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 (16.7%), 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 choice in medical treatment, steroids and steroid-sparing immunosuppressive agents, may sometimes fail to prevent to progression of the disease. Interleukin (IL)-6, which synthesizes 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.

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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 ≥ 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.