abnormalities and disturbed endocrine secretion have been reported in patients with systemic sclerosis (SSc). Additionally, some of the patients have sleep disturbances or depression, a factors known to effect the MT level. All these findings suggest possible role of MT in pathogenesis of some phenomena seen in SSc patients.

Objectives The aim of the study was to evaluate the circadian profile of serum MT levels in patients with SSc.

Methods Six women with definite SSc, aged 32.4±1.8 yrs (symptoms duration 4–7 yrs) and six age-matched healthy women were investigated. Blood samples were drawn at 8.00, 12.00, 16.00, 20.00, 22.00, 24.00, 02.00, 04.00, 06.00. Light exposure of the subjects was from 07.30 to 21.00. MT was determined with ELISA method.

Results A significant circadian rhythm of serum MT was detected in all investigated subjects. Serum MT levels were lower in the SSc patients than in the controls, MESOR = 21.4 pg/ml and 47.3 pg/ml, in the patients and controls, respectively. In the SSc patients, the night increase in MT was also relatively lower when compared to the daily levels. Maximal values were found between 02.00–04.00 both in the patients and controls.

Conclusion The general nature of the circadian serum MT rhythm is maintained in the SSc patients although secretion of MT is depressed. The mechanism of impaired MT secretion remains unknown. It may be speculated that a low MT level is involved in development of immune abnormalities, neuroendocrine disfunctions and/or impaired free radical elimination. All these phenomena may contribute to development of fibrosis.

FRI0187 THE CLINICAL PROFILE OF JUVENILE SYSTEMIC SCLEROSIS (JSSc)

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Background JSSc represents the unique form of SSc characterised by variable clinical picture. JSSc has specific features maintained during adult period of life.

Objectives The aim of the study was to determine the clinical profile of JSSc.

Methods Sixty pts with JSSc aged from 14 to 54 years (mean 25.1±7.2 yrs) with the onset of the disease at the age of 1 to 16 years (mean 11.4±3.8 yrs) were analysed. Duration of disease was 13.1±7.1, range 1–39 yrs.

Results We revealed three clinical variants of JSSc. The first variant (55% of pts) is characterised by the combination of localised scleroderma (morphea, hemiform-typed) and systemic manifestation such as Raynaud's phenomenon (78%), arthritis (41%), lungs involvement (66%), heart disease (58%), gastrointestinal tract involvement (60%). The visceral signs were moderate and founded by careful clinical instrumental investigation. In the other variant (38% pts) the clinical manifestation of JSSc were similar to adult ISSc with the prevalence of vascular disorders and digital ischaemic changes, limited skin lesion, visceral organ involvement. Musculoskeletal manifestations (arthralgia, arthritis, flexion contractures) were also seen in this group. The pts had slow progressive form of disease. The third variant (17% of pts) may be regarded as overlap syndrome SSc with polymyositis or/and rheumatoid arthritis. The state in these pts mainly depended on the prevailing disease process.

Conclusion The definition of the clinical variants of JSSc may provide the correct diagnosis and adequate treatment.

FRI0188 DIASTOLIC DYSFUNCTION IN PATIENTS WITH SCLERODERMA: A TISSUE DOPPLER STUDY

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Background Systemic sclerosis may involve myocardium and brings about myocardial fibrosis.

Objectives In our study, left ventricular diastolic functions were evaluated with the tissue Doppler echocardiography, a newer and more sensitive method than others.

Methods Standard and tissue Doppler echocardiography were applied to 11 patients with systemic sclerosis. Systolic and valve functions were evaluated with standard echocardiography and then the signals obtained from the lateral section of mitral annulus were analysed with pulse wave tissue Doppler echocardiography. Left ventricular diastolic functions were assessed by calculating early wave (E), atrial wave (A) and E/A ratio of these signals.

Results The impairment of left ventricular diastolic functions was determined in 4 patients. This finding was not related to the duration of disease and the spread of involvement. All patients have normal ejection fractions.

Conclusion Left ventricular diastolic dysfunction is a newly defined special feature for cardiac involvement of systemic sclerosis. This finding may be due to myocardial ischemia, fibrosis, or both of them. However, larger studies are necessary to evaluate the prognostic importance of this finding.

FRI0189 PROGNOSTIC SIGNIFICANCE OF LIMITED AND DIFUSSE FORM OF SKIN CHANGES IN SYSTEMIC SCLEROSIS

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Background

Objectives To assess the difference in frequency of pulmonary and renal involvement, as well as lungs insufficiency and renal failure between patients (pts) with limited cutaneous Systemic Sclerosis (lcSSc) and difusse cutaneous Systemic Sclerosis (dcSSc). **Methods** Group of 138 pts with Systemic Sclerosis (SSc), 127 females and 11 males, age 28–75 years (median 53), were followed for 3–26 (median 13) years period. Extent of skin involvement was recorded, and pts were examined for radiographic signs of pulmonary fibrosis, carbon monoxide transfer (TCO) factor (done in 28 pts), and renal involvement and failure (urine sediment, creatinin clearance, arterial tension). Difference of visceral involvement, pulmonary insufficiency and renal failure between pts with lcSSc and dcSSc was correlated.

Results Among 138 pts, 105(76%) had lcSSc, and 33(24%) dcSSc. Pulmonary lesion were found in 40/105 pts with lcSSc, and in 25/33 pts with dcSSc ($p = 0,004\text{-}\chi^2$ test). Renal lesion was detected in 11/105 pts with lcSSc, and in 8/33 pts with dcSSc ($p = 0,04\text{-}\chi^2$ test). Signs of lung insufficiency developed in 30/105 pts with lcSSc, and in 18/33 pts with dcSSc ($p = 0,04\text{-}\chi^2$ test). Lower creatinin clearance and high arterial blood

pressure were detected in all patients with recorded renal lesion during the follow up period (in 11/105 pts with lcSSc, and in 8/33 pts with dcSSc). Renal failure was significantly more frequent in dcSSc pts ($p = 0,04$ - χ^2 test). In 7(21%) pts with dcSSc no criteria for pulmonary and renal involvement were present during the follow up period. On the other hand, in 14(13%) pts with lcSSc pulmonary insufficiency or renal failure were recorded in first 5 years of disease duration.

Conclusion Difuse cutaneous lesions were prognostic sign of lungs and kidneys involvement, as well as pulmonary insufficiency and renal failure in most of the patients, but about 20% of the dcSSc patients were without severe visceral lesions during the 13 years of follow up period. On the other hand, 13% of patients with limited cutaneous lesions had bad prognosis and developed pulmonary insufficiency or renal failure in first 5 years of disease duration.

FRI0190 SURGICAL TREATMENT FOR GASTROESOPHAGEAL REFLUX DISEASE IN SYSTEMIC SCLEROSIS

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Background

Objectives To describe the experience with the surgical treatment for gastroesophageal reflux disease (GRD) in systemic sclerosis (SSc).

Methods From 202 patients with the diagnosis of SSc attended at the Scleroderma outpatient clinic in the period 1991–2000, ten patients (5%) who presented GRD, with severe chronic esophagitis and stricture, were submitted to surgical treatment. All the patients were female and eight were Caucasian, with seven presenting limited SSc (mean age 53 years and mean disease duration 17 years) and three diffuse SSc (mean age 44 years and mean disease duration 6 years).

Results The correction of the gastroesophageal reflux was performed through videolaparoscopy in nine patients. Seven patients underwent modified Nissen technique and three patients Lind technique. There was satisfactory clinical response, with significant improvement in the heartburn and dysphagia in all the patients. After a mean followup of 34 months (12–76 months), one patient presented clinical relapse (after 54 months) and another patient needed a surgical resection of a paraesophageal hernia (Y-Roux).

Conclusion The surgical treatment of GRD represents an efficient therapeutic option in SSc patients with severe esophagitis and stricture.

FRI0191 SKIN SCORE IN SYSTEMIC SCLEROSIS

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Background

Objectives To report the experience with the use of the Total Skin Score (TSS), in the original (OR-TSS) and modified (MR-TSS) Rodnan methods, in systemic sclerosis (SSc), analysing its clinical significance in the model of two (diffuse and limited) and three (diffuse, intermediate and limited) subgroups.

Methods Prospective study analysing skin thickness (assessed using the original and modified Rodnan TSS methods) in 56 patients with SSc. TSS results were compared with SSc clinical variants (divided in the model of two and three subgroups), pathological findings observed in standardised skin biopsies and cutaneous (calcinosis, telangiectasias, pigmentary abnormalities), systemic (articular, vascular, esophageal, pulmonary, cardiac, renal) and laboratory (antinuclear antibody, anticentromere antibody, anti-topoisomerase I antibody) manifestations. Data were analysed on the basis of whether patients had a low TSS ($= < 20$) or a high TSS (> 20), in both methods. Logistic regression analysis was performed to determine the independent variables that influenced TSS (in the two methods) and death (after a 5-year follow-up).

Results There was statistical association between TSS $= < 20$ and limited SSc (both methods, in the SSc model of two subgroups), grade I skin biopsy (OR-TSS) and calcinosis (MR-TSS). The TSS > 20 was associated with diffuse SSc (in the SSc models of two and three subgroups), pigmentary abnormalities and articular involvement (both methods) and grade II and III skin biopsy (OR-TSS). There was a statistical trend between TSS $= < 20$ and calcinosis and anticentromere antibody (OR-TSS), as well as between TSS > 20 and pulmonary restrictive disease (MR-TSS) and anti-topoisomerase I antibody (OR-TSS). The multivariate logistic regression analysis indicated that the clinical variant was the most important factor that influenced significantly TSS (in both methods). In the SSc model of two subgroups, the diffuse variant was associated with TSS > 20 (both TSS methods) and pigmentary abnormalities, with a statistical trend related to pulmonary restrictive disease and anti-topoisomerase-I antibody, whereas the limited variant was associated with TSS $= < 20$, calcinosis and anticentromere antibody. In the SSc model of three subgroups, the diffuse variant was associated with TSS > 20 (both TSS methods) and pigmentary abnormalities, whereas the limited variant was associated with grade I skin biopsy; the intermediate variant was associated with calcinosis and telangiectasias. After a 5-year follow-up, there were 20 deaths (35.7%); the multivariate logistic regression analysis indicated that the vascular involvement influenced significantly death, despite the TSS score.

Conclusion TSS, in both original and modified Rodnan methods, represents an useful tool in the management of the skin involvement in SSc, permitting to differentiate distinct patterns in the clinical spectrum of the disease.

FRI0192 LIPID PROFILE IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background Systemic sclerosis (SSc) is an inflammatory connective tissue disorder characterised by collagen proliferation, vascular damage and internal organ involvement. It is speculated that vascular changes in SSc patients may be at least partially secondary to impaired lipid metabolism. Hypothyroidism has been reported in patients with SSc and thyroid involvement may additionally effect lipid metabolism.

Objectives The aim of the present study was to evaluate the serum lipid profile in patients with SSc with regard to their thyreometabolic state.