

# Juvenile systemic sclerosis

Ivan Foeldvari, MD

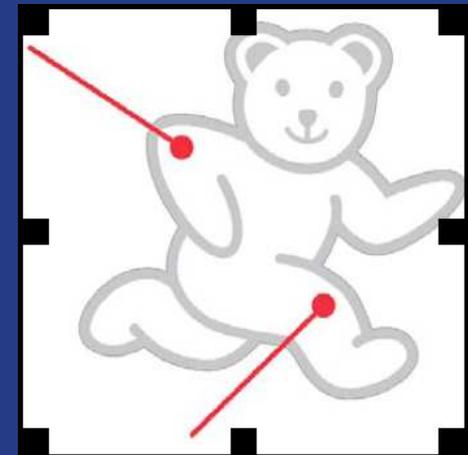
Hamburger Zentrum für Kinder- und  
Jugendrheumatologie

Hamburg, Germany

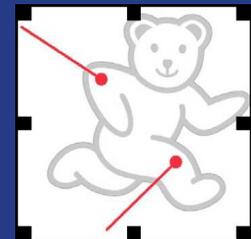
[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)

[www.sklerodermie.org](http://www.sklerodermie.org)

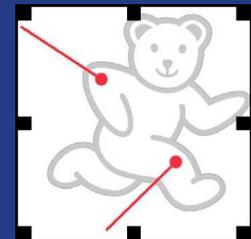
[www.uveitis-kindesalter.de](http://www.uveitis-kindesalter.de)



- Epidemiology
- Classification
- Special issues in assessment of organ involvement
- Organ involvement and outcome



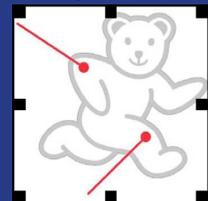
- **Epidemiology**
- Classification
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# Epidemiology and demographic of jSSc



- ◆ Data regarding incidence and prevalence is rare
- ◆ According to study from Finland
  - ◆ Incidence: 0.5 /Million
  - ◆ Prevalence: ?//  
(Pelkonen et al. J Rheumatol 1994;21:2143-6)
- ◆ According a current study by Herrick et al. Arthritis Care 2010;62:213)
  - ◆ Incidence rate 0.27 (95% CI 0.1-0.5) per million children

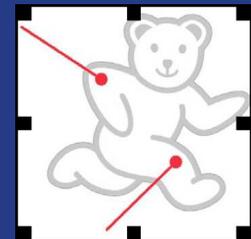


# Age at onset of juvenile systemic sclerosis

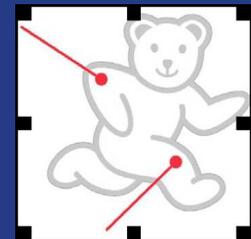
- ◆ 1.2 to 9% of all patients develop disease before age of 16 years
- ◆ Current data show that mean age at disease onset is 8.8–9.1 years\*

\*Foeldvari et al. Rheumatology 2000;  
39:556

\*Martini et al. Arthritis Rheum 2006;  
54:3971-8



- Epidemiology
- **Classification**
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# Proposed classification criteria for juvenile systemic scleroderma

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

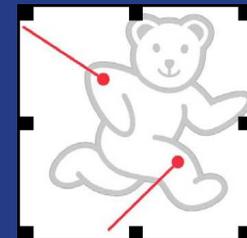
Ist and IInd International Workshop on Juvenile Scleroderma  
June 2001 and 2004 Padua, Italy

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

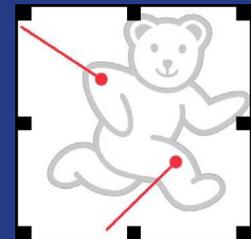
- Major criteria
  - ◆ Sclerosis\* / induration\*
- Definite disease
  - 1 major and 2 minor criteria
- ◆ Minor criteria
  - ◆ Vascular changes\*
  - ◆ Pulmonary involvement\*
  - ◆ Gastrointestinal involvement\*
  - ◆ Renal involvement\*
  - ◆ Cardiovascular involvement\*
  - ◆ Musculoskeletal involvement\*
  - ◆ Neurologic involvement\*
  - ◆ Serology\*

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\* Per definition typical for SSc

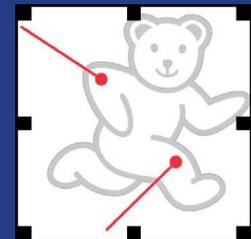


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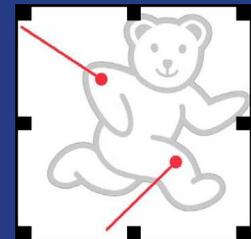
*What do we know about the applicability of standardised assessment in jSSc ?*

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# Skin - Modified Rodnan Skin Score (MRSS)

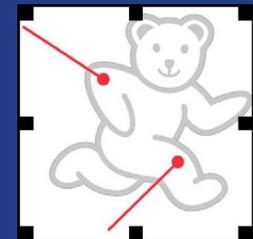
- MRSS is validated according OMERACT criteria for adults with systemic sclerosis, and fulfils all OMERACT criteria
- *Does it work the same way in children?*



# Methods

Foeldvari et al. Rheumatology 2006;45:76-8

- Evaluation of MRSS with standardised pinching method in consecutive patients, under age of 16 years, in paediatric rheumatology outpatient clinic between 1st of February and 31st of March 2004
- Exclusion criteria
  - Any sign of connective tissue disease or skin disorder influencing the score (e.g. psoriasis, ectopic dermatitis...)



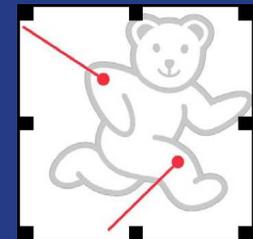
# Patients without SSc

Foeldvari et al. Rheumatology 2006;45:76-8



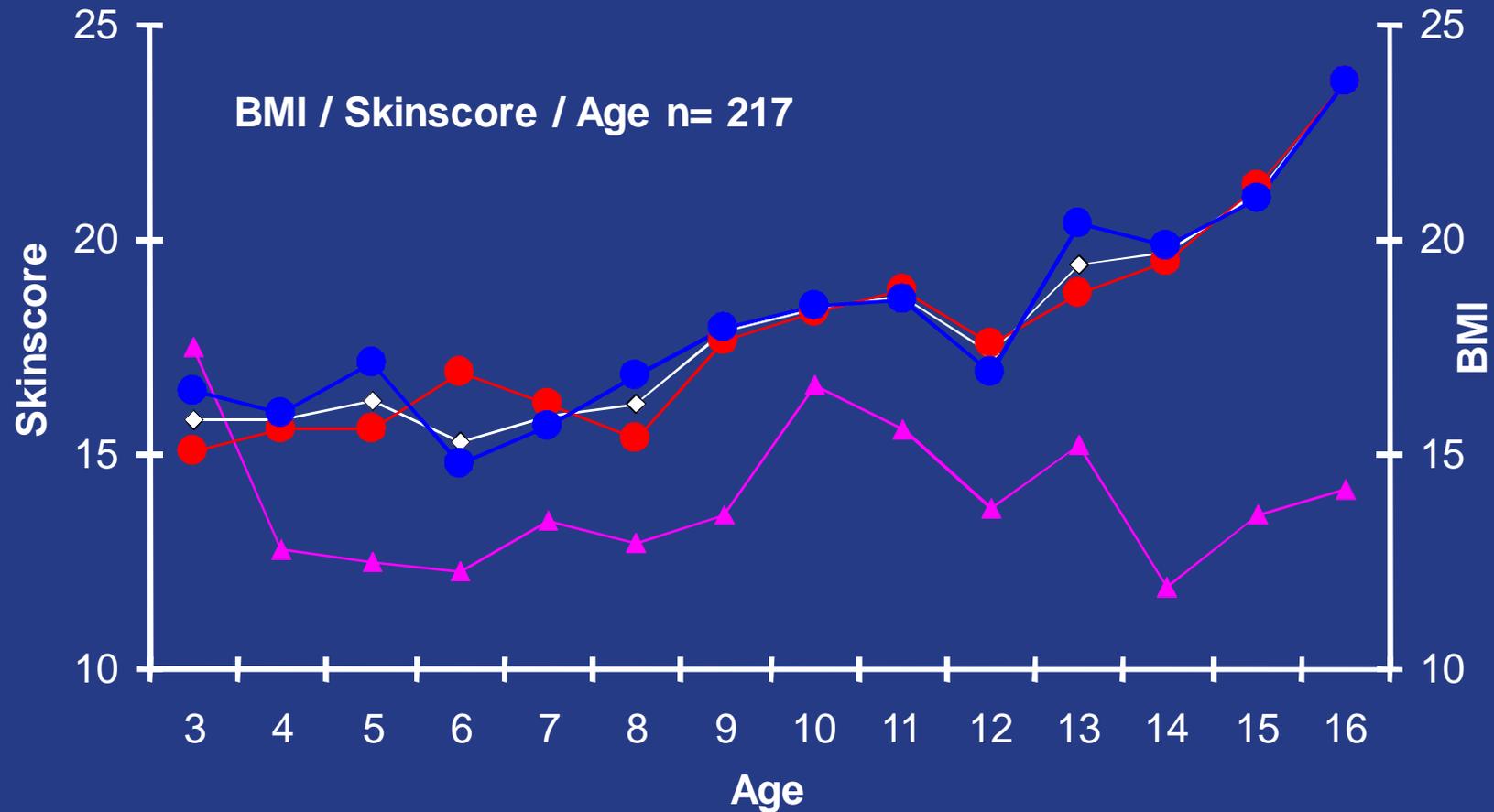
- 217 consecutive patients
- 100 female / 117 male
- Mean age: 10.5 years (range 2.9-16)
- Mean BMI: 18.3 (9.3-35.7)  
(50th percentile for age range: 15.5-20.5)
- Mean MRSS: 13.92 (range 4-25)

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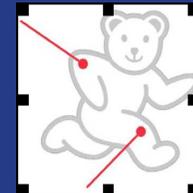
# Correlation between BMI and MRSS

Foeldvari et al. Rheumatology 2006;45:76-8



[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)

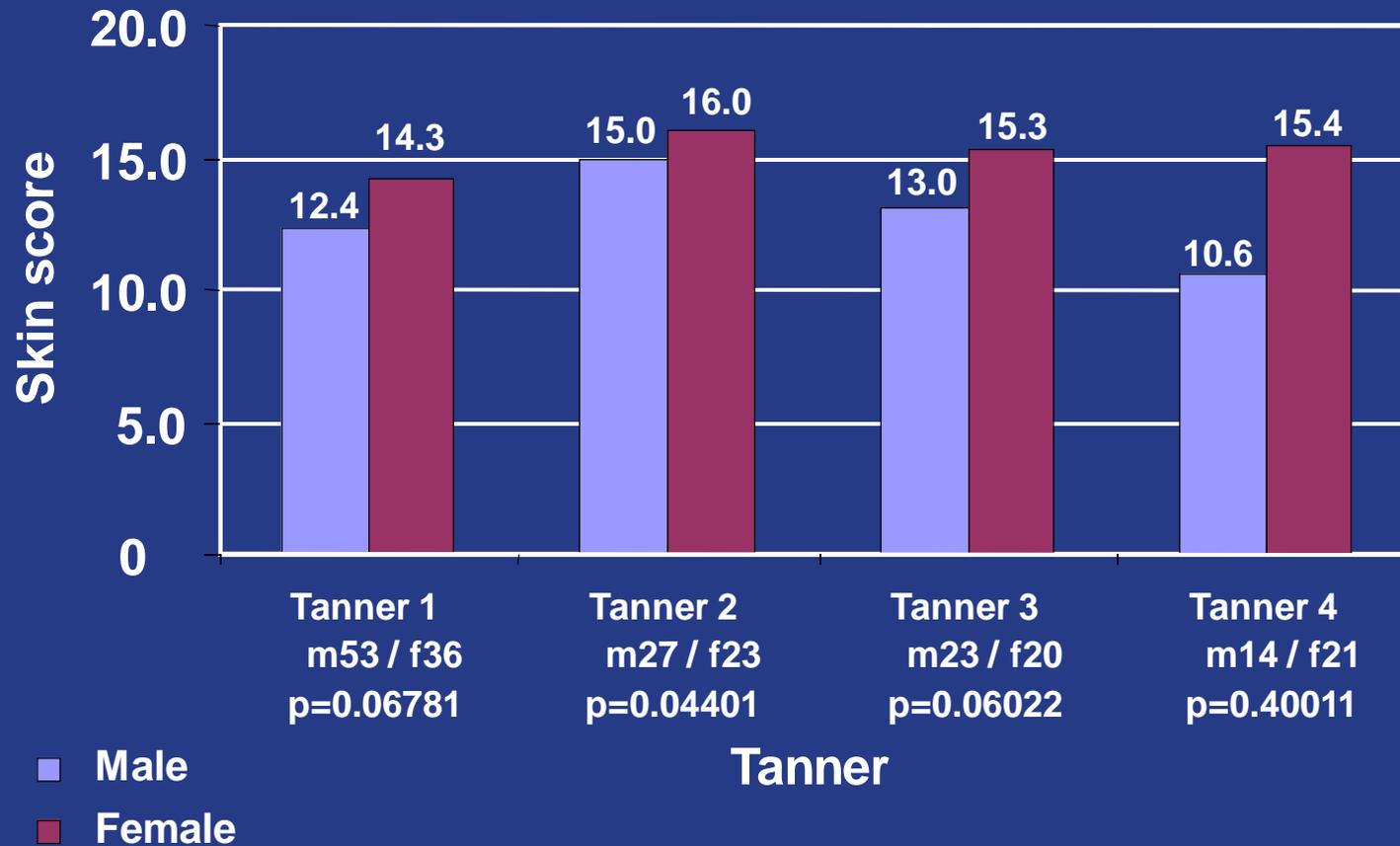
▲ Skinscore    ◆ BMI ges.  
● BMI fem.    ● BMI mal.



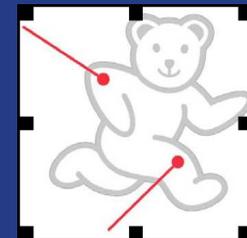
# Correlation between Tanner score and MRSS

Foeldvari et al. Rheumatology 2006;45:76-8

male n=117 / female n=100 / Tanner

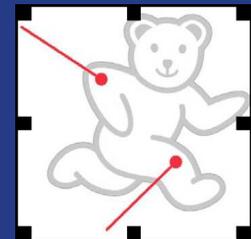


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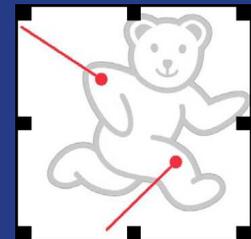
# Suggestion for adaptation of MRSS in paediatric patients

- MRSS should be corrected for
  - Tanner Stage
  - BMI of patient compared to normal BMI for age and sex
- In a multinational approach, MRSS should be tested for validity in a cross sectional juvenile SSc cohort

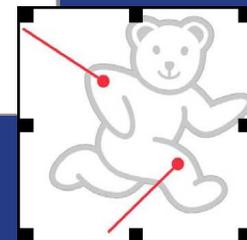
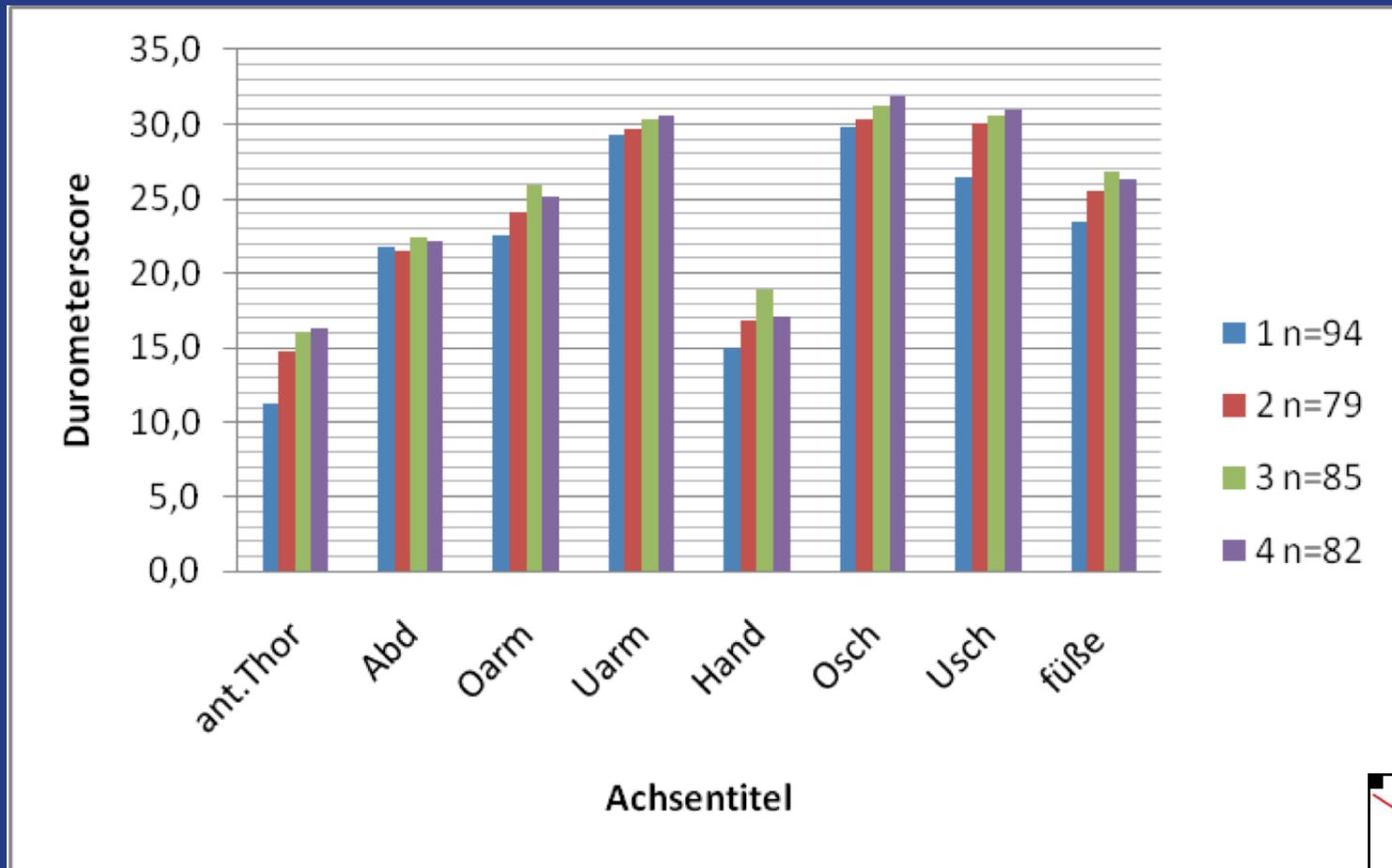


# Assessment of the Durometer as usefull tool in jSSc

- Consecutive patients of the paediatric rheumatology clinic with the diagnosis of JIA from first of November 2009 till end of Februry 2010, without a skin disease, or a skin involvement of the rheumatic disease, were prospectively evaluated for the Durometer score, with REX Gauge durometer with round head
- Aim of the study - to establish norm values for the different anatomic areas, areas with bony underlayer were excluded.
- Results - in 340 consecutive patients was the skin thicknes with the Durometer prospectively evaluated.
- The mean age of the patients was 10.7 years (rang 4.3- 17.0). 186 patients were female
- The mean values were calculated for the upper arm, for the lower arm, for the hand, for the upper leg , for the lower leg, for the back of the feet, for the abdomen and thorax.

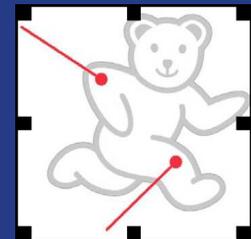


# Assessment of the Durometer as usefull tool in jSSc



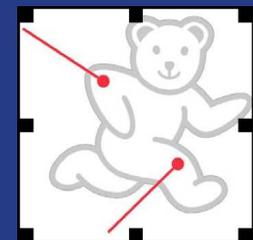
## Introduction

- ◆ Six- minute walk test (6MWT) is a frequently used indicator of functional exercise capacity.
- ◆ It was developed for patients with cardiovascular and pulmonary disease
- ◆ There are existing guidelines for the conduction of the test ( Am J Respir Crit Care 2002,166:111.7)
- ◆ It is the one of the approved primary endpoint by the Food and Drug Administration (FDA) in prospective clinical trials for patients with pulmonary hypertension
- ◆ It has its limitation in patients with pulmonary hypertension and systemic sclerosis, because of the joint involvement.



# Introduction

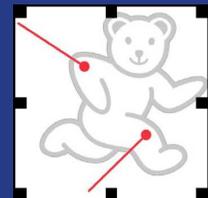
- Currently there are some studies looking at the 6MWT in children
- It is known that peak  $VO_2$  is reached around age of 14 years and it increase between age 8 and 16 years about 80% in girls and 150% in boys.
- Peak  $VO_2$  correlates with fat free body mass
- Exercise capacity is influenced by cardio-respiratory fitness, improved motor skills and movement efficiency



# Six-minute walk test in healthy children

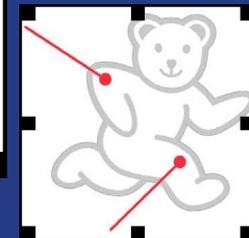
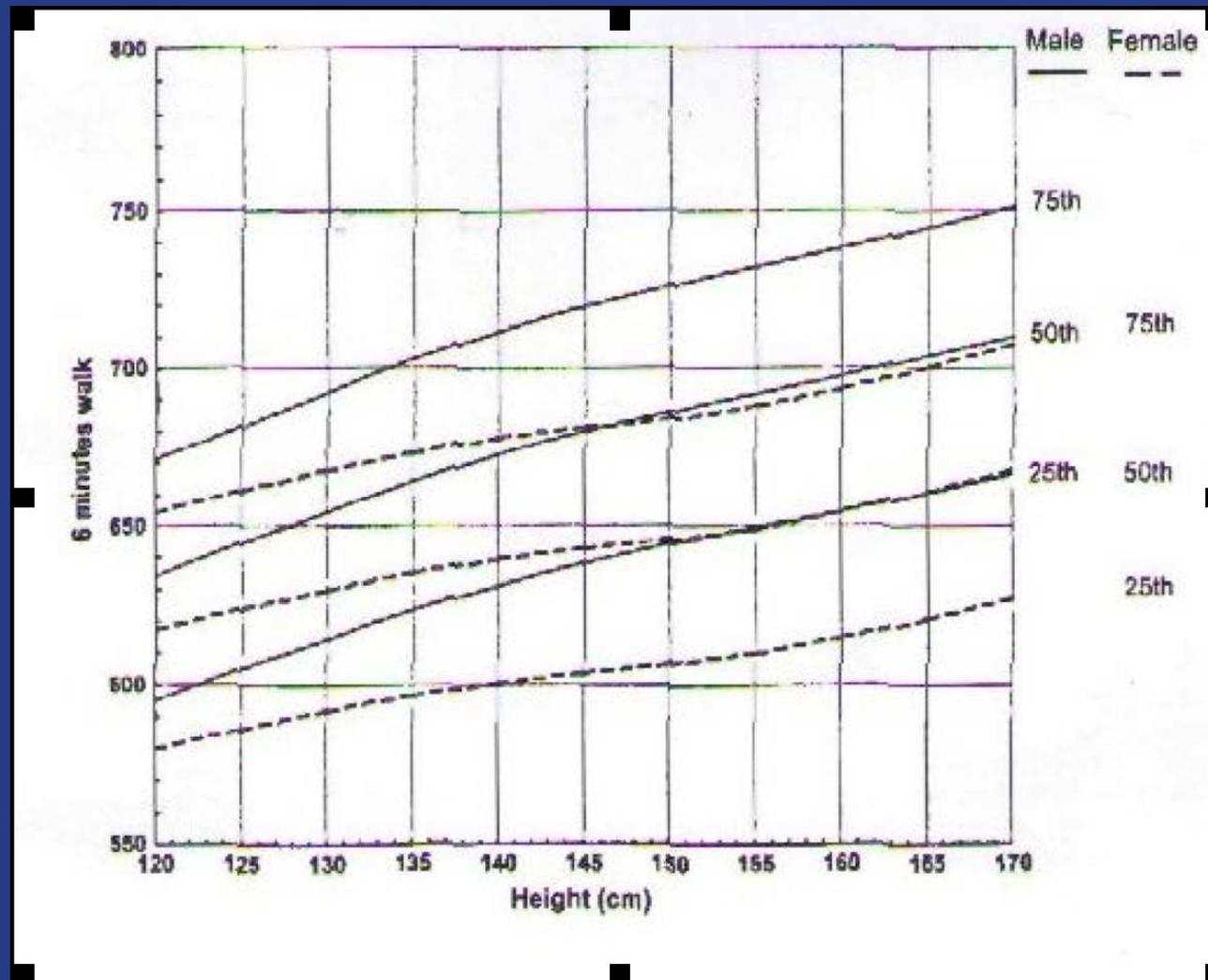
Li et al. Am J Respir Crit Care Med 2007, 176:174

- ◆ To construct height-specific standards for the 6mWT from 7 to 16 years
- ◆ 1445 **Chinese** subjects , 805 males, were studied to construct a height specific standards – which showed a normal distribution
- ◆ The mean 6MWD was 664 m +/- 65.3 m
- ◆ A percentile for the different heights and sex were established
- ◆ Height showed a better correlation than age
- ◆ Prediction equation for 6MWD
- ◆ Males =  $554.16 + (\text{difference in heart rate} \times 1.76) + (\text{height (cm)} \times 1.23)$
- ◆ Females =  $526.79 + (\text{difference in heart rate} \times 1.66) + (\text{height (cm)} \times 0.62)$



# Six-minute walk test in healthy children

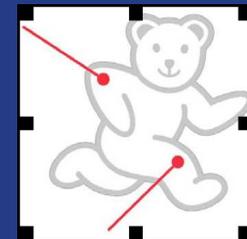
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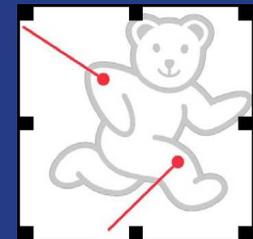
Lammers et al Arch Dis Child 2008,93:464

- ◆ Aimed to provide norm values for children between 4 and 11 years
- ◆ 378 UK primary school children , 54% males.
- ◆ Norm values for different age groups were established
  - 4 years 383 +/- 41 m
  - 5 years 420 +/-39 m
  - 6 years 463 +/-40 m
  - 7 years 488 +/-35 m ( Li et al 650 m as 50<sup>th</sup> percentile)
  - 11 years 512 +/-41m (Li et al 670 m as 50<sup>th</sup> percentile)
  - Modest increase between 7 and 11 years
  - No significant difference between boys and girls
- ◆ 6MWD correlated with age(  $r=0.64$ ), height ( $r=0.65$ ), weight ( $r=0.51$ )



Our project for 6MWT to develop  
age /height specific norm values  
supported by Actelion

- ◆ Approved by IRB and by the head office of the state school department
- ◆ 6 MWD in healthy German school children- aged 6 to 16 years
  - Measuring height, weight and **leg length**
  - **Compliance ( good / sufficient / insufficient)**
  - **Survey for physical fitness**
  - **(Tanner)**
- ◆ Development of norm values- percentiles for height / age

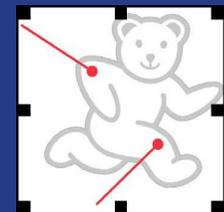


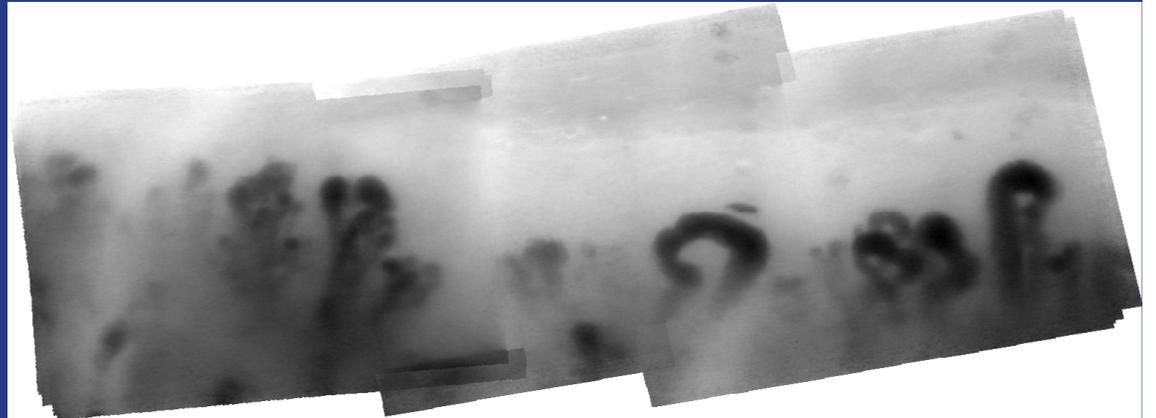
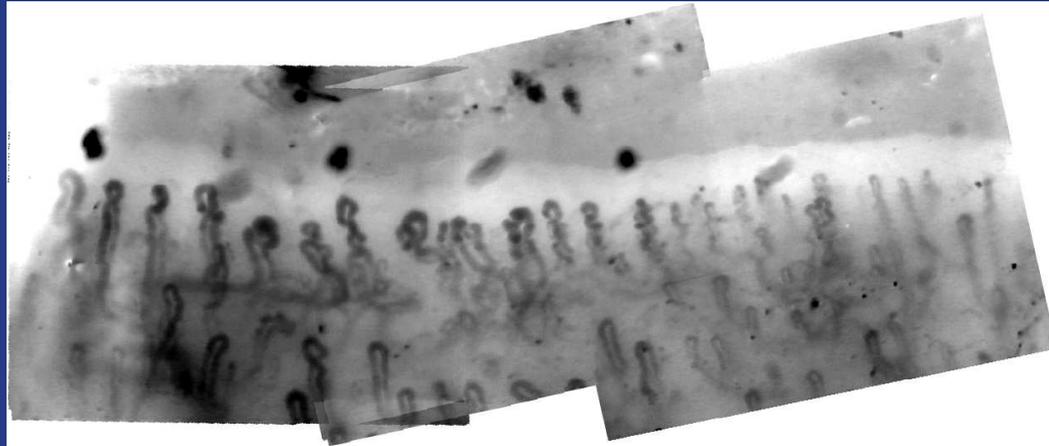
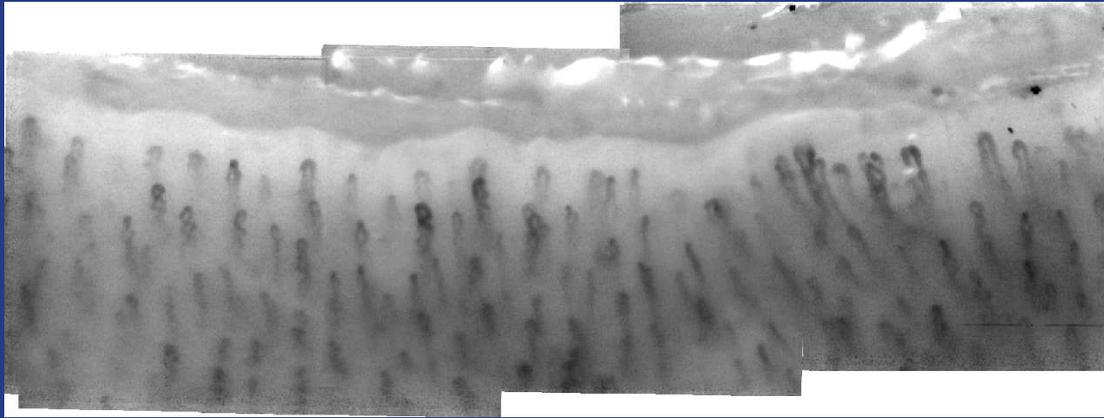
# Our project for 6MWT to develop age /height specific norm values

supported by Actelion

- Up till now 354 students participated from the age 7 to 10 years.
- The mean 6 Minute Walk Distance
- 22 in the age group of 6 years                      449.1 m
- 49 in the age group of 7 years                      470.0 m (Li et al 650 m as 50<sup>th</sup> )
- 61 in the age group of 8 years                      484.0 m
- 64 in the age group of 9 years                      491.6 m
- 50 in the age group of 10 years                      471.3 m
- 51 in the age group of 11 years                      571.0 m (Li et al 670 m as 50<sup>th</sup> )
- 57 in the age group of 12 years                      502.3 m

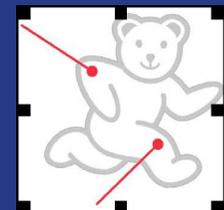
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# Technical issues

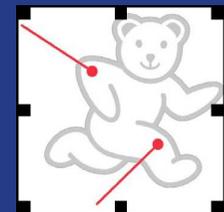
- Procedure takes 5 to 15 minutes
- Children (especially under 5s) may have problems keeping still!



# Influence of age on nailfold capillary dimension in childhood

Herrick et al. J Rheumatol 2000;27:797-800

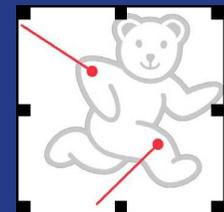
- Capillary density (number of capillaries in a 3 mm length of distal row) and capillary dimensions were measured in 110 healthy children (6 – 15 years) using nailfold microscopy technique
- In this cross-sectional study there was a significant trend for arterial and venous dimension to rise with age; this was not present for apical and loop diameters. Results did not differ between males and females
- Using capillary dimension as an outcome measure, the results should be age adjusted



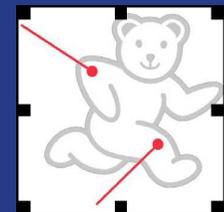
# Nailfold capillary microscopy in healthy children and those with connective tissue disease

Dolezalova et al. Ann Rheum Dis 2003;62:444-9

- Colour digital capillaroscopy was used in 26 children with CTD (3 SSc, 8 SLE, 8 JDM, 4 MCTD, 3 other), 9 with Raynaud's disease, and 17 healthy
- Capillary density and width was age related
  - Younger children having fewer (for all children: 6.9 (0.9) capillaries/mm) and wider (for all children: 3.1 [2.2-9.4] mm) capillaries than older children
- Healthy children partly had tortuous, bizarre shaped capillaries; tortuosity index median: 29% (5-49)
- CTD group had lower linear density (4.9 [1.7] capillaries/mm) and increased capillary width (10.7 (7.3) mm), and had more than two abnormal capillaries in at least two nailfolds
- Avascularity was a specific finding for CTD

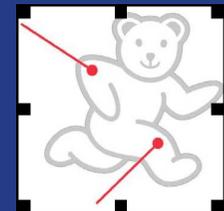


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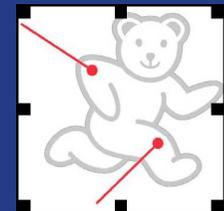
# What kind of data do we have regarding organ involvement and outcome in jSSc?

- Retrospective
  - Multinational surveys
  - Scleroderma cohorts
    - Paediatric cohort - Pittsburg cohort
    - Adult cohort with juvenile onset patients  
EUSTAR cohort / Pittsburg cohort
  - Case reports / case series
- Prospective
  - ?
  - Multinational Inception cohort project  
[www.juvenile-scleroderma.com](http://www.juvenile-scleroderma.com)  
(Foeldvari et al. - the first 8 patients are included)



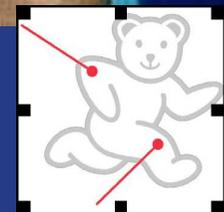
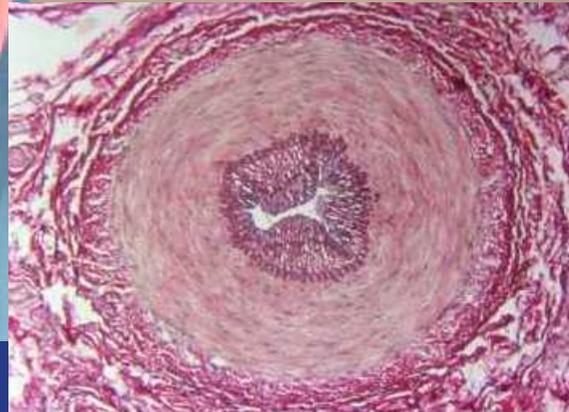
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# Organ involvement and outcome of jSSc patients

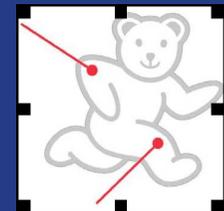
Foeldvari et al. Rheumatology 2000;39:556-9



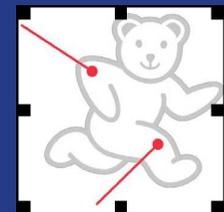
# Characteristics of the 135 patients

Foeldvari et al. Rheumatology 2000;39:556-9

<b>Sex (female / male)</b>	<b>100 / 35</b>
<b>Ethnic origin (Caucasian / non-Caucasian)</b>	<b>122 / 135</b>
<b>Mean age at disease</b>	
<b>Onset (years)</b>	<b>8.8 (<math>\pm</math> 3.3)</b>
<b>Mean disease duration at last follow-up</b>	
<b>(years)</b>	<b>5.0 (<math>\pm</math> 3.3)</b>

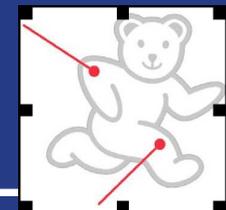


Organ involvement	Non-fatal outcome n= 127 (%)	Fatal outcome n= 8 (%)
Skin	127 (100)	8 (100)
Joints	100 (79)	6 (75)
GI tract	82 (65)	6 (75)
Only oesoph.	61 (48)	2 (25)
Pulmonary	62 (49)	6 (75)
Cardiovasc.	52 (41)	8 (100)
CNS	18 (14)	3 (38)
Renal	13 (10)	4 (50)
Muscular	13 (10)	0
Raynaud's	91 (72)	6 (75)
Calcinosis	34 (27)	2 (25)
Sjögren's syndr.	5 (4)	2 (25)
CREST	1	-



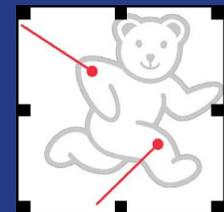
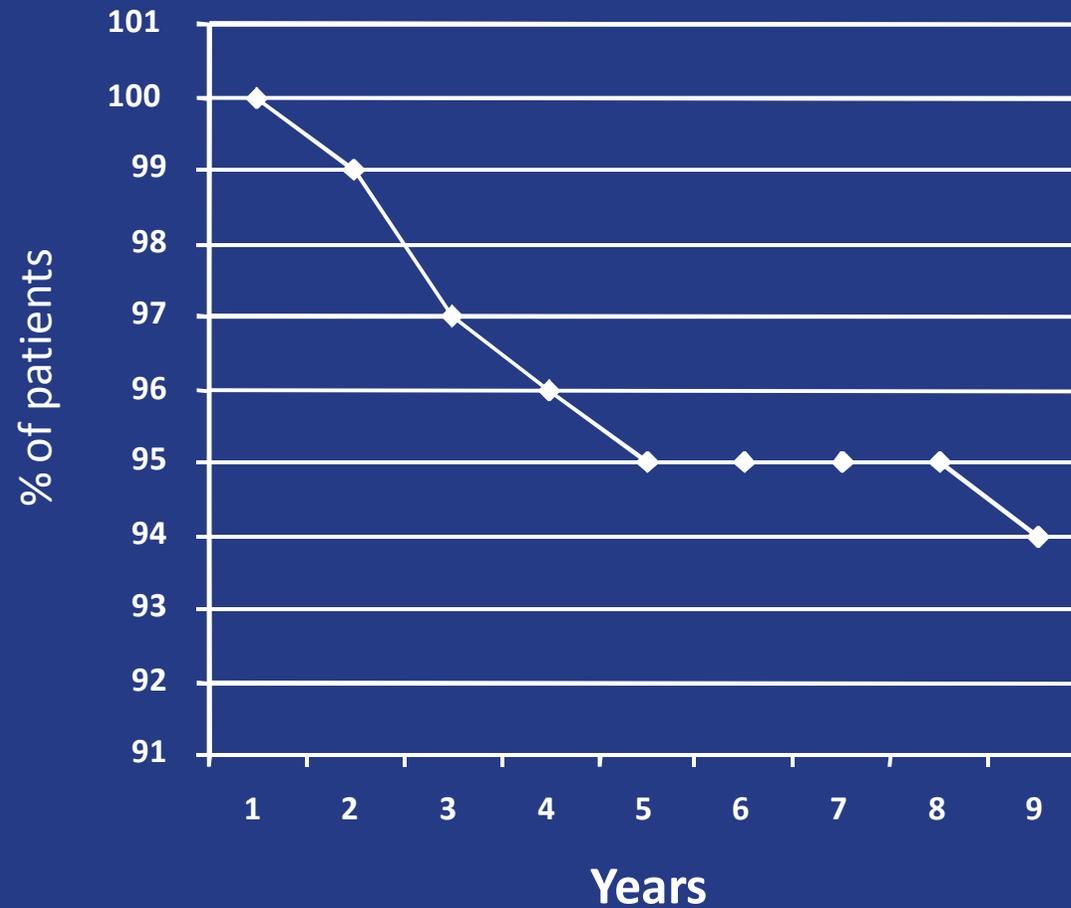
# Characteristics of patients with fatal outcome

Characteristics	n= 8
Sex (female / male)	4 / 4
Ethnic origin (Caucasian / Non-Caucasian)	6 / 2
Mean at disease onset (years)	10.5 (6.7-15.8)
Mean disease duration (years)	2.0 (1-8)
Number of patients with ANA positivity	7 / 7 (1 ND)
Number of patients with anti-Sci-70 positivity	2 / 4 (4 ND)



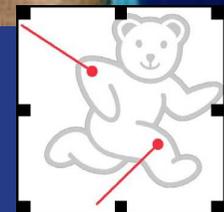
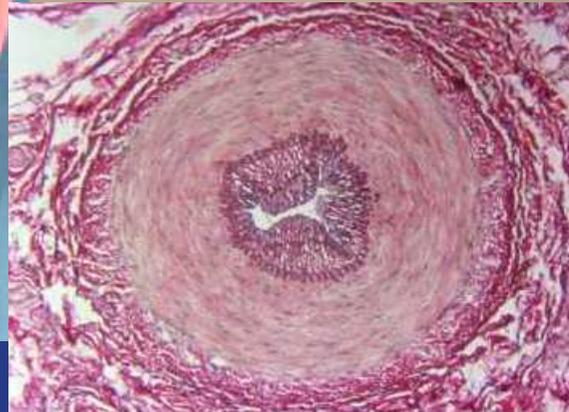
# Survival of juvenile SSc patients

Foeldvari et al. Rheumatology 2000;39:556-9



# Organ involvement and outcome of jSSc patients

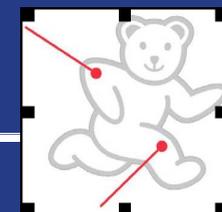
Martini et al. Arthritis Rheum 2006;54:3971-8



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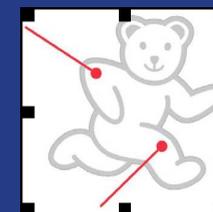
Characteristics	n= 153
Sex (female / male)	120 / 33
Mean age at disease onset (years)	8.8 (0.3–15.6)
Mean time interval between first symptom to Diagnosis	1.9 (0-12.3)
Juvenile systemic sclerosis - <i>diffuse subtype</i>	138 (90%)
Juvenile systemic sclerosis - <i>limited subtype</i>	15 (10%)
ANA positive	120/150
ENA positive	51/120
Anti-Sc1-70 positive	36/106

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Organ involvement	Foeldvari et al n= 135 (%)	Martini et al n= 153 (%)
Skin	135 (100)	116 (75.8)*
Joints	106 (79)	97(63.5)
GI tract	88 (65)	106(69)
Only oesoph.	63 (47)	47(31)
Pulmonary	68(50)	64(41.8)
Cardiovasc.	60 (44)	44(28.8)
CNS	21 (16)	4 (3)
Renal	17 (13)	15 (9.8)
Muscular	13 (10)	37(24.2)
Raynaud's	97 (72)	128(83.7)
Calcinosis	36 (27)	28(18.3)
Sjögren's syndr.	7 (5)	?
CREST	1	?

\* 75.8% skin induration; 66% sclerodactyly; 44.1% oedema

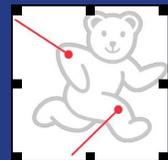


# Mortality in this patient group

Martini et al. Arthritis Rheum 2006;54:3971-8

Martini et al. Rheumatology 2009;48:119-122

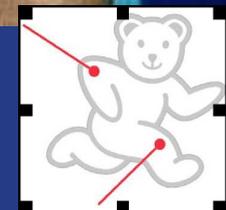
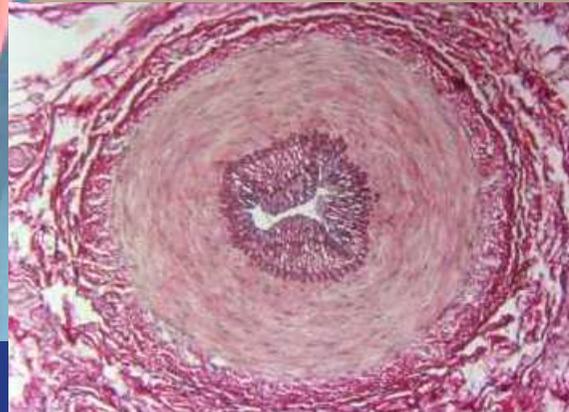
- Outcome is known in 127 of 153 patients
  - 15 of 127 patients died (11.8%)
- Cause of death
  - 10 cardiac problems (2 of them with PAH)
  - 2 renal involvement
  - 2 respiratory insufficiency
  - 1 sepsis
- Difference between survivals and non-survivals: time interval until diagnosis was 23 months compared to 8.8 months ( $p < 0.001$ )
- Mean time until death was 4.6 years after disease onset (range 0.3 to 18.8 years)
- Patients with fatal outcome had higher rate of pulmonary, GI and cardiac involvement



# Organ involvement of jSSc patients compared with the diffuse subtype SSc patients from EUSTAR cohort

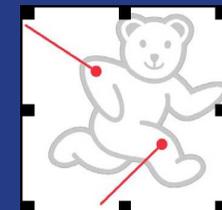
Martini et al. Arthritis Rheum 2006;54:3971-8

Walker et al. Ann Rheum Dis 2007;66:754-63



# Comparison of largest paediatric cohort with EUSTAR adult cohort

	Martini et al	EUSTAR-diffuse subtype
<b>Skin</b>		
Oedema	43.8	
Sclerodactily	66.0	
Skin induration	75.8	100
Calcinosis	18.3	
<b>Peripheral vascular system</b>		
Raynaud's phenomenon	83.7	96
Digital infarcts	28.6	43
Digital pitting	37.9	
Abnormal nailfold capillaries	39.9	
Positive capillaroscopy	51.0	
<b>Respiratory system</b>		
Dyspnoea	17.7	45
Abnormal chest x-rays	28.8	53
Abnormal chest HRCT	23.5	
Reduced DLCO	27.5	64
Reduced FVC	41.8	
<b>Cardiac involvement</b>		
Pericarditis/arrythmias	9.8	13
Heart failure	7.2	17
Pulmonary hypertension	7.2	22



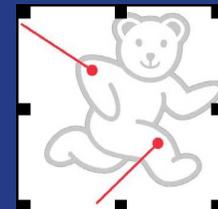
# Comparison of largest paediatric cohort with EUSTAR adult cohort

	Martini et al	EUSTAR-diffuse subtype
<b>Musculo-skeletal system</b>		
Muscle weakness	24.2	37
Arthritis	27.5	21
Arthralgia	36.0	
Tendon friction rubs	10.5	22
<b>GI system</b>		
Dysphagia	24.2	68***
Gastroesophageal reflux	30.1	68***
Diarrhoea	10.5	
Weight loss	27.5	
<b>Renal system</b>		
Raised creatinine/proteinuria	4.6	9
Renal crisis	0.7	4
Hypertension	2.6	19
<b>Nervous system</b>		
Seizures	2.6	
Peripheral neuropathy	1.3	
Abnormal brain MRI	2.6	



\*\*\* Paper states GI involvement, most common oesophagus Gthese summed

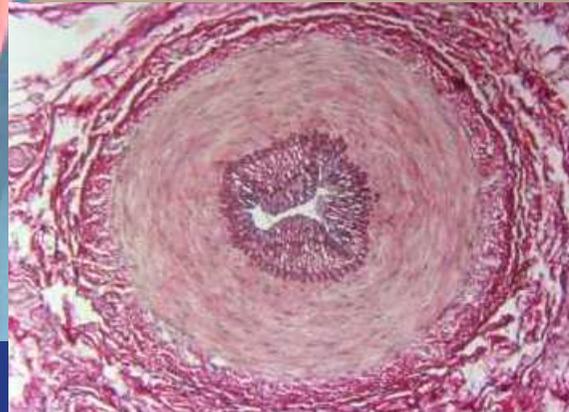
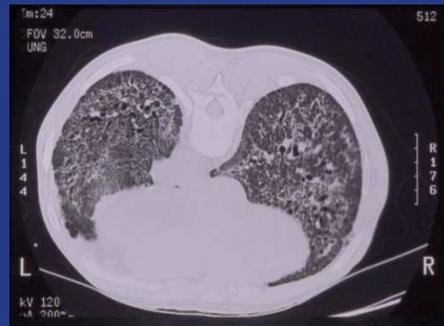
[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)



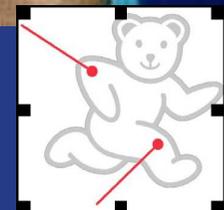
# Organ involvement of jSSc patients compared with the diffuse subtype SSc patients from EUSTAR cohort

Foeldvari et al. *Curr Opin Rheumatol* 2008;20:608-12

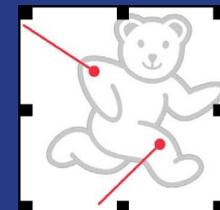
Walker et al. *Ann Rheum Dis* 2007;66:754-63



[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)

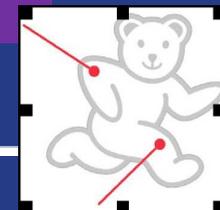


	Martini et al n=153 jSSc according to referring phys	Aoyama et al n=61 used criteria?	Russo et al n=23 according to jSSc criteria	Mista et al n=23 ACR criteria	EUSTAR aSSc n=3656 ACR criteria
<b>Gender</b>					
F:M ratio	3.6 : 1	2.75 : 1	10.5 : 1	2.1 : 1	6.7 : 1
<b>Ethnical background</b>	Mostly Caucasian	Japanese	South- American	East- Indian	Mostly Caucasian
<b>Subtype n(%)</b>					
Limited	14 (9.1)	4 (7.7)	0	9 (39)	2101 (57)**
Diffuse	139 (90.9)	48 (92.3)	23 (100)	14 (61)	1349 (37)*
Overlap					206 (5,6)
<b>Age at onset</b>					
(years) Mean	8.1	8.3	6	12	42.9
Median	9.3				
Range	0.4-15.6		(1-14)	(5-16)	
<b>Disease duration</b>					
at dg. (years)					
Mean	1.9	ND	1	4	7.4 (diffuse*)
Median	1				
Range	0 – 12.2		0.5-7	(0.2-26)	
<b>Follow-up</b>					
(years) Mean	3.9	ND	5	2,8	
Median	2.5				
Range	0.2-18.1		1-11	0-13	
<b>ANA–positiv</b>	123/150(80.7)	41/48(85)	17/23(74)	15/19(78)	92*/91**
<b>Anti-centro. +</b>	6/84(7.1)	0/15 (0)	1 (5)		6*/47**
<b>Anti-topo I +</b>	36/120(34)	21/36(60)	2 (9)		60.8*/23**
<b>Survival</b>	90% (5 years)	ND	94% (5 years)	?	



# Organ involvement of paediatric jSSc cohorts and EUSTAR adult SSc cohort

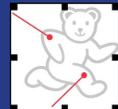
	<i>Martini et al</i>	<i>Aoyama et al</i>	<i>Russo et al</i>	<i>Misra et al</i>	<i>EUSTAR-diffuse subtype</i>
<b>Skin</b>					
Oedema	43.8		52		
Sclerodactily	66.0		91		
Skin induration	75.8		100		100
Calcinosis	18.3		48		
<b>Peripheral vascular system</b>					
Raynaud's phenomenon	83.7		83	83	96
Digital infarcts	28.6			60	43
Digital pitting	37.9		65		
Abnormal nailfold capillaries	39.9		74		
Positive capillaroscopy	51.0				
<b>Respiratory system</b>					
Dyspnoea	17.7		26		45
Abnormal chest x-rays	28.8		41(		53
Abnormal chest HRCT	23.5	23.7	27 (6/11)	65	
Reduced DLCO	27.5		14		64
Reduced FVC	41.8		65		
<b>Cardiac involvement</b>					
		29.4			
Pericarditis/arrythmias	9.8		20		13
Heart failure	7.2				17
Pulmonary hypertension	7.2		10	4	22



# Organ involvement of paediatric jSSc cohorts and EUSTAR adult SSc cohort

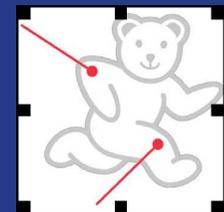
	<i>Martini et al</i>	<i>Aoyama et al</i>	<i>Russo et al</i>	<i>Misra et al</i>	<i>EUSTAR-diffuse subtype</i>
<b>Musculo-skeletal system</b>					
Muscle weakness	24.2		35		37
Arthritis	27.5		35	35	21
Arthralgia	36.0		61		
Tendon friction rubs	10.5		14		22
<b>GI system</b>					
Dysphagia	24.2		39	30	68***
Gastroesophageal reflux	30.1	42.4	18	35	68***
Diarrhoea	10.5		9		
Weight loss	27.5		48		
<b>Renal system</b>					
Raised creatinine/proteinuria	4.6		9	0	9
Renal crisis	0.7	3.8	0	0	4
Hypertension	2.6				19
<b>Nervous system</b>					
Seizures	2.6		5		
Peripheral neuropathy	1.3				
Abnormal brain MRI	2.6				

\*\*\* Paper states GI involvement, most common oesophagus Gthese summed together with dysphagia and reflux



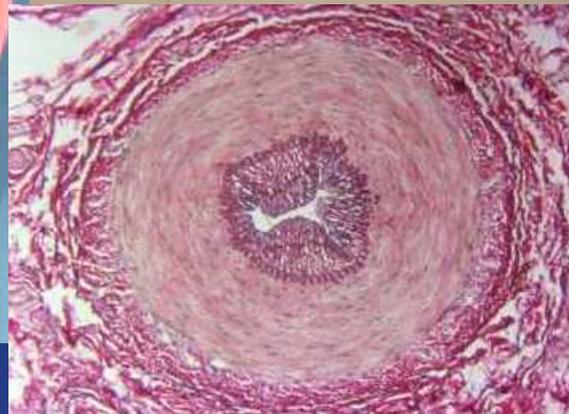
# What kind of data do we have regarding organ involvement and outcome in jSSc?

- Retrospective
  - Multinational surveys
  - Scleroderma cohorts
    - Paediatric cohort - Pittsburg cohort
    - Adult cohort with juvenile onset patients  
EUSTAR cohort / Pittsburg cohort
  - Case reports / case series
- Prospective
  - ?
  - Multinational Inception cohort project  
[www.juvenile-scleroderma.com](http://www.juvenile-scleroderma.com)  
(Foeldvari et al. - the first 20 patients are included)

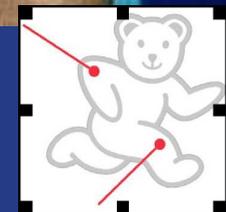


# Organ involvement and outcome of jSSc patients. Data from EUSTAR / Royal Free and Pittsburgh cohort

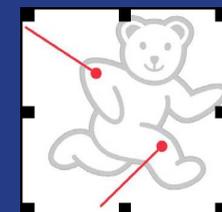
Foeldvari et al. Arthritis Rheum 2007;56(Suppl):A1,S53



[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)



	Juvenile systemic scleroderma cohort in % (100% equals n= 60)	Adult systemic scleroderma cohort in % (100 equals n= 910)
Raynaud phenomenon	95	95.1
Digital ulcers	35	41.2
Pulmonary hypertension	13.3	14.1
Pulmonary fibrosis	23.3	35.1
Renal hypertension	8.3	11
Renal crisis	0	2
Proteinuria	3.3	5.8
Oesophageal involvement	60	65
Gastric involvement	16.7	25.9
Intestinal involvement	15	22.3
Synovitis	10	15.4
Joint contractures	30	36.7
Muscle weakness	20	24.2
Tendon friction rub	8.3	12.2



# Pittsburgh cohort\* in comparison with EUSTAR\*\*, Royal Free\*\*\* and the PRES cohort\*\*\*\*

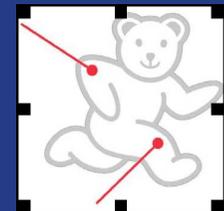
\*Retrospective Evaluation of the Pittsburgh Databank with 111 jSSc and 2559 SSc patients recruited between 1960 and 2003  
Scalapino et al. J Rheumatol 2006;33:1004-13

\*\*Foeldvari. Curr Opin Rheum 2008;20:608-13

\*\*\*Foeldvari et al. Arth Rheum 2008;Suppl

\*\*\*\*Martini et al. Arthritis Rheum 2006;54:3971-8

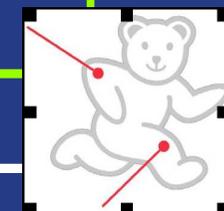
[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)



	jSSc in EUSTAR (n= 60)	jSSc Pittsburgh (3) (n= 57)	jSSc Royal Free H. (n= 46)	jSSc PRESS (5) (n= 153)
Mean age at disease onset	12.4 (2-15.9)	?	13.06 (5 to16)	8.1 (0.4-15.6)
Disease duration	17.64 (1.8-54.8)	17.2	21.15 (3 to 58)	3.9 (0.2-18.1)
Sex: male /female (ratio)	5/55 (11)	19/92 (4.8)	11/35 (3.2)	33/120 (3.6)
Disease subtype diffuse (%)	40	35	39	90.9
Disease subtype limited (%)	46.7	40	61	9.1
Overlap features (%)			43.5	
Outcome – alive (%)	59 (98%)	89% 5y / 74% 20y	97% 15y / 93%	112/127(88)
- died (%)	-		20y / 83% 25y	15/127(12)
- lost to follow up (%)	1 (2%)		-	26 (17%)



	jSSc in EUSTAR (n= 60)	jSSc Pittsburgh (3) (n= 57)	jSSc Royal Free H. (n= 46)	jSSc PRESS (5) (n= 153)
ANA positive (%)	90	97		80
Anti-Scl 70 positive (%)	40	23	26	34
Anti-centromere positive (%)	5	0	6.5	7.1
Raynaud's phenomenon (%)	95	96	MD	83.7
Pulmonary hypertension (%)	13.3	3.6	15	7.2
Pulmonary fibrosis (%)	23.3	9	47	23.5
Renal crisis (%)	0	3.6	2	0.7
No org inv beside vascular in %	MD	MD	15	MD





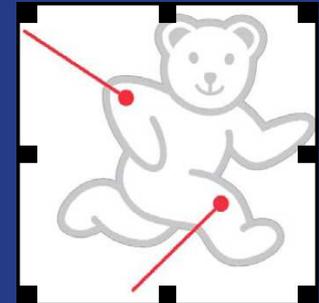
**Scleroderma Working Group**

# Update on the Inception cohort study for Juvenile systemic Scleroderma

Dr. Ivan Foeldvari

Hamburger Zentrum für Kinder- und  
Jugendrheumatologie

Kompetenz-Zentrum für Sklerodermie im Kindes-  
und Jugendalter



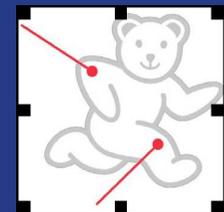


# Short summary of the project

## Inclusion criteria

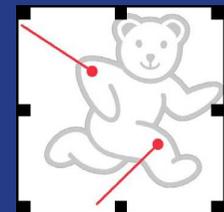
- Patients with juvenile systemic sclerosis according the proposed paediatric criteria\*, with a disease course of less then 18 months after the first non- Raynaud organ involvement

\*Zullian et al, Arthritis and Rheumatism 2007, 57: 203-12.



# Method

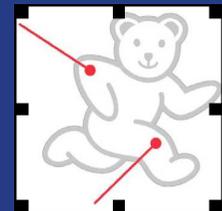
- Prospective standardized assessment of the organ involvement- every 6 months
- follow the patients over 36 months to assess the standardized outcome measures according the OMERACT filter process
- See detailed protocol under [www.juvenile-scleroderma.com](http://www.juvenile-scleroderma.com)
- Need for IRB approval depends on the country



# www.juvenile-scleroderma.com

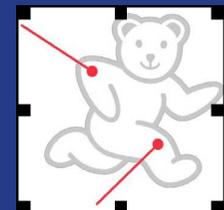
- ◆ If you have any questions please contact me-  
or the coordinating study nurse
- ◆ E-mail:
  - [sprechstunde@kinderrheumatologie.de](mailto:sprechstunde@kinderrheumatologie.de)
  - [studynurse@kinderrheumatologie.de](mailto:studynurse@kinderrheumatologie.de)
- ◆ Study protocol, consent and assent forms are on the homepage of the project- access possible after the registration for the project

[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)



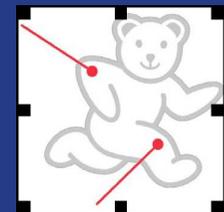
## Summary of the current data from the cohort

- 16 patients – with a mean age of first non Raynaud symptoms of 12.4 years ( range 6.9- 16)
- Subset distribution:
  - 9 diffuse
  - 3 limited
  - 4 overlap features
- Mean modified skin score at entry into the registry 16.5( range 2 to 51)
- Raynaud in 14 of 16 patients, with nailfold changes in 10 of 16.
- Cardiovascular involvement in 7 patients
- GI involvement in 9 patients
- Musculoskeletal involvement in 14



# Future...

- All the current data is retrospective data, without standardised assessment of the organ involvement or quality of life
- In the currently running prospective Inception cohort project, [www.juvenile-scleroderma.com](http://www.juvenile-scleroderma.com), a homogenous paediatric patient population will be created; we will learn about the evolvement of the certain organ involvements and about the effect of our current therapies on this organ involvement



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



[www.kinderrheumatologie.de](http://www.kinderrheumatologie.de)

