EULAR/ EUSTAR: agreement with 2017 guidelines

Physician personal data

1. In which geographical area are you currently working?

- ○ Africa
- ○ Asia
- ○ Australia
- ○ Europe
- ○ North America
- ○ South America

2. What is your speciality?

- ○ Dermatology
- ○ Internal Medicine
- ○ Rheumatology
- ○ Other

3. Is your center affiliated to EUSTAR?

- ○ Yes
- ○ No

4. Are you a member of EUSTAR?

- ○ Yes
- ○ No

5. Are you a member of an official body (national networks, SCTC, …)?

- ○ Yes
- ○ No

6. Since how long are you involved in treatment patients with SSc?

- ◯ 1-5 years
- ◯ 6 - 10 years
- ◯ more than 10 years

7. How many patients with SSc have you seen during the past 6 months?

8. How many patients with a new diagnosis (< 1 year) of SSc have you seen during the past 6 months?

9. Are you involved in clinical trails in SSc?

- ◯ yes
- ◯ no

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SSc-related Raynaud's phenomenon (RP)

Please indicate, with a mark between 0 and 10, for the statements below, your level of agreement, with 0 = disagree, and 10 = fully agree.
For questions regarding availability we kindly ask you to answer in accordance with availability of the drug in your current situation.

10. *Recommendation: Dihydropyridine-type calcium antagonists, in particular oral nifedipine, should be considered as first-line therapy for SSc-related RP.*
Do you agree that dihydropyridine-type calcium antagonists should be used for the treatment of SSc-related RP?

0 = disagree; 10 = fully agree

0 10

11. Are dihydropyridine-type calcium antagonists available for the treatment of SSc-related RP in your country/practice?

- yes
- no

12. In your opinion, is this recommendation useful in every day clinical practice?

0= not useful; 10 = very useful

0 10

13. *Recommendation: Phosphodiestherase-5 inhibitors (PDE-5) should be considered for the treatment of SSc-related RP.*
Do you agree that PDE-5 inhibitors should be used for the treatment of SSc-related RP?

0 = disagree; 10 = fully agree

0 10

14. Are PDE-5 inhibitors available for the treatment of SSc-related RP in your country/ practice?

- ◯ yes
- ◯ no

15. In your opinion, is this recommendation useful in every day clinical practice?

0 10

16. *<u>Recommendation:</u> Fluoxetine should be considered for the treatment of SSc-related RP.*
Do you agree that fluoxetine should be considered for the threatment of SSc-related RP?

0 = disagree; 10 = fully agree

0 10

17. Is fluoxetine available for the treatment of SSc-related RP in your country/practice?

- ◯ yes
- ◯ no

18. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

19. *<u>Recommendation:</u> Intravenous prostanoids (in particular, iloprost) should be considered for the treatment of SSc-related RP after failure on oral therapy.*
Do you agree that intravenous prostanoids (in particular, iloprost) should be considered for the treatment of SSc-related RP after failure on oral therapy?

0 = disagree; 10 = fully agree

0 10

20. Are intravenous prostanoids (in particular, iloprost) available for the treatment SSc-related RP in your country/practice?

- ◯ yes
- ◯ no

21. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 — 10

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SSc-related digital vasculopathy (digital ulcers, DUs)

Please indicate, with a mark between 0 and 10, for the statements below your level of agreement, with 0 = disagree, and 10 = fully agree. For questions regarding availability we kindly ask you to reflect availability of the drug in your current situation.

22. <u>*Recommendation:*</u> *Intravenous prostanoids (in particular, iloprost) should be considered in the treatment of SSc patients with active DUs.*
Do you agree that intravenous prostanoids (in particular, iloprost) should be considered in the treatment of SSc patients with active DUs?

0 = disagree; 10 = fully agree

0 — 10

23. Are intravenous prostanoids (in particular, iloprost) available for the treatment of SSc-related active DUs in your country/practice?

- ○ yes
- ○ no

24. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 — 10

25. <u>*Recommendation:*</u> *PDE-5 inhibitors should be considered for the treatment of SSc-related DUs.*
Do you agree that PDE-5 inhibitors should be considered for the treatment of SSc-related DUs?

0 = disagree; 10 = fully agree

0 — 10

26. Are PDE-5 inhibitors available for the treatment of SSc-related DUs in your country/practice?

- ○ yes
- ○ no

27. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

28. <u>*Recommendation:*</u> *Bosentan should be considered for the reduction of the number of new DUs in patients with SSc, especially in patients with multiple DUs despite the treatment with dihydropyridine-type calcium antagonists, PDE-5 inhibitors and/or iloprost.*
Do you agree that bosentan should be considered for the reduction of the number of new DUs in patients with SSc, especially in patients with multiple DUs despite treatment with dihydropyridine-type calcium antagonists, PDE-5 inhibitors and/or iloprost?

0 = disagree; 10 = fully agree

0 10

29. Is bosentan is available as treatment for the reduction of new DUs in patients with SSc in your country/practice?

- yes
- no

30. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

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SSc-related pulmonary arterial hypertension (PAH)

In most countries, prescription of PAH specific drugs is allocated to tertiairy care centers that dispose of PH- specialists. Therefore, not all specialists involved in SSc care might be directly involved in the decision to start PAH-specific drugs.
This page starts with 3 general questions on treatment of PAH (q 31-33); if you do not feel comfortable to answer questions regarding PAH treatment, you can skip questions 34 to 48, and continu on page 5.
For questions 34-48:
Please indicate, with a mark between 0 and 10, for the statements below your level of agreement, with 0 = disagree, and 10 = fully agree. For questions regarding availability we kindly ask you to reflect availability of the drug in your current situation.

31. Are you prescribing PAH-specific drugs yourself?

- ◯ yes
- ◯ no

32. Do you have to refer a patient for treatment of PAH to a pulmonologist/ cardiologist/ other?

- ◯ yes
- ◯ no

33. Do you feel comfortable to answer the survey about PAH-specific drugs?

- ◯ yes
- ◯ no

In case your answer is 'no', please continue on page 5, about SSc-related skin fibrosis and interstitial lung disease.

34. *Recommendation: Endothelin receptor antagonists (ERA),including ambrisertan or bosentan or macitentan) should be considered for the treatment of SSc-related PAH.*
Do you agree that ERA (including ambrisertan or bosentan or macitentan should be considered for the treatment of SSc-related PAH?

0 = disagree; 10 = fully agree

0 — 10

35. Are ERA (including ambrisertanor bosentan or macitentan) available for the treatment of SSc-related PAH in your country/practice?

- yes
- no

36. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

37. *Recommendation: PDE-5 inhibitors (including sildenafil or tadalafil) should be considered for the treatment of SSc-related PAH.*
Do you agree that PDE-5 inhibitors (including sildenafil or tadalafil) should be considered for the treatment of SSc-related PAH?

0 = disagree; 10 = fully agree

0 10

38. Are PDE-5 inhibitors available for the treatment of SSc-related PAH in your country/practice?

- yes
- no

39. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

40. *Recommendation: Riociguat should be considered for treatment of SSc-related PAH.*
Do you agree that Riociguat should be considered for the treatment of SSc-related PAH?

0= disagree; 10 = fully agree

0 10

41. Is Riociguat available for the treatment of SSc-related PAH in your country/practice?

- yes
- no

42. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

43. <u>Recommendation:</u> *Intravenous epoprostenol should be considered for the treatment of SSc patients with severe PAH (functional class III and IV).*
Do you agree that intravenous epoprostenol should be considered for the treatment of SSc patients with severe PAH (functional class III and IV)?

0 = disagree; 10 = fully agree

0 10

44. Is intravenous epoprostenol available for the treatment of SSc patients with severe PAH (functional class III and IV) in your country/practice?

- yes
- no

45. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful;10 = very useful

0 10

46. <u>Recommendation:</u> *Prostacyclin analogues including iloprost or treprostinil should be conidered for the treatment of SSc-related PAH.*
Do you agree that Prostacyclin analogues including iloprost or treprostinil should be considered for the treatment of SSc-related PAH?

0 = disagree; 10 = fully agree

0 10

47. Are Prostacyclin analogues including iloprost or treprostinil available for the treatment of SSc-related PAH in your country/practice?

- yes
- no

48. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

EULAR/ EUSTAR: agreement with 2017 guidelines

SSc-related skin fibrosis and interstitial lung disease (ILD)

Please indicate, with a mark between 0 and 10, for the statements below your level of agreement, with 0 = disagree, and 10 = fully agree. For questions regarding availability we kindly ask you to reflect availability of the drug in your current situation.

49. *Recommendation: Methotrexate should be considered for the treatment of skin fibrosis in early diffuse SSc.*
Do you agree that methotrexate should be considered for the treatment of skin fibrosis in early diffuse SSc?

0 = disagree; 10 = fully agree

0 10

50. Is methotrexate available for the treatment of skin fibrosis in early diffuse SSc in your country/practice?

- yes
- no

51. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

52. *Recommendation: Cyclophosphamide should be considered for the treatment of SSc-related ILD.*
Do you agree that cyclophosphamide should be considered for the treatment of SSc-related ILD?

0 = disagree; 10 = fully agree

0 10

53. Is cyclophosphamide available for the treatment of SSc-related ILD in your country/practice?

- yes
- no

54. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

55. *<u>Recommendation:</u> Hematopoietic Stem cell transplantation (HSCT) should be considered for the treatment of patients with rapidly progressive SSc at risk of developing of organ complications.*
Do you agree that HSCT should be considered for the treatment of patients with rapidly progressive SSc at risk of developing organ complications?

0 = disagree; 10 = fully agree

0 10

56. Is HSCT available for the treatment of patients with rapidly progressive SSc at risk of developing organ complications in your country/practice?

- yes
- no

57. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

EULAR/ EUSTAR: agreement with 2017 guidelines

Scleroderma renal crisis

Please indicate, with a mark between 0 and 10, for the statements below your level of agreement, with 0 = disagree, and 10 = fully agree. For questions regarding availability we kindly ask you to reflect availability of the drug in your current situation.

58. *Recommendation: Angiotensin converting enzyme (ACE) inhibitors should be used for the treatment of scleroderma renal crisis.*
Do you agree that ACE-inhibitors should be used for the treatment of scleroderma renal crisis?

0 = disagree; 10 = fully agree

0 10

59. Are ACE-inhibitors available for the treatment of scleroderma renal crisis in your country/practice?

- yes
- no

60. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

61. *Recommendation: SSc patients on steriods should be monitored carefully for blood pressure and renal function.*
Do you agree that SSc patients on steriods should be monitored carefully for blood pressure and renal function?

0 = disagree; 10 = fully agree

0 10

62. In your opinion, is this recommendation useful in every day clinical practice?

0 =not useful; 10 = very useful

0 10

EULAR/ EUSTAR: agreement with 2017 guidelines

SSc-related gastrointestinal disease

Please indicate, with a mark between 0 and 10, for the statements below your level of agreement, with 0 = disagree, and 10 = fully agree. For questions regarding availability we kindly ask you to reflect availability of the drug in your current situation.

63. *Recommendation: Proton pump inhibitors (PPI) should be used for the prevention of SSc-related gastro esophageal reflux disease (GERD), esophagal ulcers and strictures.*
Do you agree that PPI should be used for the prevention of SSc-related GERD, esophageal ulcers and strictures?

0 = disagree; 10 = fully agree

0 10

64. Are PPI available as treatment for the prevention of SSc related GERD, esophageal ulcers and strictures in your country/practice?

- yes
- no

65. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

66. *Recommendation: Prokinetic drugs should be used for the management of SSc related motility disturbances (dysphagia, GERD, early satiety, bloating, pseudo-obstruction, etc.)*
Do you agree that prokinetic drugs should be used for the management of SSc related motility disturbances (dysphagia, GERD, early satiety, bloating, pseudo-obstruction, etc.)?

0 = disagree; 10 = fully agree

0 10

67. Are prokinetic drugs available for the management of SSc related motility disturbances (dysphagia, GERD, early satiety, bloating, pseudo-obstruction, etc.) in your country/practice?

- yes
- no

68. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

69. *Recommendation: In SSc patients with malabsorption caused by bacterial overgrowth, rotating antibiotics may be useful.*
Do you agree that in SSc patients with malabsorption caused by bacterial overgrowth, rotating antibiotics may be useful?

0 = disagree; 10 = fully agree

0 10

70. Are antibiotics available for the treatment of SSc related malabsorption caused by bacterial overgrowth in your country/practice?

- yes
- no

71. In your opinion, is this recommendation useful in every day clinical practice?

0 = not useful; 10 = very useful

0 10

72. Please, feel free to add any additional questions ro remarks from your side:

Thank you for your participation!

ONLINE SUPPLEMENTARY DATA

Supplementary Table 1. Recommendations for treatment of systemic sclerosis; level of agreement, perception of usefulness and availability for SSc experts worldwide

	Agreement (mean, SD)	Usefulness (mean, SD)	Availability (%)
CCB should be considered as first-line therapy for SSc-RP	8.2 (2.7)	8.0 (2.7)	98
PDE-5I should be considered in treatment of SSc-RP	7.3 (2.7)	6.7 (3.0)	59
Fluoxetine might be considered in treatment of SSc-RP attacks	4.6 (2.8)	4.6 (3.0)	62
Intravenous Iloprost should be considered for severe SSc-RP	7.9 (2.7)	7.3(3.0)	79
Intravenous iloprost should be considered in the treatment of DU in SSc-patients	8.7 (2.3)	8.0 (2.7)	82
PDE- 5I should be considered in the treatment of DU in SSc-patients	8.0 (2.5)	7.9 (2.7)	66
Bosentan should be considered for reduction of number of new DU in SSc patients	7.9 (2.8)	7.2 (3.1)	77
ERA should be considered to treat SSc-related PAH*	8.8 (2.4)	8.7 (2.4)	93
PDE-5I should be considered to treat SSc-related PAH*	8.9 (2.3)	8.7 (2.4)	98

Riociguat should be considered to treat SSc-related PAH*	7.4 (2.8)	6.8 (3.1)	60
Intravenous epoprostenol should be considered for treatment of patients with severe SSc-related PAH*	8.3 (2.5)	7.7 (2.9)	77
Prostacyclin analogues should be considered to treat SSc-related PAH*	8.0 (2.7)	7.4 (3.0)	87
MTX may be considered for skin manifestations of early diffuse SSc	7.4 (2.8)	7.4 (2.8)	94
Cyclophosphamide should be considered for treatment of SSc-ILD, in particular for patients with progressive ILD	8.0 (2.6)	7.9 (2.6)	100
HSCT should be considered for treatment of selected patients with rapidly progressive SSc at risk of organ failure	7.1 (2.9)	6.6 (3.2)	66
Experts recommend immediate use of ACEI in the treatment of SRC	9.2 (2.1)	9.1 (2.1)	100
Blood pressure and renal function should be carefully monitored in SSc patients treated with glucocorticoids	9.0 (2.2)	8.8 (2.4)	na
PPI should be used for the treatment of SSc related GERD, and prevention of oesophageal ulcers and strictures	9.0 (2.2)	8.9 (2.2)	100

Prokinetic drugs should be used for the management of SSc-related symptomatic motility disturbances	8.0 (2.4)	7.8 (2.5)	97
Intermittent or rotating use of antibiotics to treat symptomatic small intestine bacterial overgrowth in patients with SSc	8.5 (2.1)	8.2 (2.5)	99

*N= 166: of n=209 complete responders 21% did not complete the questions regarding PAH specific drugs because they stated to feel uncomfortable to answer the questions regarding guidelines on treatment of SSc-related PAH.

Abbreviations: systemic sclerosis –SSc, standard deviation – SD, Dihydropyridine-type calcium channel blockers – CCB, phosphodiestherase-5 inhibitors – PDE-5I, angiotensin converting enzyme inhibitor - ACEI, proton pump inhibitors - PPI, Raynaud's phenomenon – RP, digital ulcers – DU, endothelin receptor antagonists – ERA, pulmonary artery hypertension – PAH, methotrexate – MTX, interstitial lung disease – ILD, hematopoietic stem cells transplantation – HSCT, scleroderma renal crises- SRC, gastroesophagal reflux disease – GERD, nonsignificant – ns, not analysed - na.

Supplementary Table 2. The level of agreement and availability for EUSTAR/ EULAR recommendations for treatment of SSc among SSc experts in Europe compared to SSc-experts from other geographical areas

		Europe (n=157)	Non- Europe (n=63)	P value
CCB for RP				
	Agreement (mean, SD)	8.1 (2.7)	8.0 (2.9)	ns
	Available (percentage)	97	98	
PDE-5I for RP				
	Agreement (mean, SD)	7.3 (2.6)	7.4 (2.9)	ns
	Available (percentage)	57	67	
Fluoxetine for RP				
	Agreement (mean, SD)	4.7 (2.7)	4.3 (3.0)	ns
	Available (percentage)	59	71	
Iloprost for RP				
	Agreement (mean, SD)	8.5 (2.2)	6.5 (3.1)	<0.0001
	Available (percentage)	90	55	
Iloprost for DU				
	Agreement (mean, SD)	9.0 (1.9)	7.4 (2.7)	<0.0001
	Available (percentage)	93	59	
PDE-5I for DU				
	Agreement (mean, SD)	7.9 (2.5)	8.3 (2.5)	ns

	Available (percentage)	64	73	
Bosentan for DU				
	Agreement (mean, SD)	8.5 (2.3)	6.3 (3.2)	<0.0001
	Available (percentage)	86	57	
ERA for PAH*				
	Agreement (mean, SD)	8.8 (2.3)	8.8 (2.4)	ns
	Available (percentage)	93	96	
PDE-5I for PAH*				
	Agreement (mean, SD)	8.8 (2.3)	9.0 (2.2)	ns
	Available (percentage)	97	100	
Riociguat for PAH*				
	Agreement (mean, SD)	7.4 (2.7)	7.4 (2.9)	ns
	Available (percentage)	62	57	
Epoprostenol for PAH*				
	Agreement (mean, SD)	8.3 (2.4)	8.3 (2.6)	ns
	Available (percentage)	81	72	
Other prostacyclin analogues for PAH*				
	Agreement (mean, SD)	8.1 (2.5)	7.8 (2.9)	ns
	Available (percentage)	91	79	
MTX for skin involvement in early diffuse SSc				
	Agreement (mean, SD)	7.9 (2.7)	6.8 (3.0)	<0.05
	Available (percentage)	95	92	

Cyclophosphamide for SSc-ILD				
	Agreement (mean, SD)	8.3 (2.4)	7.4 (2.9)	<0.05
	Available (percentage)	99	100	
HSCT for rapidly progressive SSc				
	Agreement (mean, SD)	7.3 (2.7)	6.4 (3.2)	<0.05
	Available (percentage)	66	66	
ACEI for SRC				
	Agreement (mean, SD)	9.1 (2.1)	9.3 (2.2)	ns
	Available (percentage)	99	100	
Blood pressure and renal function monitoring in SSc patients on glucocorticoids				
	Agreement (mean, SD)	8.9 (2.2)	9.3 (2.2)	ns
PPI for SSc related GERD, prevention of oesophageal ulcers and strictures				
	Agreement (mean, SD)	9.0 (2.1)	9.0 (2.4)	ns
	Available (percentage)	98	100	
Prokinetic drugs for SSc-related motility disturbances				
	Agreement (mean, SD)	8.0 (2.4)	7.9 (2.5)	ns
	Available (percentage)	97	95	
Intermittent or rotating use of antibiotics for small intestine bacterial overgrowth in SSc				

Agreement (mean, SD)	8.5 (2.1)	8.6 (2.3)	ns
Available (percentage)	98	100	

*N= 115 for Europe; n= 51 for geographical areas outside Europe

Abbreviations: European League Against Rheumatism -EULAR, European Scleroderma Trials and Research group -EUSTAR, systemic sclerosis –SSc, standard deviation – SD, Dihydropyridine-type calcium channel blockers – CCB, phosphodiestherase-5 inhibitors – PDE-5I, angiotensin converting enzyme inhibitor - ACEI, proton pump inhibitors - PPI, Raynaud's phenomenon – RP, digital ulcers – DU, endothelin receptor antagonists – ERA, pulmonary artery hypertension – PAH, methotrexate – MTX, interstitial lung disease – ILD, hematopoietic stem cells transplantation – HSCT, scleroderma renal crises- SRC, gastroesophagal reflux disease – GERD, nonsignificant – ns, not analysed - na.

Supplementary Table 3. Agreement and availability for EUSTAR members compared to EUSTAR non members

		EUSTAR members (n=148)	EUSTAR non members (n=69)	P value
CCB forRP				
	Agreement (mean, SD)	8.0 (2.9)	8.5 (2.1)	ns
	Available (percentage)	98	97	
PDE-5I for RP				
	Agreement (mean, SD)	7.2 (2.8)	7.8 (2.2)	ns
	Available (percentage)	59	60	
Fluoxetine for RP				
	Agreement (mean, SD)	4.4 (2.8)	5.1 (2.8)	ns
	Available (percentage)	58	71	
Iloprost for RP				
	Agreement (mean, SD)	8.1 (2.7)	7.8 (2.5)	ns
	Available (percentage)	84	69	
Iloprost for DU				
	Agreement (mean, SD)	8.0 (2.8)	8.3 (2.1)	ns
	Available (percentage)	87	75	
PDE-5I for DU				
	Agreement (mean, SD)	7.9 (2.6)	8.4 (1.9)	ns

	Available (percentage)	66	67	
Bosentan for DU				
	Agreement (mean, SD)	8.2 (2.7)	7.4 (2.5)	<0.05
	Available (percentage)	85	65	
ERA for PAH*				
	Agreement (mean, SD)	8.9 (2.3)	8.9 (2.0)	ns
	Available (percentage)	94	94	
PDE-5I for PAH*				
	Agreement (mean, SD)	8.9 (2.3)	9.1 (1.7)	ns
	Available (percentage)	97	100	
Riociguat for PAH*				
	Agreement (mean, SD)	7.5 (2.8)	7.5 (2.5)	ns
	Available (percentage)	56	73	
Epoprostenol for PAH*				
	Agreement (mean, SD)	8.4 (2.4)	8.3 (2.2)	ns
	Available (percentage)	79	77	
Other prostacyclin analogues (iloprost, trepostinil) for PAH*				
	Agreement (mean, SD)	8.2 (2.6)	7.9 (2.5)	ns
	Available (percentage)	89	83	
MTX for skin involvement in early diffuse SSc				
	Agreement (mean, SD)	7.3 (2.8)	7.6 (2.5)	ns

Available (percentage)	95	91	
Cyclophosphamide for SSc-ILD			
Agreement (mean, SD)	8.1 (2.6)	8.0 (2.3)	ns
Available (percentage)	100	99	
HSCT for rapidly progressive SSc			
Agreement (mean, SD)	7.2 (2.9)	7.0 (2.6)	ns
Available (percentage)	65	71	
ACEI for SRC			
Agreement (mean, SD)	9.2 (2.3)	9.4 (1.2)	ns
Available (percentage)	99	100	
Blood pressure and renal function monitoring in SSc patients on glucocorticoids			
Agreement (mean, SD)	8.9 (2.3)	9.4 (1.4)	ns
PPI for SSc related GERD, prevention of oesophagal ulcers and strictures			
Agreement (mean, SD)	9.1 (2.1)	9.2 (1.8)	ns
Available (percentage)	98	100	
Prokinetic drugs for SSc-related motility disturbances			
Agreement (mean, SD)	8.0 (2.4)	8.1 (2.2)	ns
Available (percentage)	97	96	

Intermittent or rotating use of antibiotics for small intestine bacterial overgrowth in SSc			
	Agreement (mean, SD)	8.5 (2.2)	8.7 (1.6)	ns
	Available (percentage)	99	99	

Abbreviations: European League Against Rheumatism -EULAR, European Scleroderma Trials and Research group -EUSTAR,systemic sclerosis –SSc, standard deviation – SD, Dihydropyridine-type calcium channel blockers – CCB, phosphodiestherase-5 inhibitors – PDE-5I, angiotensin converting enzyme inhibitor - ACEI, proton pump inhibitors - PPI, Raynaud's phenomenon – RP, digital ulcers – DU, endothelin receptor antagonists – ERA, pulmonary artery hypertension – PAH, methotrexate – MTX, interstitial lung disease – ILD, hematopoietic stem cells transplantation – HSCT, scleroderma renal crises- SRC, gastroesophagal reflux disease – GERD, nonsignificant – ns, not analysed - na.

Supplementary Table 4. Agreement and availability stratified for years of clinical experience in the field of SSc

		>10 years of experience (n=140)	<10 years of experience (n=79)	P value
CCB for RP				
	Agreement (mean, SD)	7.9 (3.0)	8.6 (2.3)	ns
	Available (percentage)	97	99	
PDE-5I forRP				
	Agreement (mean, SD)	7.2 (2.9)	7.4 (2.3)	ns
	Available (percentage)	61	57	
Fluoxetine for RP				
	Agreement (mean, SD)	4.5 (2.8)	4.9 (2.7)	ns
	Available (percentage)	64	60	
Iloprost for RP				
	Agreement (mean, SD)	7.7 (2.8)	8.3 (2.4)	ns
	Available (percentage)	78	82	
Iloprost for DU				
	Agreement (mean, SD)	8.5 (2.4)	8.7 (2.2)	ns
	Available (percentage)	82	85	
PDE-5I for DU				
	Agreement (mean, SD)	8.1 (2.5)	7.8 (2.4)	ns

	Available (percentage)	65	68	
Bosentan for DU				
	Agreement (mean, SD)	7.8 (2.9)	8.1 (2.4)	ns
	Available (percentage)	77	79	
ERA for PAH*				
	Agreement (mean, SD)	8.9 (2.3)	8.5 (2.6)	ns
	Available (percentage)	93	96	
PDE-5I for PAH*				
	Agreement (mean, SD)	9.0 (2.2)	8.6(2.4)	ns
	Available (percentage)	98	98	
Riociguat for PAH*				
	Agreement (mean, SD)	7.7 (2.7)	6.8 (2.9)	ns
	Available (percentage)	65	49	
Epoprostenol for PAH*				
	Agreement (mean, SD)	8.5 (2.4)	7.8 (2.6)	ns
	Available (percentage)	81	74	
Other prostacyclin analogues (iloprost, treprostinil) for PAH*				
	Agreement (mean, SD)	8.2 (2.6)	7.6 (2.7)	ns
	Available (percentage)	86	91	
MTX for skin involvement in early diffuse SSc				

Agreement (mean, SD)	7.2 (2.9)	7.7 (2.8)	ns
Available (percentage)	94	93	
Cyclophosphamide for SSc-ILD			
Agreement (mean, SD)	8.1 (2.7)	8.0 (2.4)	ns
Available (percentage)	99	100	
HSCT for rapidly progressive SSc			
Agreement (mean, SD)	7.0 (3.0)	7.2 (2.5)	ns
Available (percentage)	67	63	
ACEI for SRC			
Agreement (mean, SD)	9.1 (2.1)	9.0 (2.2)	ns
Available (percentage)	99	100	
Blood pressure and renal function monitoring in SSc patients on glucocorticoids			
Agreement (mean, SD)	9.0 (2.3)	9.1 (2.0)	ns
PPI for SSc related GERD, prevention of oesophageal ulcers and strictures			
Agreement (mean, SD)	9.1 (2.1)	8.9 (2.1)	ns
Available (percentage)	98	100	
Prokinetic drugs for SSc-related motility disturbances			
Agreement (mean, SD)	8.0 (2.4)	8.0 (2.3)	ns

Available (percentage)	96	99	
Intermittent or rotating use of antibiotics for small intestine bacterial overgrowth in SSc			
Agreement (mean, SD)	8.5 (2.3)	8.5 (1.9)	ns
Available (percentage)	99	97	

Abbreviations: systemic sclerosis –SSc, standard deviation – SD, Dihydropyridine-type calcium channel blockers – CCB, phosphodiestherase-5 inhibitors – PDE-5I, angiotensin converting enzyme inhibitor - ACEI, proton pump inhibitors - PPI, Raynaud's phenomenon – RP, digital ulcers – DU, endothelin receptor antagonists – ERA, pulmonary artery hypertension – PAH, methotrexate – MTX, interstitial lung disease – ILD, hematopoietic stem cells transplantation – HSCT, scleroderma renal crises- SRC, gastroesophagal reflux disease – GERD, nonsignificant – ns, not analysed - na.

Supplementary Table 5. The level of agreement stratified for availability of the drug/treatment option

	Drug/ treatment option available	Drug/ treatment option not available	P value
PDE-5I for RP			
Agreement (mean, SD)	7.9 (2.5)	6.4 (2.7)	<0.0001
n	129	88	
Fluoxetine for RP			
Agreement (mean, SD)	4.9 (2.9)	4.1 (2.5)	<0.05
n	138	81	
Iloprost for RP			
Agreement (mean, SD)	8.3 (2.6)	6.8 (2.7)	<0.001
n	170	44	
Iloprost for DU			
Agreement (mean, SD)	8.8 (2.2)	7.5 (2.6)	<0.0001
n	176	37	
PDE-5I for DU			
Agreement (mean, SD)	8.2 (2.6)	7.7 (2.3)	ns
n	142	70	
Bosentan for DU			
Agreement (mean, SD)	8.3 (2.4)	6.5 (3.0)	<0.0001
n	165	48	

Riociguat for PAH*			
Agreement (mean, SD)	8.1 (2.4)	6.6 (3.0)	<0.0001
n	99	66	
Epoprostenol for PAH*			
Agreement (mean, SD)	8.4 (2.4)	8.1 (2.5)	ns
n	127	35	
HSCT for rapidly progressive SSc			
Agreement (mean, SD)	7.4 (2.8)	6.5 (2.8)	<0.05
n	136	71	

Abbreviations: systemic sclerosis –SSc, standard deviation – SD, Raynaud's phenomenon – RP, digital ulcers – DU, pulmonary artery hypertension – PAH, hematopoietic stem cells transplantation – HSCT, nonsignificant – ns