

**TABLE 1 • COMPARISON OF EPIDEMIOLOGICAL, CLINICAL, SEROLOGICAL DATA IN SCLERODERMA PATIENTS WITH AND WITHOUT HIGH ASCA LEVELS.**

	ASCA IgG			ASCA IgA		
	Positive N=21	Negative N=53	p	Positive N=12	Negative N=62	p
Mean age (years)	51.4±12.7	52.0±12.1	0.84	52.08±13.8	51.6±12.1	0.92
Gender (male /female)	4/32 (12.5%)	13/39 (33.3%)	1.00	0/12 (12.9%)	8/62 (12.9%)	0.33
Median Disease duration (IQR)	6.5 (4.2-14.0)	9.0 (3.5-12.0)	0.83	9.0 (6.5-17.5)	7.0 (3.0-11.5)	0.15
Tobacco exposure	5/32 (14.2%)	4/31 (12.9%)	0.72	1/12 (8.3%)	8/62 (12.9%)	1.00
Afrodescent/caucasians	18/30 (60%)	4/42 (9.5%)	< 0.0001	6/11 (54.5%)	25/58 (43.1%)	0.48
Median BMI (Kg/m <sup>2</sup> ) (IQR)	24.01 (22.04-28.4)	26.02 (23.1-30.9)	0.09	24.9 (22.1-29.3)	25.4 (22.3-28.6)	0.84
Median Medsger index (IQR)	4 (3.0-8.0)	4.0 (2.5-6.0)	0.98	4.0 (1.0-4.0)	5.0 (3.0-8.0)	0.05
Median Rodnan m (IQR)	4 (2.0-10.5)	6 (2.5-14.0)	0.44	3 (0-10)	6.0 (3.0-13.0)	0.13
Raynaud (100%)	32/32 (100%)	40/42 (95.2%)	0.50	12/12 (100%)	60/62 (96.7%)	1.00
Telangiectasias	6/32 (18.7%)	16/42 (38.09%)	0.07	3/12 (25%)	19/62 (30.6%)	1.00
Digital scars	14/32 (43.7%)	19/42 (45.2%)	0.89	3/12 (25%)	30/62 (48.3%)	0.20
Pneumonitis (40.6%)	13/32 (40.6%)	25/42 (59.5%)	0.10	4/12 (33.3%)	34/62 (54.8%)	0.21
Calcinosis (9.3%)	3/32 (9.3%)	2/41 (4.8%)	0.72	1/12 (8.3%)	4/61 (6.5%)	1.00
Arthritis (34.3%)	11/32 (34.3%)	8/42 (19.04%)	0.13	3/12 (25%)	16/62 (25.8%)	1.00
Myositis (9.3%)	3/32 (9.3%)	4/42 (9.5%)	1.00	1/12 (8.3%)	6/62 (9.5%)	1.00
Esophageal dysmotility (63.3%)	19/30 (63.3%)	23/40 (57.5%)	0.62	6/12 (50%)	36/58 (62.0%)	0.43
Pulmonary hypertension (15.3%)	4/26 (15.3%)	8/32 (25%)	0.51	1/10 (10%)	10/46 (21.7%)	0.67
Anti centromere (36.4%)	8/30 (36.4%)	22/39 (56.4%)	0.01	4/10 (40%)	26/59 (42.3%)	1.00
Anti Scl-70 (6.4%)	2/31 (6.4%)	9/41 (21.9%)	0.10	0/11 (0)	11/61 (18.03%)	0.19
Anti Ro (22.5%)	7/31 (22.5%)	4/40 (10%)	0.19	4/12 (33.3%)	7/59 (11.8%)	0.08
Anti La (12.9%)	4/31 (12.9%)	3/40 (7.5%)	0.69	2/12 (16.6%)	5/59 (8.4%)	0.33
Median ESR (mm) (IQR)	32.5 (18.2-48.5)	25.0 (14.0-41.0)	0.30	32.0 (14.2-55.2)	27 (14.0-41)	0.49
Median C reactive protein (mg/dL) (IQR)	8 (2.4-14.2)	9.5 (4.0-17.5)	0.64	9.0 (5.7-14.7)	8.8 (1.07-16.7)	0.60
Median hemoglobin (g/dL) (IQR)	13 (12-13.5)	13.0 (12.0-14.1)	0.68	13.2 (12.8-13.4)	13.0 (12.0-13.7)	0.34
Scleroderma subset:			0.36			0.13
Limited	19/32(59.3%)	18/42(42.8%)		7/12 (58.3%)	31/62 (50%)	
Generalized	11/32(34.3%)	21/42 (50%)		3/12 (25%)	28/62 (42.1%)	
Others	02/32 (6.2%)	03/42 (7.1%)		2/12 (16.6%)	3/62 (4.9%)	

IQR= interquartile rate; ESR= erythrocyte sedimentation rate; BMI= Body mass index.

negatively with anticentromere antibodies ( $p=0.013$ ); ASCA IgA had a negative association with Medsger score ( $p=0.05$ ). In multivariate analysis IgG ASCA associated independently only with African ethnic background.

**Conclusions:** Positivity for IgG and IgA ASCA are higher among scleroderma patients than controls. African descendants have more positivity for IgG ASCA and ASCA IgA were less commonly seen in patients with more severe disease.

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## Basic science in paediatric rheumatology

### AB0187 LONG-TERM ADMINISTRATION OF FENSPIRIDE HAS NO NEGATIVE IMPACT ON BONE MINERAL DENSITY AND BONE TURNOVER IN YOUNG GROWING RATS

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**Background:** In young organisms intensive bone turnover is observed and it

allows the skeleton to achieve proper size, shape and weight of bones. It is extremely important to assess the influence on various drugs on growing bones. Fenspiride is registered for therapy of acute and chronic respiratory tract infections in children and adolescence. It decreases the synthesis of proinflammatory cytokines, blocks H1 receptors and has bronchodilatory properties.

**Objectives:** The aim of the study was to assess the influence of long-term administration of fenspiride on bone mineral density and selected markers on bone turnover in young growing rats.

**Methods:** The experiment was carried out on 18 young (8-week-old) male Wistar rats receiving standard diet containing 1.2% of calcium and 0.7% of phosphate. Rats were randomly assigned to one of two groups (9 animals in each group): group F – rats receiving fenspiride (15 mg/kg) in saline solution (4ml/kg), and group C (control group) – rats receiving saline solution (4ml/kg). Saline solution and fenspiride were given intragastrically once daily for 90 days (from day 3 to day 92). On day 1 and 93 blood samples for serum isolation were collected. Markers of bone turnover were assessed with commercial ELISA kits according to producers' instruction. Bone mineral density (BMD) was measured by dual-energy X-ray absorptiometry (DXA) with Hologic DXA equipment (Hologic Discovery W 81507) using a small animal software. The experiment was performed with the approval of the First Local Ethics Committee for Experiments on Animals in Wrocław.

**Results:** On Day 1 there was no difference in age and body weight between groups. On Day 1 no difference in total body bone mineral density (BMD) ( $0.160\pm 0.0065$  g/cm<sup>2</sup> vs.  $0.1608\pm 0.0056$  g/cm<sup>2</sup>), lower limbs BMD ( $0.232\pm 0.0267$  g/cm<sup>2</sup> vs.  $0.2274\pm 0.0314$  g/cm<sup>2</sup>), serum levels of bone turnover markers (osteocalcin:  $1000.921\pm 109.0705$  pg/ml vs.  $952.5777\pm 178.5306$  pg/ml; bCTX:  $280.089\pm 54.4298$  pg/ml vs.  $292.8979\pm 116.0042$  pg/ml; osteoprotegerin:  $3.5\pm 0.6338$  pg/ml vs.  $3.7963\pm 0.6894$  pg/ml; RANKL:  $0.167\pm 0.4099$  pg/ml vs.  $1.0218\pm 1.2717$  pg/ml) was detected.

On Day 93 there was no difference in body weight, total body BMD and lower limbs ( $0.212\pm 0.0104$  g/cm<sup>2</sup> vs.  $0.2035\pm 0.0242$  g/cm<sup>2</sup>;  $0.264\pm 0.0159$  g/cm<sup>2</sup> vs.  $0.2520\pm 0.0271$  g/cm<sup>2</sup>, respectively) between groups. On Day 93 no difference between groups in serum bone turnover markers was detected (OC:  $422.758\pm 92.3316$  pg/ml vs.  $429.2071\pm 83.0520$  pg/ml; bCTX:  $307.748\pm 77.6733$  pg/ml vs.  $285.3486\pm 79.16334$ ; OPG:  $5.466\pm 0.7815$  pg/ml vs.  $5.3520\pm 1.6458$  pg/ml; RANKL:  $0.647\pm 0.8457$  pg/ml vs.  $0.5630\pm 0.8608$  pg/ml).

**Conclusions:** Long-term administration of fenspiride has no negative impact on bone mineral density and bone turnover in young growing rats

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### AB0188 EFFECT OF MIR-19A AND MIR-21 ON THE JAK/STAT SIGNALING PATHWAY IN THE PERIPHERAL BLOOD MONONUCLEAR CELLS OF PATIENTS WITH SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

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**Background:** Overexpression of the components of the Janus kinase/signal transducer and activator of transcription (JAK/STAT) signaling pathway are key factors of the pathogenic mechanisms underlying systematic juvenile idiopathic arthritis (SJIA).

**Objectives:** The present study aimed to investigate the association between microRNA (miR)-19a, miR-21 and the JAK/STAT signaling pathway.

**Methods:** A total of 20 patients with SJIA were included in the study, and peripheral blood mononuclear cells (PBMCs) from 20 normal controls were also collected. RNAiso was used to extract total RNA, and the RNA was then reverse transcribed into cDNA. Primers were designed to detect the mRNA of miR-19a and miR-21, and U6 was set as the internal parameter. In addition, the mRNA of STAT3, suppressor of cytokine signaling 3 (SOCS3), interleukin-6 (IL-6) and tumor necrosis factor- $\alpha$  (TNF- $\alpha$ ) was detected, and  $\beta$ -actin was set as the internal parameter. Reverse transcription-quantitative polymerase chain reaction was performed to detect the expression levels of these proteins in patients with SJIA and control subjects, and non-parametric tests were used to analyze the statistical differences in  $2^{-\Delta\Delta Cq}$  between the two groups.

**Results:** The expression levels of miR-19a and miR-21 were significantly lower in the SJIA group compared with the control group ( $P<0.05$ ). SOCS3, TNF- $\alpha$  and STAT3 were shown to be the target genes of miR-19a and miR-21, as determined by Targetscan. The expression levels of STAT3, SOCS3, TNF- $\alpha$  and IL-6 mRNA were significantly higher compared with those of the control group ( $P<0.05$ ).

**Conclusions:** In the PBMCs of the patients with SJIA, miR-19a and miR-21 expression levels were lower compared with those of the control group, and the JAK/STAT signaling pathway was activated, which indicated that miR-19a and miR-21 may participate in the activation of the JAK/STAT signaling pathway.

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