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AB0728 OSTEOGENESIS IMPERFECTA: ABOUT 12 CASES

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Background: Osteogenesis imperfecta (OI), is a rare hereditary disease characterized by bone fragility and low bone mass. The clinical presentation is various with varying severity skeletal signs and inconstant extra-skeletal signs. Type 1 is the most common form (60% of cases).

Objectives: Our objective is to describe the various clinical features observed over a period of 15 years.

Methods: This is a retrospective descriptive study including 12 patients followed for OI, hospitalized in the Rheumatology Department at Fattouma Bourguiba Hospital Monastir TUNISIA between 2006 and 2019. Files were collected and analyzed.

Results: They are 9 boys and 3 girls with an average age of 14.9 ± 8.6 years. Consanguinity was reported in 25% of cases. The reason leading to consultation was, recurrent fractures (75%), blue sclera (16.7), and bone deformity (8.3%). The number of previous fractures was on average of 5, all of which were caused by a low energy trauma. Similar family cases were noted in 41.6%. The mean age of the first fracture was 4.41 ± 3.2 years. The most frequent fracture sites were respectively: femur (7/12), leg (6/12), tibia (3/12), humerus (4/12), ankle (2/12), and forearm (2/12). A deformity was noted in 58.3% of the cases: lumbar kyphosis (2), exaggerated dorsal kyphosis (2), femurs in parenthesis (2), and an anarctic deformity of 2 lower limbs (1). Imperfect dentinogenesis was found in 8.3% of cases, while ENT examination revealed conductive and sensorineural hearing loss in 2 patients each. The main radiological abnormalities were diffuse bone demineralization (9 patients), cortical thinning (5 patients), vertebral compression (3 patients), and fracture (2 patients). The bone densitometry showed a mean Z score of 3.49 ± 1.4 in the lumbar spine. The average serum calcium level was 2.38 ± 1.15 , alkaline phosphatases were elevated in all cases with an average of 756 ± 624.9 . The vitamin D level was deficient in all cases with an average of 22.75 ± 5.3 . All patients received in addition to the vitamin-calcium supplementation, pamidronate intravenously at a dose of 9mg/kg/year with a mean number of 6 cures. The main side effects observed during the infusion were abdominal pain, polyarthralgia and asthenia (1 patient), chest pain (1 patient) and fever and chills (1 patient). The control bone densitometry showed a mean Z score of 1.81 ± 1.2 in the lumbar spine.

Conclusion: Despite advances in the OI diagnosis and treatment, more research is needed. Bisphosphonate treatment decreases long-bone fracture rates, but such fractures are still frequent. New antiresorptive and anabolic agents are being investigated but efficacy and safety of these drugs, especially in children, need to be better established before they can be used in clinical practice.

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AB0729 CLINICO-EPIDEMIOLOGICAL PROFILE OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) PATIENTS IN KENYA; DATA FROM THE KENYA PEDIATRIC RHEUMATOLOGY (KAPRI) REGISTRY

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Background: Pediatric rheumatic diseases are chronic illnesses that impart a significant disease burden upon societies (1-3). Determination of the burden and clinical characteristics of these diseases is a critical first step to improving access to care and optimizing use of existing health systems for the well-being of these patients (4-6). A pediatric rheumatology registry is critical in defining the spectrum, clinical characteristics, outcomes and responses of various interventions for pediatric rheumatic diseases. Given that none exists in Kenya, the Kenya Pediatric Rheumatology Registry (KAPRI) registry offers a platform to generate this much needed data in sub-Saharan Africa.

Objectives: Our objective was to determine the baseline patient characteristics, clinical features and outcomes of the Juvenile Idiopathic Arthritis (JIA) patients assessed at the Aga Khan University Medical College East Africa who were enrolled into the KAPRI registry from inception in March 2019 to December 2020.

Methods: All patients with an ICD 10M code diagnosis of Juvenile Arthritis were selected from the KAPRI registry database. Age, gender, laboratory and clinical features at diagnosis and treatment options offered were extracted from the database. A further detailed chart review was undertaken to determine the proportion of patients who achieved remission or minimally active diseases.

Results: Among the 207 patients enrolled thus far, 16 (7.7%) were diagnosed to have JIA. Majority of the patients were females (75%; n=12) with a mean age of 7 years and 3 months (Range: 1 year – 13 years 7 months).

All patients had joint pain and swelling as the initial presenting complaints. Majority of the patients had polyarticular JIA (75%, n=12). The other 4 patients were oligoarticular (n=2) and systemic JIA (n=2). Among the polyarticular JIA patients (n=12), only 3 (25%) were rheumatoid factor (RF) positive and 1 was antinuclear antibody (ANA) positive. The oligoarticular and systemic JIA patients were all negative for antinuclear antibody, rheumatoid factor and cyclic citrullinated peptide antibodies (anti-ccp). Seven patients (43.8%) required biological therapies; tocilizumab (n=2: systemic JIA), adalimumab (n=2: polyarticular JIA), etanercept (n=2: polyarticular JIA) and tofacitinib (n=1: polyarticular JIA). One patient with systemic JIA on tocilizumab developed herpes simplex which was successfully managed with oral acyclovir. All the other patients did not develop any infections, allergic reactions or any other untoward events as adverse outcomes following the use of biological therapies. Five patients have attained remission as illustrated in the Table 1 below. Two patients have been lost to follow up.

Conclusion: Seronegative polyarticular JIA was the predominant form of JIA observed with a predilection to affect more girls and boys. Over a period of 2 years, remission has been attained among 31.25% of the patients (5 of 16) with use of synthetic disease modifying anti-rheumatic drugs and biological therapies.

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AB0730 HIP INVOLVEMENT IN JUVENILE IDIOPATHIC ARTHRITIS

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Background: Juvenile idiopathic arthritis (JIA) is a heterogeneous group of diseases that have in common an onset before the age of 16 years, more than 6-month duration, and the presence of arthritis for at least 6 weeks with no identifiable cause. Hips are the commonly affected joints in severe destructive JIA. Hip disease develops in 20% to 50% of children with JIA.

Objectives: We aimed to analyze the epidemiological, clinical, and radiological aspects of hip involvement in Tunisian JIA.

Methods: A retrospective study including 35 patients was conducted between 2010 and 2019. The patients enrolled met the ILAR criteria for the diagnosis of JIA. Clinical, biological and radiological parameters relating to the hip involvement were collected.

Results: Thirty-five patients were enrolled. The mean age of the disease onset was 9 years [3-15]. The mean age of the patients at the time of the study was 37.8 years [17-69]. The mean duration of the disease was 27 years [2-56]. These patients were assigned to discrete JIA categories: rheumatoid factor positive polyarthritis (43.5%), rheumatoid factor negative polyarthritis (21.7%), enthesitis-related arthritis (17.4%), oligoarthritis (13%) and psoriatic arthritis (4.4%). A biological inflammatory syndrome was noted in 52% of patients. Hip involvement was noted in 43.5% of patients. Coxitis occurred on average 15 years after the JIA onset [4-37]. Thirty four percent of the patients had bilateral hip involvement. The mean Lequesne index was 14. Joint space narrowing and erosions were noted in respectively 68% and 34% of cases. The majority of patients (82.6%) received medical treatment combining nonsteroidal anti-inflammatory drugs (NSAID) and rehabilitation. In the other cases, a total hip replacement was necessary. Coxitis was significantly correlated with rheumatoid factor positive polyarthritis subtype ($p=0.02$) and with the presence of biological inflammatory syndrome ($p=0.03$).