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Objective: A questionnaire form was prepared by the Turkish Pediatric Hematology Society- Subcommittee of Hemostasis, Thrombosis and Hemophilia to determine the current approaches in the diagnosis and treatment of childhood ITP in our country. Our aim was to share the results of this study, and to do new, national, multicenter prospective studies. **Methodology:** This form, which consists of twenty questions with multiple choices, but a brief explanation is requested when there is a different approach other than the options given, was sent to all pediatric hematologists via e-mail. **Results:** The response was obtained from 55 hematologists experienced in ITP from 47 centers in total. Due to space constraints, this summary could not present the survey questions and answers. **Conclusion:** In conclusion, the approaches for diagnosis and management of childhood ITP differ between centers.

https://doi.org/10.1016/j.htct.2021.10.1070

PP 61

THE CLINICAL PICTURE AND LABORATORY WORK-UP OF GLANZMANN THROMBASTHENIA

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Case report: We present the clinical picture and laboratory work-up of Glanzmann thrombasthenia, based on a group of 7 patients. Bleeding history was significant in all patients and included both mucosal and postsurgery bleeds. Laboratory analysis revealed decreased or absent platelet aggregation (< 10%) with all physiologic agonists (ADP, collagen, epinephrin, arachidonic acid) together with normal agglutination response to ristocetin. In three patients diagnosis was confirmed by flow cytometry.

https://doi.org/10.1016/j.htct.2021.10.1071

PP 62

THE USE OF ROMIPLOSTIM IN AN INFANT

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Case report: Immune thrombocytopenia (ITP) is the most common platelet disorder in children, peaking between the

ages of 1-7.The first line therapy consists of intravenous immunoglobulin, anti-D immunoglobulin or corticosteroids. Second-line treatment options are immunosuppressive therapy, Rituximab.Thrombopoietin receptor agonists are used, which increase platelet production in the bone marrow. Our case report on a child with refractory chronic ITP, who failed the first and second line therapy.

https://doi.org/10.1016/j.htct.2021.10.1072

PP 63

ESSENTIAL THROMBOCYTOSIS IN CHILDREN

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Objective: Sporadic essential thrombocytosis is a very rare disease in the childhood age group and its frequency has been reported as 1/1,000,000. WHO 2008 essential thrombocytosis diagnostic criteria; high platelet count for more than one year (>450 × 109/l), exclusion of reactive or secondary causes of thrombocytosis (iron deficiency, megaloblastic anemia, acute phase reactants, trauma, operation), no family history of myeloproliferative neoplasm and thrombocytosis, and WHO It can be summarized as the absence of myeloid neoplasm criteria. **Methodology:** In this study, seven cases diagnosed as sporadic essential thrombocytosis in our Pediatric Hematology clinic are presented. Six of the patients were girls and one was a boy. The median age at presentation was 13 years (the youngest 5 months, the oldest 15 years old). Application complaints: Headache, vertigo and tinnitus in adolescent children were not present in young children, they were detected incidentally. Thrombus was not detected in any patient. The median platelet count at diagnosis was 1442 × 109/l (range 963- 2438). **Results:** An increase in megakaryocytes was detected in bone marrow aspiration, no cytogenetic anomalies were found. Jak-2 (V617F) mutation was detected in one case and CALR mutation in two cases. No MPL (W515L) mutation was found in any case. In one case with a CALR mutation, a known type 2 mutation was detected, and in the other a new, previously unidentified mutation was detected. In the other four cases, no clonality was detected. Three cases with mutations and two cases with no mutations are being followed up with hydroxyurea therapy. The other two cases are using low-dose aspirin. Follow-up periods range from six months to nine years. No complications developed. **Conclusion:** Thrombocytosis is a common problem in childhood. Reactive and secondary causes are usually identified. Essential thrombocytosis is a diagnosis that should be considered after excluding other causes. Mutation studies should be performed in pediatric patients who meet the WHO 2008 criteria. While Jak-2 (V617F), CALR and MPL (W515L) mutations are seen in 90% of cases in adults, these three mutations are only seen in 25% of the childhood age group. The high number of cases with no mutations indicates that new candidate genes should be sought and studied.

https://doi.org/10.1016/j.htct.2021.10.1073

RED BLOOD CELL DISORDERS

PP 64

GARDNER DIAMOND SYNDROME: A CASE REPORT

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Case report: Gardner Diamond Syndrome (GDS) is a rare autoimmune disorder also known as autoerythrocyte sensitization syndrome represented with skin lesions. These lesions mostly occur after a triggering factor. The pathophysiology of the disease is not completely understood yet. In this case report, the characteristic features of GDS is presented; furthermore, our aim is to emphasize the effect of emotional stress during the disease.

https://doi.org/10.1016/j.htct.2021.10.1074

IMMUNODEFICIENCIES / NEUTROPHIL DISEASES

PP 65

MYELOPEROXIDASE DEFICIENCY: SINGLE CENTER EXPERIENCE

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Objective: Myeloperoxidase (MPO) is a hemoprotein expressed in azurophilic granules of neutrophils and lysosomes of monocytes. It is caused by mutations in the MPO gene on chromosome 17 and is estimated to affect 1:2000-4000 people. It is the most common inherited defect of phagocytes. Microbial killing is impaired in patients with MPO deficiency, but most patients are asymptomatic, except for diabetic patients. In this article, we aimed to present our patients diagnosed with primary MPO deficiency. **Methodology:** During the investigation for the etiology of neutropenia in the hematology department of our hospital, patients who were diagnosed with MPO deficiency were examined. In the evaluation of the patients, it was observed that the neutrophil count in the hemogram printout and the counted neutrophil count in the peripheral smear were inconsistent. We performed MPO staining with FCM from the peripheral blood samples of the patients and we found that the neutrophils were MPO negative. **Results:** A 1-day-old male patient has no additional disease (c.608A>C H mutation). C.578G>C mutation was detected in the follow-up due to ANA+ in a 6.5-year-old female patient. A c.2031-2A>C mutation was found in the 18-year-old patient who was being followed up with the diagnosis of Granulamatous Polyangiitis and his sister. A c.493del mutation was detected in an 11-year-old patient who was diagnosed with ITP 5 years ago. The noval mutation was detected in the patient followed up with the diagnosis of