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Disease activity states, reasons for discontinuation and adverse events in 1038 Italian children with juvenile idiopathic arthritis treated with etanercept

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Introduction: The advent of biologic medications has increased considerably the potential for treatment benefit in juvenile idiopathic arthritis (JIA), with clinical remission being now achievable in a substantial proportion of patients. However, there is a need of data from the real world of clinical practice to evaluate thoroughly the efficacy and safety profile of the biologic agents currently approved.
Objectives: To evaluate the outcome of etanercept (ETN) therapy in Italian children with JIA.
Methods: This is a multicenter, observational study that includes all children with JIA who were given ETN at Italian pediatric rheumatology centers after January 2000. Based on the status of ETN therapy at study start, patients were classified in 2 groups: patients still receiving ETN (Group 1); patients discontinued from ETN (Group 2). Patients in Group 1 underwent both retrospective and cross-sectional assessments, patients in Group 2 only retrospective assessments. Outcome of ETN therapy was assessed as follows: a) in Group 1, by evaluating the state of disease activity at cross-sectional visit through formal definitions or JADAS10 and cJADAS10 cut-offs; b) in Group 2, by evaluating reasons for ETN discontinuation. ETN-related side effects were recorded in both groups.
Results: A total of 1038 patients, 422 in Group 1 and 616 in Group 2 were enrolled in the study. Median treatment duration was 2.5 years in Group 1 and 2.6 years in Group 2. In group 1, the frequency of inactive disease (ID) by Wallace criteria was 41.8% and the frequency of low (or minimal) disease activity (LDA) by Magni-Manzoni criteria was 63.6%. The frequency of ID, LDA, moderate disease activity, and high disease activity on JADAS10 was 46.4%, 17.5%, 24.4%, and 11.6%, respectively. The frequency of the same disease activity states on cJADAS10 was 48.6%, 9.7%, 27.7%, and 14%. In Group 2, reasons for ETN discontinuation included disease remission (52.4%), lack of efficacy (29%), and side effects (21.4%).Serious adverse events were seen in 18 patients of the entire study population and included inflammatory bowel disease (10 pts), tuberculosis (1 pt), varicella complicated by purpura fulminans (1 pt), CMV hepatitis (1 pt), acute pancreatitis (1 pt), papilledema (1 pt), bladder carcinoma (1 pt), thyroid carcinoma (1 pt); 1 patient died of streptococcal sepsis.
Conclusion: Around half of the patients treated with ETN achieved clinical remission and another 10-15% had low disease activity. Serious adverse events were rare and were mostly represented by the development of inflammatory bowel diseases.
Disclosure of Interest
None Declared.

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Successful treatment refractory pediatric takayasu's arteritis with anti-interleukin 6 receptor monoclonal antibody (tocilizumab): a case report

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Introduction: The goals of medical therapy of patients with Takayasu arteritis are to control active inflammation and to normalize clinical and laboratory parameters while preventing further vascular damage. Corticosteroids and conventional immunosuppressive agents are not always safe or efficacious. The complex formed by interleukin-6 (IL-6) and soluble IL-6 receptor appears to play a pivotal role in the pathogenesis of TA.
Objectives: Herein, we report a child with TA to share the efficacy and safety of tocilizumab.
Methods: Case report
Results: A- 14-year old girl who fulfilled the classification criteria for TA was identified. The interval from first symptom onset to diagnosis was 7 months. PVAS at presentation was 13; DEI.Tak was 10 and ITAS2010 was 10. She consisting of thickening of the aortic arch, descending aorta and superior mesenteric artery wall, obstruction of the right main carotid artery and superior mesenteric artery failed to respond to corticosteroids, methotrexate, and cyclophosphamide. After 1 year, she had severe manifestations (blurred vision and severe headache) and elevated both CRP and ESR the disease, and stenosis of the left vertebral artery in MRI angio which was considered as relapse. The patient started TCZ infusions (8 mg/kg for 2 weeks), and a rapid clinical remission was observed, associated with a drastic reduction of inflammatory markers. Corticosteroids were withdrawn, the patient's weight and height improved. 2 months later, TCZ infusions were extended, with no significant side effects. Ischemic manifestations resolved, and new lesions were not observed in MRI during 8 months on TCZ. At the last follow-up, PVAS was 0, ITAS2010 was 0 and DEI.Tak was 0. ESR was 5 mm/hr, and CRP was 3 mg/dL.
Conclusion: TA in children is a rare but potentially life-threatening condition. The treatment of TA in children is weak, it is essential to treat TA aggressively as soon as the diagnosis is secured to reduce mortality and morbidity. TCZ appears to be effective in the management of patients with TA, in particular in patients refractory to corticosteroids and/or conventional immunosuppressive drugs.
Trial registration identifying number: -
Disclosure of Interest
None Declared.

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Efficacy of rituximab in childhood-onset systemic lupus erythematosus: a retrospective study of 16 patients

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Introduction: Systemic lupus erythematosus in children (juvenile-onset SLE, jSLE) has an unpredictable course and a more severe phenotype compared to adults. There are no standards for treatment of jSLE now. Several studies of B-cell targeted therapy with rituximab (RTX) in adults have demonstrated its high efficiency in various rheumatic diseases. Robust data on the use of rituximab in jSLE are still lacking and dependent on small cohort studies.
Objectives: To evaluate the efficacy and safety of rituximab in children with jSLE.