enlarged gland. MRI was performed in 10, CT in 4, scintigraphy in 2 and biopsy of minor salivary glands in 1. The median level of C-reactive protein and erythrocyte sedimentation rate was 4 (range 0.2-55.5 mg/l, $n=30$) and 7 (0.5-76 mm in 1st hour, $n=20$), respectively. The median blood leukocyte count $8.13 \times 10^9/L$ (range 3.73-25.4 $\times 10^9/L$, $n = 36$). 20 (50%) patients had serum immunoglobulins measured, 6 of them having hypergammaglobulinaemia. One patient had a marginally low serum Immunoglobulin A (50; normal variation 55-210). Antibody screening was performed in 18 patients (45%), with 10 with positivity for antinuclear antibodies, 3 anti-SSA, 2 anti-SSB, 2 anti-RNP and 2 rheumatoid factor. One of the children had sialolithiasis in the parotid gland and other had an abscess with microaerophilic streptococcus isolated in culture of pus. Among all patients, 5 (12.5%) had a final diagnosis of a rheumatic immune-mediated disease: 3 with primary Sjögren Syndrome (SS) and 2 with mixed connective tissue disease. An idiopathic etiology was determined in 33 (82.5%). These were treated with analgesics and nonsteroidal antiinflammatory drugs with complete resolution of symptoms and the previous ones with immunosuppressants/immunomodulators.

Conclusion: The clinical, laboratory and imaging profile of our sample is in accord with the literature, except for sex predilection. Although recurrent juvenile parotitis is more common, primary juvenile SS should be considered. A high suspicion is crucial mainly because of a lower incidence of xerostomy and xerophthalmia in this population.

REFERENCES

- [1] De Luca R, et al. Juvenile Recurrent Parotitis: Could be a Primary Pediatric Sjogren Syndrome? *SM Otolaryngol*. 2017; 1(3): 1013.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.5124

SAT0501

THE IMPACT OF MORNING STIFFNESS ON THE DEFINITION OF INACTIVE DISEASE IN JUVENILE IDIOPATHIC ARTHRITIS

Maria Francesca Gicchino¹, Gabriella Giancane², Alessandra Alongi², Silvia Rosina², Jessica Tibaldi², Marta Mazzoni², Angelo Ravelli², Alessandro Consolaro². ¹Università degli Studi della Campania L. Vanvitelli, Naples, Italy; ²IRCCS Gaslini, Genova, Italy

Background: Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood. Morning stiffness is a major symptom of JIA, and is usually associated with active disease. The 2004 preliminary criteria for inactive disease (ID) in JIA did not include the assessment of morning stiffness, whereas the 2011 revision of the criteria has allowed the presence of morning stiffness (MS) lasting ≤ 15 minutes. MS was included in 2011 revision based on the consideration that MS of a short duration (i.e., ≤ 15 minutes) can represent residua of previously active disease without current active disease. However, it is unclear whether the disease status of children with ID who have or do not have morning stiffness is comparable.

Objectives: To compare the disease status of children with JIA who met the 2004 and 2011 revised criteria for ID in relation to the presence or absence morning stiffness.

Methods: A database of 1208 Italian children included in 2 multicenter studies (1,2) who underwent a total of 3380 visits was examined to identify all visits in which the patients fulfilled the 2004 or 2011 criteria for ID. In case a patient met the ID criteria in more than 1 visit, only the first visit was retained. For each visit with ID, the duration of morning stiffness was categorized as ≤ 15 min or > 15 min. Clinical assessments included demographic features and parent-reported outcomes

Results: A total of 668 visits in which patients met the criteria for ID were identified. Absence of morning stiffness was reported in 564 (84.4%) visits, whereas in 104 visits (15.5%) there was morning stiffness. Among the visits with morning stiffness, in 55 (8.2%) duration was ≤ 15 min, and in 49 (7.3%) duration was > 15 min. The table shows the comparison of disease duration and parent-reported outcomes between patients with absence or presence of morning stiffness.

	Patients meeting 2004 ID criteria			p-value
	No MS N = 564	MS ≤ 15 min N = 55	MS > 15 min N = 49	
Median [IQR] disease duration	2.2 [0.6, 6.3]	2.0 [0.8, 4.0]	3.4 [0.4, 7.3]	0.859
JAFS total score > 0	107 (19.0)	24 (43.6)	38 (77.6)	< 0.001
PRQL PhS > 1 SD*	61 (10.9)	22 (40.7)	38 (79.2)	< 0.001
PRQL PsS > 1 SD*	32 (5.8)	9 (17.0)	16 (33.3)	< 0.001
VAS DA > 0	170 (30.1)	44 (80.0)	43 (87.8)	< 0.001
VAS well-being > 0	211 (37.4)	42 (76.4)	44 (89.8)	< 0.001
VAS pain > 0	140 (24.8) 484	39 (70.9) 31	35 (71.4) 20	< 0.001
Remission	(87.5)	(59.6)	(41.7)	< 0.001

MS: morning stiffness; IQR: interquartile range, *above the mean of healthy children (2)

Conclusion: Among patients who met the 2011 criteria for ID, those with morning stiffness ≤ 15 min had worse parent-reported outcomes than those without morning stiffness. This finding suggests that parents may not perceive their child's disease state as true remission when lower degrees of morning stiffness are present. Notably, a sizeable proportion (7.3%) of children meeting the 2004 ID criteria had morning stiffness lasting > 15 min. The removal of the criterion "Duration of morning stiffness of ≤ 15 minutes" to "Absence of morning stiffness" in the definition for ID should be considered.

REFERENCES

- [1] Filocamo, et al. A new approach to clinical care of juvenile idiopathic arthritis: the Juvenile Arthritis Multidimensional Assessment Report. *J Rheumatol* 2011

[2] Verazza, et al. Disease status, reasons for discontinuation and adverse events in 1038 Italian children with juvenile idiopathic arthritis treated with etanercept. PROJ 2016

Disclosure of Interests: Maria Francesca Gicchino: None declared, Gabriella Giancane: None declared, Alessandra Alongi: None declared, Silvia Rosina: None declared, Jessica Tibaldi: None declared, Marta Mazzoni: None declared, Angelo Ravelli Grant/research support from: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Consultant for: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Speakers bureau: Angelini, AbbVie, Bristol-Myers Squibb, Johnson & Johnson, Novartis, Pfizer, Reckitt Benkiser, and Roche, Alessandro Consolaro Grant/research support from: AbbVie, Pfizer, DOI: 10.1136/annrheumdis-2019-eular.7784

SAT0502

EFFICACY AND SAFETY OF ADALIMUMAB IN JUVENILE IDIOPATHIC ARTHRITIS – 10 YEAR EXPERIENCE USING DATA OF THE BIKER REGISTRY

Gerd Horneff^{1,2}, Kisten Minden¹, Toni Hospach¹, Ivan Foeldvari¹, Peter Haas¹, Angelika Thon¹, Betina Rogalski¹, Gerd Ganser¹, Frank Weller-Heinemann¹, Andreas Urban¹, Markus Hufnagel¹, Prasad Oommen¹, Ariane Klein¹. ¹BIKER-Registry, Sankt Augustin, Germany; ²Paediatric Rheumatology Centre, General Paediatrics, Sankt Augustin, Germany

Background: Since 2008, Adalimumab is approved for polyarticular juvenile idiopathic arthritis (JIA) and approval has been extended to other JIA categories and for uveitis.

Objectives: To evaluate efficacy and safety of Adalimumab in clinical practice in JIA patients in comparison to a biologic-naïve JIA cohort using the German Biologics registry (BiKeR).

Methods: Baseline demographics and disease activity parameters have been documented. Efficacy was determined using the JADAS10. Safety assessments were based on adverse events (AE) reports processed according to MedDRA.

Results: Until October 1, 2018, 951 JIA patients treated with Adalimumab were registered in BiKeR, representing 1519 patient-years (PY) of exposure. The total observation time (from date of first dose until last follow-up, censored, if another biologic was started) was calculated with 1709 PY.

At baseline, the cohort treated with Adalimumab had experienced disease with a disease duration of 5.4+/-3.9 years (mean+/-SD). 849 patients (89.3%) were pretreated with methotrexate, 525 (54.2%) with Etanercept. Concomitantly, 584 (61.4%) received methotrexate, 529 (55.6%) NSAIDs and 255 (26.8%) systemic corticosteroids. In the control cohort, 1517 biologic naïve JIA patients started methotrexate.

Upon treatment, the median (IQR1-3) JADAS10 score decreased from 9.8 (4.7-15.5) at baseline to 3.9 (0.9-9.7) at the last follow-up. With respect to patients with ongoing treatment, approximately 43% achieved a JADAS defined minimal disease activity while 25% reached a JADAS defined remission at last follow-up.

904 AE have been observed during exposure or up to 90 days follow-up (58.4/100 exposure years (EY) [95% CI 54.7-62.3]). 66 qualified as serious AE (SAE) (4.3/100 EY [3.3-5.4]). These figures were compared to 1294 AE reported in the control cohort (34.8/year [32.9-36.8]) and 52 SAE (1.4/100 EY [1.1-1.8]).

Adverse events of special interest were serious/medically important infection (n=33), opportunistic infection (n=6, all H. zoster), malignancy (n=2), anaphylaxis/hypersensitivities (n=3), thrombotic disorders (n=2), autoimmune diseases (n=91, including 12 cases with psoriasis and 53 reports of uveitis), bleeding (n=3), cytopenias (n=4), pregnancies (n=2). Macrophage activation syndrome, demyelination, cardiac or cerebral infarction, or death were not observed. However, 2 malignancies were reported in patients who had ever been exposed to Adalimumab before. Both events were judged as unrelated.

A total of 449 patients (48.5%) discontinued Adalimumab, 188 (19.8%) due to lack of efficacy, 111 (11.7%) due to remission and 78 (8.2%) because of intolerance.

Conclusion: The current analysis adds to the established safety profile of Adalimumab and demonstrates that the rate of SAEs was comparable and consistent with the overall AE profile in paediatric patients. As expected, infections were the most frequent SAE. Uveitis, as well as psoriasis, are likely associated with JIA. Of notice, only one patient developed a chronic inflammatory bowel disease. No new safety signals specific to the paediatric population were identified in this large cohort of JIA patients.

Efficacy measure: JADAS 10

REFERENCES

- [1] Klein A, Becker I, Minden K, Foeldvari I, Haas JP, Horneff G. Adalimumab versus adalimumab and methotrexate for the treatment of juvenile idiopathic arthritis: long-term data from the German BIKER registry. *Scand J Rheumatol*. 2018 Nov 9:1-10.

Acknowledgement: The authors acknowledge all patients, parents and contributors to the BIKER registry and Abbvie, Germany, for financial support for parts of BIKER.

Disclosure of Interests: Gerd Horneff: None declared, Kisten Minden Grant/research support from: Pfizer, Abbvie, Toni Hospach Speakers bureau: Chugai, Roche, Novartis, Ivan Foeldvari Consultant for: Chugai, Novartis, Peter Haas Grant/research support from: Pfizer, Angelika Thon: None declared, Betina Rogalski <: None declared, Gerd Ganser: None declared, Frank Weller-Heinemann: None declared, Andreas Urban: None declared, Markus Hufnagel: None declared, Prasad Oommen: None declared, Ariane Klein: None declared

DOI: 10.1136/annrheumdis-2019-eular.1457

SAT0503

DEVELOPMENT OF MALIGNANCIES IN JIA PATIENTS EXPOSED TO BIOLOGIC AGENTS: A SINGLE CENTRE RETROSPECTIVE STUDY

Oya Koker¹, Sezgin Sahin¹, Amra Adrovic¹, Mehmet Yildiz¹, Kenan Barut¹, Rukiye Eker Omeroglu², Ozgur Kasapcopur¹. ¹Istanbul University-Cerrahpasa, Cerrahpasa Medical School, Pediatric Rheumatology, Istanbul, Turkey; ²Istanbul University, Istanbul School of Medicine, Pediatric Rheumatology, Istanbul, Turkey

Background: Over the last two decades, the usage of biological agents in the treatment of Juvenile Idiopathic Arthritis (JIA) has been a successful and promising approach in controlling the disease activity and preventing chronic sequelae. However, since Food and Drug Administration (FDA) has drawn attention to the possible association between the use of biological agents and the development of malignancy in 2008, there are ongoing concerns about the long-term safety data and side effect profile of the drugs with the contradictory study results.

Objectives: We aimed to present preliminary data on the incidence of malignancy in patients with JIA treated with biological agents versus the general population rates in Turkey.