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REVIEW ARTICLE

Nationwide and international registries on scleroderma. Past, present, and future

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Supplementary Tables

Suppl. Table 1. National and International Systemic sclerosis registries

	UK (UKSSG) 1995	Germany (DNSS) 2003	EUSTAR 2004	Canada (CSRG) 2004	Spain (RESCLE) 2006	Australia (ASIG) 2007	SPIN 2012	US (CONQUER) 2013	US (GRASP) 2013	Italy (SPRING) 2015	INSYNC 2018
First paper	1998	2008	2005	2008	2012	2011	2012	2020	2017	2020	2018
Type of data	Prospective	Prospective	Prospective	Prospective	Retrospective/Prospective	Prospective	Prospective	Prospective	Retrospective/Prospective	Prospective	Prospective
Sample size	> 2500	> 3500	> 11,500	1750	2238	1670	1700	151	1200	1708	638
Sites	37	40	234	15	40	13	43	12	23	38	31
Biobank	Yes	Yes	Yes	Yes	No	Yes	No	Yes	Yes	No	No
Data centre	Royal Free Hospital, London	University of Cologne	University Hospital Zurich	Jewish General Hospital, Montreal	Vall d'Hebron University Hospital, Barcelona	St. Vincent's Hospital, Melbourne	Jewish General Hospital McGill University, Montreal	University of Utah University of Texas Health Science Center, Houston	Johns Hopkins University, Baltimore	University of Modena and Reggio Emilia University of Florence	St. Vincent's Hospital, Melbourne
Coordinator	C. Denton	N. Hunzelmann	O. Distler	M. Baron K. McKenna	C.P. Simeón V. Fonollosa	S. Proudman M. Nikpour	B. Thombs	T. Frech S. Assassi	F. Wigley F. Boin	C. Ferri M. Matucci	M. Nikpour
Website	https://royalfree.nhs.uk/services/services-a-z/scleroderma/scleroderma-study-group/	https://dermatologie.uk-koeln.de/forschung/studienregister/details/studienregister/dnss-e-v---deutsches-netzwerk-fuer-sklerodermie/	https://eustar.org/	https://canadiansclerodermaresearchgroup.org	https://registros.shmedical.es/rescle/	https://rheumatology.org.au/patients/asig.asp	https://spinsclero.com	https://conquerssc.org	https://srfcure.org/grasp-2/	https://reumatologia.it/progetto-spring	NA

ASIG: Australian Scleroderma Interest Group; INSYNC: International Systemic Sclerosis Inception Cohort; CONQUER: Collaborative, National Quality and Efficacy Registry for Tracking Disease Progression in Systemic Sclerosis Patients; CSRG: Canadian Scleroderma Research Group; DNSS: German Network for Systemic Scleroderma; EUSTAR: EULAR Scleroderma Trials and Research group; GRASP: Genome Research in African American Scleroderma Patients; NA: not available; RESCLE: Spanish Scleroderma Registry; SPIN: Scleroderma Patient-centered Intervention Network; UKSSG: UK Scleroderma Study Group.

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Suppl. Table 2. Clinical comparison among registries

	UK (UKSSG)	Germany (DNSS)	EUSTAR	Canada (CSRG)	Spain (RESCLE)	Australia (ASIG)	SPIN	US (CONQUER)	US (GRASP)	Italy (SPRING)	INSYNC
Females %	82.1%	83.4%	83.6%	86%	86.9%	85.9%	87.3%	83.4%	84%	89.9%	81%
Age mean at SSc diagnosis, years mean (SD)	NA	55.7 (13.7)	54.3 (13.8)	55.8 (12.2)	51.2 (15.1)	57.6 (12.5)	55.6 (12.1)	50.6 (14.2)	42.4 (13.5)	58.4 (14.2)	54 (NA)
Time from 1st symptom, years	NA	11.4 (NA)	NA	14.7 (12.3)	6.3 (8.9)	11.6 (10)	11.6 (8.7)	NA	9.9 (8.7)	NA	NA
Skin subsets %											
dcSSc	42.2%	32.7%	37.1%	38%	26.5%	29.3%	40.9%	59.6%	57%	16.2%	44.6%
lcSSc	57.8%	45.5%	58.5%	59%	61.8%	70.7%	59.1%	40.4%	43%	66.9%	NA
Sine scleroderma	0	1.5%	4.4%	4%	7.5%	0	0	0	0	16.8%	NA
Overlap	0	10.9%	0	0	NA	0	0	0	0	0	NA
Visceral involvement %											
Esophageal involvement	NA	60%	67.3%	NA	57.4%	82.9%	86.9%	NA	NA	45.6%	NA
SRC	6.7%	NA	2.1%	4.3%	2.6%	3.7%	NA	NA	7%	0.9%	4.8%
ILD	15%	34.5%	39.5%	33%	45.6%	66.4%	36.2%	NA	68%	55%	26.2%
PAH	20%	15.8%	21.1%	5%	17.6%	11.1%	10.4%	NA	18%	NA	3.6%
Autoantibodies %											
ANA +	96%	94.2%	93.4%	95%	92.2%	95.4%	92.9%	86.1%	94%	97%	95%
ACA +	24.2%	35.9%	32.3%	36%	39.1%	49.6%	32.8%	8.6%	8%	46.4%	32%
ATA I +	20.4%	30.1%	36.8%	15%	19%	15%	24.8%	33.8%	30%	31.4%	20%
Survival											
From onset (RP)	NA	NA	99.6/1 years 97.4%/3 years 94.9%/5 years 86.5%/10 years 77.6%/15 years	NA	64.4%/10 years 31.3%/20 years 12.3%/30 years	63.2%/10 years 28.6%/20 years 10.6%/30 years 3.6%/40 years	NA	NA	NA	NA	NA

ASIG: Australian Scleroderma Interest Group; INSYNC: International Systemic Sclerosis Inception Cohort; CONQUER: Collaborative, National Quality and Efficacy Registry for Tracking Disease Progression in Systemic Sclerosis Patients; CSRG: Canadian Scleroderma Research Group; DNSS: German Network for Systemic Scleroderma; EUSTAR: EULAR Scleroderma Trials and Research group; GRASP: Genome Research in African American Scleroderma Patients; NA: not available; RESCLE: Spanish Scleroderma Registry; SPIN: Scleroderma Patient-centered Intervention Network; UKSSG: UK Scleroderma Study Group; ANA: antinuclear antibody; ACA: anticentromere antibodies; ATA I: antitopoisomerase I antibodies; SRC: scleroderma renal crisis; ILD: interstitial lung disease; PAH: pulmonary arterial hypertension; dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous systemic sclerosis; SSc: systemic sclerosis; RP: Raynaud's phenomenon.

Suppl. Table 3. Registries on SSc-related pulmonary arterial hypertension

	FR (ItinérAIR-Sclérodermie) 2002	US/CAN (PHAROS) 2006
First paper	2004	2011
Type of data	Prospective	Prospective
Sample size	599	558
Sites	21	19
Biobank	No	No
Data center	Hôpital Antoine Béclère, Clamart	Georgetown University
Coordinator	M. Humbert	V. Steen
Website	-	-

Suppl. Table 4. Registries on SSc-related pulmonary arterial hypertension

	FR (ItinérAIR-Sclérodermie)	US/CAN (PHAROS)
Females %	84.1%	89.3%
Age mean at PH diagnosis, years mean (SD)	54.9 (13)	60 (10.9)
Time from 1st symptom, years mean (SD) or median [range] RP Non-RP	 14.6 (12.4) 8.8 (8.1)	 11.5 [0.3-54.4] 8.6 [0-43.2]
Skin subsets % dcSSc lcSSc Sine scleroderma Overlap	 27.5% 62.5% - -	 28.9% 71.1% - -
Visceral involvement % Esophageal involvement SRC ILD PH	 - - 15.8% 3%	 - - - 30.8%
Autoantibodies % ANA + ACA + ATA I +	 - 51.9% 28%	 96.2% 42.7% 3.8%
Survival (from the diagnosis of PAH)	91.4%/3 years	95%/1 year 75%/3 year 63%/5 year 49%/8 year

ANA: antinuclear antibody; ACA: anticentromere antibodies; ATA I: antitopoisomerase I antibodies; SRC: scleroderma renal crisis; ILD: interstitial lung disease; PH: pulmonary hypertension; dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous systemic sclerosis; SSc: systemic sclerosis; RP: Raynaud's phenomenon.