study data were analyzed using analysis of covariance models. Results reported as least squares mean (LSM) change from baseline±standard error (SE). Integrated data of COAST-V and -W were analyzed for inferential statistics due to small sample sizes in the individual studies. ADA was included in COAST-V as an active reference arm only, and the study was not powered to allow comparisons between the IXE and ADA arms.

Results: Overall, 365 (64.5%) PBO- and IXE-treated patients reported paid work (part-time or full-time) at baseline in COAST-V and COAST-W. Based on integrated data, Absenteeism was significantly lower in IXEQ2W treated patients than PBO and Presenteeism, Work Productivity Loss, and Activity Impairment were significantly reduced in IXEQ2W and IXEQ4W treated patients versus PBO (Table 1). WPAI-SpA outcomes for biologic-naïve and TNFi-experienced were similar with some exceptions (Table 2). The active reference arm (ADA) exhibited numerical improvements for presenteeism, work productivity, and activity impairment versus PBO.

Table 1. Integrated WPAI-SpA results for patients with r-axSpA

	PBO	IXEQ2W	IXEQ4W
Absenteeism	-1.7±1.7 n=107	-7.3±1.8* n=95	-1.5±1.7 n=104
Presenteeism	-14.0±2.3 n=104	-24.0±2.4*** n=99	-22.0±2.3* n=99
Work Productivity Loss	-14.7±2.4 n=104	-24.9±2.5*** n=99	-22.0±2.4* n=99
Activity Impairment	-13.3±1.7 n=105	-22.4±1.7*** n=179	-20.8±1.7*** n=192

IXEQ2W = ixekizumab every 2 weeks; IXEQ4W = ixekizumab every 4 weeks; n = number of patients in the analysis category; PBO = placebo

Table 2. WPAI-SpA results for biologic-naïve and TNFi-experienced patients with r-axSpA

	PBO	IXEQ2W	IXEQ4W	ADA
Biologic Naïve (COAST-V)				
Absenteeism	-1.3±2.3 n=50	-6.7±2.3 n=49	1.2±2.2 n=57	-1.2±2.0 n=65
Presenteeism	-17.7±3.1 n=50	-23.3±3.1 n=47	-22.7±2.9 n=55	-20.9±2.8 n=60
Work Productivity Loss	-17.8±3.3 n=50	-24.1±3.3 n=47	-21.4±3.1 n=55	-21.4±2.9 n=60
Activity Impairment	-14.1±2.3 n=86	-23.4±2.3 n=83	-23.0±2.4 n=80	-21.1±2.2 n=88
TNFi Experienced (COAST-W)				
Absenteeism	-1.2±2.8 n=57	-6.9±3.1 n=46	-4.7±2.9 n=47	
Presenteeism	-8.9±3.6 n=54	-22.2±4.1 n=43	-19.5±3.9 n=44	
Work Productivity Loss	-9.8±3.7 n=54	-23.5±4.2 n=43	-21.0±4.0 n=44	
Activity Impairment	-10.1±2.6 n=99	-18.4±2.7 n=96	-16.5±2.4 n=112	

IXEQ2W = ixekizumab every 2 weeks; IXEQ4W = ixekizumab every 4 weeks; n = number of patients in the analysis category; PBO = placebo

Conclusion: In this integrated analysis of patients with active r-axSpA, IXE treatment for 16 weeks yielded significant improvements in work productivity and activity impairment versus PBO. The effect is similar in biologic-naïve and TNFi-experienced patients. A limitation for evaluations of work productivity in the individual studies was the relatively low number of employed patients.

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SSc and Myositis – novel therapeutic options_____

OP0239

PROGRESSIVE LUNG FIBROSIS IN PATIENTS WITH SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE IN THE EUSTAR DATABASE

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Background: Some patients with systemic sclerosis-associated interstitial lung disease (SSc-ILD) develop progressive ILD, but the prevalence is unknown and no validated algorithms for early detection exist.

Objectives: To assess the frequency of, and parameters associated with, progressive SSc-ILD in the EUSTAR database.

Methods: Patients from the EUSTAR database registered after 2010, ≥18 yrs old, fulfilling SSc classification criteria, with serial lung function, HRCT assessments and disease duration recordings were eligible. Progressive ILD, assessed as absolute changes in %predicted, was defined as: significantly progressive (FVC decline >10% or FVC decline 5–10% and DL_{CO} decline ≥15%); moderately progressive (FVC decline 5–10%); or stable (<5% FVC change in either direction). Lung fibrosis changes from baseline to 12±3 months, disease course and cumulative frequency of progressive ILD during the 5-yr follow-up were assessed. Candidate predictors of significant progressive ILD were selected by expert opinion, and logistic regression applied. Time to event was evaluated by Kaplan-Meier estimates.

Figure: Time to first SSc-ILD progression events of A. Moderately progressive ILD and B. Significantly progressive ILD in the EUSTAR database over a 5-year follow-up period

Results: 826 SSc-ILD patients were included; 219/826 (26.5%) showed moderate/significant ILD progression at 12±3 months' follow-up. Baseline FVC, erythrocyte sedimentation rate, modified Rodnan skin score (mRSS), GERD and disease duration were significantly associated with significant progressive fibrosis in uni- and multivariable modelling (area under curve 0.67). Serial lung function data with median follow-up of 5 yrs were available in 411/826 (49.8%) patients. In 128/411 with initial progressive ILD at 12±3 months, 59/128 (46.1%) showed further progression, while 69/128 (53.9%) were stable. Baseline FVC, mRSS and disease duration were significantly associated with further ILD progression. The incident cumulative frequency of moderate and significant ILD progression was 22.9% (n=189/826) and 28.5% (n=235/826) during the median 5-yr follow-up and time to first progressive event (Figure).

Conclusion: This study provides novel insights into the frequency, severity and association of progressive fibrosis in patients with SSc-ILD in the EUSTAR database.

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OP0240

ASSESSMENT OF PULSE WAVE VELOCITY IN SYSTEMIC SCLEROSIS: POTENTIAL BENEFICIAL EFFECTS OF BOSENTAN ON FOREARM ARTERIAL STIFFNESS? AN EXPLORATORY STUDY

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Background: Systemic sclerosis (SSc)-related vasculopathy is generally thought to occur on a microvascular level. However, some observations also suggest involvement of arterial vessels. Macrovascular involvement (e.g., aorta or upper extremity) can be non-invasively assessed by measuring pulse wave velocity (PWV). Although, several studies have assessed aortic and upper extremity PWV in SSc, studies have not reached a consensus regarding this matter. Furthermore, we hypothesized that the endothelin antagonist bosentan may improve arterial stiffness by its direct endothelial and potential anti-fibrotic effects.

Objectives: The aim of this exploratory study was two-fold. First, we aimed to compare arterial stiffness in patients with SSc and age and sex-matched healthy controls (HC). Secondly, we will investigate the effect of bosentan on both short-term (three month) and long-term (one year) PWV.

Methods: Baseline differences between HC and SSc patients were studied in a case-control design. The follow-up of SSc patients was a randomized, prospective, 2-arm parallel group, open-label, (usual care with bosentan versus usual care only), blinded endpoint, intervention study. PWV (Sphygmocor) in meters/second, was measured to assess arterial stiffness in the aorta (carotid-femoral PWV), upper arm (carotid-brachial PWV), and forearm (brachial-radial PWV), adjusted for mean arterial pressure.

Results: Baseline characteristics are shown in table 1. No significant differences were observed in PWV (at all sites) between HC and SSc patients. No effect of bosentan on aortic, and upper arm PWV was found. The change in forearm PWV was different between the groups, with a decrease (e.g. lowering arterial stiffness) in the bosentan group (figure 1).

Table 1. Baseline characteristics

	Healthy controls (n=19)	Systemic sclerosis patients (n=19)	Bosentan group (n=9)	Usual care group (n=7)
Female, n (%)	13 (69)	13 (69)	5 (56)	6 (86)
Age in years, median (IQR)	53 (47–63)	50 (44–54)	52 (49–66)	44 (43–54)
Pack years, median (IQR)	0 (0–7.5)*	2.5 (0–30.6)*	17.3 (0.7–34.1)	0.8 (0–2.75)

Body mass index in kg/m ² , median (IQR)	25 (23–27)	25 (21–29)	25 (22–27)	26 (20–37)
Systolic blood pressure in mm Hg, median (IQR)	130 (121–137)	127 (123–134)	126 (117–142)	127 (123–132)
Diastolic blood pressure in mm Hg, median (IQR)	81 (72–88)	85 (79–89)	84 (76–87)	83 (79–85)
Raynaud's phenomenon duration in years, median (IQR)	N/A	8.0 (4.0–13.0)	1.0 (1.0–7.0)	13.0 (6.0–16.0)
Systemic sclerosis disease duration since first non-Raynaud symptom in years, median (IQR)	N/A	2.0 (1.0–7.0)	1.0 (1.0–7.5)	5.0 (2.0–14.0)

*difference between groups with a p<0.05

Figure 1. The median (IQR) of the brachial-radial pulse wave velocity over time of the bosentan and usual care group

Conclusion: This small study shows that aortic, upper arm, and forearm arterial stiffness does not appear to be increased in patients with SSc, as compared to age- and sex-matched healthy controls. To the best of our knowledge, this is the first study to investigate the concept of potential effects of an endothelin receptor antagonist on macrovascular involvement in SSc. Although the results demonstrate no effects on aorta and upper arm arterial stiffness, they may indicate a beneficial effect on the stiffness of the smaller arteries of the forearm. Future studies are needed to further investigate the potential effect of bosentan on these smaller arteries.

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OP0241

SAFETY AND EFFICACY OF LENABASUM IN AN OPEN-LABEL EXTENSION OF A PHASE 2 STUDY OF LENABASUM IN REFRACTORY SKIN-PREDOMINANT DERMATOMYOSITIS (DM) SUBJECTS

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Background: Lenabasum is a synthetic, non-immunosuppressive, selective cannabinoid receptor type 2 agonist that activates resolution of innate immune responses. Lenabasum had acceptable safety and tolerability and improved efficacy outcomes in the initial 16-week double-blind, randomized, placebo-controlled Part A of Phase 2 trial JBT101-DM-001 (NCT02466243) in dermatomyositis (DM) subjects with refractory, skin-predominant involvement.

Objectives: To provide long-term safety and efficacy data in DM subjects in this study.

Methods: Subjects who completed Part A were eligible to receive oral lenabasum 20 mg BID in an open-label extension (OLE) that assessed safety and efficacy at 4 weeks, then every 8 weeks.

Results: 20/22 (90.9%) eligible subjects received open-label lenabasum, following a mean interval of 31 weeks from end of Part A, during which when they received only standard-of care, to start of OLE during which lenabasum 20 mg BID was added. 17/20 (85.0%) subjects were on stable baseline