KI.14

Application of CRISS score, revised CRISS score and RCID score in patients with Juvenile systemic sclerosis

Jens Klotsche[1], Ivan Foeldvari[2], Kathryn Torok[3], Francesco Del Gado[4], Daniel E. Furst[5], Ozgur Kasapcopur[6], Amra Adrovic[6], Brian Feldman[6], Maria Teresa Terreri[6], Ana Paula Sakamoto[6], Flavio Sztajnbok[6], Jordi Anton[6], Maria Katsicas[6], Valda Stanevicha[6], Simone Appenzeller[6], Tadey Avcin[6], Sindhu Johnson[6], Mikhail Kostik[6], Hana Malcova[6], Edoardo Marrani[6], W.-Alberto Sifuentes-Giraldo[6], Raju Khubchandani[6], Dana Nemcova[6], Maria Jose Santos[6], Dieneke Schonenberg[6], Christina Battagliotti[6], Lillemor Berntson[6], Blanca Bica[6], Juergen Brunner[6], Despina Eleftheriou[6], Liora Harel[6], Gerd Horneff[6], Tilmann Kallinich[6], Thomas Lehman[6], Kirsten Minden[6], Monika Moll[6], Susan Nielsen[6], Anjali Patwardhan[6], Vanessa Smith[6], Nicola Helmus[2]

[1]German Rheumatism Research Center, Berlin, Deutschland
[2]Hamburger Zentrum für Kinder- und Jugendrheumatologie, Hamburg, Deutschland
[3]University of Pittsburgh, Pittsburgh, United States
[4]University of Leeds, Leeds, Vereinigtes Königreich
[5]University of California, Los Angeles, United States
[6]jSSc collaborative group, Hamburg, Deutschland

Einleitung:

Juvenile systemic sclerosis (jSSc) is a rare disease in childhood. To date, no composite response index exists to assess treatment effect in jSSc patients. ACR CRISS score (probability of improvement ranging from 0 to 1 based on mRSS, FVC%, PtGA, MDGA and HAQ-DI) and revised ACR CRISS (rCRISS, proportion of patients who improve in ≥ 3/5 ACR CRISS core items by a certain percentage, e.g. 30%, except 5% for FVC) were developed by experts in the field as outcome measures in adult patients with SSc. In addition, the Ranked Composite Important Difference (RCID) score was recently introduced as anchor to the ACR CRISS.

Methoden:

Data from the international jSSc inceptions cohort were used for this analysis. The ACR CRISS, rCRISS and RCID were calculated between baseline and 12-months follow-up according to the scoring algorithms. Missing values in the core items were estimated by multiple imputation by chained equations. Here we aimed to determine the value of the response measures to detect clinically change defined by the anchor questions about change (much better or little better versus almost the same, little worse or much worse) in patients overall health due to scleroderma since the last visit provided by the treating physicians and parents or patients (aged > 12 years).

Ergebnisse:

We included 95 jSSc patients with diffuse cutaneous subtype with available baseline and 12-months visit. Seventy-nine percent were female, the mean age at enrollment was 13.0 (3.8) and the mean disease duration was 3.1 (2.8) years. Among 95 patients, 57% were treated with steroids, 47% with methotrexate, 27% with MMF and 3% with a biological at baseline. ACR CRISS showed a ceiling effect (>.998) in 51% and a floor effect (<0.005) in 26% of patients. Patients who reported at least moderate improvement had a median ACR CRISS of 0.99 and in mean 2.6 (1.3) core items that improved by ≥20% from baseline to 12-months follow-up. The rCRISS 20/30/50 responses were 59%/49%/33% in patients who reported improvement (table 1) and 25%/25%/8% in patients with worsening. The RCID was approximately normal distributed (mean 20.7, SD 43.4). Mean (SD) RCID for patients who reported worsening was -10.5 (38.6) vs RCID of 20.7 (45.2) for patients who reported improvement. RCID scores for physician reported anchors of worsening or improvement were 6.5 (44.2) and 18 (45.4), respectively. The concordance between a positive RCID score and rCRISS 20/30 was moderate (rCRISS 20 and RCID, 43%, kappa=0.43; rCRISS 30 and RCID, 38%, kappa=0.36).

Table 1: ACR CRISS, rCRISS and RCID score by patients and physicians ratings about scleroderma disease course

	Worsening/ no improvement reported by patients (n=12)	Improvement reported by patients (n=49)	P value	Worsening/ no improvement reported by physicians (n=14)	Improvement reported by physicians (n=50)	P value
Median ACR CRISS score (IQR)	0.0 (0 to 0.75)	0.99 (0 to 1.0)	0.007	0.35 (0 to 0.99)	0.99 (0 to 1.0)	0.037
rCRISS response 20%	3(25%)	29 (59%)	0.034	5 (36%)	29 (58%)	0.140
rCRISS response 30%	3 (25%)	24 (49%)	0.134	2 (14%)	27 (54%)	0.008
rCRISS response 50%	1 (8%)	16 (33%)	0.092	0 (0%)	18 (36%)	0.008
Mean RCID score (SD)	-10.5 (38.6)	20.7 (45.2)	0.031	6.5 (44.2)	18 (45.4)	0.411

CRISS = Composite Response Index in Systemic Sclerosis; RCID=Ranked Compsoite Important Difference; rCRISS = revised Composite Response Index in Systemic Sclerosis; SD = standard deviation

Schlussfolgerung:

Our data confirmed the presence of a ceiling and floor effect of ACR CRISS as shown in studies of adult SSc patients. The CRISS, rCRISS and RCID response distinguished between patients who rated their disease course since last visit as worsened or improved. Future studies should focus on the determination of specific pediatric weights for the CRISS and RCID components rather than extrapolation from adult SSc. In general, the RCID offers a meaningful tool in order to determine response to therapy in future clinical trials in jSSc patients.