Table 1. Summary of ANN scores. A) Global Still's disease evaluation. B) Immune system component. ANN scores mean the probability of the resulted relationship is true positive: +++ correspond to values >78% (p-value<0.05); ++ correspond to values > 59% (p-values<0.15) and; + correspond to values > 38% (p-value<0.25)

A) Still's disease molecular definition	Biologics				Non-biologics	
	Anakinra	Canakinumab	Sarilumab	Tocilizumab	Methotrexate	Prednisone
Still's disease	+++ (81%)	+++ (86%)	+++ (85%)	+++ (85%)	- (5%)	++ (70%)
Systemic profile	+++ (80%)	+++ (88%)	+++ (85%)	+++ (85%)	- (4%)	++ (70%)
Rheumatic profile	+++ (87%)	+++ (92%)	+++ (81%)	+++ (81%)	- (9%)	++ (64%)
B) Immune system components	Biologics					
Innate immune system deregulation	Anakinra	Canakinumab	Sarilumab	Tocilizumab	Non-biologics	
Adaptive immune system	++ (71%)	++ (71%)	++ (55%)	++ (55%)	Methotrexate	Prednisone
T-cell response activation	+ (45%)	- (37%)	++ (71%)	++ (71%)	- (10%)	++ (65%)
Defective immune regulation	- (19%)	- (37%)	+ (47%)	+ (47%)	- (25%)	+ (47%)
					- (15%)	+ (50%)

Figure 1. Kaplan-Meier Estimates of a Combined JADAS27 MDA and CHAQ-DI=0 Response by Age Cohort

Number of pts with the event of this combined response at baseline: 1
Pts without combined JADAS27 MDA and CHAQ-DI=0 response were censored at the date of the last combined JADAS27 MDA and CHAQ-DI=0 response assessment
N at Mo 0 is number of treated pts with combined JADAS27 MDA and CHAQ-DI=0 response assessments at Day 1
Pts who had the event of this combined response at baseline were excluded from the analysis
CHAQ-DI=Childhood Arthritis Disease Activity Score 27; MDA=Minimal Disease Activity; pt=patient

Figure 2. Kaplan-Meier Estimates of a Combined JADAS27 MDA and Pain VAS <35 mm Response by Age Cohort

Number of pts with the event of this combined response at baseline: 3
Pts without combined JADAS27 MDA and Pain VAS <35 mm response were censored at the date of the last combined JADAS27 MDA and Pain VAS <35 mm response assessment
N at Mo 0 is number of treated pts with combined JADAS27 MDA and Pain VAS <35 mm response assessments at Day 1
Pts who had the event of this combined response at baseline were excluded from the analysis
JADAS27=Juvenile Arthritis Disease Activity Score 27; MDA=Minimal Disease Activity; pt=patient; VAS=visual analogue scale

THU0498

PATIENT-REPORTED TREATMENT BURDEN AND ITS IMPACT ON QUALITY OF LIFE IN JUVENILE IDIOPATHIC ARTHRITIS: RESULTS FROM THE PHARMACHILD REGISTRY

A. Alongi¹, A. Consolaro², G. Vijatov-Djuric³, G. Filocamo⁴, O. Vougiouka⁵, A. N. Olivier⁶, C. Herrera Mora⁷, W. Emminger⁸, A. Ravelli², N. Ruperto².
¹Università Degli Studi di Genova, Genoa, Italy; ² Istituto Giannina Gaslini,

Genoa, Italy; ³University of Novi Sad, Novi Sad, Serbia; ⁴Fondazione IRCCS Ca' Granda-Ospedale Maggiore Policlinico, Milan, Italy; ⁵P a A Kyriakou" Children's Hospital, Athens, Greece; ⁶Second University of Naples F.Fede, Naples, Italy; ⁷Hospital de Niños Roberto Gilbert Elizalde, Guayaquil, Ecuador; ⁸University Children's Hospital, Vienna, Austria

Background: Juvenile Idiopathic Arthritis (JIA) patients experience impaired health and wellbeing due to multiple causes of physical and psychosocial distress, including treatment burden. Despite emerging evidence of its relevance [1], the contribution of treatment adverse events to patient-reported outcomes (PROs) in JIA has been poorly explored.

Objectives: To evaluate and rank the impact of patient-reported adverse events (AEs) on overall wellbeing, health-related quality of life (HRQoL), school problems and self-reported medication adherence using data from Pharmachild, a large international JIA pharmacovigilance registry.

Methods: Registry entries on 5340 prospective visits of 2251 patients enrolled till December 2018 were analyzed; all included patients were treated with at least one DMARDS or Biologic agent at the time of visit. In the Juvenile Arthritis Multidimensional Assessment Report (JAMAR), patients and parents compiled a checklist of treatments, side effects, self-reported adherence, administration difficulties and disease-related school problems occurred in the previous 4 weeks. Evaluated outcomes included patient acceptable symptom state (PASS), VAS-measured patient assessment of overall wellbeing (PGA) and HRQoL, assessed through the physical health (PhH) and psychosocial health (PsH) subscales. The relationships between AEs and PROs were tested through generalized linear models, accounting for disease activity and symptoms levels. Bayesian Networks were used to explore the causal effects of specific AEs on outcomes to disentangle the confounding role of disease status.

Results: AEs were reported in 22.9% of visits. For similar levels of physician global assessment (MD global), patient-assessed disease activity, pain and function, patients reporting AEs had worse PGA, PsH, and lower probability of reaching PASS (fig. 1, all p-values <0.001). The impact of AEs on PGA was small but not trivial (effect size η^2 0.031) and appears to be mediated by effects on PsH and school problems (p <0.001). Non-linear regression modelling revealed a significant moderating effect of MD global levels < 2.5 on the relationship between AEs and PGA (p 0.003), indicating that the impact of AEs is higher for lower disease activity states. AEs predicted self-reported medication adherence (p<0.001), even when adjusted for the number of administered treatments. In the Bayesian network model, mood swing and sleep problems emerged as the most influential items affecting PsH, (respectively, total effect 2.62 and 1.25, both p< 0.001). Fig. 2 shows the total standardized effect of specific AEs on mean PsH levels. Nausea had the highest impact on treatment adherence (total effect -0.0541, p <0.001), being the only AE directly linked to drug refusal.

Conclusion: AEs have a measurable effect on the wellbeing and psychosocial health of JIA patients, particularly when disease activity is low, and significantly affect school activity and medication adherence. Mood swings and sleep problems show the strongest influence on HRQoL. Addressing AEs appears important to reduce disease impact, improve patients' satisfaction and therapeutic compliance.

References: :
[1] Weitzman, Elissa R., et al. Journal of patient-reported outcomes 2.1 (2018): 1.

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THU0499

IS THERE A DIFFERENT PRESENTATION OF JUVENILE SYSTEMIC DIFFUSE AND LIMITED SUBSET? DATA FROM THE JUVENILE SCLERODERMA INCEPTION COHORT. WWW.JUVENILE-SCLERODERMA.COM

I. Foeldvari¹, J. Klotzsche², O. Kasapcopur³, A. Adrovic³, K. Torok³, M. T. Terrier³, A. P. Sakamoto³, V. Stanevich³, F. R. Sztajnbock³, J. Anton³, B. Feldman³, E. Alexeeva³, M. Katsikas³, V. Smith³, E. Marrani³, M. Kostik³, N. Vasquez-Canizares³, S. Appenzeller³, M. Janarthanan³, M. Moll³, D. Nemcova³, A. Patwardhan³, M. J. Santos³, S. Sawhney³, D. Schonenberg³, C. Battagliotti³, L. Berntson³, B. Bica³, J. Brunner³, P. Costa Reis³, D. Eleftheriou³, L. Harel³, G. Horneff³, D. Kaiser³, D. Lazarevic³, K. Minden³, S. Nielsen³, F. Nuruzzaman³, Y. Uziel³, N. Helmus¹. ¹Hamburg Centre for Pediatric and Adolescence Rheumatology, Hamburg, Germany; ²German Rheumatism Research Center, Berlin, Germany; ³JSSc Collaborative Group, Hamburg, Germany

Background: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence of 3 per 1 000 000 children. There are limited data regarding the clinical presentation of jSSc. The Juvenile Systemic Sclerosis Inception Cohort

(JSSIC) is the largest multinational registry that prospectively collects information about jSSc patients.

Objectives: Evaluation of the jSSc patients at the time of inclusion in the JSSIC. **Methods:** Patients were included in the JSSIC if they fulfilled the adult ACR/EULAR classification criteria for systemic sclerosis, if they presented the first non-Raynaud symptom before 16 years of age and if they were younger than 18 years of age at time of inclusion. Patients' characteristics at time of inclusion were evaluated.

Results: Until 15th of December 2019 hundred fifty patients were included, 83% of them being Caucasian and 80% female. The majority had the diffuse subtype (72%) and 17% of all jSSc had overlap features. The mean age of first presentation of Raynaud's phenomenon was 9.8 years in the diffuse subtype (djSSc) and 10.7 years in the limited subtype (ljSSc) (p=.197). The mean age at first non-Raynaud's symptoms was 10.0 years in the djSSc and 11.2 years in the ljSSc (p=0.247). Mean disease duration at time of inclusion was 3.4 years in the djSSc and 2.4 years in the ljSSc group.

Significant differences were found between the groups regarding mean modified Rodnan skin score, 18.2 in the djSSc vs 6.2 in the ljSSc (p=0.02); presence of Gottron's papulae (djSSc 30% vs ljSSc 13%, p=0.43); presence of telangiectasia (djSSc 42% vs 18% ljSSc, p=0.01); history of ulceration (djSSc 42% vs 18% ljSSc, p=0.008); 6 Minute walk test below the 10th percentile (djSSc 85% vs ljSSc 54%, p=0.044), total pulmonary involvement (djSSc 49% vs ljSSc 31%, p=0.045), cardiac involvement (ljSSc 17% vs djSSc 3%, p=0.002). djSSc patients had significantly worse scores for Physician Global Assessment of disease activity compared to ljSSc patients (VAS 0-100) (40 vs 15) (p=0.001) and for Physician Global Assessment of disease damage (VAS 0-100) (36 vs 17) (p=0.001).

There were no statistically significant differences in the other presentations. Pulmonary hypertension occurred in approximately 6% in both groups. No systemic hypertension or renal crisis was reported. ANA positivity was 90% in both groups. Anti-Scl70 was positive in 35% in djSSc and 36% in the ljSSc group. Anticentromere positivity occurred in 3% in the djSSc and 7% in the ljSSc group.

Conclusion: In this unique large cohort of jSSc patients there were significant differences between djSSc and ljSSc patients at time of inclusion into the cohort regarding skin, vascular, pulmonary and cardiac involvement. According to the physician global scores the djSSc patients had a significantly more severe disease. Interestingly the antibody profile was similar in both scleroderma phenotypes.

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