Conclusion: The most commonly prescribed biologic drugs were IL-1 inhibitors especially for patients with IL-1-mediated AID (FMF, CAPS, and SJIA). The biologic treatment in AID is effective and there were no serious side effects.

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AB0961

ADVERSE EVENTS ASSOCIATED WITH ANTI-TNF- α THERAPY IN PEDIATRIC RHEUMATIC DISEASES

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Background: In recent years, the biologic drugs has led to a dramatic change in the management of rheumatic diseases. The most commonly used molecules are TNF- α antagonists among biologic treatments in pediatric age (1). The anti-TNF- α agents used in childhood are; etanercept (a fusion protein of the TNF- α receptor), infliximab (a chimeric monoclonal antibody) and adalimumab (completely human monoclonal antibody). As a result of the increasing use of anti-TNF- α agents in recent years, adverse events reports have also increased (2). We assessed the prevalence of adverse events (AEs) in a single pediatric referral center for chronic rheumatic diseases.

Objectives: The aim of this study was to evaluate the adverse events that associated with anti-TNF- α therapy in children with rheumatic disease.

Methods: This was cross-sectional study conducted University of Health Sciences, Umraniye Training and Research Hospital, Department of Pediatric Rheumatology, in Turkey. We retrospectively reviewed the patients with a diagnosis any of pediatric rheumatic disease whom treated at least 3 months with an anti-TNF-alpha agents (etanercept, infliximab, adalimumab), between June 2016 and January 2019. Adverse events that develop after anti-TNF- α treatment were recorded. AEs were categorized and graded based on the Common Terminology criteria for AEs (CTCAE). Grades 3-5 were considered severe AEs.

Results: We evaluated 131 patients who were treated with anti-TNF- α drugs; 110 with juvenile idiopathic arthritis (JIA) (27 of with uveitis) 83%, 10 with idiopathic uveitis 7%, 5 with Behçet's uveitis 4%, 4 with adenosine deaminase deficiency 3%, and 2 with juvenile sarcoidosis 2%.

The study included 74 females and 57 males (F/M: 1.29/1) and the mean age of the patients was 12.8 years. Of the patients, 106 had used only 1, 21 had two different and 4 had 3 different anti-TNF-alpha biologic treatments. A total of 160 different anti-TNF-alpha experiences had in 131 patients.

A total of 333.4 patient-years (PYs) were included: 136 PYs-63 patients for etanercept (2.15 patient/year), 134 PYs-68 patients for adalimumab (1.97 p/y) and 63.4 PYs- 29 patients for infliximab (2.18 p/y). During follow-up, 44 patients (33.5%) experienced at least one AEs. 14 of them (10%) were recorded as SAEs. 17 of the AEs were show up after adalimumab, 16 were etanercept and 11 were infliximab. The most common AEs were found; local reactions, increased infection frequency and PPD positivity in follow up, respectively. The SAEs that observed were; anaphylactoid reactions (n=5), uveitis (n=3), pneumonia (n=3), TAILS (n = 2) and vasculitis (n=1).

Conclusion: Here in, we presented safety data of anti-TNF-alpha drugs in pediatric patients. Although the high prevalence of AEs was observed, when anti-TNF-a discontinuation, AEs were mostly not persistent and not mortal.

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UPDATE FOR THE CLINICAL PRACTICE: INTEGRATED, EVIDENCE-BASED APPROACH FOR THE MANAGEMENT OF JUVENILE SPONDYLOARTHRITIS

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Background: Juvenile onset spondyloarthritis (SpA) is a heterogeneous group of human leukocyte antigen (HLA) -B27 associated inflammatory syndromes that affect children and adolescents under the age of 16 years. No specific recommendations for the treatment of juvenile spondyloarthritis have been established. Important differences exist between spondyloarthritis in children and adults, supporting the need for pediatric-specific recommendations.

Objectives: To set recommendations for the management of children and adolescents with spondyloarthritis.

Methods: Searching Medline for Juvenile spondyloarthritis management was done. A systematic literature search was conducted to collect the existing recommendations, guidelines involved line of treatment for Juvenile SpA & adult SpA. These included 2011 ACR recommendations for the treatment of JIA and the 2016 update of the ASAS-EULAR management recommendations for axial spondyloarthritis.

Results: NSAIDs have been shown to improve symptoms, reduce inflammatory lesions and may slow spinal radiographs progression with continuous use. Glucocorticoid injections directed to the local site of musculoskeletal inflammation may be considered. Sulfasalazine may be considered in patients with peripheral arthritis. Initiation of a (tumor necrosis factor inhibitor (TNFi) was recommended for patients with active sacroiliac arthritis who have received an adequate trial of NSAIDs. Also, it should be initiated to those who fail to respond to synthetic disease modifying anti-rheumatic drugs (sDMARD). If TNFi therapy fails, switching to another TNFi should be done. If a patient is in sustained remission, tapering of a biological DMARD can be considered.

Conclusion: These guidelines provide up-to-date guidance on the management of patients with juvenile SpA, based on combining evidence and expert opinion.

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ROLE OF THYROID HORMONES IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: The frequency of autoimmune thyroid disease have been reported in adults with systemic autoimmune diseases. However, little is

known about the association between those diseases and rheumatic disorders during childhood, such as juvenile idiopathic arthritis (JIA).

Objectives: Our objective was to evaluate the correlation between the thyroid function and the clinical presentation in children with juvenile idiopathic arthritis.

Methods: Ninety-seven children (56 female, 41 male, median age 9.57 years, range 1.1–17.5 years) with juvenile idiopathic arthritis were evaluated. All patients underwent thyroid function tests (TSH, free T₄ and free T₃), antithyroglobulin (TgA) and antiperoxidase (TPOA) antibodies. All patients had a thyroid high-resolution sonography examination. Patients with nodules on US examination, low-TSH level and/or high thyroid hormone levels or positive TPOA and/or TgA values were referred to thyroid scintigraphy. The gained data was correlated with disease activity indices. Thyroid autoimmunity was evaluated by fluorescence enzymatic immunoassays of TgA and TPOA antibodies, considering TgA >50 IU/ml and TPOA>100 IU/ml as positive values.

Results: Fifty-one of the cases were classified as oligoarticular, thirty-one as seronegative polyarticular, two as seropositive polyarticular, eleven as systemic, and two as enthesitis related- juvenile idiopathic arthritis.

No cases of overt clinical and biochemical hypothyroidism were found among children with juvenile idiopathic arthritis. On opposite, mean thyroid free T₃ levels were higher in 36.4% of cases and free T₄ in 29.7% cases. Anti-TPO antibodies were found in 5 of 97 patients (5.15%) and anti-TG antibodies were found in 2 patients (2.06%). All patients with thyroid antibodies positives were females. The ultrasound examination of thyroidal gland revealed abnormalities in 41% cases, most of them cystic changes (33.7%) and hypo-echogenicity (28.57%). In those patients with thyroid nodules on US examination, the thyroid scintigraphy did not revealed pathological activity or other processes. Although, 2 patients presented mean thyroid volume above 2SDS according their age reference values. An increased vascular flow pattern on Doppler examination of thyroidal gland was found in 26% cases. Correlation and regression analysis showed low age at diagnosis and longer duration of the disease to be predictors for those thyroid disorders.

Conclusion: These data seem to suggest careful monitoring of thyroid function and thyroid autoantibodies just in children with juvenile idiopathic arthritis presenting clinical symptoms of thyroid involvement.

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AB0964 DIAGNOSIS OF CROHN'S DISEASE IN PATIENTS WITH PRIMARY RHEUMATOLOGICAL MANIFESTATIONS

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Background: Sometimes Crohn's disease (CD) manifests initially with rheumatological symptoms. Therefore, there's an important for a rheumatologist to recognize this pathology correctly and timely.

Objectives: To present cases of CD, referred to V.A. Nasonova Research Institute of Rheumatology.

Methods: Case series of 7 pts (5 boys, 2 girls) with initial rheumatological condition who developed CD. All pts were subjected to standard rheumatological examination. CD diagnosis was suggested by a rheumatologist and confirmed in gastroenterology hospital.

Results: Mean age at CD onset was 12.7±3.98 yrs, varying from 5 to 17 yrs. In 4 pts gastrointestinal (GIT) symptoms manifested simultaneously with rheumatological, in 2 pts GIT symptoms were delayed for 4 and 5 years. The time-interval from the onset of first symptoms until establishing CD diagnosis varied from 2 months till 6 years, averaging to 96±38 months. The list of initial rheumatological diagnoses included the following entities: juvenile idiopathic arthritis (JIA) with oligoarticular onset – in 5 pts, and systemic onset JIA – in 2 pts. In all cases arthritis manifested initially as an oligo-involvement of a lower limb joints. High active sacroiliitis with deep bone oedema was detected by MRI in 5 pts. Fever

was present in 5 pts, uveitis – in 1, cutaneous psoriasis – in 1. Other documented symptoms included: weight loss (3), hepatosplenomegaly (1), lymphadenopathy (1), aphthous stomatitis (1), nodular erythema (1), erythematous rash (1). HLA B27-antigen positivity was established in 3 pts out of 5 examined. All pts had significant increase in CRP and ESR levels and WBC counts; (additionally 3 examined for calprotectin cases demonstrated significant increase of its' concentration (>1000 µg/g).

Conclusion: IBD, including CD, should not be missed and ruled out in all pts with oligoarticular and systemic JIA onset. Fecal calprotectin should be checked in suspected CD cases among individuals with rheumatologic symptoms. Special attention should be given to cases manifesting during puberty, accompanied by weight loss, fever, GIT symptoms, increased levels of acute phase inflammatory markers. MRI stir lesion may be as suspected of IBD especial CD.

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AB0965 PAEDIATRIC SARCOIDOSIS: A PROFESSIONAL IMITATOR

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Background: Sterile granulomatous inflammation is a hallmark of disorders covered by the term „paediatric sarcoidosis“, a rare disease classified among autoinflammatory conditions. While the classical triad of arthritis, uveitis and dermatitis associated with the NOD2 gene mutation is typical for Early-onset sarcoidosis (EOS), mutation-negative disease has extremely variable phenotype in children and can mimic various other conditions.

Objectives: To illustrate variability of clinical presentation of sarcoidosis in a cohort of children from 2 collaborating paediatric rheumatology units

Methods: Retrospective case analysis.

Results: Seven Caucasian patients (2 boys) followed for a median of 1 (0.2-12) year were identified (Table). Patients 1 and 3 presented with typical rash (with biopsy-proven granuloma), polyarthritis and severe, treatment-resistant uveitis which has never been fully controlled. Patient 2 presented with recurrent self-limited febrile episodes and was investigated for periodic fever syndrome. Within 3 months she developed erythema nodosum-like rash, granulomatous uveitis and arthritis. In 4 patients with disease onset after 10 years of age two major organ systems were affected: renal and gastrointestinal (GIT). In patients 4 and 5 renal biopsy performed for insufficiency showed tubulointerstitial nephritis with granulomas. Patients 6 and 7 had been initially treated for “atypical inflammatory bowel disease” with granulomas on biopsy. In both of them finding of granulomatous uveitis eventually lead to the diagnosis of sarcoidosis. Additional symptoms are listed in the Table. Presence of granulomatous inflammation was confirmed in all patients. Chronic uveitis was present in 6/7 patients and was an important diagnostic clue in EOS as well as adult-type of sarcoidosis while respiratory tract was affected subclinically in 2 older patients only. Apart from combinations of clinical symptoms elevation of chitotriosidase significantly aided diagnostic process and when available, it also reflected disease activity. (1)

Pt – patient, F – female, M – male, Dx – diagnosis, CS – corticosteroids, MTX – Methotrexate, ADA – Adalimumab, MMF – Mycophenolate mophetil

Pt	Sex	Age (yrs) at Onset/ Dx	NOD2 gene mutation	Manifestations	Chitotriosidase N 4.4-89 nmol/ml/h	Therapy
1	F	0,25/4	R334Q	Rash, fever, uveitis, arthritis, meningitis	299	MTX, MMF, ADA, MTX
2	F	0,6/1	Negative	Rash, fever, uveitis, arthritis, panniculitis	433	ADA, CS, MTX
3	F	1/3	Variants S178S, V955I, N289S	Rash, fever, panuveitis, arthritis, liver granuloma	N/A	CS, MTX, ADA, Infliximab
4	M	4/15	Pending	Fever, arthralgia, lymphadenopathy, granulomatous nephritis	N/A	CS, MMF
5	F	13/13	Negative	Uveitis, granulomatous nephritis, hearing loss	604	CS, MTX, ADA