<0.01) while a lower level of QoL was associated with more intense pain ($r = 0.542$, $p < 0.01$) and poor current emotional state ($r = 0.401$, $p < 0.05$). The activity of the disease in children was not significantly related to any determinant of the QoL. In contrast to children, parents of children with JIA-U estimated current emotional state of their children significantly worse ($t = 2.05$, $p < 0.05$) and the overall level of functioning significantly lower than parents whose children did not have uveitis ($t = 2.03$, $p < 0.05$).

Conclusion: Since children with JIA evaluated the QoL equally well, whether they had uveitis or not, we can conclude that they are psychologically well adapted to their health status and cope well with different levels of pain. Unlike children with uveitis, their parents evaluated the current emotional state and overall functionality significantly worse compared to parents of children who did not have uveitis. These results support a need for the development of new uveitis specific questionnaires that will enable us better identification of patient's requirements. It is necessary to implement a holistic and multidisciplinary approach to assess the emotional functioning of the children and their families, and to devise appropriate psychosocial interventions for continuing support for ill children and their families.

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SAT0485

THE FIRST 18 MONTHS OF A PEDIATRIC RHEUMATOLOGY SERVICE AT A TERTIARY REFERRAL HOSPITAL IN KENYA

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Background: Pediatric rheumatic diseases impart a significant disease burden upon children and their families with the potential to cause diminished quality of life and significant direct and indirect costs [1-3]. Determination of the burden of disease is a prerequisite to improving access to care and optimizing use of the healthcare ecosystem for the well-being of these patients [4-5].

Objectives: We thus set out to describe what clinical cases were referred to the Pediatric Rheumatology Service at Aga Khan University Hospital Nairobi, Kenya (AKUHN) for the first 18 months since its establishment.

Methods: A prospective chart review of all patients referred to the pediatric rheumatology service at AKUHN was undertaken from October 2016 to March 2018. Patients were followed up for at least 3 months to determine the course of their disease. Data on bio-demographics and diagnosis was extracted from medical records of these patients and documented in a standard study tool. Frequencies were calculated for each disease category and sub-category.

Results: A total of 113 patients attended the pediatric rheumatology service of which 61.06% (69 of 113) were female and 53.15% (59 of 113) were aged above 10 years. Most patients attending the service were Kenyan accounting for 95.58% (108 of 113). Among the Kenyan patients seen, 77.88% (88 of 113) were from the capital, Nairobi. Out-patients accounted for 84.96% (96 of 113) of the patients compared to 15.04% (17 of 113) who were seen as in-patients. Inflammatory musculoskeletal conditions accounted for 36.3% of all cases (41 of 113) compared to 52.2% (59 of 113) of non-inflammatory musculoskeletal conditions.

Among the inflammatory conditions diagnosed, reactive arthritis accounted for 29.3% (12 of 41), juvenile idiopathic arthritis (JIA) accounted for 14.6% (6 of 41) and vasculitis accounted for 12.2% (5 of 41) of the cases as demonstrated in graph 1. The non-inflammatory musculoskeletal conditions included mechanical joint pain 42.4% (25 of 59), conversion disorder 13.6% (8 of 59), benign joint hypermobility 8.5% (5 of 59) among others as illustrated in graph 1. The other medical conditions diagnosed at the pediatric rheumatology service were two cases of acute lymphoblastic leukemia and one case of neurofibromatosis, serum sickness, sturge weber syndrome, diabetes, eczema, epidermolysis bullosa and a case of suspected griscelli syndrome. There were four patients lost to follow up; two children with prolonged fever and two others with unexplained painless unilateral limb swelling. There were three mortalities; two lupus patients and the systemic scleroderma patient, all of whom succumbed to sepsis.

Conclusion: Non inflammatory mechanical musculoskeletal complaints are the commonest complaints noted at AKUHN pediatric rheumatology clinic. The three commonest inflammatory musculoskeletal conditions are reactive arthritis, JIA and vasculitis. Sepsis was the main cause of mortality among the pediatric rheumatology patients. Further studies are required to better describe the burden and natural history of these diseases in Kenya and Sub-Sahara Africa at large.

Graph 1: Distribution of inflammatory cases reviewed at AKUHN's Pediatric Rheumatology Service

Graph 2: Distribution of non-inflammatory cases reviewed at AKUHN's Pediatric Rheumatology Service

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SAT0486

SERIOUS ADVERSE EVENTS OF TOCILIZUMAB IN CHILDREN WITH RHEUMATIC DISEASES – A REAL-WORLD EXPERIENCE

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Background: Tocilizumab (TCZ) is a humanized monoclonal antibody that blocks interleukin 6. In children, TCZ has been approved for the

treatment of systemic-onset and polyarticular course juvenile idiopathic arthritis (JIA), and is used off-label in other treatment refractory inflammatory diseases. Real-world data regarding tolerance of TCZ in children are scarce.

Objectives: To assess the incidence rate and type of serious adverse events in children treated with TCZ.

Methods: A single-center retrospective review of all consecutive patients receiving TCZ in the pediatric rheumatology division from 01/2007 to 12/2018 was performed. Inclusion criteria were a diagnosis of inflammatory rheumatic disease and treatment with at least one dose of TCZ. Patients with missing data were excluded. Serious adverse events (SAE) were defined as a life-threatening event and/or an event requiring hospital admission, leading to permanent disability or treatment discontinuation.

Results: A total of 70 children (39 females) were included. Thirty children had systemic JIA, 25 polyarticular course JIA, 14 systemic inflammatory disorders such as autoinflammatory syndromes and one patient severe refractory idiopathic uveitis. Median age at diagnosis was 4.2 years (IQR 2.0 – 7.3). The majority of children had received prednisone (n=56), and/or at least one biologic agent (n=63) prior to TCZ initiation. At TCZ start, children had a median age of 8.9 years (IQR 4.9 – 12.3) and disease duration of 3.2 years (IQR 0.9 – 7.4). TCZ was initiated at a 2-weekly interval in 44 children, 26 children received TCZ every 4 weeks. Fifty-eight children received concomitantly another immunosuppressive agent (prednisone (n=47) and/or methotrexate (n=20) and/or various other drugs (n=5)). A median of 26 TCZ doses were given per child (IQR 7 – 48), TCZ exposure was 146.8 patient years. Twenty-two SAE were observed in 18 (26%) children (SAE 14.9/100 patient years): 8 severe infusion reactions, 7 serious infections, 3 cutaneous reactions (toxic reaction, cutaneous vasculitis, scleroderma-like skin thickening with fasciitis), two macrophage activation syndromes, and one abnormal hepatic function and severe dizziness each. Infusion reactions occurred most frequently during the first few infusions (median during 4th infusion, IQR 3 – 5). All patients with infusion reactions had either systemic JIA (n=4) or another autoinflammatory disease (n=4). At time of infusion reaction, all children (8/8) had active disease, 7/8 had a concomitant treatment with prednisone and 2/8 additionally with cyclosporine.

At last follow up, 35 (50%) children were still on TCZ, the other 35 had discontinued TCZ: 4 for remission, 13 because of inefficacy, 12 because of side effects, 3 because of both inefficacy and side effects, and 3 for other reasons. No death occurred during TCZ treatment; two patients with systemic inflammatory disease, in whom TCZ had been inefficient, died from uncontrollable disease 13 and 15 months after TCZ discontinuation, respectively.

Conclusion: In this cohort, serious adverse events were observed in a quarter of children with half of them having severe infusion reactions. Higher incidence rates of severe infusion reactions were observed in children with systemic JIA or autoinflammatory diseases. Ongoing careful monitoring of patients treated with TCZ, especially those with important systemic inflammation is required.

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SAT0487 THE OUTCOMES OF JUVENILE IDIOPATHIC ARTHRITIS (JIA) IN CHILDREN MANAGED WITH CURRENT TREATMENTS: THE SINGAPORE EXPERIENCE

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Background: JIA remains one of the most common rheumatic diseases afflicted children in SE Asia¹. Despite the improvement in our treatment, outcomes of children treated with current therapy are unknown.

Objectives: To describe the clinical outcomes of children with JIA in a large, single center cohort in Singapore

Methods: Children diagnosed with JIA with a minimum follow-up of 6 month duration were recruited from our registry, from 1997 to 2015, at KK Women's and Children's Hospital, Singapore. Kaplan-Meier survival analyses were used to estimate the probability of ever attaining inactive disease (ID), remission with (CRM) or off (CR) medication according to modified Wallace criteria². Revised Steinbrocker functional classification

was used to assess functional status of JIA patients at their last follow-up visit. Non-parametric descriptive statistics including median (IQR) were used to described data.

Results: In a cohort of 251 children (62% male and 71% Chinese; 35% ERA, 34% oligoarthritis and 12% RF-negative polyarthritis were among the most common JIA subtypes encountered locally), 43% used biologics and 65% used methotrexate. Median follow-up was 35 months (IQR 13 – 70). Of all subtypes, 89%, 71% and 21% achieved ID, CRM and CR at least once during the course of follow-up, respectively. Over 85 – 100% of all JIA children could experience ID, including RF-positive polyarthritis (Fig. 1, 2). The probabilities of attaining ID within 1 and 5 years were 30 – 81% and 80 – 100%. The probabilities of attaining remission within 1 and 5 years were 10-59% and 67-100% across JIA subtypes. All of our psoriatic arthritis and systemic onset JIA reached remission on medication within 3 and 5 years, respectively. Only 33.6% of children could totally stop their medication at 1 year, but 19.3 – 87.0% could do so at 5 years. However, none of the RF-positive polyarthritis and undifferentiated JIA children had CR during the course of their diseases. Functional classes were achieved in 96%, 4%, 0.4% and 0% for class I – IV, respectively.

Fig 1. Cumulative proportion of children attaining clinical inactive disease and clinical remission on medication

Fig 2. Proportion of children achieving at least one time of clinical inactive disease and remission in each JIA category during the follow-up period.

Conclusion: Majority of children with JIA managed by our current therapy attained inactive disease in 2 years. Despite significant improvement in JIA therapy, most of them remained on medications to maintain their inactive disease status. Only 20% were able to totally withdraw treatment. Almost all of our JIA children had no disability.

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