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POS1331

LONG-TERM OUTCOME IN CHILDREN WITH JUVENILE DERMATOMYOSITIS FROM SOUTHERN INDIA

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Background: There is paucity of data regarding long-term outcome and cumulative damage in children with juvenile dermatomyositis (JDM) from the Indian subcontinent.

Objectives: To assess the long-term outcome and cumulative damage in children with JDM receiving treatment at a tertiary hospital in southern India.

Methods: Retrospective review of records and cross-sectional assessment of outcome and damage in 29 patients with JDM at a tertiary hospital in Kochi, India. The disease course was categorized as monocyclic, polycyclic and chronic progressive. Cumulative damage was assessed using the IMACS myositis damage index (MDI).

Results: Twenty-nine patients (male-16) diagnosed with definite or probable juvenile dermatomyositis based on the Bohan and Peter criteria and having a minimum follow-up period of 3 years each were enrolled. Of these, 20 children were diagnosed and initiated on treatment at our institute (inceptional) and 9 were diagnosed elsewhere and referred to our centre for further management (non-inceptional). The mean age at disease onset was 7.01 ± 3.34 years (range 1.0 to 13.5 years). The median interval from onset to diagnosis was 3 months (range- 3 weeks to 8.75 years). Delayed diagnosis defined as interval from onset to diagnosis exceeding 6 months was noted in case of 8 children. Among patients in the non-inceptional group, six were considered to have not received standard of care treatment prior to referral to our centre. Standard of care treatment was defined as initiation of a treatment regimen comprising of a combination of glucocorticoids with an immunosuppressive agent within 4 weeks of diagnosis. A total of 11 children had a delayed diagnosis and/or had not received standard of care treatment prior to referral.

At our centre, all patients received oral steroids and subcutaneous methotrexate as standard therapy. Pulse steroids were used to induce remission in 12 patients and as a rescue for relapses in 2 patients. Intravenous immunoglobulin was used in 10 children with severe myositis, oropharyngeal weakness, refractory cutaneous disease including calcinosis and concomitant infection, where affordable with good results. Hydroxychloroquine was added in 15 patients with dominant cutaneous manifestations. Mycophenolate mofetil, azathioprine and tacrolimus were used in patients refractory or intolerant to methotrexate. Cyclophosphamide and rituximab were used in 4 patients each with refractory disease and extra-muscular manifestations such as interstitial lung disease. Seven patients with refractory calcinosis received pamidronate infusions.

The total follow-up duration was 121.06 patient-years. A monocyclic course was observed in 11/29 (37.9%), chronic progressive course in 16/29 (55.2%) and polycyclic course in 2/29 (6.9 %). Nearly half of the patients (14/29) had damage in at least one organ using the MDI (MDI ≥ 1). 38% of the patients (11/29) had at least one sign of cutaneous damage, the most common being calcinosis (n=11) and lipodystrophy (n=4). This was followed by skeletal damage (n=6, joint contractures-5, osteoporosis with fracture-1), muscle and pulmonary (pulmonary

fibrosis) damage in 3 patients each, endocrine damage in 2 patients (diabetes mellitus-1, growth failure and delayed development of secondary sexual characteristics-1) and chronic infection in one patient. All patients with damage in at least one organ had a chronic progressive course, indicating damage accrual. Delayed diagnosis and lack of standard of care treatment prior to referral (i.e., longer duration of untreated/ sub-optimally treated disease) were associated with an increased risk of cumulative damage ($p < 0.05$).

Conclusion: Nearly half of the patients had damage in at least one organ using the MDI. Cutaneous damage was the most common, followed by skeletal, muscle, pulmonary and endocrine damage. Longer duration of untreated/ sub-optimally treated disease significantly increases the risk of cumulative damage, highlighting the need for an early diagnosis and referral to pediatric rheumatology services.

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POS1332

ETANERCEPT-ASSOCIATED NEW ONSET UVEITIS IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS UNDER BIOLOGICAL THERAPY: SINGLE CENTER EXPERIENCE

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Background: Biological agents (BA), especially TNF inhibitors, are high efficacy options for current therapy for patients (pts) with juvenile idiopathic arthritis (JIA). They are successfully used not only for the arthritis but also for JIA-associated uveitis, however, development of uveitis *de novo* in pts treated with BA is a well-established paradoxical phenomenon.

Objectives: to evaluate the frequency of new onset (*no*-) uveitis, occurring under BA therapy in JIA pts, to establish clinical features, which may be associated with development of such effects.

Methods: retrospective cohort study involved all JIA pts (1136) who were treated with BA in our clinic from 2004 to 2020. All cases of *no*-uveitis were collected for the describing of their clinical features in disease onset and course, activity level, JIA category, exposure to Methotrexate (MTX) and BA, presence of ANA, HLA B27. **Results:** among of 1136 pts treated with different BA we identified 36 (3.3%) pts (19 female/17 male) with *no*-uveitis under BA. Mostly during etanercept (ETA) therapy (34 cases from 488 ETA courses, 7%), 1/166 - in abatacept (ABA) and 1/372 - in adalimumab (ADA). 30 pts (83%) with *no*-uveitis developed it on the 1st line of BA treatment (29 ETA vs 1 ADA). 4pts (11%) developed *no*-uveitis on 2nd line (3 ETA vs 1 ABA). 2 pts (6%) on third line (all ETA, both pts had also psoriasis). There are no cases of *no*-uveitis under other BA. Frequency of *no*-uveitis was much higher in ETA group. ETA exposure was 26.8 ± 28.8 months (mo). It means there are no "safe" period of therapy from paradoxical phenomenon of *no*-uveitis. JIA subtypes were as follows: RF-neg polyarthritis 9 (25%), persistent oligoarthritis 3 (8%), extended oligoarthritis 21 (59%), enthesitis-related arthritis (ERA) - 3 (8%). Average age at JIA onset was 4.6 ± 3.9 yrs. 20/36 patients had high laboratory activity (CRP 54 ± 23 mg/l; ESR 41 ± 19 mm/h) and severe arthritis before BA initiation. However most of pts (25/36) achieved 90-100% ACRpedi-response by the uveitis development. 23/36 pts were ANA-positive, 17/36 pts had HLAB27, including 7 pts who had the both features. Uveitis was occurred earlier in ANA plus HLAB27-positive pts (mean exposure - 15.3 mo) than in only ANA-positive or HLAB27-positive pts (27.7 mo and 27.6 mo accordingly). 29/36 (81%) of pts received methotrexate (MTX) in mean dosage $11.5 \text{ mg/m}^2/\text{week}$. There are no differences in time of uveitis development depending of MTX. In all cases of *no*-uveitis BA was switched.

Conclusion: Our study suggested that new onset of uveitis is rare adverse event during BA therapy in JIA. Uveitis can develop despite the excellent effect of therapy on joint manifestation. The most typical development of *no*-uveitis is under ETA therapy, especially in the predisposed cases (certain variants of JIA, ANF positivity, HLAB27 presence) and in patients with very high disease activity at the time of the start of biological therapy.

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POS1333

ANKLE ARTHRITIS IN JUVENILE IDIOPATHIC ARTHRITIS: DON'T MISS TENOSYNOVITIS. CLINICAL AND SONOGRAPHIC FEATURES OF ANKLE TENOSYNOVITIS IN JIA

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Background: Tenosynovitis can occur in patients with Juvenile Idiopathic Arthritis (JIA) and may be clinically difficult to distinguish from joint synovitis. The role of musculoskeletal ultrasound (msk-US) is still discussed in the management of JIA but recent studies supported the utility of msk-US especially in detection of tenosynovitis. There is no consensus treatment for tenosynovitis in children with JIA and almost all studies focused on tendon sheath injection with glucocorticoids.

Objectives: The aims of the study were: i) to define the prevalence of tenosynovitis of the ankle in JIA patients and ankle swelling; ii) to describe the clinical characteristics of patient with tenosynovitis and to analyze different response to treatment.

Methods: We conducted an observational cross-sectional study of a group of patients with JIA followed at the Rheumatology Service of the Maternal and Child Institute "Burlo Garofolo" of Trieste. We enrolled all the patients who reported a swelling of the ankle at least once during the follow-up period and, among these patients, we included only those who underwent msk-US. Based on both clinical and sonographic examination, we identified patients with tenosynovitis and we described their demographical and clinical characteristics as well as the therapeutic approach undertaken in this group of patients.

Results: On December 31st 2019, 56 swollen ankles of 48 patients were assessed with msk-US: 22 ankles showed sonographic signs of synovitis (39%), 16 ankles of both synovitis and tenosynovitis (28%), 14 ankles of tenosynovitis only (25%). Overall, tenosynovitis was detected on twenty-seven (56%) out of 48 children with at least a swollen ankle. The majority of patient were females (70%) and the most affected tendon was the tibialis posterior (66%). Twenty-five patients with tenosynovitis (92%) achieved clinical and radiological remission: seven out of 26 patients (26%) treated with methotrexate achieved clinical and radiological remission without the addition of other therapies; fifteen out of seventeen patients (88%) treated with a biological drug responded to the therapy, of which eleven (73%) were in combination therapy with methotrexate.

Conclusion: We observed that more than 50% of the patients with a swelling of the ankle presented a tenosynovitis and among these patients about 50% did not show sonographic sign of synovitis. Msk-US was decisive in order to identify tenosynovitis and to characterize ankle swelling in JIA patients. Among patients with tenosynovitis biological therapy alone or in association with immunomodulating therapy showed effectiveness in inducing disease remission.

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POS1334 RHEUMATOLOGY PATIENT ATTENDANCE TO THE PAEDIATRIC EMERGENCY DEPARTMENT

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Background: A large proportion of patients present to the paediatric emergency department (PED) with non-urgent problems [1]. With growing pressures on services, there is a need to reduce unnecessary attendances. Children with rheumatological conditions may attend PED with flares of their condition or unrelated presentations. These patients may be immunosuppressed due to steroid, disease-modifying or biologic therapy; PED attendance may therefore pose a serious infection risk.

Objectives: To evaluate why children with rheumatological conditions attend the PED and whether these attendances are avoidable.

Methods: This study was run in Royal Manchester Children's Hospital, a UK hospital with a PED and tertiary paediatric rheumatology services. Patients under rheumatology who attend PED are usually seen directly. A dataset of all patients referred directly to paediatric rheumatology between 01/01/19 and 31/12/19 was

obtained from electronic records (n=59). The age, sex and reason for attendance were collected, as well whether they were admitted/discharged. Further data on management was collected from discharge letters for those with joint pain/swelling.

Results: This cohort had 36 females and 23 males, with an average age of 11.9 years (range 1-18). 28 were discharged (47.5%) and 30 were admitted (50.8%). One patient left before being seen. The average waiting time was 3.97 hours.

Table 1 summarises reasons for attendance to PED and final outcome. The most common reason for attendance was joint pain/swelling (n=14, 23.7%). Of these, nine were discharged with advice on analgesia or steroid dosing and for early review in clinic. There was no documentation on whether there had been any attempt to access the available rheumatology helpline prior to attending PED. Non-rheumatology-related reasons for attendance are also shown in Table 1, and notably include 20 patients with infections, 75% of whom were admitted to hospital. Chicken pox was the second most common reason for attendance (n=5, 8.5%); all patients attending with chicken pox or shingles (n=6) were admitted.

Table 1. Reasons for attendance to PED and final outcome

Reason for attendance	Number admitted	Number discharged
Joint pain/swelling	5	9
Chicken pox	5	0
Viral URTI	1	4
Rash	0	4
Pneumonia	3	0
SLE flare	3	0
Influenza A	2	0
GPA flare	2	0
Asked to attend due to blood results from clinic	2	0
Drug reaction	0	2
Minor injury	0	2
Shingles	1	0
Oral herpes simplex	1	1
Orbital inflammation	1	0
Urinary tract infection	1	0
Dysphagia	1	0
Gastroenteritis	1	0
Cerebral infarcts	1	0
Constipation	0	1
Chest pain	0	1
Abdominal pain	0	1
Enlarged lymph node	0	1
Picking up prescription	0	1
Unresponsive episode	0	1

Conclusion: Our study shows that attendance to PED with joint pain/swelling is usually avoidable in patients known to paediatric rheumatology services. Appropriate alternative services are needed to reduce hospital attendances; nurse-led helplines are beneficial, but still face some challenges in both availability and accessibility [2]. It is also important that patients and parents are given action plans for acute flares, which would ideally involve early clinic review rather than PED attendance. Our data also showed that patients are spending a long time waiting in PED, which needs to be avoided particularly in the context of immunosuppression. This is especially poignant now in light of the Covid-19 pandemic. Direct referral to rheumatology with non-rheumatological problems likely reflects a desire for specialist advice in the context of complex conditions and medications, for example with regards to infections whilst on immunosuppressive medications, or uncertainty about whether new symptoms are related to the underlying condition. Expansion of helpline services and new approaches such as remote consultations should be explored.

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Other orphan diseases

POS1335 CLINICAL CHARACTERISTICS OF THE PULMONARY ARTERY INVOLVEMENT IN PATIENTS WITH BEHÇET'S SYNDROME: A SINGLE-CENTRE EXPERIENCE OF 61 PATIENTS

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