Methods: Five hundred and sixty five (565) children were diagnosed with KD during the period from January 1993 to December 2016 in Pediatric Rheumatology Clinic, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India. Nine children had pulmonary presentation of KD. A retrospective case review with respect to clinical presentation, radiological findings and treatment was done.
Results: Pulmonary presentation of KD was seen in 1.6% patients. Mean age at diagnosis of KD was 2.9years (range 9months – 4 years). 77.8% patients had no features suggestive of KD either on history or at presentation. First sign of KD was noted at a mean duration of 17.6 days (range 6–28 days) from the onset of symptoms. Periungual desquamation was the most common clinical sign seen in 66.6% patients followed by erythematous rash and perianal desquamation in 33.3% patients each.
Persistent fever, thrombocytosis and elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were seen in all patients. Microbiological investigations showed evidence of infection in only 2 patients – methicillin sensitive staphylococcus aureus in pus and positive mycoplasma agglutinin titre in one patient each. Parenchymal consolidation was the most common radiological finding (100%) followed by pleural effusion (55.5%), empyema (33.3%) and pneumothorax (11.1%). Coronary artery abnormalities were evident on echocardiography in 22.2% patients with dilatation of right coronary artery and left main coronary artery in 1 patient each.
All patients received intravenous antimicrobials for pneumonia. Intravenous immunoglobulin (IVIG) at 2g/kg was given after a mean duration of 24 days of onset of fever. Fever, which was unresponsive to intravenous antimicrobials, responded to IVIG therapy in all except 2 patients which required a second dose of IVIG. 55.5% patients required intercostal drainage tube (ICDT) insertion, 2 patients required streptokinase and 1 patient each required video assisted thoracoscopic surgery (VATS) and decortication. Mean follow up period was 15.9 months (range 0–62 months).
Conclusions: Pulmonary involvement in patients with KD is uncommon and is less commonly recognized. Unresolving pneumonia in a child who continues to be febrile despite adequate antimicrobials with elevated inflammatory markers can be a clue towards the diagnosis of KD. Early recognition can prevent delays in diagnosis and shorten the hospital stay.
Disclosure of Interest: None declared
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THU0497 RHEUMATOLOGICAL MANIFESTATIONS OF X-LINKED AGAMMAGLOBULINEMIA: PROFILE OF 17 CASES FROM A TERTIARY CARE CENTER IN NORTH INDIA

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Background: X-linked agammaglobulinemia (XLA) is characterized by recurrent bacterial sino-pulmonary infections. Arthritis can be seen in 10–30% patients of hypogammaglobulinemia (1) and is usually a mono- or oligoarthritis of large joints, but polyarthritis has also been defined.
Objectives: We describe our experience of managing children with XLA who presented with rheumatological manifestations.
Methods: Sixty two children were diagnosed with XLA during the last 10 years in Primary Immunodeficiency Clinic, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India. Seventeen children had rheumatological manifestations. A retrospective case review with respect to clinical presentation, musculoskeletal findings and treatment was done.
Results: Rheumatological manifestations were seen in 27.4% patients of XLA. Thirteen out of 17 patients had a proven mutation in *Btk gene*. Mean age at symptom onset was 3.3 years (range 6 months – 13 years) and mean age at diagnosis of XLA was 5.6 years (range 1.5- 10 years). Rheumatological manifestations were seen at a mean age of 8.7 years (range 1.5- 20 years). In 2 patients, arthritis preceded the diagnosis of XLA while 10 patients developed rheumatological manifestations after the diagnosis of XLA. Arthritis as an initial presentation of XLA was seen in 5 patients.
Oligoarthritis was the most common presentation seen in 15 patients. Knee was the most commonly involved joint (11 patients) followed by ankle joint in 5 patients and shoulder, wrist and hip arthritis in 2 patients each. One patient each had arthritis involving proximal interphalangeal joints of both hands, clinical evidence of sacroiliitis and spondylodiscitis involving L4–5 and L5-S1 vertebrae. Three patients manifested as septic arthritis (fever, redness, warmth and pus in joint) involving shoulder, ankle and knee joint respectively.
Joint aspiration and/or arthrotomy had been performed in 8 patients, however cultures were sterile in all. Microbiological investigations for mycoplasma infection (viz. culture and polymerase chain reaction) in joint fluid/pus could not be performed due to unavailability. All patients received replacement intravenous immunoglobulin (IVIG - 400 mg/kg body weight) and co-trimoxazole prophylaxis. Eight patients received macrolide antimicrobials (erythromycin or azithromycin) for clinical suspicion of mycoplasma arthritis.
One patient expired during the course of his illness while four patients were lost to follow up. Arthritis showed clinical improvement after IVIG replacement therapy in remaining 12 patients with resolution of arthritis after first dose in 11 patients and after 6 months of replacement therapy in 1 patient. None of the patients had recurrence of joint symptoms on follow up, however radiological evidence of spondylodiscitis persisted in one patient.
Conclusions: Arthritis is a relatively frequent manifestation in patients with hypogammaglobulinemia. Family history and history of recurrent infections in a child with arthritis may provide a clue towards the diagnosis of primary immunodeficiency. Prompt institution of regular IVIG replacement therapy results in resolution of arthritis and prevents its recurrence.
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THU0498 PFAPA SYNDROME IN LARGE PEDIATRIC POPULATION: A SINGLE CENTER EXPERIENCE

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Background: Periodic fever, aphthosis, pharyngitis, and adenitis (PFAPA) syndrome is an auto-inflammatory condition of unknown etiology. It is the second most common auto-inflammatory disease in our country, following familial Mediterranean fever. Previous studies showed that tonsillectomy represents efficient treatment options.
Objectives: Our aim was to explore the main clinical features, response to tonsillectomy and long–term outcome of PFAPA pediatric patients in a single cohort. We assessed association of MEFV gene mutation with disease characteristics and treatment response.
Methods: We reviewed medical records of patients who were diagnosed with PFAPA syndrome between the January 2010 and June 2016. All of the recorded 562 patients were called by the telephone and 365 (65%) of them were reached. Demographic, clinical and therapeutically features were taken from the patients' medical records. Data on clinical course and the disease outcome were collected by using a structured questionnaire which was fulfilled during the phone conversation between investigator and patients parents.
Results: A total of 365 patients with PFAPA were examined: 154 (42%) of them were female. The mean age at disease onset, at diagnosis and at the investigation was 22.79±18.8, 41.7±21.7 and 77.14±31.35 months, respectively. The most common disease feature at the disease onset was: recurrent fever in 365 (100%), cryptic tonsillitis in 365 (100%) and aphtous stomatitis in 317 (88%). Sixty three (17%) patients met the criteria for both PFAPA and FMF. MEFV gene mutation analysis was performed in 93 (25%) patients and 51 of them (54%) had a heterozygous mutation in exon 10. Surgical treatment was performed in 158 (43%) patients. Complete clinical remission was achieved in 127 (80.3%) patients. Six (3%) showed no response to surgical treatment while 25 (15.8%) patients had a partial response. In patients with partial clinical response, frequency of fever attacks decreased significantly from 17.5 to 7.3 attack per year ($p<0.05$). Among patients who did not respond to tonsillectomy,11 (52.4%) were carrier of MEFV heterozygous mutation in exon 10. There was a statistically significant difference between patients with and without coexistence of FMF features, according to surgical treatment response ($p<0.05$).The mean age of resolution of PFAPA symptoms in patients who underwent tonsillectomy was 52±22.4 months and in patients without tonsillectomy 66±22.6 months.
Conclusions: Although PFAPA symptoms usually resolve before age of eight, some patients' complaints persist. FMF should be considered in tonsilloadenoidectomy unresponsive PFAPA patients, especially in endemic regions like Turkey. Tonsilloadenoidectomy seems to be an effective treatment option for pediatric PFAPA patients.
References:
[1] Vanoni F, Theodoropoulou K, Hofer M. PFAPA syndrome: a review on treatment and outcome. Pediatr Rheumatol Online J 2016;14:38.
[2] Batu ED, Kara Eroğlu F, Tsoukas P, et al. Periodic Fever, Aphthosis, Pharyngitis, and Adenitis Syndrome: Analysis of Patients From Two Geographic Areas. Arthritis Care Res (Hoboken) 2016;68:1859–1865.
[3] Celiksoy MH, Ogur G, Yaman E, et al. Could familial Mediterranean fever gene mutations be related to PFAPA syndrome? Pediatr Allergy Immunol. 2016;27:78–82.
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THU0499 ANTI-MULLERIAN HORMONE IN A COHORT OF YOUNG ADULT WOMEN WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile Idiopathic Arthritis (JIA) represents one of more common chronic disease of the childhood, affects young people before sixteen and persists into their reproductive years. It is reported in one study that fertility was not impaired in JIA women but potentially it can be compromised by pharmacological treatments such as the prolonged immunosuppressive therapy used in young women patients (1). The Anti-Müllerian Hormone (AMH) is secreted from granulosa ovary cells and serum levels of Anti-Müllerian Hormone are used as a measure of ovarian reserve, reflecting the number of primary follicles.

Objectives: The aims of this study were to evaluate AMH serum levels in a cohort of young adult women affected from JIA, to compare these levels between patients and healthy controls and to assess whether the presence of the disease and the influence of previous exposure to disease-modifying antirheumatic drugs (DMARDs) and of other disease parameters may affect the ovarian reserve.
Methods: Forty women with a diagnosis of JIA, aged 18 to 25 years and with regular menses, and 20 healthy women age-matched were evaluated. Anti-Müllerian Hormone serum levels were measured according to a 2-stage enzyme-linked immunosorbent assay (ELISA) technique using a commercially available kit (AMH Gen II ELISA; Beckman Coulter). Clinical and demographic characteristics, disease duration, previous and current therapies disease activity score on 44 joints (DAS), health assessment questionnaire (HAQ) were performed at the time of blood sample.
Results: JIA patients had a mean age of 21.4±3.2 years, a disease duration of 11.5±6.6 years, a DAS of 1.22±0.58, and 12 (30%) were smokers. No significant differences were found in our cohort of JIA and healthy subjects in AMH serum levels (5.6±0.4 vs 6.6±0.6 ng/ml, respectively, p=0.5).
Considering the JIA cohort, 23 patients (57.4%) were treated with methotrexate (MTX) for a mean period of 2.1±3.1 years and 20 (50%) with anti-TNF drugs for 4.3±2.3 years. Twelve JIA women (30%) were treated with both MTX and anti-TNF. No correlations were found between AMH serum levels and patients age (p=0.6), disease duration (years) (p=0.67) and duration of therapy with MTX (p=0.5) or anti-TNF (p=0.11).
Dividing JIA patients according to MTX use, no differences were observed between MTX users and non users patients in AMH levels (6.0±0.5 vs 5.1±0.5 ng/ml, respectively, p=0.22), age, disease duration and other clinical characteristics. Patients expose to anti-TNF had AMH serum levels tendentially higher then non users patients (6.3±0.6 vs 4.9±0.5 ng/ml, respectively p=0.07).
Conclusions: In our JIA group of young adult JIA women, ovarian reserve seems not be changed by the presence of the disease, the long disease duration and the use of immunosuppressive drugs. These findings could be important for adult JIA patients.
References:
[1] Ostensen M et al. J Rheumatology 2000;27:1783–7.
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THU0500 PHOTOBIOSTIMULATION OF CHRONIC CERVICAL PAIN IN JUVENILE POLYARTHRITIS

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Background: Chronic pain management in juvenile arthritis constitutes a special provocation not only for the medical doctors, but also for the patients and parents. Despite the extensive use of the biological agents with high efficacy, combined with the multimodal therapies, chronic pain still remains an important issue for the public health, with implications on the activities of daily living and the scholar performances. Lasers could be used for transmitting biological messages and initiating metabolic changes within living cells: no more pain, much more energy, will contribute to muscular and joints repair.
Objectives: Aim of the present study was to evaluate the effects of photobiostimulation combined with DMARDs in patients with juvenile polyarthritis and chronic cervical pain.
Methods: 62 patients diagnosed with juvenile polyarthritis (ILAR criteria), 11.4 years mean age were randomly divided in Group I (42 patients treated with local laser biostimulation and methotrexate), comparatively with a Group II - control (20 patients) treated with methotrexate and *placebo* laser, for a period of 9 months. Group I received local laser biostimulation with a dose of 2.5 J/cm^2 in 14 latero-cervical painful points, corresponding to the 7 cervical vertebrae, 1 point on the insertion of sternocleido-mastoidian muscle on the styloid processus, and 1 point on each loco-regional submandibular lymphatic ganglions, using a GaAlAs laser probe of 670 nm, 25 mW output power and a modulation frequency of 10 Hz. 45 joules were applied daily as laser treatment, 10 sessions per month, repeated 3 times, in the 9 months. For all the patients, the main medication was methotrexate in a dose of 0.6 mg/kg (maximum 20 mg) per week, steroids and symptomatic drugs, when necessary. Tutors and the older children have signed the informed consent. Measurement of the subjective pain was on VAS (0 – 100 mm), and objective pain was assessed with an electronic device, the Algometer Commender. The range of motion for flexion/extension, and the rotation to the left/right of the cephalic extremity was measured with Dualer IQ Inclinometer.
Results: In the end of study, the level of chronic pain estimated with Commander Algometer decreased by 74.6% in Group I, compared with only 41% in the control Group, and the range of motion in the affected cervical segment assessed with Dualer Inclinometer increased by 66% in the first Group, compared with only 34% in *placebo* group.
Conclusions: Laser biostimulation proved to be an effective method for the complex management of chronic pain in juvenile polyarthritis.
References:
[1] Ravelli A, Martini A. Juvenile idiopathic arthritis. Lancet. 2007;369:767–778.
[2] Ravelli A, et al. Oral versus intramuscular methotrexate in juvenile chronic arthritis. Clin Exp Rheumatol. 1998;16:181–83.
[3] Ailioaie LM, Litscher G, Weber M, Ailioaie C, Litscher D, Chiran DA. Innovations and Challenges by Applying Sublingual Laser Blood Irradiation in Juvenile Idiopathic Arthritis. International Journal of Photoenergy 2014(2):1–8, June 2014. DOI: 10.1155/2014/130417.
[4] Ailioaie C, Lupusoru-Ailioaie LM. Beneficial effects of laser therapy in the early stages of rheumatoid arthritis onset. Laser Therapy.1999;Vol. 11–2, 79–87.
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THU0501 EFFICACY AND SAFETY OF METHOTREXATE AS MAINTENANCE THERAPY FOR CHILDREN WITH ANTI–N-METHYL-D-ASPARTATE RECEPTOR (NMDAR) ENCEPHALITIS: EXPERIENCE OF A SINGLE CENTER

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Background: Autoimmune-mediated encephalitis (A-ME) in children remains as a diagnostic and therapeutic challenge (1). These patients have a 12% risk of relapse, which is usually more severe (2). We previously proposed the therapy with methotrexate (MTX) for this condition (3), and we are offering now additional data on its potential benefits.
Objectives: To describe the outcome of children with A-ME receiving MTX for at least one year after stabilization of symptoms.
Methods: In this retrospective study we recruited 11 patients (7 females) with A-ME, a mean age of 7.5 years (range 8 months - 14 years), and with a median follow up of 22 months. In all cases, anti-NMDAR antibodies (subunit NR1) were detected in the CSF. Data from these patients were collected by consulting medical records. Relapse of encephalitis was defined as new onset of symptoms occurring after at least 2 months of remission, in the absence of other CNS disease.
Results: Patients presented with seizures (n=10), behavioral changes (n=11), psychosis (n=11), speech problems (n=10), and autonomic/breathing dysregulation (n=9). Patients were initially treated with methylprednisolone pulses (n=11), rituximab (n=6), intravenous immunoglobulins (n=4), cyclophosphamide (n=3) and MTX (n=11). Complete remission was observed in all cases, and maintenance therapy with MTX (10 mg/m^2 BSA) was started in all them, with gradual tapering until it was stopped. Interestingly, no relapses have been observed in any case during the mean follow up. One patient had mild oral ulcers and other showed mild elevation of liver enzymes; both events remitted after discontinuing the treatment for a couple of weeks.
Conclusions: Since relapses in patients with A-ME are a relatively frequent, the immunosuppressive therapy to prevent them is fully justified (4). Moreover, MTX therapy in pediatric patients is safe and usually well tolerated (3,5,6). The recommended dose is less than 15 mg/m^2 BSA or 1 mg/kg, with a maximal dose of 40 mg and with folic acid supplementation. In this regard, our study suggests that MTX administration (10 mg/m^2 BSA) during at least one year is a viable and effective therapy for maintenance treatment of A-ME. Accordingly, we did not detect relapses in the 11 patients studied with a median follow up time of 22 months and with an acceptable safety profile.
References:
[1] AlHakeem AS, et al. Neurosciences (Riyadh). 2017;22:67–68.
[2] Titulaer MJ, et al. Lancet Neurol. 2013;12:157–165.
[3] Bravo-Oro A, et al. Rev Neurol. 2013;57:405–410.
[4] Lin JJ, et al. Pediatr Neurol. 2014;50:574–80.
[5] Weibel L, et al. Br J Dermatol. 2006;155:1013–20.
[6] Li SC, et al. Arthritis Care Res. 2012;64:1175–1185.
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THU0502 EFFICACY AND SAFETY OF CANAKINUMAB IN PATIENTS WITH STILL'S DISEASE: A POOLED ANALYSIS OF SJIA DATA BY AGE GROUPS

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Background: Still's disease presents in paediatric and adult patients (pts) as a disease continuum with similar symptoms and pathophysiology.[1,2]
Objectives: To evaluate the efficacy and safety of canakinumab (CAN), a selective human anti-IL1 β monoclonal antibody, in SJIA pts from pooled data across 3 age groups (grps): children, adolescent and adults (the latter representing adult-onset Still's disease [AOSD] population).
Methods: Data of CAN treated pts were pooled from 4 SJIA studies (NCT00426218, NCT00886769, NCT00889863, NCT00891046). CAN was ad-