Juvenile localised scleroderma and juvenile systemic scleroderma
FESCA session

Ivan Foeldvari, MD

Hamburg Centre for Pediatric and Adolescence Rheumatology, Germany

Centre for Treatment of Scleroderma and Uveitis in Childhood and Adolescence

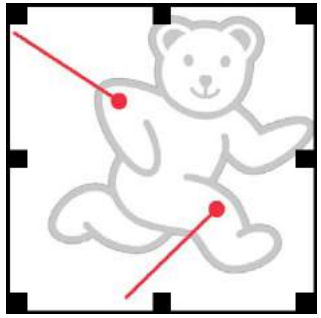
Teaching Unit of the Asklepios Campus of the Semmelweis Medical School, Budapest, Hungary

www.kinderrheumatologie.de

www.sklerodermie.org

www.uveitis-kindesalter.de

www.orphan-diseases-in-pediatric-rheumatology.de



Introduction

- My name is Ivan Foeldvari
- I am a pediatric rheumatologist
- Head of the Hamburg Center of Pediatric and Adolescent Rheumatology
- My major clinical and research interests are juvenile localized and systemic scleroderma
- Currently I am head of the Juvenile Scleroderma Working Group of the Pediatric Rheumatology European Society
- To mention some research projects:
  - I am the lead investigator of the juvenile scleroderma inception cohort- www.juvenile-scleroderma.com , the largest prospective cohort
  - Development of outcome measures for juvenile systemic and localised scleroderma
  - ....

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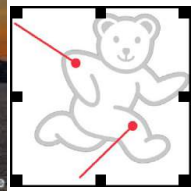

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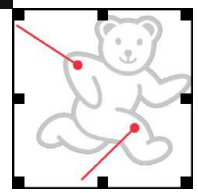
First part : Juvenile Localised Scleroderma

- Definition and classification
- How often does it occur?
- How is it diagnosed?
- How is it followed up?
- How is it treated?
- What is the long term prognosis?



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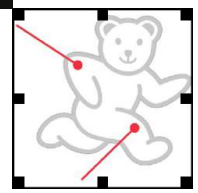

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Definition and Classification

- It as an autoimmune disease („allergy against own components of the body“), involving mainly the skin
- but it has extracutaneous manifestations, like
  - „White uveitis“ (higher risk in patients with head and face involvement)
  - Arthritis (inflammation of joint)
  - Central nervous system involvement (mostly in patients with involvement on the head and face)
- The lesions crossing joints can cause decreased range of motion in the joints
- Linear lesions on the extremities can cause length discrepancies
- Lesions in the face can cause cosmetic dysfiguration
- IT IS NOT DEVELOPING INTO SYSTEMIC SCLERODERMA

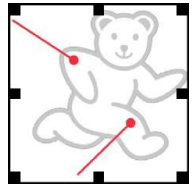

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Table 1 Preliminary proposed classification of juvenile localized scleroderma

Main group	Subtype	Description
(1) Circumscribed morphoea	(a) Superficial	Oval or round circumscribed areas of induration limited to epidermis and dermis, often with altered pigmentation and violaceous, erythematous halo (lilac ring). They can be single or multiple
	(b) Deep	Oval or round circumscribed deep induration of the skin involving subcutaneous tissue extending to fascia and may involve underlying muscle. The lesions can be single or multiple. Sometimes the primary site of involvement is in the subcutaneous tissue without involvement of the skin
(2) Linear scleroderma	(a) Trunk/limbs	Linear induration involving dermis, subcutaneous tissue and, sometimes, muscle and underlying bone and affecting the limbs and the trunk
	(b) Head	*En coup de sabre* (ECDS). Linear induration that affects the face and the scalp and sometimes involves muscle and underlying bone.
		Parry Romberg or progressive hemifacial atrophy loss of tissue on one side of the face that may involve dermis, subcutaneous tissue, muscle and bone. The skin is mobile
(3) Generalized morphoea		Induration of the skin starting as individual plaques (four or more and larger than 3 cm) that become confluent and involve at least two out of seven anatomic sites (head–neck, right upper extremity, left upper extremity, right lower extremity, left lower extremity, anterior trunk, posterior trunk)
(4) Panclerotic morphoea		Circumferential involvement of limb(s) affecting the skin, subcutaneous tissue, muscle and bone. The lesion may also involve other areas of the body without internal organs involvement
(5) Mixed morphoea		Combination of two or more of the previous subtypes. The order of the concomitant subtypes, specified in brackets, will follow their predominant representation in the individual patient [i.e. mixed morphoea (linear-circumscribed)]

Associated conditions: lichen sclerosus et atrophicus (LSA) and atrophoderma of Pasini and Pierini (APP) can be associated with the previous subtypes but are not included in the above classification. Source: Consensus conference, Padua, Italy, 2004.

Diagnosis in localised scleroderma

All children with suspected localised or systemic scleroderma should be referred to a specialized center).

Recommendation strength D.

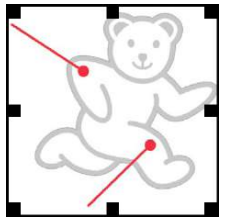
Vote 10 for, 0 against.

Ann Rheum Dis 2019;**78**:1019–1024.

OPEN ACCESS

Consensus-based recommendations for the management of juvenile localised scleroderma

Francesco Zulian,[1] Roberta Culpo,[1] Francesca Sperotto,[1] Jordi Anton,[2] Tadej Avcin,[3] Eileen M Baildam,[4] Christina Boros,[5] Jeffrey Chaitow,[6] Tamàs Constantin,[7] Ozgur Kasapcopur,[8] Sheila Knupp Feitosa de Oliveira,[9] Clarissa A Pilkington,[10] Ricardo Russo,[11] Natasa Toplak,[3] Annet van Royen,[12] Claudia Saad Magalhães,[13] Sebastiaan J Vastert,[12] Nico M Wulffraat,[12] Ivan Foeldvari[14]



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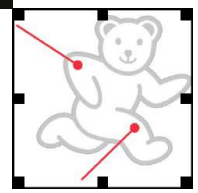

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Estimated Prevalence of juvenile localised scleroderma using the USA claims data

Timothy Beukelman1, Fenglong Xie2 and Ivan Foeldvari3

Table 1. The estimated prevalence of juvenile localised scleroderma in the United States.

Year	Total children (N)	Diagnosis code for localised scleroderma (N)	No diagnosis code for systemic sclerosis or mixed connective tissue disease (N)	Use of methotrexate	Estimated prevalence per 10,000 children [95% CI]
2010	5,894,628	2064	2006/2064	75/2006	3.4 [3.3–3.6]
2011	6,231,475	2269	2222/2269	86/2222	3.6 [3.4–3.7]
2012	6,278,118	2198	2154/2198	68/2154	3.4 [3.3–3.6]
2013	4,950,018	1732	1692/1732	61/1692	3.4 [3.3–3.6]
2014	4,933,523	1620	1588/1620	71/1588	3.2 [3.1–3.4]

CI: confidence interval.

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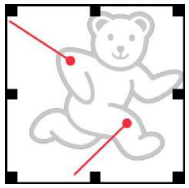

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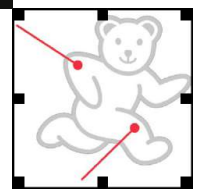

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How is it diagnosed?

- **It is mostly a clinical diagnosis**
- The modified Rodnan Skin score helps to assess the skin involvement
- Whole body joint exam: looking for range restrictions, joint swelling, sign of enthesitis
- Magnetic resonance imaging can help to assess deeper involvement under the skin
- Ultrasound with Doppler can assess increased blood flow in the involved area compared to the healthy side
- Skin biopsy is rarely needed, only in non typical cases

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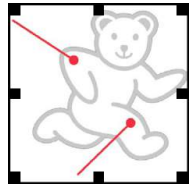

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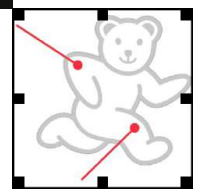

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RHEUMATOLOGY

Rheumatology 2010;49:373–381
doi:10.1093/rheumatology/kep361
Advance Access publication 14 December 2009

Original article

Development and initial validation of the Localized Scleroderma Skin Damage Index and Physician Global Assessment of disease Damage: a proof-of-concept study

Thaschawee Arkachaisri[1,2], Soamarat Vilaiyuk[1], Kathryn S. Torok[1] and Thomas A. Medsger Jr[3]

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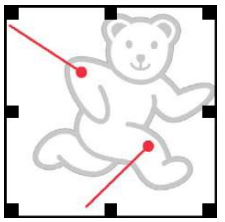

Diagnosis in localised scleroderma
Agreed: 10/10

LoSSI, that is part of LoSCAT, is a good clinical instrument to assess activity and severity in JLS lesions and is highly recommended in clinical practice.

Level of evidence 3, strenght of recommendation level C.

Vote: 9 for, 0 against

Ann Rheum Dis 2019;**78**:1019–1024.

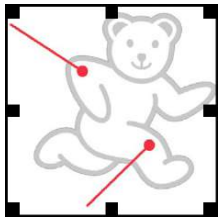

FIG. 4 LoSCAT.

Localized Scleroderma Cutaneous Assessment Tool

		mLoSSI Localized Scleroderma Skin **Activity** Index			LoSDI Localized Scleroderma Skin **Damage** Index		
Site		**New/ enlarge (within 1 mo)** **0 = none** **3 = N/E**	**Erythema** **0 = none** **1 = pink** **2 = red** **3 = dark red/ violaceous**	**Skin Thickness** **0 = none** **1 = mild** **2 = moderate** **3 = marked**	**Dermal Atrophy** **0 = none** **1 = shiny** **2 = visible vessel** **3 = obvious 'cliffdrop'**	**Subcutaneous Atrophy** **0 = none** **1 = flat** **2 = concave** **3 = marked atrophy**	**Dyspigmentation (hypo/hyperpig)** **0 = none** **1 = mild** **2 = moderate** **3 = marked**
Scalp/ face							
Neck							
Chest							
Abdomen							
Upper back							
Lower back							
RT	arm						
	forearm						
	hand						
	thigh						
	leg						
	foot						
LT	arm						
	forearm						
	hand						
	thigh						
	leg						
	foot						

Total Score: **mLoSSI (Activity)** _____ **LoSDI (Damage)** _____

PLEASE MARK WITH A STRAIGHT LINE:

Physician Global Assessment of Disease Activity

0 **100**
Inactive **Markedly active**

Physician Global Assessment of Disease Damage

0 **100**
No damage **Markedly damage**

Comment:

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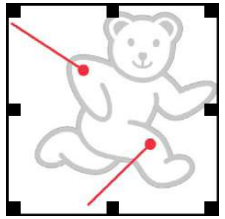
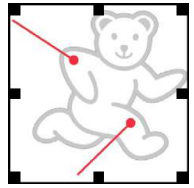


How is it followed?

- Assessment of the LoSCAT
- Whole joint count is needed at every follow up, including temporomandibular joint (jaw)
- Assessment for muscle strength
- Assessment for length discrepancy is needed
- Screening for uveitis every 6 to 12 months is needed
- Orthodentic assessment for patients with facial involvement
- Assessment for central nervous system at base line in case of head and facial involvement and only if there is a clinical sign
- Assessment of quality of life



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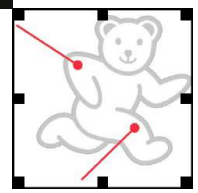

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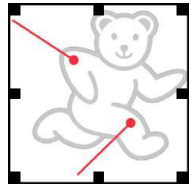
How is it treated?

- Patients with lesions, which are crossing a joint or lead to potential cosmetic deformaty, should be treated with a systemic medication.
- First choice is Methotrexate (15 mg/m2 body surface/week)
- In case of methotrexate-intolerance mycophenolate is the first choice
- In case of non-response to Methotrexate or Mycophenolate , or a combination of Methotrexate and Mycophenolate, a biologic agent can be added (tocilizumab, abatacept...)
- Physiotherapy in case of joint restriction
- Autologous fat cell transplantation to correct facial lesions
- Psychosocial support, if needed
- ....



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How is it treated?

Pediatric Drugs
https://doi.org/10.1007/s40272-019-00363-5

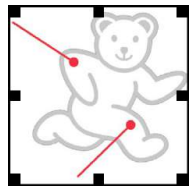
REVIEW ARTICLE

Check for updates

Update on the Systemic Treatment of Pediatric Localized Scleroderma

Ivan Foeldvari[1]

© Springer Nature Switzerland AG 2019



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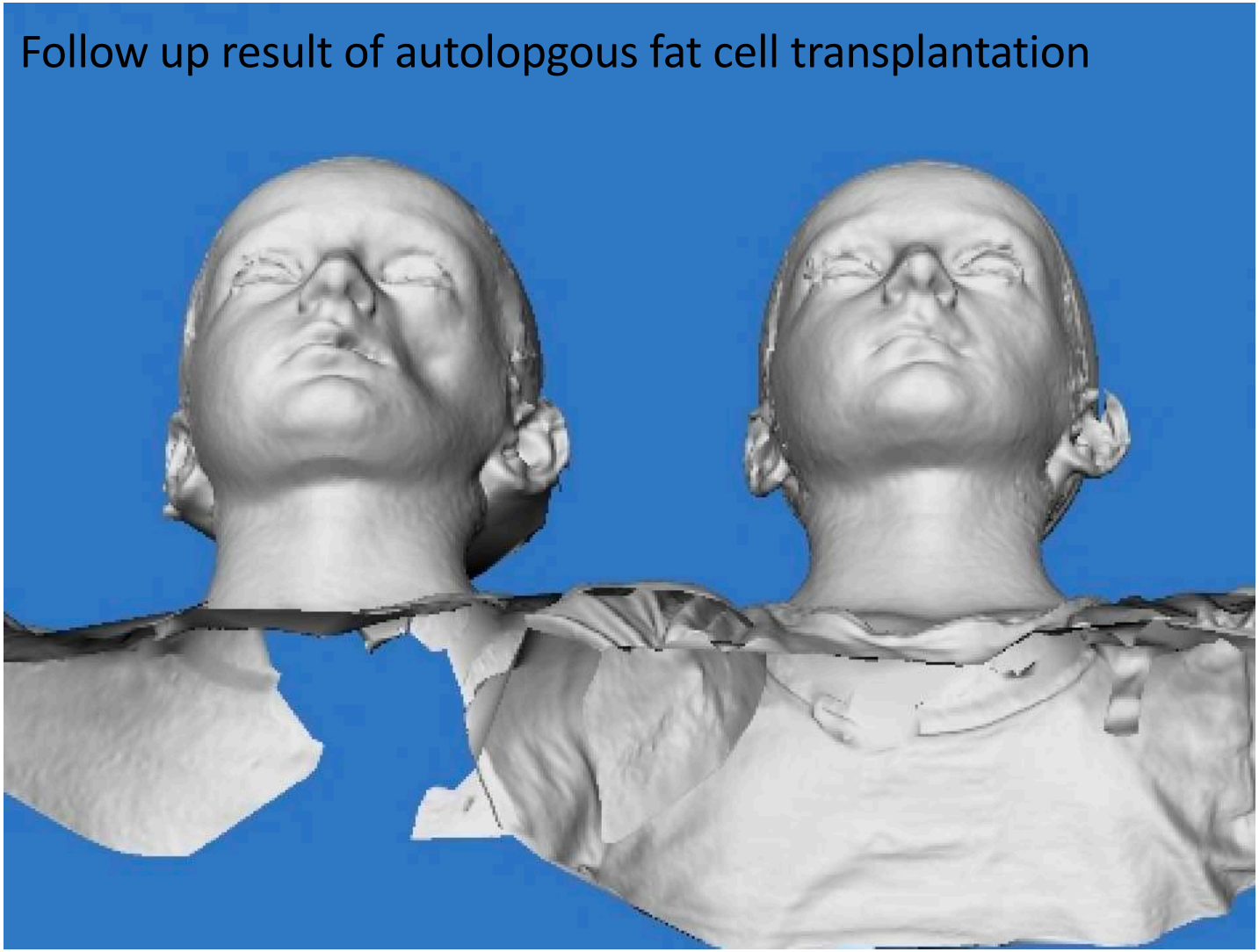
Follow up result of autolopgous fat cell transplantation



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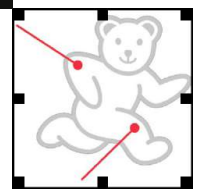

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What is the long term prognosis?

- It is a well treatable disease, if the treatment starts, when damage is not significant !!!
- Early recognition and diagnosis is a key with a follow up in cooperation with pediatric rheumatology/pediatric dermatology
- The use of the „therapeutic window“ is very important
- The disease can be active even after 30 years

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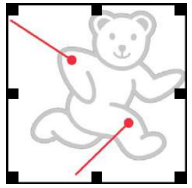

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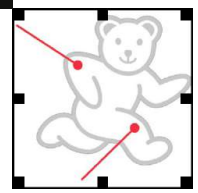

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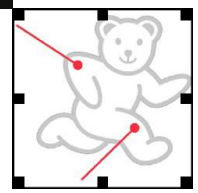

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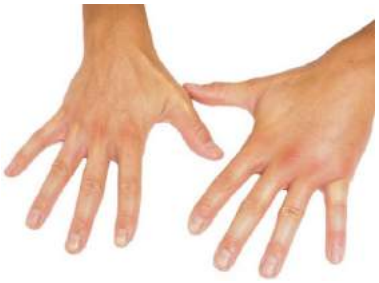
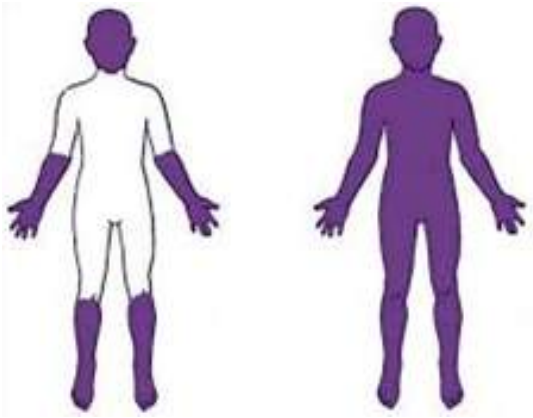
Systemic sclerosis (SSc)

- Scleroderma
  - thickened, hardened skin

JUVENILE
Juvenile SSc (JSSc)
Subtypes
Limited cutaneous SSc (jlcSSc)
Diffuse cutaneous SSc (jdcSSc)



www.medicalnewstoday.com



The courtesy of Nicole Bundy, MD

Proposed classification criteria for juvenile systemic scleroderma

Zulian et al. Arthritis Rheum 2007;57:203-12

First and Second International Workshop on Juvenile Scleroderma
June 2001 and 2004 Padua, Italy

***Steering Committee*: F. Zulian (Padua), I. Foeldvari (Hamburg), J. Harper (London), A. Peserico (Padua), N. Ruperto**

- Major criteria
  - Sclerosis* / induration*
- Definite disease
- 1 major and 2 minor criteria

- Minor criteria
  - Vascular changes*
  - Pulmonary involvement*
  - Gastrointestinal involvement*
  - Renal involvement*
  - Cardiovascular involvement*
  - Musculoskeletal involvement*
  - Neurologic involvement*
  - Serology*

  * Per definition typical for SSc

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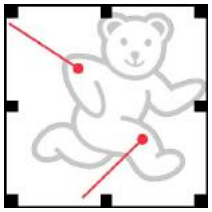
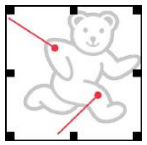


New Classification of Systemic Sclerosis

Arth Rheum 2013,65: 2737-47

Item	Sub-item(s)	Weight/score†
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints *(sufficient criterion)*	–	9
Skin thickening of the fingers *(only count the higher score)*	Puffy fingers	2
	Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	4
Fingertip lesions *(only count the higher score)*	Digital tip ulcers	2
	Fingertip pitting scars	3
Telangiectasia	–	2
Abnormal nailfold capillaries	–	2
Pulmonary arterial hypertension and/or interstitial lung disease *(maximum score is 2)*	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Raynaud's phenomenon	–	3
SSc-related autoantibodies (anticentromere, anti–topoisomerase I [anti–Scl-70], anti–RNA polymerase III) *(maximum score is 3)*	Anticentromere	3
	Anti–topoisomerase I	
	Anti–RNA polymerase III	



* These criteria are applicable to any patient considered for inclusion in an SSc study. The criteria are not applicable to patients with skin thickening sparing the fingers or to patients who have a scleroderma-like disorder that better explains their manifestations (e.g., nephrogenic sclerosing fibrosis, generalized morphea, eosinophilic fasciitis, scleredema diabeticorum, scleromyxedema, erythromyalgia, porphyria, lichen sclerosis, graft-versus-host disease, diabetic cheiroarthropathy).

† The total score is determined by adding the maximum weight (score) in each category. Patients with a total score of ≥9 are classified as having definite SSc.

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New Classification of Systemic Sclerosis

Arth Rheum 2013,65: 2737-47

- The maximum possible score is 19
- Patients with a score of ≥9 are classified as having SSc.
- The definitions of the items used in the criteria are defined in the publication.

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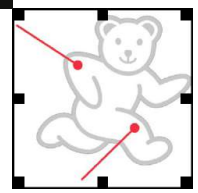

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Estimated Prevalence of juvenile systemic scleroderma using the USA claims data
T. Beukelman, F. Xie, I. Foeldvari. JSRD 2018, 3: 189-190

Year	N of Total Children	Diagnosis Code for Systemic Sclerosis	No Diagnosis Code for Localized Scleroderma	Use of Methotrexate, Mycophenolate Mofetil, or Cyclophosphamide	Estimated Prevalence per 1,000,000 Children [95% CI]
2010	5,888,868	254	186	23	3.9 [2.5-5.9]
2011	6,231,475	249	185	22	3.5 [2.2-5.3]
2012	6,278,116	217	170	26	4.1 [2.7-6.1]
2013	4,950,018	175	120	17	3.4 [2.0-5.5]
2014	4,933,522	138	91	14	2.8 [1.6-4.8]

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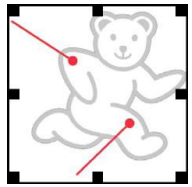
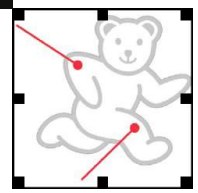


Second part : Juvenile Systemic Scleroderma

- Definition and classification
- How often does it occur?
- How is it diagnosed and followed?
- How is it treated?
- What is the long term prognosis?



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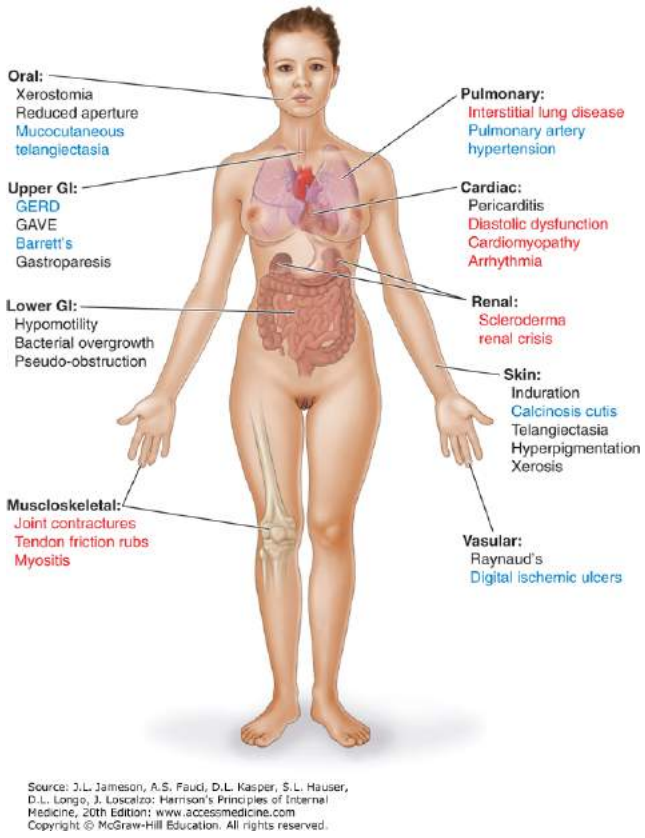
Organ involvement in SSc

SKIN
PULMONARY
CARDIOVASCULAR
RENAL
MUSCULOSKELETAL
GASTROINTESTINAL
NEURAL



Source: J.L. Jameson, A.S. Fauci, D.L. Kasper, S.L. Hauser, D.L. Longo, J. Loscalzo: Harrison's Principles of Internal Medicine, 20th Edition: www.accessmedicine.com
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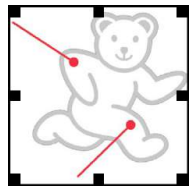
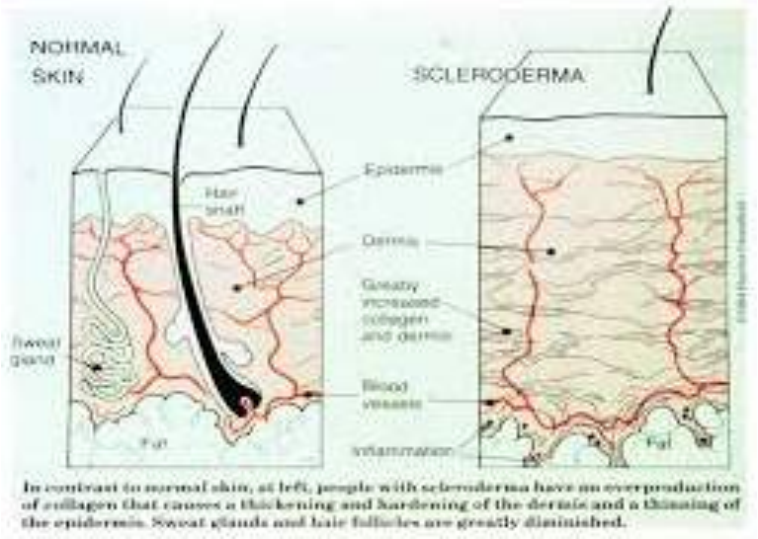
Jameson JL (www.accesmedicine.com)



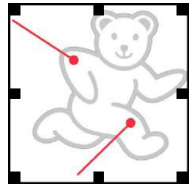
2

Organ involvement in SSc

SKIN
Thickening and hardening
Scleordactyly
Raynaud phenomenon
Digital ulcers
Telangiectasia
Calcinosis cutis



In contrast to normal skin, at left, people with scleroderma have an overproduction of collagen that causes a thickening and hardening of the dermis and a thinning of the epidermis. Sweat glands and hair follicles are greatly diminished.



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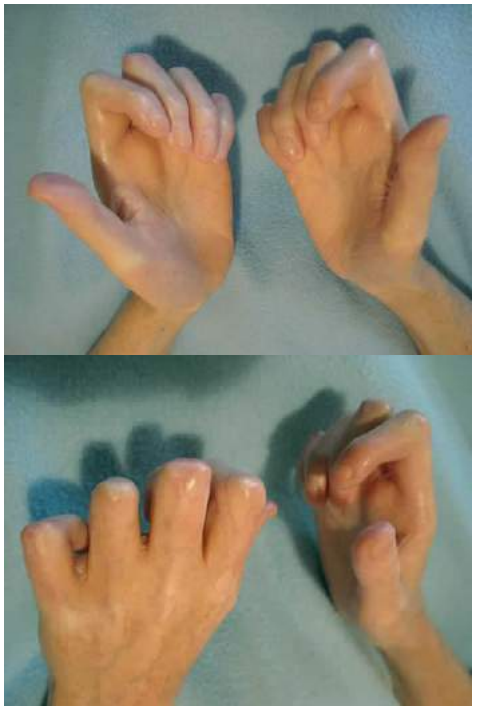
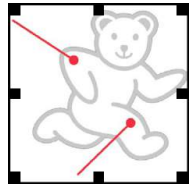


www.sclero.org



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2

Organ involvement in SSc

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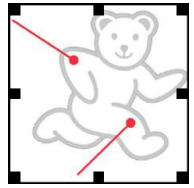
Scleordactyly
Raynaud phenomenon
Digital ulcers
Telangiectasia
Calcinosis cutis

Shabir Bhimji, MD (www.eMedicineHealth.com)



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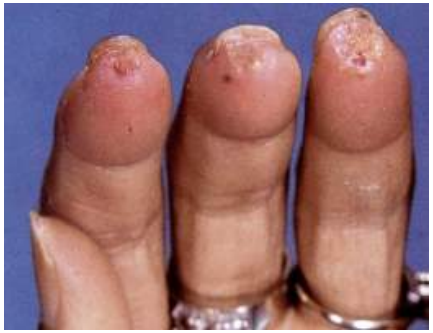
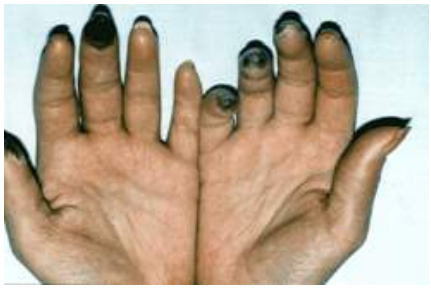
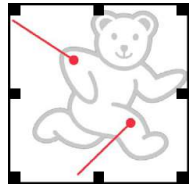


www.rheumnow.com



www.medscape.com



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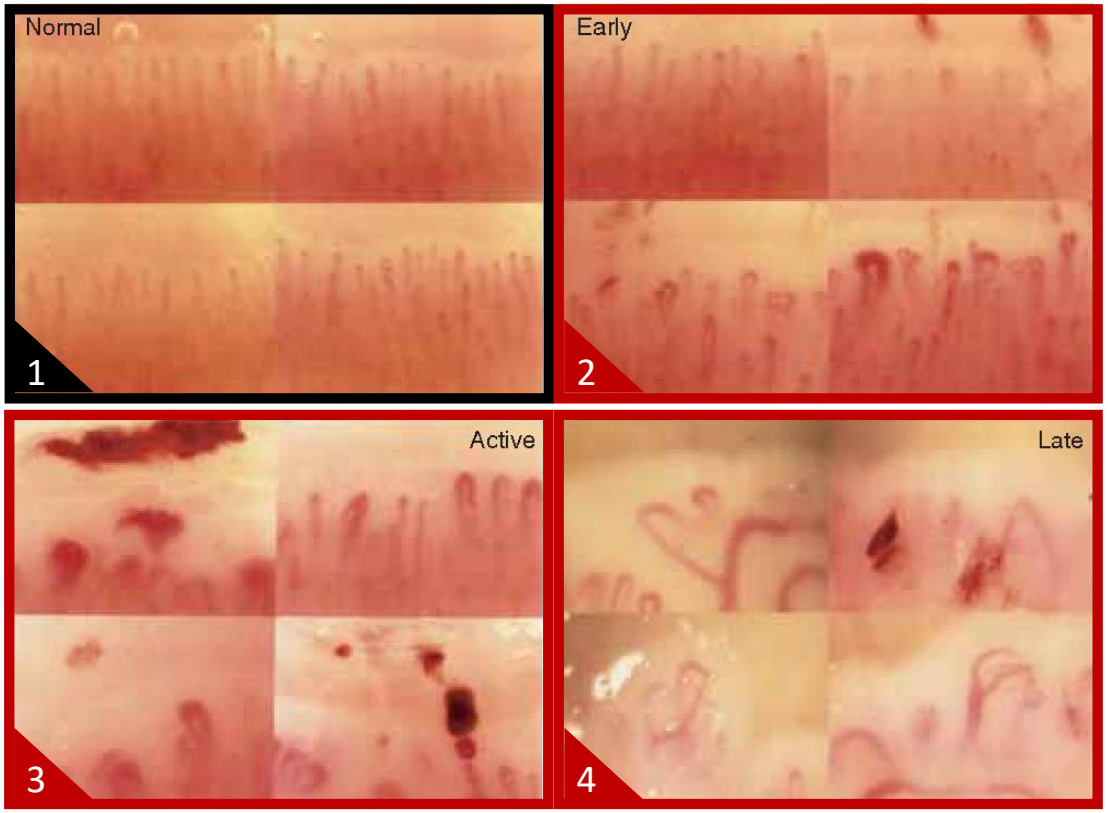
Scleroderma pattern (SSc pattern)



Cutolo et al. (Rheumatology 2000)

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2

Organ involvement in SSc

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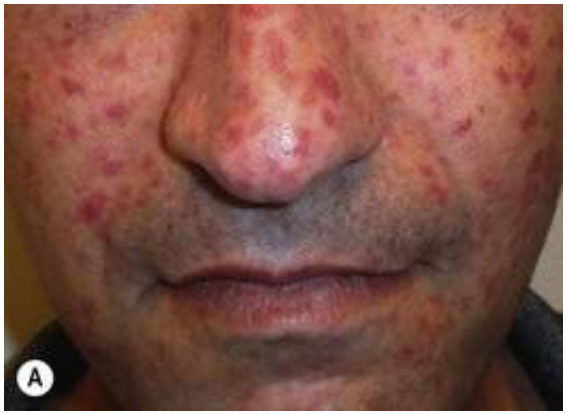

www.clinicalgate.com

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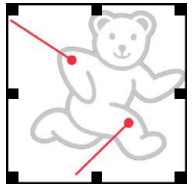

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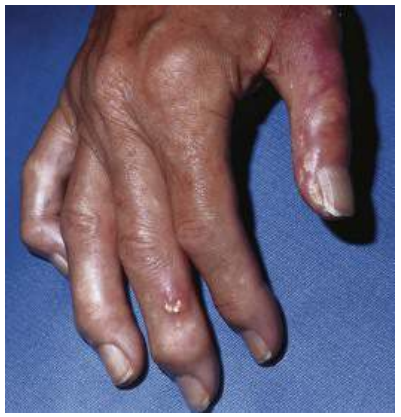

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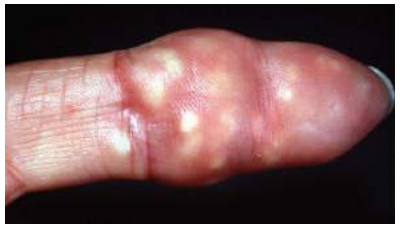
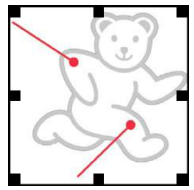


www.healthline.com



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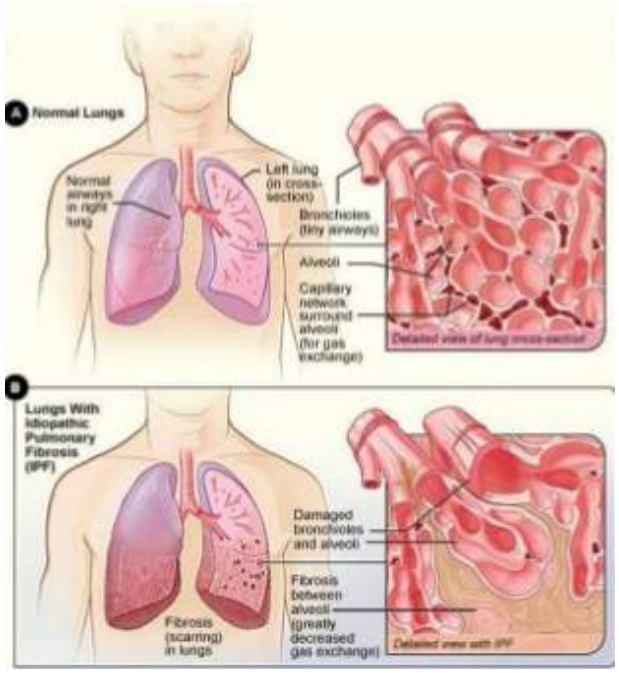
2

Organ involvement in SSc

PULMONARY
Interstitial Lung Disease (ILD)
Pulmonary Arterial Hypertension (PAH)



Jane Dematte MD (Scleroderma Foundation)

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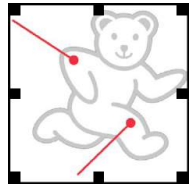

2

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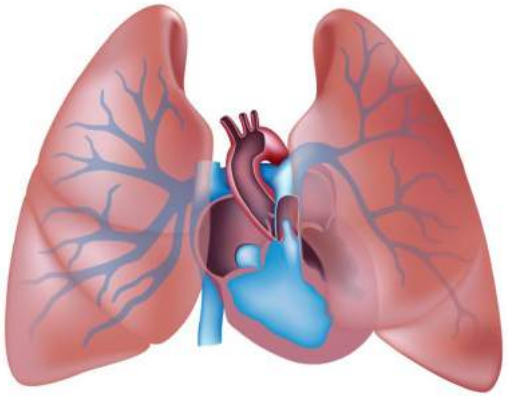
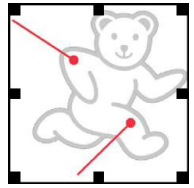


Laura Stiles (ACR 2017 San Diego - Coverage)



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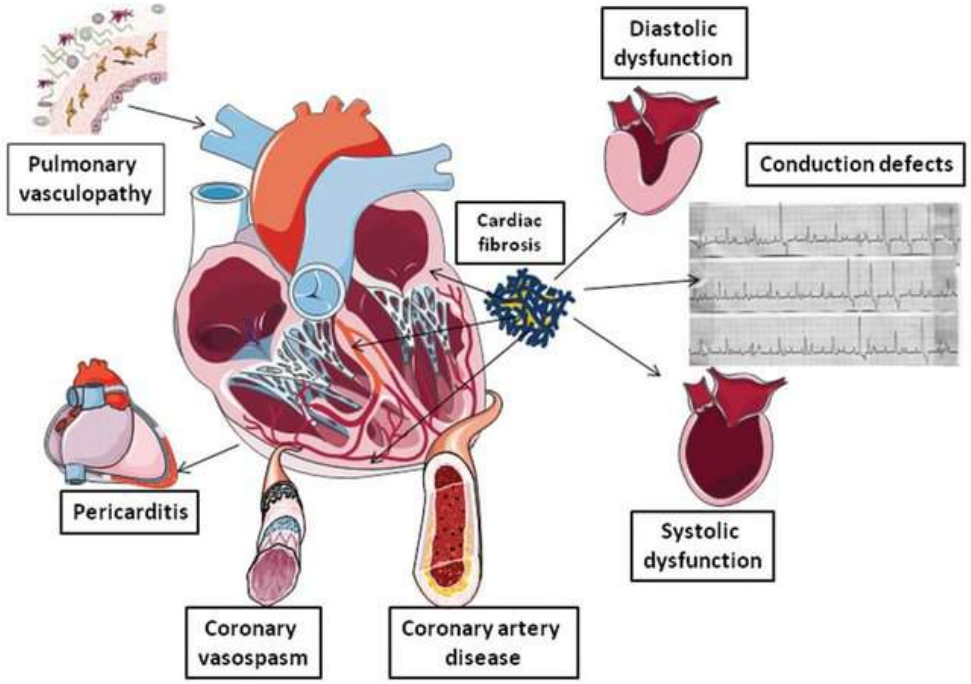
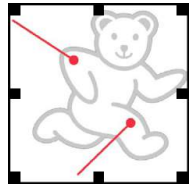
Organ involvement in SSc

CARDIOVASCULAR
Myocardial disease
Pericardial disease
Arrhythmias and conduction abnormalities



Theodoros Dimiroulas (www.researchgate.net)



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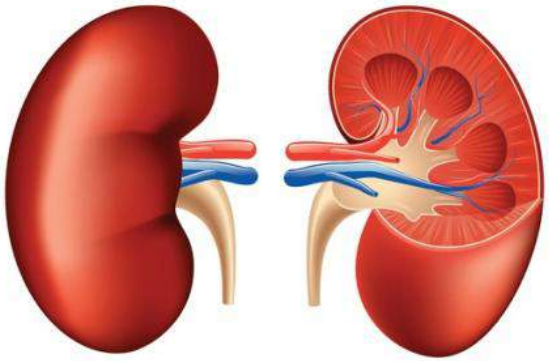
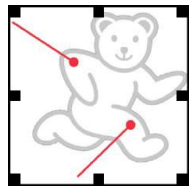
2

Organ involvement in SSc

RENAL
SSc renal crisis
MUSCULOSKELETAL
Arthritis, myopathy
Contractures
GASTROINTESTINAL
NEURAL
Autonomic neuropathy



www.downtoearth.org.in



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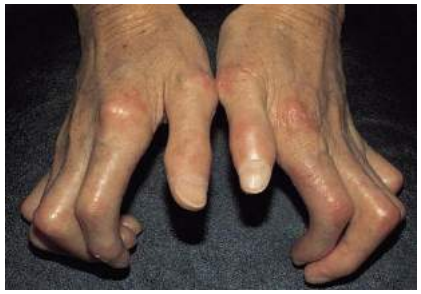
Organ involvement in SSc

RENAL
SSc renal crisis
MUSCULOSKELETAL
Arthritis, myopathy
Contractures
GASTROINTESTINAL
NEURAL
Autonomic neuropathy

Robert H. Schmerling, MD
(www.health.harvard.edu)



Nevares AM, MD (www.merckmanuals.com)

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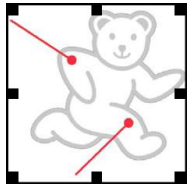

Organ involvement in SSc

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SSc renal crisis
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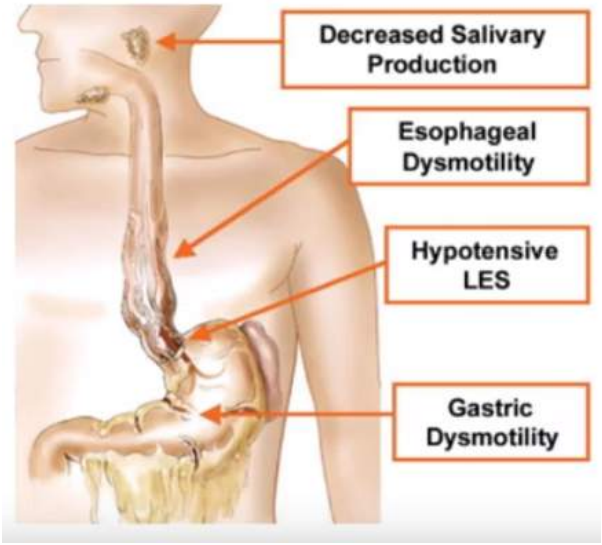
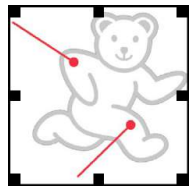


Dr Elizabeth Harrison (www.youtube.com)



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JSRD Journal of Scleroderma and Related Disorders

Original Research Article

Are diffuse and limited juvenile systemic sclerosis different in clinical presentation? Clinical characteristics of a juvenile systemic sclerosis cohort

Journal of Scleroderma and Related Disorders
1–13
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Ivan Foeldvari[1], Jens Klotsche[2], Kathryn S Torok[3], Ozgur Kasapcopur[4], Amra Adrovic[4], Valda Stanevicha[5], Maria Teresa Terreri[6], Ekaterina Alexeeva[7], Maria Katsicas[8], Rolando Cimaz[9], Mikhail Kostik[10], Thomas Lehman[11], Walter-Alberto Sifuentes-Giraldo[12], Vanessa Smith[13], Flavio Sztajnbok[14], Tadej Avcin[15], Maria Jose Santos[16], Monika Moll[17], Dana Nemcova[18], Cristina Battagliotti[19], Despina Eleftheriou[20], Mahesh Janarthanan[21], Tilmann Kallinich[22], Jordi Anton[23], Kirsten Minden[2,22], Susan Nielsen[24], Yosef Uziel[25] and Nicola Helmus[1]

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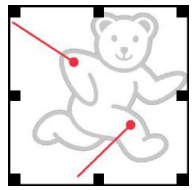

Table 1. Clinical characteristics of the patients at the time of inclusion into the cohort: demographic, subtype distribution, antibody profile and distribution of cutaneous and vascular involvement.

	Whole group (N=80)	Diffuse subtype (N=58)	Limited subtype (N=22)	p value between diffuse and limited
Female-to-male ratio	4.3:1 (65/15)	4.8:1 (48/10)	3.4:1 (17/5)	0.667
Ethnicity				
Caucasian	71 (89%)	51 (88%)	20 (91%)	0.710
African	4 (5%)	4 (7%)	0 (0%)	
Indian	3 (4%)	1 (2%)	2 (9%)	
Mean disease duration (years), mean (SD)	3.5 (3.1)	3.7 (3.2)	3.0 (2.5)	0.590
Mean age of onset of Raynaud's symptoms (years), mean (SD)	9.4 (4.0), 8 non-Raynaud	9.0 (3.8), 5 non-Raynaud	10.4 (4.3), 3 non-Raynaud	0.446
Mean age of onset of non-Raynaud's symptoms (years), mean (SD)	9.9 (4.1)	9.4 (3.7)	10.9 (4.6)	0.300
Autoantibody positivity				
ANA	78% (60/77)	79%* (44/56)	76%* (16/21)	0.937
Anti-Scl-70	31% (24/77)	30% (17/56)	33% (7/21)	0.856
Anticentromere	9% (4/46)	6% (2/33)	15% (2/13)	0.363
Inflammatory markers				
ESR elevated (>20 mm/h)	26% (20/76)	30% (17/57)	16% (3/19)	0.344
CRP elevated (>5 mg/L)	16% (11/70)	17% (9/52)	11% (2/18)	0.590
Cutaneous				
Mean modified Rodnan skin score	15.7 (0–51); n=79	18.2 (0–51); n=57	9.1 (0–24); n=22	0.004
Vascular				
Raynaud's phenomenon	90% (72/80)	91% (53/58)	86% (19/22)	0.878
Nailfold capillary changes	60% (48/80)	62% (36/58)	55% (12/22)	0.757
History of ulceration	50% (39/78)	60% (34/57)	23% (5/22)	0.068
Active ulceration	26% (10/56)	29% (10/34)	0% (0/22)	0.005

SD: standard deviation; ANA: antinuclear antibody; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein.

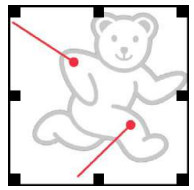

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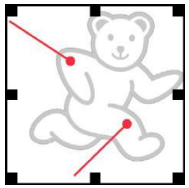
Table 2. Clinical characteristics of the patients at the time of inclusion into the cohort: cardiopulmonary involvement.

	Whole group (N = 80)	Diffuse subtype (N = 58)	Limited subtype (N = 22)	p values between diffuse and limited
Number of patients assessed for cardiopulmonary involvement	81% (65/80)	78% (45/58)	91% (20/22)	0.666
ECG done	71% (57/80)	67% (39/58)	82% (18/22)	0.605
Cardiac US done	59% (47/80)	50% (29/58)	82% (18/22)	0.206
FVC done	60% (48/80)	62% (36/58)	55% (12/22)	0.757
DLCO done	35% (28/80)	33% (19/58)	41% (9/22)	0.640
HRCT done	56% (45/80)	55% (32/58)	59% (13/22)	0.868
Pulmonary				
FVC < 80%	37% (18/48)	44% (16/36)	15% (2/12)	0.180
DLCO < 80%	53% (15/28)	53% (10/19)	56% (5/9)	0.937
6-min walk test (mean (SD))	419.3 m (138.2); n = 21	392.6 m (141); n = 16	504.6 m (85); n = 5	0.391
Interstitial lung disease Assessed by HRCT	47% (21/45)	56% (18/32)	23% (3/13)	0.128
Total pulmonary involvement	36% (29/80)	41% (24/58)	22% (5/22)	0.009
Cardiac				
Pulmonary hypertension Assessed by US	11% (5/47)	14% (4/29)	13% (1/18)	0.603
Total cardiac involvement	9% (7/80)	3% (2/58)	23% (5/22)	0.015



ECG: electrocardiography; US: ultrasound; FVC: forced vital capacity; DLCO: diffusing capacity of the lungs for carbon monoxide; HRCT: high-resolution computed tomography; SD: standard deviation.

Table 3. Clinical characteristics of the patients at the time of inclusion into the cohort: renal and gastrointestinal involvement.

	Whole group (N = 80)	Diffuse subtype (N = 58)	Limited subtype (N = 22)	p values between diffuse and limited
Renal				
Assessed by urine test	6% (5/80)	7% (4/58)	5% (1/22)	0.714
Proteinuria	4	4	0	–
Erythrocyturia	1	0	1	–
Hypertension Assessed by RR	0% (0/80)	0% (0/58)	0% (0/22)	–
Gastrointestinal				
Number of patients assessed for gastrointestinal involvement	46% (37/80)	45% (26/58)	50% (11/22)	0.803
Endoscopy done	15% (12/80)	17% (10/58)	9% (2/22)	0.425
Oesophageal scintigraphy done	9% (7/80)	7% (4/58)	14% (3/22)	0.389
Barium swallow done	26% (21/80)	24% (14/58)	32% (7/22)	0.599
Colon scintigraphy done	0% (0/80)	0% (0/58)	0% (0/22)	–
Total gastrointestinal involvement	33% (26/80)	38% (22/58)	18% (4/22)	0.212
Oesophageal involvement	69% (18/26)	68% (15/22)	75% (3 /4)	0.909
GI beside oesophageal	31% (8/26)	32% (7/22)	25% (1/4)	0.093

RR: relative risk.

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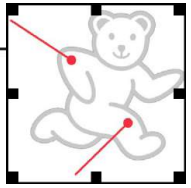
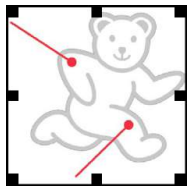


Table 4. Clinical characteristics of the patients at the time of inclusion into the cohort: musculoskeletal involvement.

	Whole group (N = 80)	Diffuse subtype (N = 58)	Limited subtype (N = 22)	p values between diffuse and limited
Musculoskeletal	62% (49/79)	58% (33/57)	73% (16/22)	0.563
Joint manifestation				
Patients with swollen joints	35% (17/49)	36% (12/33)	31% (5/16)	0.724
Number of joints with pain on motion	43% (21/49)	39% (13/33)	50% (8/16)	0.482
Patients with contractures	45% (35/77)	42% (23/55)	55% (12/22)	0.542
Muscle manifestation				
Muscle weakness	20% (9/46)	17% (6/35)	27% (3/11)	0.553
Muscle weakness and joints' contractures	13% (6/46)	11% (4/35)	18% (2/11)	0.616
Muscle weakness with no contractures	7% (3/46)	6% (2/35)	9% (1/11)	0.713
Tendon friction rub	10% (7/70)	11% (6/53)	6% (1/17)	0.515



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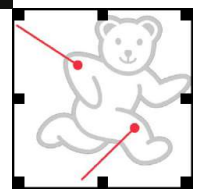
Second part : Juvenile Systemic Scleroderma

- Definition and classification
- How often does it occur?
- How is it diagnosed and followed?
- How is it treated?
- What is the long term prognosis?



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How is it treated?

- The treatment is mostly based on adult recommendations, as currently no pediatric data, beside case reports, exist
- It is a shared multidisciplinary treatment concept
- The proposed SHARE guidelines are process to be published
- Physiotherapy
- Psychosocial support
- …

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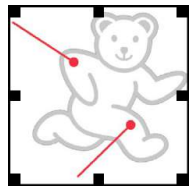

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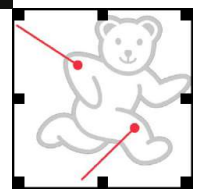

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Is there a change in organ involvement pattern after 24 months follow up in the cohorte?

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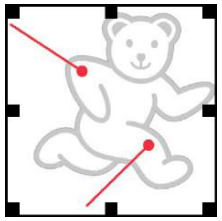

1. Demographic and Subtype Distribution

	0 month N=40	24-month follow-up N=40
Female to Male Ratio	4:1 (32/8)	
Diffuse subtype	31 (77.5%)	
Diffuse overlap	4	
Limited subtype	9 (22.5%)	
Limited overlap	4	

There is no significant change in the organ involvement distribution between time point of inclusion into the cohort and after 12 or 24 months follow up, but there are further positive changes in the patient related outcomes

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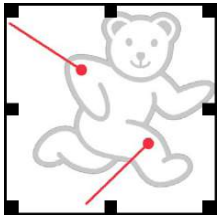
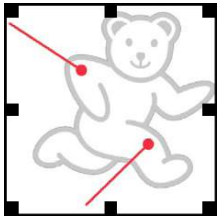


	0 months n=40	24 months follow up	P value months 0 compared 24
Physician global Disease activity	48.3 (5-80) N=22	33.2 (10-90) N=22	0.021
Physician global Disease damage	40.3 (5-80) N=21	35.7 (0-90) N=21	0.094
Patient global disease activity	49.2 (10-80) N=18	34.2 (0-90) N=18	0.001
Patient global disease damage	43.9 (10-80) N=18	34.4 (0-90) N=18	0.013
Patient Raynaud activity	26.7 (0-80) N=34	14.2 (0-70) N=34	0.045
Patient ulceration activity	19.9 (0-100) N=35	10.8 (0-60) N=35	0.069
CHAQ	0.4 (0-1.3) N=28	0.6 (0-2.625) N=19	0.791



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RHEUMATOLOGY

Rheumatology 2012;51:1832–1837
doi:10.1093/rheumatology/kes144
Advance Access publication 22 June 2012

Concise report

Juvenile and young adult-onset systemic sclerosis share the same organ involvement in adulthood: data from the EUSTAR database

Ivan Foeldvari[1], Alan Tyndall[2], Francesco Zulian[3], Ulf Müller-Ladner[4], László Czirjak[5], Chris Denton[6], Ottilia Kowal-Bielecka[7], Dominique Farge Bancel[8] and Marco Matucci-Cerinic[9,10]

The Journal of Rheumatology

The Journal of Rheumatology Volume 37, no. 11

Characteristics of Patients with Juvenile Onset Systemic Sclerosis in an Adult Single-center Cohort

IVAN FOELDVARI, SVETLANA I. NIHTYANOVA, ANGELA WIERK and CHRISTOPHER P. DENTON

J Rheumatol 2010;37;2422-2426
http://www.jrheum.org/content/37/11/2422

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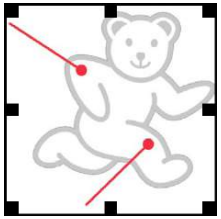


Promotion for our project!

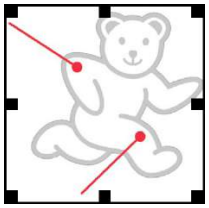
- I would like to invite you to participate on the

  Juvenile Inceptions Cohort Project

  www.juvenile-scleroderma.com

- If interested, please contact us:
  foeldvari@t-online.de or
  inceptioncohort@kinderrheumatologie.de

www.kinderrheumatologie.de



Thanks for Your interest!
I am looking forward to your questions!
☺

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