

Rheumatology Abstracts

Evaluation of immunological response in children with rheumatological disorders

Aghighi Y¹, Atarod L¹, Khedri F¹, Raeeskarami SR^{1,2}

1. Department of Pediatrics, Tehran University of Medical Sciences

2. Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences

Background: Juvenile rheumatoid arthritis (JRA) is the most common chronic pediatric arthritis and rheumatologic disease during childhood. The relation between rheumatologic disorders and autoimmune diseases has been shown. The aim of this study was to assess the immunologic responses in children with JRA.

Methods: In a case series study, 24 patients with JRA referring to Imam Khomeini pediatric rheumatologic clinic during 2005, whom has not been treated, were enrolled. In all the patients complement tests (including C3, C4 and CH50), humoral immune system tests (including IgM, IgE, IgG, IgA, ASO and ISO Hemagglutinin antibody titers) and cellular immune system tests (including lymphocyte count, Td and NBT) were performed.

Findings: The mean age was 7.15±4.43 years. The most frequent age group was 4 years old with 6 patients (25%). 54.17% were males. In 20.83%, C3 level mildly increased. In one case (4.17%), C4 level has a slight decrease. CH50 level was normal in all cases. In 3 cases, the final diagnosis was not JRA (2 with Broton disease, one with ataxia telangiectasia and one with Hyper IgM). Excluding one case, almost in all of them a decrease in three of four immunoglobulins were reported and with the rest of the patients the immunoglobulins' titer was normal or slightly increased. Decreased in ASO titer was also reported in two cases. ISO Hemagglutinin antibody titer decreased in two cases with final diagnosis of Broton disease and was normal in the rest of the patients with JRA. The lymphocyte count, PPD, Td and NBT results were normal in all the patients.

Conclusion: In patients with JRA, an increase in C3 level and alternate raise in immunoglobulins' titer, especially IgG, were reported.

Keywords: Juvenile Rheumatoid Arthritis, Complement, Humoral Immune System, Cellular Immune System

Juvenile idiopathic arthritis, yesterday, today and tomorrow

Shafaie N

Rheumatology Research Center, Tehran University of Medical Sciences

The most common chronic arthritis in children younger than 16 is now termed "Juvenile Idiopathic Arthritis" (JIA). This is an umbrella term for arthritis of unknown etiology, appeared in seven different subtypes, when other causes of arthritis are excluded.

Yesterday: Chronic arthritis in children was described as Juvenile Rheumatoid Arthritis (JRA) based on terminology of Rheumatoid Arthritis (RA), since 1800s. Heterogeneity in different pictures of inflammatory arthritis in children

and the differences with adult RA led to the consensus to call the disease "JIA".

Today: Different pictures and unknown etiology in the absence of pathognomonic laboratory/imaging diagnostic tools are challenging for the therapeutic decision-making which is the major goal for the patient management. The main aims of proper treatment include control of pain, restoration of range of motion and function and promotion of normal growth and development for the child to achieve a normal Quality of Life (QOL) as his/her peers.

Tomorrow: To achieve this goal, we need to thoroughly examine the patient and status of the disease (Health-Related Quality of Life, HRQOL) comparing to healthy subjects of the same age and sex. Different assessment tools have been developed and validated to evaluate the HRQOL of JIA patients, including generic (CHQ, CHAQ) and disease-specific (JAFAR) tools. The most recent tool is a questionnaire developed by PRINTO (Pediatric Rheumatology International Trials Organization), named JAMAR (Juvenile Arthritis Multidimensional Assessment Report), prepared in two versions (Parent & Patient). The questionnaires comprise 110 items within 16 domains. JAMAR is being internationally validated and cross-culturally adapted in 47 countries (including Iran) in an international study (EPOCA) which entitles the completion of JAMAR by JIA patients/parents and a case report form by physician for global assessment of patient's status. We recommend the JAMAR to be implemented in the routine practice for the children with JIA.

Keywords: JIA, JAMAR, Idiopathic Arthritis

Assessment of urinary function disorder in patients with hypermobility

Aghighi Y¹, Hajizadeh N¹, Tavakoli HM¹, Raeeskarami SR^{1,2}

1. Department of Pediatrics, Tehran University of Medical Sciences

2. Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences

Background: Voiding dysfunctions include a wide range of urinary signs and symptoms. It is not only restrict to urinary system, but also signs include gastrointestinal tract symptoms. Constipation is the main symptom that it plays important role in enhancement of symptoms. Generalized joint hypermobility is a multisystemic disease that its pathology is not exactly clear. It associates with a lot of symptoms and diseases such as voiding dysfunction that it is seen in these patients more than normal general population. The goal of this study was to find out prevalence of voiding dysfunction in the patients with generalized joint hypermobility (GJH) and compare that with normal group.

Methods: 43 cases of GJH and 63 normal persons were selected. The age range of persons under study were 3-12 years in both groups. In the GJH, 11 persons were males (25%) and 32 persons were females (74.4%) and in controls, there were 25 males (39.7%) and 38 females (60.3%). The ratio of F/M in GJH was 1.5 and in control group was 2.9. The principle variable examined was

voiding dysfunction. The persons that had score equal to 9 or more related to voiding dysfunction (VD) in questionnaire's questions was distinguished as VD patients.

Findings: According to gained score, 6 patients from control group (9.5%) and 17 patients from GJH (30.5%) had VD. The difference between score of these two groups statistically was significant ($P < 0.001$, odds ratio=6.2). Another variable that was different between two groups and was significant included squatting state, urgent feeling to voiding, night incontinency, disrupting during voiding and strain during voiding. Day time urinary incontinence, pain during voiding and constipation were the variables that their difference between two groups were not significant.

Conclusion: This study revealed that voiding dysfunction in GJH is more than general population, so evaluation of the dysfunctional symptoms in GJH is recommended.

Keywords: Generalized Joint Hypermobility, Voiding Dysfunction, Constipation, Night Urinary Incontinence

CGD associated with SLE

*Javadi Parvaneh V, Shiari R, Babaei D
Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences*

Background: Chronic granulomatous disease (CGD) is a primary immune deficiency that characterized with recurrent infections and granuloma formation. Although a variety of autoimmune disorders such as inflammatory bowel disease, discoid lupus erythematosus, juvenile idiopathic arthritis, autoimmune hemolytic anemia, antiphospholipid syndrome and IgA nephropathy has been associated with CGD, a few cases of systemic lupus erythematosus (SLE) have been reported.

Case Presentation: We report an adolescent girl presented with prolonged fever, malar rash, photosensitivity, generalized maculopapular rash, pancytopenia, splenomegaly, elevated antinuclear antibody and anti-double strand DNA, abnormal nitro blue tetrazolium test and flow cytometry-based dihydrorhodamine 123 assay with diagnosis of chronic granulomatous disease and systemic lupus erythematosus.

Keywords: chronic granulomatous disease, systemic lupus erythematosus, autoimmune disorders

The frequency of malignancy in children presenting with musculoskeletal manifestations

*Malek A, Aghighi Y, Raeskarimi SR, Shafaie N, Oloomi Z
Department of Pediatrics, Vali-e-Asr Hospital, Faculty of Medicine, Tehran University of Medical Sciences*

Background: Musculoskeletal (MSK) pain is a frequent complaint in children. Although most of these cases are benign, malignancy especially hematologic cancers are frequently manifesting with MSK symptoms. Hematologic malignancies and neuroblastoma have many similar clinical pictures with juvenile idiopathic arthritis (JIA). This study determines the frequency of malignancies in children with MSK presentations.

Methods: Children with MSK symptoms who referred to Vali-e-Asr Hospital, pediatric rheumatology clinic between 2006-2012 were studied. Either the first diagnosis was JIA or the child did not have any definite diagnosis all of the children were undergone bone marrow aspiration or biopsy.

Then the frequency of hematologic malignancies or JIA was evaluated and clinical and laboratory presentations were analyzed.

Findings: Overall 207 cases were entered the study. Eleven patients were diagnosed as hematologic malignancies (5.2%) and 134 cases had JIA (64%). All of malignant cases (8 boys and 3 girls) were recorded as leukemia. The incidence of malignancy was significantly higher in boys than girls ($P < 0.04$). Among malignant cases of the present study, the mean age of patients was 6.6 years (ranged between 2y, 3m to 11y) and peak age incidence was between 5 to 7 years. The most common clinical presentations were musculoskeletal pain (90%), fever (45%) and organomegaly (27%). Knee was the most involved joint (45%) and hip joint involvement was seen in 27% of patients. The most frequent laboratory findings were leukocytosis (63%) and thrombocytopenia (45%) respectively. Erythrocyte sedimentation rate 1st hour (ESR_{h1}) was between 36 to 121mm with mean of 72mm.

Conclusion: Because of high incidence of malignancy in children with MSK manifestations, especially in 5-7 year boys, bone marrow analysis is recommended in children with unusual constitutional symptoms such as fever, bone pain and abnormal cellular blood count prior to treatment of JIA.

Keywords: Musculoskeletal, Malignancy, Leukemia, Child

Osteoid osteoma; a report in children who referred to a rheumatology clinic

Mehregan FF¹, Moradinejad MH², Ziaee V^{2,3}, Panjavi B², Rahmani K¹

1. *Shahid Beheshti University of Medical Sciences*
2. *Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences*
3. *Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*

Background: Limping is a common complaint in childhood, accounting for 4 per 1000 visits in one pediatric emergency department. Limping is caused by benign and/or life-threatening conditions such as (infectious, inflammatory, neoplasm, mechanical and orthopedic causes). Osteoid osteoma is a relatively common benign tumor of bone that can present with a limping. The femur is the most common site of occurrence. Although osteoid osteoma is seen in all age groups, 70 % of cases occur in individuals < 20 years of age. Osteoid osteoma occurs twice as commonly in boys as in girls. The pain typically is nocturnal and aching, and responds briskly to NSAIDs therapy. The lesion may be visible as a lucency with a dense central nidus on plain radiographs or CT scan, or it may be apparent only on bone scan or MRI.

Case Presentation: Since 2010, five patients (4 boys and 1 girl) were referred to a rheumatology clinic in Children's Medical Center, Tehran, Iran with history of limping. The mean age of patients was 11 years. The most location of tumor was the neck of femur (4 patients) and in only 1 case, it was in proximal of tibia. All of them had history of a dull aching pain for weeks to months, which was worse at night and was relieved by aspirin or nonsteroidal anti-inflammatory drugs. Two cases were treated with NSAIDs medications. One case had operative treatment and 3 cases were treated with radiofrequency ablation under CT guide (after incomplete response to NSAIDs) with complete relief of pain up to now, without any complication.

Conclusion: Radiofrequency ablation can be considered as a safe treatment in osteoid osteoma in children.

Keywords: osteoid osteoma, limping, children, radiofrequency ablation

Autoimmunity in x-linked agammaglobulinemia; a patient with several episodes autoimmunity

Malekzadeh I¹, Moradinejad MH¹, Ziaee V^{1,2}, Malek A¹, Khalili A¹

1. *Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences*
2. *Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*

Background: Autoimmunity is very rare in X-linked agammaglobulinemia(XLA), although it is not common in primary antibody deficiency. Kawasaki, juvenile idiopathic arthritis(JIA) and macrophage activating syndrome(MAS) as presentation of XLA led us to report this case.

Case Presentation: Our patient was a 7.5 year old boy with a history of acute flaccid paralysis. The second episode of admission, when he was 3 years old, was due to fever, generalized maculopapular rash, cervical lymphadenopathy, and bilateral non-purulent conjunctivitis (Kawasaki disease). Five months after this episode, he was admitted with prolonged arthritis and fever as a systemic onset JIA case. During 2 years, almost all drugs had been tapered and he was in remission. He was admitted again in PICU with fever, dyspnea, and pancytopenia. After some paraclinical evaluation, he was treated with IVIg and methylprednisolone plus therapy with MAS diagnosis. Similar episode plus myocarditis was repeated after 9 months. Gram negative bacilli septicemia was detected in second attack. The immune system was evaluated 1 month after the first and second episodes of MAS and low immunoglobulins and 0 level of CD19 were detected in both. XLA (Bruton) was the final diagnosis and he is on IVIg, monthly. Moreover, he is on treatment with cyclosporine, methotrexate and prednisolone for polyarthritis and he is in remission on medication now.

Conclusion: XLA can be presented with various types of autoimmunity and it should be considered in a patient with several episode of autoimmunity.

Keywords: x-linked agammaglobulinemia, Bruton, autoimmunity, Kawasaki, Juvenile Idiopathic Arthritis

Presence of a rheumatologic and an immunodeficiency diseases in a child

Sadr T¹, Ziaee V^{1,2}, Moradinejad MH^{1,2}

1. *Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences*
2. *Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*

Background: Juvenile Idiopathic Arthritis (JIA) is one of the chronic diseases of childhood that its etiology and pathogenesis are unknown. JIA is the most common rheumatologic disease in children, however, its prevalence is not more than 401 in 100000 cases. Chronic Granulomatosis disease (CGD) results from a defect of phagocyte's function. With an incidence of 4-5 per one billion cases, it is known as a rare disease. The presence of these two diseases in a child ended to prepare this case report.

Case Presentation: The case was a 7 year old boy who was born from a related parents after two siblings dead because of myelomeningocele and anencephaly. He was 2 years old when suffered from bilateral hip pain and effusion that progressed to bilateral wrist and ankle pain. He was treated by methotrexate, hydroxychloroquine and prednisolone after the diagnosis of polyarticular JIA. Four years later, he presented with a cervical lymphadenitis which had not cured until after a surgical drainage was done. Two episodes of sinusitis and complicated pneumonia resulted to the impression of immunodeficiency that confirmed by the lab tests and the final diagnosis of CGD. Now, he is on medication with poor control and some deformities in wrists and ankles had occurred.

Conclusion: There is a few reports of concordance of rheumatologic diseases and immunodeficiencies. Diagnosis and evaluation of the similar cases may help us understand the exact pathogenesis of these concordances.

Keywords: Juvenile Idiopathic Arthritis, chronic granulomatosis disease, immunodeficiency

Periodic Fever: A Review on Clinical, Management and a guideline for Iranian patients

Ziaee V^{1,2}, Ahmadinejad Z², Mansori M³, Alijani N⁴

1. *Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*
2. *Department of Pediatrics, Tehran University of Medical Sciences, Tehran, Iran*
3. *Department of Infectious Diseases, Imam Khomeini Hospital, Tehran University of Medical Sciences*

Periodic fever syndromes are a group of diseases characterized by episodes of fever with healthy intervals between febrile episodes. The first manifestation of these disorders are presented in childhood and adolescence, but an infrequently it may be presented in young and middle age. Genetics base has been known for all type of periodic fever syndromes except periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA). Common periodic fever disorders are familial mediterranean fever (FMF) and PFAPA. In each patient with periodic fever, acquired infection with chronic and periodic nature should be ruled out. It is depend on epidemiology of infectious diseases. In Iran and other middle east countries, brucellosis, Malaria, infectious mononucleosis and borreliosis should be considered in differential diagnosis of periodic fever disorders especially with fever and arthritis manifestation. In children, urinary tract infection may be presented as a periodic disorders, urine analysis and culture is necessary in each child with periodic symptoms. Some malignancies such as leukemia and tumoral lesion should be excluded in each patient with periodic syndromes and weigh loss in each age and weight gain disorder or malaise in children. After exclude infection, malignancy and cyclic neutropenia, FMF and PFAPA are the most common periodic fever disorders. Similar other countries, Hyper IgD, CINCA, TRAPS and other autoinflammatory syndromes are rare causes of periodic fever disorders in our system registry of periodic fever in Iran. In this presentation, we review common periodic fever disorders regarding to the prevalence of them in Iran and present our periodic fever clinic algorithm in Children Medical Center for Iranian patients based on the epidemiology of periodic fever in Iran.

Keywords: Periodic Fever, Familial Mediterranean Fever, PFAPA, Iran

Epidemiology of Kawasaki disease in Golestan province, Iran, 2005-2012

Cheraghali F, Azadfar S, Hajimoradloo N, Roshandel G, Golsha R
Infectious Disease Research Center, Golestan University of Medical Sciences

Background: Kawasaki disease is an acute febrile systemic vasculitis with unknown etiology. The aim of this study was investigating the epidemiology of Kawasaki disease in Golestan province.

Methods: In a retrospective case series study, records of all patients with a diagnosis of Kawasaki disease from Sep 2005 to the end of Oct 2012 hospitalized in Taleghani hospital were reviewed. Data were analyzed with the SPSS 16. P values of less than 0.05 were considered significant.

Findings: 51 patients entered the study including 58/8% males and 41/17% females. Children were between 2 months and 10 years old and included 39 typical and 12 atypical Kawasaki. The mean age of typical and atypical was 32/1 and 15/2 months respectively; the mean time for the beginning symptoms before admission was 6/7 days in typical and 7/1 days in atypical. Time from admission to receive IVIG in typical and atypical was reported 25 and 116 hours. More clinical signs among typical cases were skin rash 76/9%, lips and oral mucosa change 66/6% and conjunctivitis 64/1% and in the atypical cases were rash 41/6%, cough 33/3, rhinorrhea 33/3% and diarrhea 33/3%. Among the laboratory findings in the typical group CRP was positive in 88/2%, ESR>35 in 79/5% and anemia in 63/2% cases and in the atypical group, CRP was positive in 91/7%, anemia in 91/7% and AST in 63/6%. Echocardiography was performed in 42 cases and 5 patients had abnormal findings that one case was atypical.

Conclusion: The clinical symptoms in typical and atypical Kawasaki were conjunctivitis, rash, desquamation, scaling, unilateral lymphadenopathy, lips and oral change; among laboratory findings in atypical cases, anemia was more common than typical cases. Cardiac complications were significantly lower than other studies.

Keywords: Kawasaki, children, IVIG

Periodic fever, Aphthous stomatitis, Pharyngitis, and cervical Adenitis Syndrome

Cheraghi T
Division of Pediatric allergy and clinical Immunology, Department of Pediatrics, Guilan University of Medical Sciences

Background: PFAPA syndrome is a subtype of periodic fever syndromes characterized by recurrent fever, associated with aphthous stomatitis and cervical adenitis. Some physicians think of immunodeficiency diseases as a predisposing factor. It is noteworthy that tonsilopharyngitis without other organ system infections is not characteristically a sign of primary immunodeficiency disorders. Etiology of this syndrome is unknown, but dysregulation of immune response to antigenic stimuli has been suggested.

Methods: We followed 7 patients with this syndrome. Possible differential diagnoses such as immunodeficiency syndromes including cyclic neutropenia, infectious tonsilopharyngitis, and other periodic fever syndromes, predisposing malignancy and known rheumatologic

disorders have been ruled out. We treated patients with a single dose prednisone at the onset of fever syndrome.

Findings: We could not find any immunologic or infectious cause, nor were there any other known cause for this syndrome. Our treatment with 1-2mg/kg of prednisone was associated with dramatic response and resolution of fever and other signs and symptoms.

Conclusion: when we encounter a recurrent tonsilostomatitis in a given child that is culture negative and resistant to antibiotics and acetaminophen and Non-Steroidal, Anti-inflammatory Drugs (NSAIDs), we should consider this syndrome and perform a cost-effective approach.

Keywords: PFAPA, Periodic Fever, Pharyngitis, Adenitis

Interleukin-4 single nucleotide polymorphisms in juvenile systemic lupus erythematosus

Tahghighi Sharbiani F¹, Ziaee V^{1,2}, Moradinejad MM¹, Rezaei A³, Soltani S³, Sadr M³, Aghighi Y⁴, Rezaei N^{1,3}

- 1. Growth & Development Research Center, Tehran University of Medical Sciences*
- 2. Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*
- 3. Molecular Immunology Research Center, and Department of Immunology, Tehran University of Medical Sciences*
- 4. Department of Pediatrics, Tehran University of Medical Sciences*

Background: Juvenile systemic lupus erythematosus (JSLE) is a chronic, recurrent multisystem inflammatory disease, caused by a combination of environmental events and genetic risk factors. As cytokines, including interleukin-4(IL-4), seem to have a role in the pathogenesis of JSLE, this investigation was performed to evaluate the associations of specific single-nucleotide polymorphisms (SNPs) of IL-4 gene in a case control association study.

Methods: Fifty nine patients with JSLE were recruited in this study as patients' group and compared with 140 healthy volunteers. Genotyping was performed for IL-4 gene at positions -1098, -590 and -33, using polymerase chain reaction with sequence-specific primers method.

Findings: Following alleles were found to be more common among patients with JSLE: C at -590 and -33 and T at -1098 of IL-4 gene ($P<0.001$; OR=4.6, $P<0.001$; OR=2.7, and $P<0.001$; OR=2.1, respectively). Moreover, frequency of following alleles were remarkably lower in patients with JSLE, in comparison with controls: T at -590 and -33 and G at -1098 of IL-4 gene ($P<0.001$; OR=0.2, $P<0.001$; OR=0.3, and $P<0.001$; OR=0.4, respectively). Additionally, significant positive associations for the following genotypes were recognized in JSLE cases, compared with controls: C/C and T/T at -33, C/C at -590 and T/T at -1098 of IL-4 gene ($P<0.001$; OR=5.3, $P<0.02$, $P<0.001$; OR=29.5, and $P<0.001$; OR=3.3, respectively), while following genotypes were less frequent among patients with JSLE: T/C at -33 and -590 and T/G at -1098 of IL-4 gene ($P<0.001$; OR=0.1, $P<0.001$; OR=0.03, and $P<0.001$; OR=0.3, respectively). Furthermore, we noticed an astonishing negative haplotypic association for JSLE for IL-4 (positions -1098, -509, -33) TTC, GCC, and TTT haplotypes ($P<0.001$). There was also a significant relationship between TCC haplotype (IL-4 gene at positions -1098, -590, -33) and having JSLE ($P<0.001$).

Conclusion: Cytokine gene polymorphisms may influence susceptibility to JSLE. Particular IL-4 gene variants are associated with JSLE and might have a role in pathophysiology of disease.

Evaluation of liver and renal abnormalities in Kawasaki disease

*Soleimani G, Sadeghi Bojd S, Tajik M
Zahedan University of Medical Sciences*

Background: Kawasaki disease(KD) is a vasculitis that affects multiorgan systems such as liver and kidney. The diagnosis of the Kawasaki disease is made by clinical criteria. Sterile pyuria, microscopic hematuria, and proteinuria due to renal involvement present in Kawasaki disease; liver abnormalities contain abnormal liver function tests, gallbladder hydrops, and hypoalbuminemia. The aims of this study were to define frequency of liver and renal abnormalities in patients with KD that hospitalized in Zahedan ali ebne abitaleb hospital in 2006-2013.

Methods: Paraclinical findings including serum and urine tests and gallbladder ultrasonography of 47 patients with KD hospitalized in Zahedan ali ebne abitaleb hospital from 2006 to 2013 were reviewed retrospectively.

Findings: Forty seven patients were studied; liver abnormalities were found in 22 cases(46.8%), impaired liver function tests were more common than other liver abnormalities in our study. The incidence of abnormal liver function tests in our studies was about 42%. Gallbladder hydrops were found in 9 patients(12.7%). Eighteen patients(38/3%) had renal involvement, sterile pyuria was the most common in our study, the incidence of sterile pyuria was 36/2%. Microscopic hematuria and proteinuria were rarely found in our study; also blood urea nitrogen and creatinin had normal levels in all of children.

Conclusion: Kawasaki disease should be considered in any infant or child with abnormal liver or renal function tests. Paraclinical findings of liver and renal involvement of our patients were partly similar to other articles we reviewed.

Keywords: Kawasaki disease, liver involvement, renal involvement, sterile pyuria, children

Epidemiology of Kawasaki disease in Golestan province, Iran, 2005-2012

*Cheraghali F, Azadfar S, Hajimoradloo N, Roshandel G, Golsha R
Infectious Disease Research Center, Golestan University of Medical Sciences*

Background: Kawasaki disease is an acute febrile systemic vasculitis with unknown etiology. The aim of this study was investigating the epidemiology of Kawasaki disease in Golestan province.

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Findings: 51 patients entered the study including 58/8% males and 41/17% females. Children were between 2 months and 10 years old and included 39 typical and 12 atypical Kawasaki. The mean age of typical and atypical was 32/1 and 15/2 months respectively; the mean time for the beginning symptoms before admission was 6/7 days in

typical and 7/1 days in atypical. Time from admission to receive IVIG in typical and atypical was reported 25 and 116 hours. More clinical signs among typical cases were skin rash 76/9%, lips and oral mucosa change 66/6% and conjunctivitis 64/1% and in the atypical cases were rash 41/6%, cough 33/3, rhinorrhea 33/3% and diarrhea 33/3%. Among the laboratory findings in the typical group CRP was positive in 88/2%, ESR>35 in 79/5% and anemia in 63/2% cases and in the atypical group, CRP was positive in 91/7%, anemia in 91/7% and AST in 63/6%. Echocardiography was performed in 42 cases and 5 patients had abnormal findings that one case was atypical.

Conclusion: The clinical symptoms in typical and atypical Kawasaki were conjunctivitis, rash, desquamation, scaling, unilateral lymphadenopathy, lips and oral change; among laboratory findings in atypical cases, anemia was more common than typical cases. Cardiac complications were significantly lower than other studies.

Keywords: Kawasaki, Children, IVIG, Vasculitis

Cytokine polymorphisms in juvenile systemic lupus erythematosus

*Tahghighi F¹, Rezaei N², Moradinejad MH¹, Ziaee V^{1,3}
1. Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences
2. Molecular Immunology Research Center, and Department of Immunology, Tehran University of Medical Sciences
3. Pediatric Rheumatology Research Group, Rheumatology Research Center, Tehran University of Medical Sciences*

Background: Juvenile Systemic Lupus Erythematosus (JSLE) is a multi-system autoimmune disorder of unknown origin. Given the importance of the contribution of pro-inflammatory cytokines including tumor necrosis factor- α (TNF- α) and Interleukin-6 (IL-6) toward the pathogenesis of JSLE, this investigation was performed to assess TNF- α and IL-6 gene polymorphisms in a case-control study.

Methods: Fifty nine children with SLE were enrolled in this study and were compared with 140 healthy control group. The frequency of alleles, genotypes of TNF- α single-nucleotide polymorphisms (SNPs) at positions -308 and -238 and genotypes of IL-6 SNPs at position+565 and -174 were evaluated using polymerase chain reaction with sequence-specific primers method.

Findings: The G allele was remarkably more frequent at position -238 in TNF- α promoter region in patients with JSLE than in the healthy controls($p<0.001$), while the frequency of A allele at the same position was significantly lower than among control($p<0.001$). Furthermore, a notable positive association for GG genotype as well as an outstanding negative association for GA genotype at the same position were detected in patients group compared with control subject($p<0.001$; and $p<0.001$:respectively). Frequency of GG genotype at position +565 in IL-6 promotore region in patients with JSLE was significantly lower than healthy control ($p<0.001$).

Conclusion: Certain TNF- α and IL-6 promoter polymorphisms are associated with the amount of TNF- α and IL-6 production and can be used as a genetic marker for susceptibility to JSLE.

Keywords: Systemic Lupus Erythematosus, TNF-A, Interleukin 6, Gene Polymorphisms

هیپرموبیلیتی در کودکان مبتلا به هرنی اینگوینال بررسی شیوع

رضا شیاری، شیما صالحی، محسن روزرخ، ودود جوادی
بیمارستان کودکان مفید، دانشگاه علوم پزشکی شهید بهشتی

زمینه: تحرک بیش از اندازه مفاصل (Joint Hyper Mobility) اولین بار توسط بقراط در افراد قبیله Scythians که در جنوب روسیه فعلی می زیسته اند شرح داده شد. این اصطلاح نخستین بار در اواخر قرن نوزدهم در توضیح و تشریح سندرم های مارفان و اهلرز دانلوس وارد فرهنگ پزشکی شد. هیپرموبیلیتی ژنرالیزه مفاصل در غیاب سایر بیماری ها در نسبت بالایی از افراد سالم نیز وجود دارد که بسته به سن، جنس، نژاد و متدولوژی در آمارهای مختلف از ۷ تا ۲۸/۵٪ گزارش شده است. بیماران مبتلا به هیپرموبیلیتی مفاصل برای ابتلا به مشکلات احشایی مستعدتر از سایر افراد هستند. این موضوع میتواند ناشی از ضعف ساختمان ها و بافت های نگهدارنده در بدن آن ها باشد. این موارد شامل فتق های شکمی و ورید های واریسی و ریفلاکس ادراری (RVU) می باشد. همراهی Joint Hyper Mobility با فتق هیاتال، دیورتیکولوم پیش از این گزارش شده است. از سایر مشکلات احشایی که همراه با نرمی مفاصل گزارش شده است می توان به پرولاپس های رکتوم و رحم اشاره نمود. هدف این مطالعه بررسی شیوع هیپرموبیلیتی در کودکان مبتلا به فتق اینگوینال بود.

روش: این مطالعه نوعی مطالعه توصیفی بود که در قالب یک مطالعه مورد - شاهد انجام شده است. در هر گروه ۶۷ کودک که از لحاظ سن و جنس متناسب بودند وارد مطالعه شدند. گروه مورد عبارت بود از بیماران مبتلا به هرنی اینگوینال و گروه شاهد عبارت بود از کودکانی که به صورت سرپایی به سایر درمانگاه های بیمارستان مفید مراجعه کرده و فاقد بیماری هرنی اینگوینال، روماتیسمی و ارتوپدیک بودند. به منظور تجزیه و تحلیل آماری از فراوانی و نسبت برای متغیرهای کیفی و از میانگین و انحراف معیار برای متغیرهای کمی استفاده شده است. مقایسه نسبتها با آزمون کای-دو و مقایسه میانگین ها نیز با استفاده از آزمونهای T انجام شد.

یافته ها: یافته های این مطالعه نشان داد که میانگین نمره بیگتون در گروه مورد $6/2 \pm 7/5$ با دامنه صفر الی ۹ نمره و در ۱۳ بیمار (۱۹/۴٪) کمتر از چهار و در ۵۴ بیمار (۸۰/۶٪) چهار و بیشتر بوده است. در گروه شاهد میانگین نمره بیگتون $1 \pm 1/8$ بوده است که به طور معنی داری از گروه مورد کمتر می باشد ($P < 0.001$).

نتیجه گیری: یافته های این مطالعه نشان داد که در گروه مورد ۸۰٪ بیماران نمره بیگتون ۴ و بیش از چهار بوده است و بر اساس این مطالعه دچار هیپرموبیلیتی بودند. در مقایسه با گروه شاهد این نسبت بسیار بالاتر بوده است. همچنین میانگین نمره بیگتون در بیماران گروه مورد بیشتر از گروه شاهد می باشد. این یافته ها این فرضیه را تقویت می کنند که میان هیپرموبیلیتی و هرنی اینگوینال می تواند ارتباط وجود داشته باشد به نحوی که هیپرموبیلیتی می تواند ریسک فاکتوری برای بروز هرنی اینگوینال باشد.

کلمات کلیدی: هیپرموبیلیتی مفاصل، فتق اینگوینال، نمره بیگتون، هرنی