

Juvenile- and adult-onset systemic sclerosis: similarities and differences in early disease patterns. A scoping review*N. Vasquez-Canizares, M. Twilt, Q. McCormick, P. Bhave, I.A. Dobre, S.C. Li***Supplementary Table S1.** Articles extracted for jSSc and aSSc scoping review.

Study ID	Full reference	Publication year	Multinational	Classification criteria used	Cohort name, country(ies)	Notes
Early (<3 years) Adult systemic sclerosis						
S1	Hanif I, Assassi S, Mayes MD <i>et al.</i> : Hand Swelling and Other Non-Raynaud Phenomenon Symptoms as the Initial Presentation of Systemic Sclerosis: Prevalence and Clinical Associations in Two US Cohorts. <i>Arthritis Rheumatol</i> 2025; 77(11): 15851595. https://doi.org/10.1002/art.43237	2025	No	2013 ACR/EULAR	GENISOS, CONQUER; US	Data extracted separately for the two cohorts
S2	Richard N, Gyger G, Hoa S <i>et al.</i> : Immunosuppression does not prevent severe gastrointestinal tract involvement in systemic sclerosis. <i>Clin Exp Rheumatol</i> 2021; 39 (Suppl .131): S142-8. https://doi.org/10.55563/clinexprheumatol/7683pg	2021	Yes	1980 ACR or 2013 ACR/EULAR	Australian Scleroderma Cohort Study (ASCS)/Canadian Scleroderma Research Group (CSRG); Australia, Canada	

S3	Butt SA, Jeppesen JL, Torp-Pedersen C <i>et al.</i> : Cardiovascular Manifestations of Systemic Sclerosis: A Danish Nationwide Cohort Study. <i>J Am Heart Assoc</i> 2019; 8(17): e013405. https://doi.org/10.1161/jaha.119013405	2019	No	None	Denmark	
S4	Pokeerbux MR, Giovannelli J, Dauchet L <i>et al.</i> : Survival and prognosis factors in systemic sclerosis: data of a French multicenter cohort, systematic review, and meta-analysis of the literature. <i>Arthritis Res Ther</i> 2019; 21(1): 86. Published 2019 Apr 3. https://doi.org/10.1186/s13075-019-1867-1	2019	No	1980 ACR or 2013 ACR/EULAR	France	
Late (> 5 years) Adult Systemic Sclerosis						
S5	Tonutti A, Motta F, Bajocchi G <i>et al.</i> : Elderly-onset systemic sclerosis defines a distinct clinical subset: Analysis from the SPRING registry of the Italian Society for Rheumatology. <i>J Autoimmun</i> 2025; 157: 103501. https://doi.org/10.1016/j.jaut.2025.103501	2025	No	2013 ACR/EULAR	SPRING, Italy	
S6	Fairley JL, Hansen D, Proudman S <i>et al.</i> : The burden and determinants of fatigue in incident and prevalent systemic sclerosis. <i>Clin Exp Rheumatol</i> 2024; 42(8): 1669-74. https://doi.org/10.55563/clinexprheumatol/65280d	2024	No	2013 ACR/EULAR	ASCS, Australia	

S7	Didier K, Sobanski V, Robbins A <i>et al.</i> : Impact of autoantibody status on stratifying the risk of organ involvement and mortality in SSc: experience from a multicentre French cohort of 1605 patients. <i>RMD Open</i> 2024; 10(4): e004580. Published 2024 Nov 20. https://doi.org/10.1136/rmdopen2024-004580	2024	No	2013 ACR/EULAR	France	
S8	Al-Sheikh H, Ahmad Z, Johnson SR: Ethnic Variations in Systemic Sclerosis Disease Manifestations, Internal Organ Involvement, and Mortality. <i>J Rheumatol</i> 2019; 46(9): 1103-8. https://doi.org/10.3899/jrheum.180042	2019	No	2013 ACR/EULAR	Toronto Scleroderma Program, Canada	
S9	Bacher A, Mittoo S, Hudson M <i>et al.</i> : Systemic sclerosis in Canada's North American Native population: assessment of clinical and serological manifestations. <i>J Rheumatol</i> 2013; 40(7): 1121-6. https://doi.org/10.3899/jrheum.121212	2013	No	None	CSRG, Canada	
S10	Carbonell C, Marcos M, Guillén-Del-Castillo A <i>et al.</i> : Standardized incidence ratios and risk factors for cancer in patients with systemic sclerosis: Data from the Spanish Scleroderma Registry (RESCLE). <i>Autoimmun Rev</i> 2022; 21(10): 103167. https://doi.org/10.1016/j.autrev.2022.103167	2022	No	1988 LeRoy or 2013 ACR/EULAR	RESCLE, Spain	
S11	Hughes M, Huang S, Alegre-Sancho JJ <i>et al.</i> : Late skin fibrosis in systemic sclerosis: a study from the EUSTAR cohort. <i>Rheumatology</i> (Oxford) 2023; 62(SI): SI54SI63. https://doi.org/10.1093/rheumatology/keac363	2023	Yes	2013 ACR/EULAR	European Scleroderma Trials and Research Group (EUSTAR), multinational	

S12	Aguila LA, da Silva HC, Medeiros-Ribeiro AC, Bunjes BG, Luppino-Assad AP, Sampaio-Barros PD: Is exposure to environmental factors associated with a characteristic clinical and laboratory profile in systemic sclerosis? A retrospective analysis. <i>Rheumatol Int</i> 2021; 41(6): 1143-50. https://doi.org/10.1007/s00296020-04693-3	2021	No	2013 ACR/EULAR	Brazil	
Early (<3 years) Juvenile systemic sclerosis						
S13	Kass H, Hanson V, Patrick J: Scleroderma in childhood. <i>J Pediatr</i> 1966; 68(2): 243-56. https://doi.org/10.1016/s00223476(66)80155-2	1966	No	None	University of California Los Angeles (UCLA), USA	Same study used to extract data from reported early and late strata
S14	Jacquel L, Bechara R, Terzic J <i>et al.</i> : An updated overview of Juvenile systemic sclerosis in a French cohort. <i>Pediatr Rheumatol Online J</i> 2025; 23(1): 13. Published 2025 Feb 8. https://doi.org/10.1186/s12969024-01043-6	2025	No	None	France	
S15	Cassidy JT, Sullivan DB, Dabich L, Petty RE: Scleroderma in children. <i>Arthritis Rheum</i> 1977; 20(2 Suppl): 351-4.	1977	No	None	University of Michigan, USA	Same study used to extract data from reported early and late strata
S16	Belgaumkar VA, Gokhale NR, Mahajan PM, Tolat SN, Bhokare A, Kamble S: Progressive systemic sclerosis in childhood: a report of three cases. <i>Indian J Dermatol Venereol Leprol</i> 2004; 70(2): 96-8.	2004	No	None	India	

S17	Martini G, Foeldvari I, Russo R <i>et al.</i> : Systemic sclerosis in childhood: clinical and immunologic features of 153 patients in an international database. <i>Arthritis Rheum</i> 2006; 54(12): 3971-8. https://doi.org/10.1002/art.22207	2006	Yes	None	Paediatric Rheumatology European Society (PRES), multinational	Cohort overlaps with REF S30
S18	Zhang C, Wang C, Gao S <i>et al.</i> : Application of the Scleroderma Clinical Trials Consortium Damage Index in patients with juvenile systemic sclerosis. <i>Pediatr Rheumatol Online J</i> 2025; 23(1): 53. Published 2025 May 14. https://doi.org/10.1186/s12969025-01110-6	2025	No	2013 ACR/EULAR	China	Same study used to extract data from reported early and late strata
S19	Sousa S, Fernandes S, Estanqueiro P <i>et al.</i> : Juvenile systemic sclerosis: review of 17 Portuguese patients. <i>J Scleroderma Relat Disord</i> 2017; 2(2): 121-6. https://doi.org/10.5301/jsrd.5000242	2017	No	2013 ACR/EULAR	Portugal	Same study used to extract data from reported early and late strata
S20	Foeldvari I, Klotsche J, Kasapcopur O <i>et al.</i> : Gender differences in juvenile systemic sclerosis patients: Results from the international juvenile scleroderma inception cohort. <i>J Scleroderma Relat Disord</i> 2023; 8(2): 120-30. https://doi.org/10.1177/23971983221143244	2023	Yes	2013 ACR/EULAR	Juvenile Systemic Sclerosis Inception Cohort (Hamburg), multinational	
S21	Li JC, Tadros S, Rosser F, Torok KS: Pulmonary Nodules in Juvenile Systemic Sclerosis: A Case-Series from the National Registry for Childhood Onset Scleroderma (NRCOS). <i>Diagnostics</i> (Basel) 2023; 13(12): 2103. Published 2023 Jun 17. https://doi.org/10.3390/diagnostic13122103	2023	No	2013 ACR/EULAR	National Registry for Childhood-Onset Scleroderma (NRCOS), USA	Data added for features not reported in main extracted article from two different publications reporting on the same cohort: Robinson AD, Pain CE, Morgan GA <i>et al.</i> : Distinct by Design: Unraveling the Unique Clinical and Transcriptomic Identity of Juvenile Scleromyositis Overlap Compared to

						Juvenile Systemic Sclerosis and Juvenile Dermatomyositis: Implications for Care and Pathogenesis. <i>ACR Open Rheumatol</i> 2026; 8(3): e90005. https://doi.org/10.1002/acr.2.90005. and Stefancic S, Robinson A, Havrilla HJ, Sood V, Torok KS: Gastrointestinal Impact and Tool Performance in Juvenile Systemic Sclerosis Using the University of California, Los Angeles Scleroderma Clinical Trial Consortium Gastrointestinal Tract, Version 2.0 Assessment. <i>Arthritis Care Res</i> (Hoboken) 2026; 78(4): 449-55. https://doi.org/10.1002/acr.25643
S22	Ambartsumyan L, Zheng HB, Iyer RS, Soares J, Henstorf G, Stevens AM: Relationship Between Esophageal Abnormalities on Fluoroscopic Esophagram and Pulmonary Function Testing in Juvenile Systemic Sclerosis. <i>Arthritis Care Res</i> (Hoboken) 2019; 71(11): 1444-9. https://doi.org/10.1002/acr.23778	2019	No	None	Seattle Children's Hospital, USA	

Late (> 5 years) Juvenile systemic sclerosis						
S23	Kass H, Hanson V, Patrick J: Scleroderma in childhood. <i>J Pediatr</i> 1966; 68(2): 243-56. https://doi.org/10.1016/s00223476(66)80155-2	1966	No	None	UCLA, USA	Same study used to extract data from reported early and late strata
S24	Cassidy JT, Sullivan DB, Dabich L, Petty RE: Scleroderma in children. <i>Arthritis Rheum</i> 1977; 20 (2 Suppl): 351-4.	1977	No	None	University of Michigan, USA	Same study used to extract data from reported early and late strata
S25	Jeong JE, Kim SH: Clinical characteristics of juvenile systemic sclerosis in Korea: 31year single-center study. <i>J Rheum Dis</i> 2024; 31(1): 25-32. https://doi.org/10.4078/jrd.2023.0046	2024	No	2007 PRES/ACR/EU LAR	South Korea	
S26	Vancheeswaran R, Black CM, David J <i>et al.</i> : Childhood-onset scleroderma: is it different from adult-onset disease. <i>Arthritis Rheum</i> 1996; 39(6): 1041-9. https://doi.org/10.1002/art.1780390624	1996	No	1980 ACR	United Kingdom	
S27	Russo RA, Katsicas MM: Clinical characteristics of children with Juvenile Systemic Sclerosis: follow-up of 23 patients in a single tertiary center. <i>Pediatr Rheumatol Online J</i> 2007; 5: 6. Published 2007 May 1. https://doi.org/10.1186/1546-0096-5-6	2007	No	2007 PRES/ACR/EU LAR	Hospital de Pediatría Prof. Dr. Juan P. Garrahan, Argentina	
S28	Sousa S, Fernandes S, Estanqueiro P <i>et al.</i> : Juvenile systemic sclerosis: review of 17 Portuguese patients. <i>J Scleroderma Relat Disord</i> 2017; 2(2): 121-6. https://doi.org/10.5301/jsrd.5000242	2017	No	none initially, but all fulfilled 2013 ACR/EULAR	Portugal	Same study used to extract data from reported early and late strata

S29	Martini G, Vittadello F, Kasapçopur O <i>et al.</i> : Factors affecting survival in juvenile systemic sclerosis. <i>Rheumatology</i> (Oxford) 2009; 48(2): 119-22. https://doi.org/10.1093/rheumatology/ken388	2009	Yes	None	PRES, multinational	Cohort overlaps with REF S17
S30	Adrovic A, Şahin S, Barut K, Kasapçopur Ö: Juvenile Scleroderma: A Referral Center Experience. <i>Arch Rheumatol</i> 2018; 33(3): 344-51. Published 2018 Jan 18. https://doi.org/10.5606/ArchRheumatol.2018.6578	2018	No	2007 PRES/ACR/EULAR	Turkey	Data added for features not reported in main extracted article from a different publication reporting on the same cohort: Adrovic A, Karatemiz G, Esatoglu SN <i>et al.</i> : Juvenile and adult-onset scleroderma: Different clinical phenotypes. <i>Semin Arthritis Rheum</i> 2023; 60: 152197. https://doi.org/10.1016/j.semarthrit.2023.152197
S31	Szamosi S, Maródi L, Czirják L, Ellenés Z, Szücs G: Juvenile systemic sclerosis: a follow-up study of eight patients. <i>Ann NY Acad Sci</i> 2005; 1051: 229-34. https://doi.org/10.1196/annals.1361.063	2005	No	1980 ACR	Hungary	
S32	Borowiec A, Dabrowski R, Wozniak J <i>et al.</i> : Cardiovascular assessment of asymptomatic patients with juvenile-onset localized and systemic scleroderma: 10 years prospective observation. <i>Scand J Rheumatol</i> 2012; 41(1): 33-8. https://doi.org/10.3109/03009742.2011.609489	2012	No	1980 ACR	Institute of Cardiology, Warsaw, Poland	

S33	Sampaio-Barros PD, Bortoluzzo AB, Del Rio APT, LuppinoAssad AP, Andrade DC, Marques-Neto JF: Clinical and laboratory profile of juvenile onset systemic sclerosis in a Brazilian cohort. <i>J Scleroderma Relat Disord</i> 2019; 4(1): 43-8. https://doi.org/10.1177/2397198318769796	2019	No	1980 ACR	Brazil	
S34	Hatta Y, Hasegawa M, Matsushita T, Hamaguchi Y, Fujimoto M, Takehara K: The clinical characteristics of juvenile-onset systemic sclerosis in Japanese patients. <i>Mod Rheumatol</i> Published online April 27, 2013. https://doi.org/10.1007/s10165-013-0884-1	2013	No	2013 ACR/EULAR	Kanazawa University, Japan	
S35	Zulian F, Lanzoni G, Castaldi B <i>et al.</i> : Systemic sclerosis sine scleroderma in children. <i>Rheumatology</i> (Oxford) 2022; 61(6): 2555-22. https://doi.org/10.1093/rheumatology/keab738	2022	No	2013 ACR/EULAR	University of Padova, Italy	
S36	Szymańska-Jagiello W, Rondio H, Jakubowska K: Changes in the locomotor system in progressive systemic sclerosis in children. <i>Mater Med Pol</i> 1972; 4(4): 201-9.	1972	No	None	Institute of Rheumatology and Institute of Postgraduate Medical Education, Medical Academy, Warsaw, Poland	
S37	Tsuji H, Shirakashi M, Hiwa R <i>et al.</i> : Differences in the autoantibody phenotypes and longterm outcomes between juvenile- and adult-onset systemic sclerosis. <i>Mod Rheumatol</i> 2025; 35(4): 683-90. https://doi.org/10.1093/mr/roaf005	2025	No	1980 ACR or 2013 ACR/EULAR	Kyoto University Graduate School of Medicine, Japan	

S38	Scalapino K, Arkachaisri T, Lucas M <i>et al.</i> : Childhood onset systemic sclerosis: classification, clinical and serologic features, and survival in comparison with adult-onset disease. <i>J Rheumatol</i> 2006; 33(5): 1004-13.	2006	No	1980 ACR or 1988 LeRoy	University of Pittsburgh Scleroderma Databank, USA	
S39	Panigada S, Ravelli A, Silvestri M <i>et al.</i> : HRCT and pulmonary function tests in monitoring of lung involvement in juvenile systemic sclerosis. <i>Pediatr Pulmonol</i> 2009; 44(12): 1226-34. https://doi.org/10.1002/ppul.21141	2009	No	1980 ACR or 2007 PRES/ACR/EULAR	Genova and Pavia, Italy	
S40	Suárez-Almazor ME, Catoggio LJ, Maldonado-Cocco JA, Cuttica R, García Morteo O: Juvenile progressive systemic sclerosis: clinical and serologic findings. <i>Arthritis Rheum</i> 1985; 28(6): 699-702. https://doi.org/10.1002/art.1780280615	1985	No	1980 ACR	Instituto Nacional de Rehabilitacion, and the Laboratory, Centro Medico San Luis, Argentina	
S41	Zhang C, Wang C, Gao S <i>et al.</i> : Application of the Scleroderma Clinical Trials Consortium Damage Index in patients with juvenile systemic sclerosis. <i>Pediatr Rheumatol Online J</i> 2025; 23(1): 53. Published 2025 May 14. https://doi.org/10.1186/s12969025-01110-6	2025	No	2013 ACR/EULAR	China	Same study used to extract data from reported early and late strata
S42	Rutkowska-Sak L, Gietka P, Gazda A, Kołodziejczyk B: Juvenile systemic sclerosis - observations of one clinical centre. <i>Reumatologia</i> 2021; 59(6): 367-72. https://doi.org/10.5114/reum.2021.112350	2021	No	2007 PRES/ACR/EULAR	National Institute of Geriatrics, Rheumatology, and Rehabilitation, Warsaw, Poland	

S43	Bagri NK, Raj D, Kaur J <i>et al.</i> : Juvenile systemic sclerosis: experience from a tertiary care center from India. <i>Rheumatol Int</i> 2017; 37(10): 1687-91. https://doi.org/10.1007/s00296-017-3793-3	2017	No	1980 ACR or 2007 PRES/ACR/EULAR	All India Institute of Medical Sciences, India	
S44	Zayed HS, Gheita TA, Hammam N <i>et al.</i> : Systemic sclerosis in children and older juvenile-onset patients: real-life map by the Egyptian College of Rheumatology. <i>Sci Rep</i> 2026; 16(1): 15542. https://doi.org/10.1038/s41598-026-52795-y	2026	No	2007 PRES/ACR/EULAR	Egypt	