Table 1. Clinical characteristics, demographics and outcome

	No ILD (n=227)	Subclinical ILD (n=67)	Clinical ILD (n=231)
Age, y (SD)	50 (15.4)	51 (14.4)	52 (15.3)
Male sex, n (%)	89 (39)	22 (33)	111 (48)
Deceased, n (%)	50 (22)	12 (18)	91 (39)
Observation period, y median (range)	13.7 (18.6)	13.9 (17.9)	11.5 (17.1)
FVC% (SD)	97 (18.6)	99 (17.9)	81 (20.9)
FVC decline% (SD)	-0.70 (11.1)	-0.81 (16.5)	-1.61 (15.9)
DLCO% (SD)	73 (19.4)	73 (16.9)	55 (17.4)
Extent of ILD% (SD)	0 (0)	2.3 (1.5)	19.3 (16.8)
ILD progression% (SD)	0.08 (1.0)	3.1 (6.2)	3.6 (9.9)
ILD progressors, n (%)	3 (2)	20 (38)	72 (51)

Figure 1. Time to ILD progression in CTD without ILD, with subclinical and clinical ILD

Conclusion: Subclinical ILD is frequently present across CTDs and progresses over time in a substantial subgroup of patients, comparable to patients with clinical ILD. Our findings question the terms sub- and preclinical ILD, which may potentially lead to a suboptimal “watchful waiting management strategy”. Monitoring all CTD patients with any ILD is of high importance to identify disease progression early.

Disclosure of Interests: Anna-Maria Hoffmann-Vold Speakers bureau: Actelion, Boehringer Ingelheim, Roche, Merck Sharp & Dohme, Lilly and Medscape, Consultant of: Actelion, Boehringer Ingelheim, Bayer, ARXX, and Medscape, Grant/research support from: Boehringer Ingelheim, Helena Andersson: None declared, Silje Reiser: None declared, Håvard Frøtheim Consultant of: Actelion, Bayer, Imon Barua: None declared, Torhild Garen: None declared, Øyvind Midtvedt: None declared, Ragnar Gunnarsson: None declared, Mike Durheim Speakers bureau: Boehringer Ingelheim and Roche, Consultant of: Boehringer Ingelheim, Grant/research support from: Boehringer Ingelheim, Trond M Aaløkken: None declared, Øyvind Molberg: None declared

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OP0175 INTERNATIONAL MULTICENTRIC PROSPECTIVE STUDY ON PREGNANCY IN SYSTEMIC SCLEROSIS (IMPRESS-2)

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Background: Data about the obstetric outcomes of pregnant women with Systemic Sclerosis (SSc) mainly derive from retrospective studies. Moreover, little evidence is available to define if pregnancy impacts on SSc course and if children of SSc mothers have a normal post-natal development.

Objectives: To assessed the obstetric, pediatric and rheumatologic outcomes of SSc pregnancies in a prospective controlled study

Methods: Prospective recruitment of three cohorts.

1) 110 pregnant women with SSc.

2) 218 control pregnancies without systemic autoimmune diseases

3) 78 non-pregnant control SSc with a matching subject in cohort-1.

SSc was characterized for disease activity, severity, cutaneous/organ involvement and therapy. Women and their offspring were followed until 21 months after enrollment (ie, 12 months after expected delivery).

Results: Gestational and neonatal outcomes

Miscarriages and fetal death occurred in 7% and 5% of SSc pregnancies. Compared to control pregnancies, SSc pregnancies had higher rates of gestational hypertension (12% Vs 4%, p=0.004), pre-eclampsia (9% Vs 1%, p=0.002), fetal growth-restriction (13% Vs 4%, p=0.004), prematurity (26% Vs 7%, p<0.001) and cesarean section (52% Vs 4%, p=0.002). Newborns from SSc mothers weighted less (2773 Vs 3243g, p<0.001), were more frequently small for gestational age (SGA, 18% Vs 12%, p=0.05) and required neonatal-intensive care unit (ICU) more frequently (12% Vs 1%, p<0.001). Rates of newborn malformation/death, and one year-pediatric outcomes were similar.

Univariate and multivariate analyses were performed for relevant outcomes. For example, pre-eclampsia positively associated with baseline skin score and its evolution during pregnancy (p=0.015 and 0.013), immunosuppressive agents, bosentan and iloprost at baseline (p=0.001, 0.041 and 0.007), and twin pregnancy (p=0.006). Multivariate logistic regression for pre-eclampsia in SSc identified baseline arterial hypertension, immunosuppressive agents or of iloprost, twin pregnancy and assisted conception as risk factors (p=0.000, 0.002, 0.0025, 0.000 and 0.027), and baseline calcium channel blockers as protective factors (p=0.001).

SSc course during pregnancy

As compared to matched non-pregnant SSc, SSc pregnant women had lower Medsger disease severity index (0.6 +/-0.9 Vs 1.0 +/-1.1, p=0.022), health assessment questionnaire (HAQ, 0.21 +/-0.38 Vs 0.40 +/-0.55, p=0.026), and lower rate of iv iloprost (21% Vs 42%, p=0.006). Pregnant and non-pregnant SSc women had a similar disease course during the 21 months of follow-up, despite a lower use of immunosuppressive agents (17% Vs 36%, p=0.014). Two scleroderma-renal-crises (SRC) occurred during pregnancy (one was a relapse of a previous SRC; the other occurred at week 33, and resolved after premature delivery of a SGA newborn and ACE-inhibitors, preventing differentiation from pre-eclampsia).

Conclusion: SSc pregnancies have generally a favorable obstetric/pediatric outcome, albeit at a higher risk of gestational hypertension, pre-eclampsia, fetal growth restriction, prematurity, delivery of SGA newborn and requirement of neonatal-ICU. Pregnancy does not impact on SSc course, although SRC during late pregnancy and pre-eclampsia might be hardly discriminated.

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OP0176 ULTRASOUND (US) EVALUATION OF BOWEL VASCULOPATHY IN SYSTEMIC SCLEROSIS (SSC)

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Background: Gastrointestinal involvement is one of the most frequent features of SSc, affecting nearly 90% of patients, with a great impact on quality of life and morbidity. One of the key pathological factors of SSc bowel involvement is vasculopathy (1), although little is known about its pathophysiology and no treatments are currently available.

Objectives: To assess with abdominal US the superior mesenteric artery (SMA) and the inferior mesenteric artery (IMA) vessel characteristics and blood flow in SSc, compared to healthy controls (HC).

Methods: we performed fasting abdominal US in SSc patients fulfilling the ACR/EULAR 2013 classification criteria and HC. Patients with a history of peripheral / coronary arterial disease were excluded. For both SMA and IMA, caliber (mm), Peak Systolic Velocity – PSV (cm/sec), Reverse Velocity – RV (cm/sec), End-Diastolic Velocity – EDV (cm/sec), Mean Velocity – mV (cm/sec), Blood-flow (cm/sec), Resistive Index – RI and Pulsatility Index – PI were measured.

Results: 28 SSc patients [25 females (89.3%), mean age 48.75 ± 12.39 years; 6 (22.22%) anti-centromere and 19 (70.37%) positive for anti-topoisomerase I antibodies] and 28 HC [18 females (64.3%), mean age 36.25 ± 12.08 years] were evaluated. In SSc, the SMA caliber was significantly smaller than in HC (5.75 ± 0.62 vs. 6.45 ± 0.60 mm, p<0.0001), while IMA dimensions did not differ.

The SMA study revealed SSc patients had a significant reduction of RV (7.25 ± 6.37 vs. 18.52 ± 6.16 cm/sec, p<0.0001) and PI (3.33 ± 0.75 vs. 4.53 ± 1.03, p<0.0001) when compared to HC. In addition, in SSc the mV of SMA was significantly lower than in HC (38.03 ± 13.90 vs. 28.32 ± 9.25 cm/sec, p=0.0035), as well as the RI (0.88 ± 0.04 vs. 0.91 ± 0.03, p=0.0034); EDV was significantly increased (16.34 ± 7.03 vs. 12.64 ± 5.46 cm/sec, p=0.0321). Similarly to SMA, also in IMA RV and PI were significantly lower than controls (RV: 2.69 ± 6.10 vs. 17.06 ± 5.75 cm/sec, p<0.0001; PI: 3.54 ± 0.95 vs. 6.08 ± 1.53, p<0.0001). Moreover SSc patients presented a significant reduction of PSV and RI of IMA (PSV: 72.27 ± 27.23 vs. 93.81 ± 25.73 cm/sec, p=0.0084; RI: 0.88 ± 0.04 vs. 0.91 ± 0.03, p=0.0132) when compared to HC. Although the HC group was significantly younger than the SSc group (p=0.0003), all the results were confirmed after adjustment for age (Table 1).