Objectives: To determine the incidence of latent tuberculosis infection and evaluate the follow-up protocol of the patients treated with canakinumab in a single pediatric rheumatology center from a TB-medium burden country.

Methods: The hospital charts of patients treated with canakinumab between 2012 and 2019 were retrospectively reviewed. The patients were screened for TB using tuberculin skin test (TST) and/or QuantiFERON-TB Gold (QFT-G) test. They had no history of active TB prior to screening and no signs or symptoms of active TB. None of the patients had recent close contact with a person diagnosed as active TB. At initial evaluation, in patients who had never been given immunosuppressive therapy, TST ≥ 15 mm (if BCG vaccinated) and TST ≥ 10 mm (if BCG unvaccinated) were considered as positive. In the case of prior immunosuppressive drug use TST cut-off limit was accepted as 5 mm. Patients with either a positive TST or QFT-G result, in whom active TB ruled out, were to receive appropriate treatment for latent TB one month prior to the first dose of canakinumab and continue throughout 9 months. Chest radiographs were performed prior to therapy and every 6 months. In case TST was anergic, both chest X-ray and TST were repeated after 3 months. Patients were evaluated for signs and symptoms of active TB by an infectious disease specialist every 3 months while on canakinumab therapy according to the "guidelines of Ministry of Health for use of biologic agents". The TST conversion was considered as a variation ≥ 5 mm between the two TSTs performed within an interval at least 3 months. If TB was suspected during the study, repeat TB testing (TST, QFT-G, and chest radiograph) was recommended.

Results: 29 Familial Mediterranean fever, 6 systemic juvenile idiopathic arthritis, 1 TRAPS and 1 hyper Ig D syndrome patients were evaluated. The mean age at canakinumab onset was 12.4 ± 4.3 years (min: 2, max: 17 years). The average duration of canakinumab use was 2.3 ± 1.4 years (min: 1 max: 6.5 years). Among 37 patients, 5 patients had positive TST and one had positive QFT-G with normal chest X-ray prior to therapy. They were given isoniazid prophylaxis for latent TB. QFT-G was checked in 6 patients prior to canakinumab, 5 were negative. Seven patients had TST conversion during follow-up. Their chest X-rays were normal. None of them had active TB symptoms, such as; cough, fever, night sweats and weight loss. The mycobacterial cultures, mycobacterium tuberculosis PCR of induced sputum (via hypertonic saline nebulization) or morning gastric aspirates were negative in these patients. QFT-G was checked in 10 patients during follow-up and all were negative. None of the patients experienced active TB during follow-up.

Conclusion: To the best of our knowledge, this is the first study investigating the frequency of latent tuberculosis infection among patients treated with canakinumab. The results of this study suggest that although frequency of latent TB infection in children treated with canakinumab may not be low in a TB-medium burden country, canakinumab seems to be a safe treatment option in terms of active tuberculosis risk. Close follow-up of children on canakinumab treatment with respect to TB by the pediatric infectious disease department is important to prevent possible complications.

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Disclosure of Interests: Balahan Makay Speakers bureau: Enzyvant, Novartis, Roche, Abbvie, ozge altug gucenmez Speakers bureau: Novartis, Abbvie, İlkur Çağlar: None declared, Süleyman Nuri Bayram: None declared, Nesrin Gülez: None declared, İlker Devrim: None declared

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THU0544 ULTRASOUND EVALUATION OF UPPER LIMBS ENTHESES AND JOINTS IN HEALTHY CHILDREN

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Background: Enthesitis in children can result from mechanical or inflammatory processes. Enthesitis is a common finding in several JIA categories, particularly in ERA. Most of the available data on the potential application of ultrasound (US) for paediatric enthesitis is currently focused on lower extremity with limited data in upper extremity entheses. To know how normal US findings of extremities by age are lead to early diagnosis and might avoid misinterpretations.

Objectives: To describe the age-related sonographic features of tendon insertions in the elbows and shoulders in healthy children.

Methods: This is a prospective cross-sectional study of 43 healthy volunteer children. Selection criteria: free of musculoskeletal symptoms, medical conditions or medication affecting the MSK system assessed by a paediatric rheumatologist. Exclusion criteria: history of trauma/surgery, family history of spondyloarthritis and more than three hours of physical activity the previous week of the evaluation. Children were grouped according to age: group 1 (3–9 years, n=22), group 2 (10–13 years, n=12) and group 3 (14–18 years, n=9).

The supraspinatus, common extensor, common flexor, and triceps tendon insertions were bilaterally examined in Power Doppler (PD) as well as grey-scale US using a GE Logic e machine with a linear transducer [8–13 MHz]. All entheses were evaluated in the longitudinal and transverse planes. Additionally, the corresponding joint was examined. Since there was no examination protocol available for scanning upper limbs entheses, a preferred US protocol for scanning children was used (1). Detailed description of entheses in grey-scale and Doppler modes was collected: echogenicity, thickness, presence of power Doppler signal (intra-enthesal at the bone/cartilage interface, peri-enthesal and within the unossified cartilage), as well as potential lesions (enthesophytes, erosions and calcifications). Measurement of tendon thickness was taken using US calipers at the site just where the tendon first contacts bone. All images were acquired by a single examiner (VM) and read by two readers. The inter-observer reliability for the tendon thickness measurements and Doppler signal was estimated between the two readers.

Results: A total of 344 entheses were evaluated in 43 healthy children. Children had a median age of 9 years (IQR 6-13) and 55% were males. All the entheses appeared normal (homogenous fibrillar pattern) on grey-scale imaging. Mean tendon measurements are reported in Table 1. In group 1, Doppler signal was seen in: supraspinatus (intra-cartilage) in one patient; common extensor (intra-enthesal) in 2 patients and triceps (intra-cartilage) in 1 patient. In group 2, common extensor (intra-cartilage) in 2 patients and common flexor in 2 patients (intra-cartilage) showed PD. None of the patients in group 3 exhibit PD signal. Children did not show either potential lesions on entheses or joint synovitis at shoulders or elbows. The ICC for enthesal measurement demonstrated high concordance 0.92 (0.84-0.93).

Abstract THU0544 – Table 1. Entheses measurement

Entheses	Group 1	Group 2	Group 3
Supraspinatus m (SD)	2.3 (0.56)	2.9 (0.35)	3.4 (0.75)
Common flexor m (SD)	2.2 (0.83)	2.44 (0.86)	2.57 (0.54)
Common extensor m (SD)	1.96 (0.6)	2.3 (0.66)	2.72 (0.48)
Triceps m (SD)	2.7 (0.89)	3.1 (0.56)	3.5 (1.2)

Abbreviations: m: mean SD: standard deviation

Conclusion: Tendon echogenicity was similar in all ages. Entheses thickness has a linear relationship with age. Detection of entheses Doppler signals at the bone/cartilage interface is rare, but its detection in the insertion to unossified cartilage may be taking account as a physiological finding in upper healthy limbs in younger children.

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THU0545 A PROSPECTIVE OUTCOME OF ALL 17 CONSECUTIVE PEDIATRIC PATIENTS WITH CHRONIC NONBACTERIAL OSTEOMYELITIS TREATED WITH INTRAVENOUS PAMIDRONATE AND FOLLOWED AT A SINGLE CENTER OVER 15 YEARS

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Background: Intravenous pamidronate (IV-PAM) has been reported to be effective in pediatric patients with severe chronic nonbacterial osteomyelitis

(CNO) in short term. Little is known about longterm outcome in CNO after IV-PAM.

Objectives: To describe a consecutive series of pediatric CNO (pCNO) patients who were prospectively followed after treatment with IV-PAM between 2003-2018 at a single center regarding: 1) The effect of IV-PAM on pain and Whole Body Magnetic Resonance Imaging (WBMRI) documented inflammation initially and after flare; 2) Relapse rate and; 3) Spinal CNO and 4) Urine N-telopeptide/creatinine ratio (UNtx/Cr) (a product of collagen 1 breakdown).

Methods:

Patients: All consecutive pCNO patients <18 years at diagnosis, with WBMRI confirmed active disease who required IV-PAM between 2003-2018. **IV-PAM:** First dose 0.5 mg/kg; subsequent doses 1 mg/kg (maximum annual dose 11.5 mg/kg). IV-PAM was administered once monthly x 9. With confirmed flare IV-PAM was repeated (max 9 doses/year). **Pain response:** Visual analogue scale for pain (VAS) with "0" indicating no pain and "10" maximum pain was administered at baseline, at each IV-PAM treatment, and at suspected flare. **Imaging:** WBMRI before 1st IV-PAM, at 6 and 12 months, at suspected flare and after retreatment. CNO resolution was documented as complete (CR) if there was 95% improvement in the WBMRI signal. Spinal x-rays were performed at baseline and annually. **UNtx/Cr** was measured at baseline, after each IV-PAM and with suspected flare.

Results: 17 patients (9F, 8M) were included. The median [range] age at CNO diagnosis was 10.3[4-15] years, and at first IV-PAM 11.6[4-20] years. The median [range] follow-up was 9.2[1-15] years. Six patients had unifocal CNO, 4 had spinal CNO (2 with baseline vertebral fractures) and 6 had multifocal non-spinal CNO. VAS was uniformly 10/10 at baseline and decreased to 0-1/10 first month after IV-PAM. CR was achieved by all at 6 months, which persisted at 12 months. Four patients, 3 unifocal and one multifocal, had no further flares. Twelve patients had WBMRI confirmed flare at previously active sites at 9-36 months and 11 received 1-9 further doses of IV-PAM. With flare, VAS ranged from 4-9/10 and decreased to 0-3/10 within first month after re-initiation of IV-PAM. On final WBMRI, 12/17 (70%) had CR and 5/17 (30%) stable mild increased signal but no clinical symptoms. No further spinal compression fractures occurred. One patient required a third course of IV-PAM. Regarding UNtx/Cr, each patient had appropriate reduction after first IV-PAM. No patients flared while UNtx/Cr remained suppressed. Three patients developed arthritis and one acne. At last follow-up, 10/17 (59%) patients were on no medications, 4/17 (24%) required prn Naproxen for CNO and 3/17 (18%) were on Naproxen and/or disease modifying medications for arthritis.

Conclusion: Long-term follow-up of pCNO patients treated with IV-PAM confirms that while several patients eventually flare, the flares are less painful and have an excellent response to IV-PAM retreatment. No further spinal fractures occurred. No flares occurred while UNtx/Cr remained suppressed, suggesting a role of osteoclasts in CNO. Further prospective multicenter studies are now required to define the long-term clinical and imaging response to IV-PAM.

Disclosure of Interests: None declared

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THU0546 RETROSPECTIVE STUDY ON ANKLE INVOLVEMENT IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA) WITH A FOCUS ON HOW TREATMENT IS LINKED TO OUTCOMES

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Background: Ankle and midfoot joints have poorer long-term outcomes in JIA compared to other joints. Esbjörnsson et al found that children with ankle involvement in the first year of disease have higher CHAQ scores and are less likely to have achieved remission by eight years [1]. Adherence to treatment has been shown to improve short term outcomes in children with JIA but there is a lack of studies which look at the long-term outcome of children who experience barriers to treatment [2].

Objectives: The aim of this study was to identify risk factors for severe ankle involvement in children with JIA. This abstract focuses on barriers to treatment; one of the main identified risk factors for adverse ankle outcomes.

Methods: This was a retrospective study which looked at JIA patients who had been treated at the Children's Hospital in Glasgow and had tibiotalar, subtalar or midfoot involvement (TT, ST and MF respectively). 680 patients with JIA were identified from clinic lists from 2002 until 2018, inclusive. The clinic lists from 2008-2010 were inaccessible meaning that anyone who was seen exclusively within those two years has not been identified. Children with systemic arthritis were excluded. Of the 680 who were identified to have JIA; 123 have been found to have TT, ST or MF involvement. Of these; 104 were included in this study and the results of 19 patients were not available for analysis. For each of the 104 patients with ankle involvement their notes were reviewed and information about their JIA disease course was noted; specifically looking at any ankle involvement or potential risk factors for an adverse outcome. Poor ankle outcomes was defined as ankle arthritis having a significant impact on the child's quality of life, persistently limited joint movement for more than 12 months or damage on imaging.

Results: Barriers to treatment was defined as anything which prevented a child from getting their medication as prescribed.

Figure 1 shows the outcomes of children who had barriers to treatment compared to the children who did not. The difference between the two groups is stark with 67% of children who faced barriers to treatment having poor ankle outcomes compared to only 25% in the group which did not experience barriers to treatment. Furthermore, children with poor ankle outcomes are almost three times more likely to need surgery if they have experienced barriers to treatment compared to those who have not. This shows that children who experience barriers to treatment are more likely to have severe long-term damage to their ankle resulting in pain and limited functionality.

Figure 1: Outcomes in Children who experienced Barriers to Treatment

Abstract THU0546 – Figure 1

Five children declined any treatment at one point in their care against the advice of their healthcare team and of these five; four (80%) had severe ankle involvement with one requiring surgery and another's ankle auto-fused.

Conclusion: Experiencing barriers to treatment has been shown to increase the risk of poor ankle outcomes. Identified barriers to treatment ranged from concerns about adherence to needle phobias in addition to side effects and medication being with-held or taken incorrectly. This study demonstrates the need for further work to explore how these barriers can be effectively minimised.

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THU0547

FIBRODYSPLASIA OSSIFICANS PROGRESSIVA IN PEDIATRIC RHEUMATOLOGY PRACTICE: LARGE SERIES EXPERIENCE OF THE SINGLE CENTER

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Background: Fibrodysplasia Ossificans Progressiva (FOP), also known as a "second skeleton disease" is extremely rare (1: 2000000) and disabling genetic disorder, caused by mutation of ACVR1 gene, a bone morphogenetic protein receptor. Among medical specialties there are no certain, capable to provide not only diagnostics, but also all medical maintenance (assessment and monitoring of extent of damages, the differentiated drug treatment, rehabilitation, contact with adjacent experts). It would be reasonable if the rheumatologist carried out a role of the main attending physician for the patient with FOP. It seems to be a lot of similarities between rheumatic diseases, especially spondyloarthritis (SpA) and FOP in the pathophysiology, clinical manifestation and the therapy approach.

Objectives: to present the single-center experience of the FOP patients and to identify similar symptoms in FOP and rheumatic disease.

Methods: The analysis of the large series of patients with FOP, who observed in the rheumatologic clinic.

Results: Our single center experience includes 26 patients with FOP. All 26 patients (13 male and 13 female) had 3 basic FOP clinical manifestations. In 23 patients molecular-genetic tests were performed and typical mutation (Arginine 206) occurred in 22 cases and one had an extremely rare mutation (Glicine 328). 22 (85%) patients had common for FOP massive heterotopic ossifications. 3 of them had formed heterotopic ossification through the X-ray negative stage to visible changes. Among typical phenotypic stigmas were: great toe malformation; thumbs malformation; peripheral osteochondromas; cervical spine abnormalities. Majority of the cases presented some similarities to SpA manifestations: ankylosis of the apophysal joints and vertebral bodies mostly in cervical spine; X-ray/CT evidence of the sacroiliitis in all patients, who were examined (n=8). Because of severe body deformity or metal details after previous surgery intervention there were no possibility to perform MRI in most patients, but we confirm typical sacroiliitis with extended bone edema on MRI in 3 patients. Recurrent episodes of the large joints synovitis were present in 5 patients. 4 patients demonstrated gradually formation of great toe ankylosis during the follow-up observation. The involution and decreasing of new FOP nodes associated with non-steroidal anti-inflammatory drug intake and/or glucocorticoids therapy were occurred in all patients.

Clinical features	Male	female	Total N of patients
Sex ratio	13	13	26 (1:1)
Age of initial manifestations (years)	<1	8	1
	1-9	6	8
	≥10	0	3
Great toe malformation	13	12	25 (96%)
Thumbs malformation	2	3	5 (19%)
Cervical spine abnormalities	12	9	21 (81%)
Peripheral osteochondromas	8	7	15 (58%)
Massive heterotopic ossifications	12	10	22 (85%)
CT/X-Ray/MRI picture of sacroiliitis	2 (of 2)	6 (of 6)	8 of 8 (100%)

Conclusion: Appearance of the similarities in FOP and SpA manifestation and the therapy approach could identify FOP as a potential rheumatic disease. Clinical observation of FOP patients could provide the important information for rheumatologist about insufficiently explored process of new bone formation.

Disclosure of Interests: None declared

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THU0548

ASSOCIATIONS BETWEEN: MARKERS OF BONE TURNOVER AND RADIOLOGICAL FINDINGS IN CHILDREN DIAGNOSED WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Although many children attending pediatric rheumatology departments are diagnosed with low bone mineral density on the basis of clinical imaging, it is difficult to determine which patients have the higher risk of developing osteoporosis as the secondary disease. Recently performed studies involving rheumatological patients have proposed

numerous serological indicators for better evaluation of the disease activity. These include markers of bone turnover, among others: bone alkaline phosphatase and osteoprotegerin (which are more specific for bone formation) and also beta isomerized carboxy terminal telopeptide of type I collagen, also known as Beta-Crosslaps (which is considered as an indicator of bone resorption).

Objectives: The main objective of the study was to evaluate the clinical usefulness of measuring serum concentrations of the selected markers of bone turnover in juvenile idiopathic arthritis (JIA) patients and assess their potential significance in prevention of osteoporosis in these children.

Methods: Study involved 59 children previously diagnosed with JIA (mean age at diagnosis: 9.0±4.3 years, mean age at study baseline: 12.7±3.9 years). All patients underwent Dual X-Ray Absorptiometry (DXA) examinations in order to assess bone mineral density. Wrist radiographs were also taken for evaluation according to the Steinbrocker classification. The presence of abnormalities in these tests was chosen as the criterion to divide patients into subgroups to perform group comparisons for the serum levels of markers of bone turnover: bone alkaline phosphatase, osteoprotegerin and Beta-Crosslaps.

Results: According to the Steinbrocker classification, 10 (16.9%) patients were staged as class I or higher. These children had significantly lower serum levels of bone alkaline phosphatase (p=0.0333) than patients with no radiological changes on wrist radiographs. The non-zero Steinbrocker subgroup had also lower Total Body Less Head Z-Score results (p=0.010) than the remaining patients.

The DXA results, expressed as Total Body Less Head and Lumbar Spine Z-Score, disclosed low bone mineral density in 10 (16.9%) and 12 (20.3%) patients, respectively. There were no significant correlations between serum levels of markers of bone turnover and bone mineral density measurements. However, bone alkaline phosphatase and osteoprotegerin were negatively correlated with DXA muscle mass (r=-0.359 p=0.040 for bone alkaline phosphatase, r=-0.372 p=0.0392 for osteoprotegerin). Similar correlation was found for bone alkaline phosphatase and DXA fat mass (r=-0.418 p<0.01).

Although Beta-Crosslaps appeared to be the only marker of bone turnover significantly (p=0.0410) associated with Juvenile Arthritis Disease Activity Score 27-Joint Count (JADAS27), it was independent from radiological findings involved in the analysis. However, serum levels of Beta-Crosslaps were higher in patients evaluated in summer than in those assessed in winter (p=0.0464). Radiological findings were not season-variable.

Conclusion: Low serum concentration of bone alkaline phosphatase seems to be a risk factor for low bone mineral density and, by extension, for osteoporosis. Therefore it should be considered as an important laboratory test in JIA patients suspected of secondary osteoporosis. Clinical significance of osteoprotegerin and Beta-Crosslaps needs to be further investigated.

Disclosure of Interests: None declared

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THU0549

SURGICAL MANAGEMENT IN JUVENILE IDIOPATHIC ARTHRITIS: A MONOCENTRIC EXPERIENCE OF 257 PROSTHESIS

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Background: Juvenile idiopathic arthritis (JIA) includes all forms of inflammatory arthritis, with onset before the 16th birthday, lasting more than 6 weeks. Although conservative management with nonsteroidal anti-inflammatory drugs and disease-modifying antirheumatic drugs can be effective, a significant number of young adults affected by JIA requires a major surgical procedure, including different types of prosthetic devices.

Objectives: This article presents a monocentric experience of an interdisciplinary team of rheumatologists and orthopedic surgeons for JIA and the outcome of patients subjected to prosthesis implantation.

Methods: Data were collected through a retrospective analysis of 583 patients attending to our transitional care center for JIA between 1999 and 2018, in the context of a once-week rheumatologic counseling with the orthopedic surgeons. Descriptive statistics were used to evaluate the baseline and follow-up data.

Results: On a total of 257 prosthesis implantations, 137 were total hip prosthesis, 101 total knee prosthesis and 19 total ankle prosthesis. The 10-year implant survival rate was 97%, with good results in term of function and comfort for the patients. Complications were observed in 9% of