thrombus in RV and showed bilateral multiple aneurysms along the pulmonary artery and its branches. According to ICB, the patient was diagnosed with BD due to having aphthous ulcers, pseudofolliculitis, and vascular involvement. IV MP (500 mg/day) for 3 days was followed by oral prednisolone 1 mg/kg/day, which was subsequently tapered. IV cyclophosphamide at a dose of 500 mg was also given every 3 weeks for a total of 6 cycles, followed by oral azathioprine (AZA). Concomitant subcutaneous IFN- α 2a was given two times per week for 6 months. Within two weeks, cough and fever disappeared, CRP values normalized. After 1 year the pulmonary artery aneurysm disappeared and cardiac thrombosis resolved. We have been following the patient with AZA for four years without recurrence.

Conclusion: We present two pediatric patients with pulmonary involvement of BD. PAI is a life-threatening condition and should be managed with more aggressive medical therapy. Early diagnosis and aggressive immunosuppressive treatment are very important in PAI. We strengthened our treatment with IFN- α 2a. There is no data in the literature regarding the use of IFN- α 2a in PAI treatment along with low dose cyclophosphamide. There were no mortality or recurrences within the 6 and 4 years follow up period. An aggressive immunosuppressive therapy leads to better prognosis in this most dreadful complication of BD.

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Disclosure of Interests: Selcan Demir: None declared, Erdal Sag: None declared, Ummusen Kaya Akca: None declared, Tuncay Hazirolan: None declared, Yelda Bilginer: None declared, Seza Özen Consultant for: Seza Ozen is receiving consultancy fees from Novartis, Speakers bureau: Roche

DOI: 10.1136/annrheumdis-2019-eular.4946

SAT0494

HOME MONITORING OF INACTIVE DISEASE IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS: PREDICTIVE VALUE OF EQ-5D-5L-Y

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Background: In recent years, juvenile idiopathic arthritis (JIA) research has shifted towards treat-to-target therapy based on clinical assessments and patient-reported outcomes (1). A well-known measurement of quality of life is the EQ-5D-5L (2). Herewith, we report preliminary results of a retrospective study using the child-friendly 'EQ-5D-5L-Y' with an E-health application (Reuma2GO) to monitor children with JIA in an outpatient setting.

Objectives: To assess the relationship between dimensions of the health-related quality of life 'EQ-5D-5L-Y' questionnaire and conventional assessments for children with JIA, including the Juvenile Arthritis Multidimensional Assessment Report (JAMAR) and active joint count (AJC), and to investigate the potential of the EQ-5D-5L-Y as instrument for outpatient management.

Methods: The study was designed as monocentric retrospective cohort study. Data from October 2017 to January 2019 were available for 70 patients with JIA. The relationships between individual dimensions of the EQ-5D-5L-Y, JAMAR and several clinical assessments were investigated. Furthermore, dimensions of the EQ-5D-5L-Y were investigated as possible predictors for binary disease activity using AJC > 0 as reference standard for active disease.

Results: Seventy patients with JIA completed 115 EQ-5D-5L-Y and JAMAR questionnaires within two weeks before a clinical visit. Moderate to high correlations were found between the EQ-5D-5L-Y and JAMAR. Moreover, the best possible EQ-5D-5L-Y score, with and without health-related visual analogue scale (EQ-VAS), demonstrated high sensitivity (81.1%) and negative predictive value (84.8%) for active disease (Table 1). The few patients who were incorrectly classified as having inactive disease (false-negatives) did not have their medication changed at the clinical visit and experienced little to no impact of disease activity on their quality of life, as indicated by the JAMAR questionnaire.

Table 1. Predictive value of EQ-5D-5L-Y dimensions for active disease using AJC > 0 as reference standard.

Cut-off for 'Active Disease'	Accuracy (% CI)	Sensitivity	Specificity	PPV	NPV
EQ-5D Mobility > 1	74.8% (65.8-82.4)	59.5%	82.1%	61.1%	81.0%
EQ-5D Self-Care > 1	76.5% (67.7-83.9)	35.1%	96.2%	81.2%	75.8%
EQ-5D Usual Activities > 1	67.8% (58.5-76.2)	73.0%	65.4%	50.0%	83.6%
EQ-5D Pain/Discomfort > 1	62.6% (53.1-71.5)	62.2%	62.8%	44.2%	77.8%
EQ-5D Anxiety/Depression > 1	73.9% (64.9-81.7)	51.4%	84.6%	61.3%	78.6%
EQ-5D EQ-VAS <85	60.0% (50.4-69.0)	64.9%	57.7%	42.1%	77.6%
EQ-5D Total Score > 5	60.0% (50.4-69.0)	81.1%	50.0%	43.5%	84.8%
EQ-5D Score > 5 & EQ-VAS <85	52.2% (42.7-61.6)	83.8%	37.2%	38.8%	82.9%

PPV: positive predictive value; NPV: negative predictive value.

Conclusion: These results demonstrate the discriminatory value of the EQ-5D-5L-Y between active and inactive disease in our cohort of patients with JIA. High negative predictive value was found for the total EQ-5D-5L-Y score, with and without EQ-VAS. In conclusion, the EQ-5D-5L-Y could be a valuable instrument for monitoring children with JIA in an outpatient setting which could aid physicians with deciding whether a clinical visit is necessary or not.

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Disclosure of Interests: Martijn J.H. Doeleman: None declared, Sytze De Roock: None declared, Nathan Buijsse: None declared, Mark Klein: None declared, Gouke J. Bonsel: None declared, V. Seyfert Shareholder of: VS is CEO and founder at MyOwnMed, Inc., Employee of: VS is CEO and founder at MyOwnMed, Inc., Nico Wulffraat: None declared, Joost F. Swart: None declared

DOI: 10.1136/annrheumdis-2019-eular.4509

SAT0495

PHENOTYPE OF PATIENTS WITH JUVENILE DERMATOMYOSITIS ASSOCIATED WITH ANTI-FACTOR H AUTO-ANTIBODY: A SEVERITY FACTOR?

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Background: Juvenile Dermatomyositis (JDM) is a rare, autoimmune and highly heterogeneous paediatric-onset myopathy, characterized by skin and muscles inflammation. Development of vasculopathy is associated with the severe extra-muscular manifestations of JDM, and portends a poor prognosis. Impaired function of JDM vasculature includes immune complex and complement deposition.

Objectives: The aim of our study is to describe the clinical and paraclinical phenotype of patients with anti-factor H autoantibody (Ab) associated with JDM and overlap myositis.

Methods: Patients with a diagnosis of JDM or overlap myositis and presence of anti-FH auto-antibody, followed in two Parisian tertiary centers over the last twelve years, were retrospectively selected. Besides demographic data, clinical features (cutaneous lesions, severity of muscle involvement, extra-muscular manifestations), treatment options and their outcome were retrieved for each patient. Biological data and presence of myositis-specific autoantibodies were collected, muscle MRI were examined, and data from muscle biopsies were assessed using a validated score tool when available.

Results: Eleven female patients were included in the study, with a median age at diagnosis of 8.8 years [2.8 - 13.1], and a median follow-