Methods: Sixty three children with newly diagnosed JIA (46 girls and 17 boys) aged 1.5-17 years and 32 healthy children as a control group appropriately matched in terms of sex and age, hospitalized in the Department of Pediatric Cardiology and Rheumatology, Medical University of Lodz due to functional disorders of the cardiovascular system were included into the study. Concentration of survivin was assessed by an ELISA test in blood and 18 matched synovial fluid samples collected from JIA patients and in sera of children from control group.

Results: Children with JIA were divided according to the subtype of the JIA. In 68.2% of patients oligoarthritis was diagnosed. The disease activity was established on the basis of JADAS-27 criteria, distinguishing three levels of rheumatoid process activity: low, medium and high. The largest group comprised children of low activity (56%). The concentration of survivin was significantly higher in JIA compared to the controls ($p=0.00041$). Higher concentration of survivin correlated positively with the presence of anti-CCP antibodies ($p=0.004$) and with higher disease activity ($p=0.0014$). The higher survivin concentration was found more often in polyarticular and systemic JIA onset. In all synovial fluid samples the concentration of survivin was higher than in matched blood ($p=0.0002$).

Conclusion: On the basis of the conducted study, it could be concluded that the higher concentration of survivin is being associated to more severe course of the JIA. Determination of survivin may be helpful in the diagnosis of JIA and may be used to identify a group of children with early arthritis and JIA with bad prognosis who should be treated more aggressively early from the disease onset.

REFERENCES:

- [1] Galeotti L, Adrian K, Berg S, Tarkowski A, Bokarewa M. Circulating survivin indicates severe course of juvenile idiopathic arthritis. *Clin Exp Rheumatol*. 2008;26(2):373-8.
- [2] Erlandsson MC, Turkila M, Siljehult F, Pullerits R, Eriksson C, Rantapää-Dahlqvist S, Bokarewa MI. Survivin improves the early recognition of rheumatoid arthritis among patients with arthralgia: A population-based study within two university cities of Sweden. *Semin Arthritis Rheum*. 2018;47(6):778-785.

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A NATIONAL SURVEY ACROSS GENERAL PAEDIATRICIANS REGARDING IMMUNIZATION PRACTICES IN CHILDREN WITH RHEUMATIC DISEASES

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Background: Robust scientific evidence on the safety and efficacy of certain vaccines in patients with rheumatic diseases is available; therefore guidelines regarding immunizations in this vulnerable group of children are published via a variety of official health organizations/societies including the Greek Rheumatology Society. Nonetheless, the uptake of specific immunizations is suboptimal. In Greece, vaccinations are mainly delivered by primary care Paediatricians.

Objectives: The purpose of this study was to describe the knowledge, attitude and current practice of General Paediatricians working in primary care regarding vaccination in children with rheumatic diseases on immunosuppressive medication across Greece and to identify barriers and facilitators that could be used to promote uptake.

Methods: This was a cross-sectional survey, conducted with an anonymous questionnaire of 25 items distributed to Paediatricians via an online platform. Data collected included demographics, questions on knowledge, perceptions and opinions as well as advice given to families. Additionally, questions addressed three specific categories: live-vaccines, non-live vaccines and annual influenza vaccine. This study was approved by the "P. & A. Kyriakou" Children's Hospital Research and Ethics Committee.

Results: Out of 400 questionnaires sent out, 256 were returned. Mean age was 48 years age (± 8.2) mean duration of working as a Paediatrician was 15 years (± 6.7). 25% of the responders worked in rural areas and the remaining in urban areas. 67% worked in the private sector. The majority (78%) of doctors felt that vaccination in children with rheumatic diseases is of pivotal importance. 50% gave specific advice on immunisations at initial diagnosis and 25% checked vaccination status at regular intervals. Responders were using a variety of guidelines in order to reach a clinical decision; still 45% were unaware of the existing national guidelines. 50% were hesitant to adhere to the national

vaccination scheme without expert input. Reasons were: not convinced from current literature that the vaccine is safe (32%), afraid to cause disease flare (43%), unable to deal with parental concerns/refusal (54%). 12% of responders felt that the rheumatic disease may have been triggered by a vaccine. The majority (95%) were pro annual influenza vaccination, while a minority (15%) was against live vaccines administration even if the patient was not on immunosuppressive treatment. 75% of doctors were keener to administer booster doses rather than primary ones, while 75% of respondents would postpone vaccinations in all cases if disease was active.

Conclusion: Variation in opinion and clinical practice exists. Overall, although Greek Paediatricians are well informed regarding efficacy and side effects of immunizations, there are steps to be made form principle to practice. Further research will allow the development of clear guidelines to aid in the management of increasing numbers of children with rheumatic diseases.

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PSTPIP1- ASSOCIATED MYELOID-RELATED PROTEINEMIA INFLAMMATORY SYNDROME/PAMI SYNDROME: CASE REPORT AND REVIEW OF THE LITERATURE

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Background: PAMI syndrome is a recently described condition, previously known as Hyperzincemia/Hypercalprotectinemia (Hz/Hc) syndrome. It is a very rare auto-inflammatory disorder characterized by a chronic systemic inflammation, cutaneous and osteo-articular manifestations, hepatosplenomegaly, anemia and neutropenia. Increased blood levels of MRP 8/14 (S100A8/A9 or calprotectin) and zinc distinguish this condition. Specific pathogenic mutations in PSTPIP1 gene (p.E250K and p.E257K) were identified as the genetic cause of this condition.

Objectives: Case presentation and review of literature

Case Presentation: We report a case of 13 months age female referred to our unit for recurrent episodes of osteoarthritis. Physical examination showed hepatosplenomegaly. Blood work revealed a systemic inflammation, a microcytic anemia and neutropenia. A complete workup for metabolic disorders, oncologic processes and uncommon infections was negative. Because of history of recurrent osteoarthritis, a whole-body MRI was performed and confirmed a multifocal osteomyelitis. Whole exome sequencing identified the missense p.E250K in the PSTPIP1 gene.

Methods: A literature search on PAMI syndrome was performed until the 15 October 2018. Pubmed was screened using a combination of the following terms: Hyperzincemia, Hypercalprotectinemia, E250K mutation, PSTPIP1 mutation, PAPA with E250K mutation.

Results: We identified 20 cases of PAMI syndrome in the literature. PAMI syndrome is an early onset inflammatory disease with a median age of 2.4 years. Clinical manifestations include Osteo-articular manifestations (80%), skin lesions (71%), splenomegaly (89%), hepatomegaly (68%), lymphadenopathy (42%), growth failure (58%) and hemorrhagic diathesis with recurrent epistaxis and/or haematoma tendency in 5 patients. All cases had relevant abnormalities in hematologic parameters: mild to severe neutropenia and anemia (100%). Thrombocytopenia (42%). Systemic inflammation was confirmed in 94% using the monitoring of CRP, ESR or SAA. Zinc and MRP 8/14 blood concentrations were markedly elevated in all tested patients. Genetic analyses of PSTPIP1 gene revealed the two specific identified mutations (p.E250K and p.E257K) in all patients. Response to the treatment was variable with no consistently effective therapy. Most common therapeutic options were AINS, Corticosteroids (n=9), Anakinra (n=9), Anti-TNF (n=6) and Cyclosporine A (n=4).

Conclusion: PAMI syndrome is a rare auto inflammatory condition which should be considered in patients with undefined systemic inflammation and neutropenia, even without skin or osteo-articular manifestations. Zinc and serum MRP 14/8 measurement may be helpful tools for the diagnostic orientation in these cases.

REFERENCES:

- [1] HOLZINGER D, FASSL SK, de JAGER D, et al.: Single amino acid charge switch defines clinically distinct proline-serine-threonine phosphatase- interacting protein 1 (PSTPIP1)- associated inflammatory diseases.
- [2] E. BELELLI, C.PASSARELLI, M.PARDEO, et al: Haematological involvement associated with a mild autoinflammatory phenotype, in two patients carrying the E250K mutation of PSTPIP1