REFERENCES:

- [1] Foster H, et al. Outcome in adults with juvenile idiopathic arthritis: A quality of life study. *Arthritis & Rheumatism*. 2003;48(3):767–775
- [2] Kontodimopoulos N, et al. Validity of SF-12 summary scores in a Greek general population. *Health Qual Life Outcomes*. 2007 Sep 28;5:55
- [3] Minden K, et al. Long-term outcome in patients with juvenile idiopathic arthritis. *Arthritis & Rheumatism*. 2002;46(9):2392–2401
- [4] Anink J, et al. Long-term quality of life and functional outcome of patients with juvenile idiopathic arthritis in the biologic era: a longitudinal follow-up study in the Dutch Arthritis and Biologicals in Children Register. *Rheumatology*. 2015;54(11):1964–1969

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6604

AB1096

OBSERVATIONAL SAFETY STUDY OF GOLIMUMAB IN TREATMENT OF POLYARTICULAR JUVENILE IDIOPATHIC ARTHRITIS USING THE GERMAN BIOLOGICS JIA REGISTRY

G. Horneff^{1,2}, A. Klein¹, on behalf of BIKER registry working group. ¹ASKLEPIOS, Sankt Augustin; ²University of Cologne, Cologne, Germany

Background: Golimumab (GLM) received European marketing authorisation for treatment of polyarticular JIA (pJIA). The long-term safety of GLM in clinical practice has not been characterised.

Objectives: The aim of the present project is to conduct a post-authorisation safety study to monitor long-term safety of subcutaneous GLM in the treatment of pJIA in routine clinical practice setting.

Abstract AB1096 – Table 1

Cohort	¹ GOL	² concurrent TNFi (ETA/ADA)	³ concurrent MTX	⁴ historic TNF	⁵ historic MTX
n	18	53	13	2103	1517
Age, median [IQR]	13.9 [12.8;15.2]	11.6 [7.9;14.8]	10.5 [4.6;12.6]	12.7 [8.9;15.5]	10.2 [5.6;13.9]
Disease duration, median [IQR]	8.0 [2.5;11.4]	2.6 [0.9;6.5]	0.2 [1.1;0.6]	3.0 [1.1;5.8]	0.9 [0.4;2.8]
Pretreatment					
NSAID, n(%)	16(89)	44(83)	11(85)	1886(90)	1293 (85.2)
Steroids systemic, n(%)	10(56)	27(51)	5 (38)	1030 (49)	315 (20.7)
MTX, n(%)	18(100)	49(92)		1794(85)	
other DMARDs, n (%)	7 (39)	4 (8)	0	636 (30)	143 (9.4)
Biologics	14 (78)	17 (36)	0	226 (11)	0
Concomitant Treatment					
NSAID, n(%)	8 (44)	38 (72%)	12(92)	1544(73)	1394 (91.9)
Steroids systemic n(%)	0	11 (21%)	4 (31)	619 (29)	359 (23.7)
MTX, n(%)	13(72)	39(74)	13(100)	1389(66)	1517 (100)
other DMARDs, n (%)	0	0	0	233 (11)	57 (4)
Baseline Disease Activity					
Active joints, median [IQR]	5 [2;6]	4 [2;9]	9 [7;13]	3 [1;7]	3 [2;7]
Phys Global, median [IQR]	37 [19;56]	33 [24;49]	59 [43;75]	48 [26;71]	43 [24;67]
JADAS_10, median [IQR]	12 [6;16]	12 [8;16]	21[18;24]	13 [8;18]	13 [8;18]

Methods: Monitoring of GLM in 200 pts with polyarticular JIA (cohort 1) in clinical practice will be performed by the German BiKeR Registry compared to a concurrent cohort 2 with 400 pts newly treated with alternative (approved) TNF-inhibitor, a concurrent cohort 3 with 500 pts newly starting methotrexate, a historic cohort 4 of patients treated with TNF-inhibitors and a historic cohort 5 never exposed to biologics but treated with methotrexate. Efficacy will be assessed by single disease activity markers and JADAS10, safety will be assessed by adverse event reporting and monitoring with a special interest on serious infections including opportunistic infections and TB, malignancies, autoimmune processes and exposure during pregnancy.

Results: Recruiting of three new cohorts 1–3 started in July 2017. Historic control cohorts 4 and 5 were obtained from the BIKER data base. Cohort 4 was recruited from 2006 to 2016, cohort 5 from 2005 to 2011. Baseline patients' characteristics

are outlined in table 1. So far, patients of the GLM cohort 1 were older, had a much longer disease duration and received pre-treatment with other biologics more often. All patients in this cohort received combination with methotrexate. Since so far GLM is approved for children with body weight of at least 40 kg and in combination with methotrexate, differences were expected. Baseline disease activity indicators are within the range of alternative TNF inhibitors and the historic cohort 4 while patients of the concurrent methotrexate cohort receiving their first treatment approach had the highest baseline disease activity.

Conclusions: The BiKeR registry has been collecting data from JIA patients treated with approved biologics in routine clinical practice since 2001. To provide context for interpreting long-term safety and effectiveness of data for GLM, analysis will also include data from contemporary pJIA patients treated with alternative TNF inhibitors and methotrexate.

Acknowledgements: This project of the BIKER registry is supported by an unrestricted grant from MSD, Germany

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2165

AB1097

PROGRESSIVE PSEUDORHEUMATOID DYSPLASIA (PPRD) IN SIBLINGS MIMICKING INFLAMMATORY ARTHROPATHY

S. Khan¹, Z. Afridi², I. Malik¹. ¹Rheumatology; ²Pediatrics, Lady Reading Hospital, Medical Teaching Institution, Peshawar, Pakistan

Background: Progressive pseudorheumatoid dysplasia is a rare condition with an incidence of 1 in 1 million.

Objectives: Keeping in mind rare disorders when dealing with children in rheumatology clinics.

Methods: A 12 years old girl was referred to rheumatology by general practitioner. Her main complaint was difficulty walking, standing up from sitting position, short stature and nodulosis of interphalangeal joints. Initially she had muscle weakness which was thought to be contributing to her gait problem and difficulty standing.

Investigation revealed myopathic changes on electromyography though her CK was entirely normal. Other investigations including routine bloods and inflammatory markers were normal. Immunology was negative. X-rays revealed reduced joint spaces in interphalangeal joints and epiphyseal dysplasia in elbows and hips. On examination there was no evidence of any synovitis. The child was of short stature i.e less than the 3rd percentile on growth chart. She had a waddling gait. There was nothing in the history to suggest previous or current inflammatory arthropathy.

Interestingly this child had another younger sibling, 7 years old girl who had similar changes in her hands.

Results: Based on history, investigations, examination and familial characteristic we felt that these two kids (siblings) had PPRD. It usually happens at the age of 3 to 8 years. Walking pattern, fatigue, weakness and intermittent episodes of stiffness are the common symptoms. Other feature is reduced joint spaces in knees and hips. At birth kids are usually of normal height but by adulthood they are short statured. They also can have calcium deposition around the joints. PPRD is caused by mutation in WISP3 gene which is responsible for bone growth and cartilage maintenance and is inherited in an autosomal recessive pattern.

Abstract AB1097 – Figure 1