Results: The study group consisted of 12 juvenile BD patients with CVST. At the time of CVST diagnosis, the most common symptom was headache (%100), followed by vomiting (25%), blurred vision (16.6-7%), and disturbances in eye movements (16.7%). Six (50%) patients presented with sinus venous thrombosis as an initial symptom. Transverse sinus was the most frequently affected sinus (9/12, 75%) followed by superior sagittal sinus (8/12, 66.6%) and sigmoid sinus (1/12, 8.3%). The median (minimum-maximum) BDCAF was 6 (5-8). Four children (33.3%) had another venous thrombosis apart from CVST. All patients received pulse methylprednisolone for three consecutive days continued with oral prednisolone. Steroid treatment was tapered and discontinued minimum in six months. Eleven patients received azathioprine concomitant to steroid treatment at the time of CVST. All the patients received anticoagulant therapy concomitantly. Only one patient had relapse. Median (min-max) follow-up period was 4 years (1-10). In the literature review, we identified nine articles, describing 35 pediatric CVST patients associated with BD. Thirty patients achieved remission, while five patients had residual neurologic deficit.
Conclusion: Further multicenter studies with more patients and prospective follow-up may help us to understand the whole spectrum in these patients.

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AB0959 EXCELLENT RESPONSE TO TOCILIZUMAB IN PEDIATRIC ONSET REFRACTORY TAKAYASU ARTERITIS: CASE SERIES OF THREE PATIENT

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Background: Takayasu arteritis (TA) is a idiopathic and rarely seen chronic systemic vasculitis that affect the aorta and its major branches. Stenosis, occlusion and aneurysms may develop in large arteries in the setting of granulomatous panarteritis (1). The first purpose in patients with TA after diagnosis is to prevent the progression of vascular lesions with medical treatment. Used as a first choise in medical treatment, steroids and steroid-sparing immunosuppressive agents, may sometimes fail to prevent to progression of the disease. Interleukin (IL)-6, which synthesize from activated dendritic cells, is one of the main cytokine in the development of panarteritis in TA. Tocilizumab (TCZ), an anti-IL-6 receptor antibody, has been shown to be used as an effective treatment in many refractory adult TA patients (2).
Objectives: We have presented our experience with TCZ treatment in three children with refractory TA.
Methods: We reviewed three cases of childhood TA diagnosed between 2016-2018. These patients were successfully treated with TCZ that started due to the refractory disease.
Results: The first patient, 15 year-old girl, presented with fever, headache and abdominal pain. Physical examination showed systolic murmur in interscapular area and abdominal tenderness. The C-reactive protein (CRP) level was 4.7 mg/dL (n: <0.5 mg/dL) and the erythrocyte sedimentation rate (ESR) was 78 mm/h (n: <20 mm/h). Doppler ultrasound imaging showed wall thickening in bilateral carotid arteries. The angio-CT imaging revealed that 70% stenosis in the bilaterally renal arteries and partial stenosis in the mesenteric artery. Methylprednisolone treatment was started (30 mg/kg/d x 3 days). Maintenance treatment consisted of prednisone (1 mg/kg/d), methotrexate and azathioprine. Her complaints improved rapidly and the acute phase reactants decreased. However, the patient relapsed while tapering the corticosteroid.
The second patient was diagnosed with TA in 2012 after the complaint of chest pain and finding of hypertension. MRI angiography revealed wall irregularities and stenosis in the descending aorta and dilatation in the ascending aorta. CRP and ESR levels found high. The patient was followed up with glucocorticoid and azathioprine treatment. After four years of follow-up, relapse of the disease was observed with symptoms of fever, chest and back pain.
The third patient presented with fever, abdominal pain, arthralgia in the left elbow and arthritis of the left knee, and was diagnosed with TA two years ago. His CRP and ESR levels were found elevated. The angio-CT imaging revealed that 70% stenosis in the 4 cm segment of the abdominal aorta, 70% stenosis in the right renal artery, 50% stenosis in the left renal artery and 90% stenosis in the origin of celiac trunk and superior mesenteric artery. Despite the given bolus methylprednisolone and methotrexate therapy, there had not seen any improvement in disease activity.
Tocilizumab was started at 8 mg/kg monthly in all three patient and they achieved complete clinical and laboratory remission.
Conclusion: These cases demonstrate that the TCZ treatment has been shown to be a successful option in TA patients resistant to conventional immunosuppressive therapies.
Disclosures: The authors have declared no conflicts of interest.
Informed consent form was obtained from patients and their legal guardians

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AB0960 THE HELIOS (HACETTEPE UNIVERSITY ELECTRONIC RESEARCH FORMS) REGISTRY: USE OF BIOLOGIC DRUGS IN AUTOINFLAMMATORY DISEASES

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Background: Autoinflammatory diseases (AID) are characterized by a dysregulation of innate immunity leading to uncontrolled inflammation. The treatment in AID is critical to control the disease activity, to prevent complications, and to improve the health-related quality of life. Biologic drugs have revolutionized the treatment and outcomes in AID.
Objectives: Herein we aim to present the clinical characteristics of children to whom biologic drug therapy was initiated for the management of AID.
Methods: A web-based registry called the Helios Registry (**Hacettepe univErsity eLectronIc research fOrmS) has been formed to evaluate the data of all children on biologic treatment. We have been enrolling patients since August 2018 retrospectively and prospectively. We have analyzed the data about the general characteristics of the patients, treatment, the biologic drug used, and adverse effects. Only the patients with the following diagnoses were included: systemic juvenile idiopathic arthritis (SJIA), familial Mediterranean fever (FMF),** cryopyrin associated periodic syndrome (CAPS), and chronic recurrent multifocal osteomyelitis (CRMO).
Results: Of 60 patients included, 19 had FMF (31.7%), 24 had sJIA (40%), 10 had CAPS, (16.7%), and 7 had CRMO (11.7%). Their median age was 10.7 (2-20) years old and disease duration was 2.8 (0-6) years, at the time of biologic drug initiation. 58.3% were currently on canakinumab, 20% anakinra, 10% tocilizumab, 10% etanercept, and 1.7% adalimumab. 63.3% of our patients had previously used at least one other biologic drug. The rate of glucocorticoid use before biologic treatment was 56.6%. The median duration of glucocorticoid treatment after initiating biologic drugs was 7.4 months. 56 (93%) patients achieved remission on biologic therapy. There were 15 patients (25%) who received tuberculosis prophylaxis due to positive tuberculin skin test (diameter$\geq$10 mm) and there was no Quantiferon test positivity. Thirteen adverse events (AE) had been noted. 2 of them were serious events as anaphylaxis due to tocilizumab infusions. The rest of the adverse events were mild thrombocytopenia (n=2), varicella infection (n=1), and local side effects (n=8). The median number of the infections per year was one and there were no death or malignancy.

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-ALPHA 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-a 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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AB0962 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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AB0963 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