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POS1299 LONG-TERM NEURODEVELOPMENTAL OUTCOME OF CHILDREN BORN TO SYSTEMIC SCLEROSIS WOMEN: PEDIATRIC NEUROPSYCHIATRIC ASSESSMENT THROUGH A SET OF VALIDATED TOOLS

Keywords: Pregnancy and reproduction, Quality of life, Systemic sclerosis

E. Pedretti[1,2], J. Galli[2,3], L. Moschetti[1,2], C. Nalli[1], M. Bendoni[2,3], L. Visconti[2,3], A. Molinaro[2,3], F. Franceschini[1,2], A. Tincani[1,2], P. Airò[1], L. Andreoli[1,2], E. Fazzi[2,3], M. G. Lazzaroni[1,2]. *[1]Rheumatology and Clinical Immunology Unit, ASST Spedali Civili of Brescia, Brescia, Italy*; *[2]Department of Clinical and Experimental Sciences, University of Brescia, Brescia, Italy*; *[3]Unit of Child and Adolescent Neuropsychiatry, ASST Spedali Civili of Brescia, Brescia, Italy*

Background: The neurodevelopmental (ND) outcome of children born to women with autoimmune disorders might be affected by factors related to maternal disease (*e.g.* the transplacental passage of maternal antibodies/drugs), as well as by mothers' coping with their disabilities during growth. These aspects have been studied in rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE) [1]; no data are available for Systemic Sclerosis (SSc).
Objectives: To evaluate the long-term ND outcome of children born to SSc mothers.
Methods: A pediatric neuropsychiatric evaluation was proposed to SSc mothers with at least one child ≤18 years (August 2021-June 2022). Evaluations, performed by a child neuropsychiatrist, included the assessment of the following domains:

- Developmental/Cognitive skill (Griffiths Mental Development Scales III -GMDS-III- and Wechsler Scale for corrected age)
- Adaptive behavior (Vineland Adaptive Behavior Scales-II -VABS-II-)
- Social/Behavioral problems (Child Behavior CheckList and Youth Self Report -YSR- for children ≥11 years)
- Quality of life (Pediatric Quality of Life Inventory 4.0 -PedsQL-)

A set of questions to investigate the child's ND milestones were submitted to mothers. Data about maternal disease were retrieved from medical records.
Results: 20 children (median age 8 years; F/M 1:1) born to 12 SSc mothers were evaluated: 4 were born before and 16 after SSc onset. Four preterm births (<37 gestational weeks -GW-) (one twin pregnancy) and no small for gestational age babies were recorded. One child already had a diagnosis of autism spectrum disorder (ASD) at enrollment. All the children showed a normal score for developmental or intelligence quotient (IQ) and in all VABS-II domains, except for the child affected by ASD. YSR, administered to 6 children, highlighted an impairment in social abilities (*e.g.* extra-academic/social activities). In CBCL, externalizing problems (*e.g.* aggressive behavior, hyperactivity) were reported in 3 children (15%), while internalizing problems (*e.g.* anxiety, somatic complaints) in 2 (10%). The lowest scores at PedsQL were observed in the emotional domain. Different types of sleep disorders were reported in 45% of children. No significant differences were found in maternal characteristics upon the comparison between children with/without ND abnormalities. Children and mothers' features are shown in Table 1.
Conclusion: Children born to SSc mothers displayed normal intellectual functioning. The reported impairment in social abilities, suggesting an "adult profile", possibly reflect the effect of coping with maternal disabilities. Our data did not show a specific pattern of ND alterations in the offspring of SSc mothers; however the inclusion of a child neuropsychiatrist in the multidisciplinary team dedicated to SSc patients and their families seems to be important to detect and correct any possible ND problems at an early stage.
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POS1300 NAILFOLD CAPILLAROSCOPY FOR STANDARD CLINICAL PRACTICE: ARE TWO FINGERS AS ACCURATE AS EIGHT?

Keywords: Systemic sclerosis

Y. El Miedany[1], S. Ismail[2], M. Fawzy[3], M. Elgaafary[4], M. H. Abu-Zaid[5]. *[1]Canterbury Christ Church University Medway Campus, Rheumatology, Gillingham, United Kingdom*; *[2]National Research Center, Rheumatology and Rehabilitation, Cairo, Egypt*; *[3]Faculty of Medicine Kasr Al-Ainy, Cairo University, General Medicine, Cairo, Egypt*; *[4]Faculty of Medicine, Ain Shams University, Community and Public health, Cairo, Egypt*; *[5]Faculty Of Medicine - Tanta University, Rheumatology and Rehabilitation, Cairo, Egypt*

Background: Nail-fold capillaroscopy (NFC) is a non-invasive, in vivo diagnostic tool for the assessment of microvasculature of the proximal nail fold. The comprehensive approach based on assessment of four finger each hand excluding the thumbs (in total eight fingers are examined) is the gold standard for NFC evaluation and scoring. The patient is identified to have abnormal nailfold capillaries, if abnormality was present in any finger/nailbed. However, this process is time consuming and is not applicable in standard clinical practice. Therefore, a question was raised can a concise approach based on assessment of a smaller number of fingers be implemented in standard practice. The challenge to this hypothesis is that there may be marked variability in the nailfold morphology between different fingers, as abnormalities may only be seen in some but not all fingers.
Objectives: 1. To evaluate the comparability of the concise to the comprehensive approach (the gold standard) represented as the percentage of agreement of the NFC patterns recorded from each approach. 2. To assess the statistical difference on comparing the concise approach score to the gold standard score.
Methods: This was a prospective, cross sectional multicenter study. 164 Patients were recruited for this study. The patients' cohort included systemic sclerosis,

Table 1. Characteristics of SSc women and their offspring

Mother	Antibodies	Cutaneous involvement	Digital Ulcers	ILD	Child	GW at birth, pregnancy complications	Cognitive domain	Adaptive behavior	Social and behavioral domain	Sleep disorders
1	Scl-70	lcSSc	1	0	M, 12 yr	40	N	N	A	0
					M, 10 yr	40	N	N	N	0
2	ANA+	lcSSc	0	0	M, 7 yr*	40	A	A	A	1
					F, 5 yr	40	N	N	N	1
3	Th/To	lcSSc	1	0	M, 13 yr	39	N	N	A	0
					F, 9 yr	39	N	N	N	1
4	Scl-70	dcSSc	1	1	*F, 11 yr	39	N	N	A	1
					M, 5yr	39	N	N	N	0
5	Scl-70	dcSSc	0	1	*F, 11 yr	39	N	N	A	1
					M, 2 yr	35°	N	N	N	1
6	ACA	lcSSc	0	0	M, 3 yr	41	N	N	N	1
7	Scl-70	lcSSc	0	0	*F, 17 yr	32^	N	N	B	0
					*F, 13 yr	39	N	N	N	0
					M, 9 yr	39	N	N	N	0
8	ACA	lcSSc	0	0	F, 1 yr	39	N	N	N	0
9	Scl-70	dcSSC	1	1	F, 8yr$	31°	N	N	N	1
					M, 8yr$	31°	N	N	N	0
10	U1-RNP	lcSSc	0	0	F, 5 yr	38	N	N	N	0
11	Scl-70	dcSSc	0	0	M, 3yr	39	N	N	N	1
12	U1-RNP	lcSSc	1	0	F, 1 yr	40	N	N	N	0

ANA: anti-nuclear antibodies, ACA: anti-centromere antibodies, GW: gestational weeks; lcSSc: limited cutaneous SSc, dcSSc: diffuse cutaneous SSc, ILD: interstitial lung disease, 0: absent, 1: present, N: normal, B: borderline, A: abnormal*ASD diagnosis, *born before SSc onset $ Twins, °Gestational diabetes, ^Preeclampsia/IUGR

rheumatoid arthritis, psoriatic arthritis, systemic Lupus erythematosus, Behcet's disease, Sjogren's syndrome and mixed connective tissue diseases. Imaging sessions were carried following the national standard for NFC [1]. NFC with 200x magnification was used to capture panoramic nailfold videocapillaroscopy images. Images were recorded from 4-fingers each hand, excluding the thumbs. For each finger 5 parameters were assessed: density, architecture, hemorrhage, neoangiogenesis and dimension. Semiquantitative scoring system was implemented to score each parameter [2].

Results: Considering the correct diagnosis using the gold standard score was 100%, the concise approach based on mean of the scores recorded from the middle and ring fingers both hands gave the correct pattern in 155/164 (94.5%) of the cases. There was no statistical difference between the comprehensive approach total score (3.5+0.76 [CI 95% 1.918 - 5.082]), when compared to the concise total score from 2 fingers both hands which was (3.25+2.87 [1.873 - 4.626], p= 0.18. There was no significant difference on comparing the right to the left hand fingers.

Conclusion: Examining the NFC from middle and ring fingers in each hand, can be time saving and not inferior to the comprehensive NFC assessment. Concise approach can be used in the standard clinical practice and, in the meantime, are able to give an overall NFC picture reflecting the real state of the nailfold capillaries.

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POS1301 MORTALITY AND CAUSES OF DEATH AMONG FINNISH PATIENTS WITH SYSTEMIC SCLEROSIS

Keywords: Systemic sclerosis, Epidemiology

S. Kortelainen[1], M. Käyrä[2], T. Rissanen[3], J. Paltta[1], L. Pirilä[1], J. Huhtakangas[4]. *[1]Turku University Hospital, Rheumatology and Clinical Immunology, Turku, Finland*; *[2]Oulu University Hospital, Rheumatology, Oulu, Finland*; *[3]University of Turku, Biostatistics, Turku, Finland*; *[4]Kuopio University Hospital, Rheumatology, Kuopio, Finland*

Background: Systemic sclerosis (SSc) is a rare rheumatic autoimmune disease, characterized by vasculopathy and fibrosis of the skin and internal organs [1]. Mortality and morbidity of systemic sclerosis is significant. The studies assessing changes in the mortality of SSc are controversial, only minor sign of improvement of prognosis is seen. Among SSc-related causes of death scleroderma renal crisis has become rare and cardiopulmonary causes have become more common. Most of the studies are conducted using either the ACR 1980-criteria or the LeRoy and Medsger -criteria.

Objectives: Aim of this study was to examine mortality and basic causes of death among Finnish patients with systemic sclerosis (SSc) during years 2000-2020 using the ACR/EULAR 2013 criteria for SSc diagnosis.

Methods: Patients with a diagnostic code of systemic sclerosis (ICD-10 codes beginning with M34) that appeared at least once in their medical records during the years 1996–2018 were identified from the hospital discharge registers of Turku and Oulu University hospitals. False diagnoses and typing errors were excluded. Using ACR/EULAR 2013 criteria and clinical findings, the diagnoses were reclassified and divided into different subsets of the disease. These subsets were diffuse cutaneous systemic sclerosis (dcSSc), limited cutaneous systemic sclerosis (lcSSc), over-lap SSc (diagnosis of other rheumatic disease fulfilled simultaneously) and sine scleroderma (SSc without skin affision). The clinical data was collected to the end of year 2020. The death certificates including the basic and imminent causes of death were obtained from Statistics Finland until August 2021. By examining patient records and death certificates the basic cause of death for each subject was re-evaluated. The study data were collected and managed using REDCap electronic data capture tools hosted at the University of Turku.

Results: Among 313 SSc patients, 91 deaths occurred between 4/2000- 9/2020. 35 deaths were caused by SSc, 20 by atherosclerosis, 18 by cancer and 18 by other causes. Majority of SSc-related deaths were due to interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH), 13 and 11 deaths respectively. Four deaths were due to scleroderma renal crisis (SRC), three deaths due to gastrointestinal tract involvement, two due to SSc-related myocardial involvement and two due to other SSc-related causes, those were vasculopathy complications. The mean age at dead of patients who died of a SSc-related cause was 65.6 years (SD 12.7, CI= 61.2-70.1) in contrast to patients who died due to other causes was 74.2 years (SD 9.6, CI= 71.5-76.9), the difference was statistically significant (p=0.0006). The median time from SSc diagnosis to death was 4.4 years [IQR 1.5, 11.8] in the SSc-related death group and 10.8 years [IQR 4.7, 17.1] for others, the difference was statistically significant (p=0.0061). Death due to renal crisis occurred fastest, in two months. An ILD related death occurred 11.8 years after and a PAH related death 3.7 years after the SSc diagnosis on average. In females, PAH and ILD were the most common causes of death, both nine of 25 subjects. In males, ILD was the most common cause of death, in four of ten subjects. In the group of dcSSc, the most common cause of death was ILD with five deaths out of twelve. In the group of lcSSc, PAH and ILD were the most common causes of death with both eight of 19 deaths. Four deaths due to SRC were all in the group of dcSSc. Twelve of 23 patients with dcSSc (52%) and nineteen of 60 patients with lcSSc (32%) died due to SSc.

Conclusion: The main finding in our study is the fact, that SSc disease itself is the major cause of death among Finnish SSc patients. Among the SSc related deaths, ILD and PAH were the leading causes of death. The patients who died due to SSc were significantly younger and the time from SSc diagnosis was significantly shorter compared to those who died due to other causes.

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POS1302 SPECKLE-TRACKING GLOBAL LONGITUDINAL STRAIN AS A PREDICTOR OF CLINICAL OUTCOMES IN SYSTEMIC SCLEROSIS PATIENTS

Keywords: Prognostic factors, Systemic sclerosis

D. Benfaremo[1,2], G. Stronati[3], C. Dichiara[4], Y. Ferraioli[4], C. Tana[4], I. Paterno[4], S. Bigoni[4], S. Contegiacomo[4], M. Sistoni Pepparoni[4], N. Pacenti[4], A. Dello Russo[3], F. Guerra[3], G. Moroncini[1,2]. *[1]Marche Polytechnic University, Department of Clinical and Molecular Sciences, Ancona, Italy*; *[2]Marche University Hospital, Clinica Medica, Ancona, Italy*; *[3]Marche University Hospital, Cardiology Clinic, Ancona, Italy*; *[4]Marche Polytechnic University, Internal Medicine Residency Program, Ancona, Italy*

Background: Primary cardiac involvement occurs in up to 40% of patients affected by systemic sclerosis (SSc), and although it may be asymptomatic is associated with negative outcomes. Speckle-tracking derived global longitudinal strain (GLS) has been proven to be a cost-effective tool in the detection of left ventricular (LV) and right ventricular (RV) dysfunction in patients with SSc and no overt cardiac disease [1].

Objectives: The aim of this study was to assess whether GLS can predict clinical outcomes in patients with SSc.

Methods: We conducted a prospective observational study enrolling all consecutive patients referred to our Scleroderma Unit between June 2016 and January 2022. All patients had a definite diagnosis of SSc according to ACR/EULAR criteria and no overt cardiac disease, pulmonary arterial hypertension, or atrial fibrillation at the time of enrollment. For each patient, echocardiogram and GLS calculations were performed at baseline and at each follow-up. We also collected all data regarding clinical history, hospitalizations or adverse events and ECGs.

Results: 164 patients (148 female, 58±14 years) were enrolled. Overall, 19 (11.6%) patients died during a median follow-up of 3.2 years for mainly non-cardiovascular deaths (7.3%) while cardiovascular deaths were lower (3% non-sudden, 1.3% sudden). Left GLS at first visit was associated with all-cause death, with a 1% left GLS worsening associated with a 19% increased risk of death after adjusting for age, gender, and LVEF (adjusted HR 1.19; 95% CI 1.05-1.35; p=0.007). Similarly, right GLS at first visit was associated with all-cause death, with a 1% right GLS worsening associated with a 12% increased risk of death after adjusting for age, gender, and TAPSE (adjusted HR 1.12; 95% CI 1.03-1.21; p=0.005). Patients with a left GLS worse (i.e. higher) than -20% had a 3.5-fold increased risk of death when compared to patients with better left GLS (HR 3.55; 95% CI 1.28-9.88; p=0.015; Figure 1a). Similarly, patients with a right GLS worse than -20% had a 4.5-fold increased risk of death when compared to patients with better left GLS (HR 4.47; 95% CI 1.4-13.74; p=0.009; Figure 1b).

Conclusion: We demonstrated that GLS is associated with clinical outcomes in patients with SSc. GLS is a reproducible and cost-effective echocardiographic