EMIRATES DERMATOLOGY SOCIETY
2024
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8th YEAR
08-10 November 2024
InterContinental Dubai - Festival City
United Arab Emirates

EMIRATES DERMATOLOGY SOCIETY

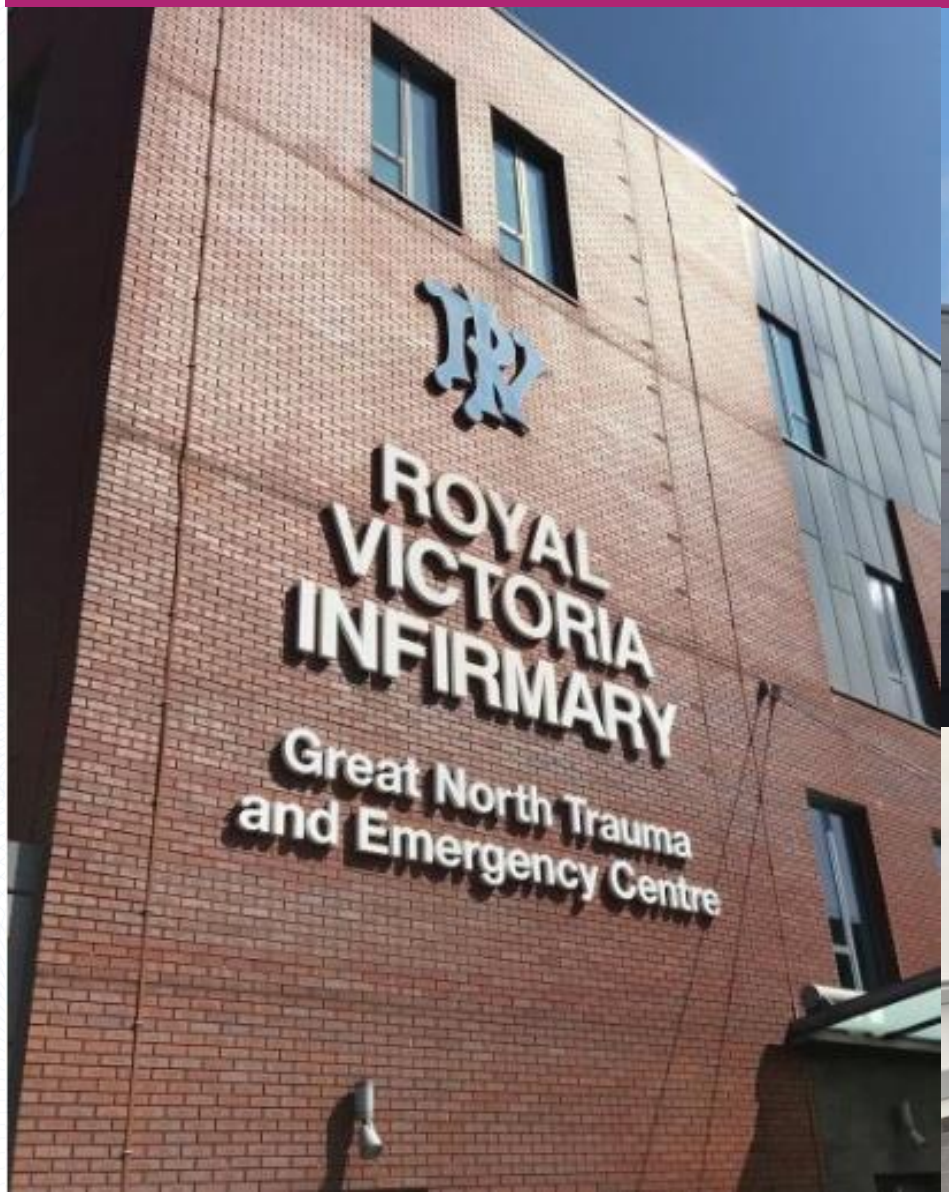
# Morphoea in children, experience from a joint dermatology rheumatology clinic

Dr Charlotte Goodhead

27TH WORLD CONGRESS OF DERMATOLOGY 2031
DUBAI - CANDIDATE CITY

# Children's rheumatology

We provide specialist care for children with inflammatory diseases,





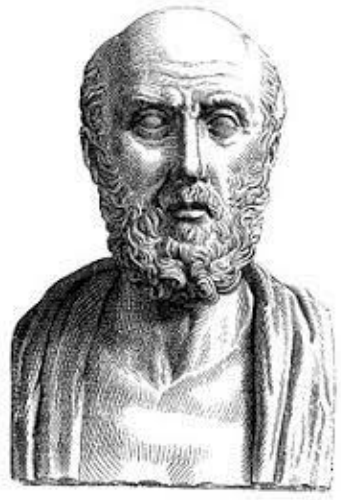
The Newcastle upon Tyne Hospitals NHS
NHS Foundation Trust

# CONFLICT OF INTEREST

# Learning Objectives

- Clinical features
- Scoring tools
- Management
- Case examples
- Psychosocial impact

# Morphoea, Morphea and Scleroderma….



‘Scleroderma’ was probably first used by Giovambattista Fantonetti in 1836 to describe a 30-year-old patient with skin swelling, pustules and progression to hard brown skin said to be ‘as firm as leather’

- **Scleroderma** hard skin
- **Morphoea** form and structure

> “In order to avoid unnecessary confusion on the part of both patients and doctors of other specialties, the authors postulate that localized scleroderma should be called morphea” (Dańczak-Pazdrowska 2021)

(De Silva 1994)

# Classification

**Tuffanelli and Winkelmann classification 1961**

**Morphea**

Circumscribed, sclerotic plaques with an ivory-coloured centre and surrounding violaceous halo.

**Linear scleroderma**

Linear, band-like distribution

Frontal or frontoparietal linear scleroderma (en coup de sabre) is characterized by atrophy and a furrow or depression that extends below the level of the surrounding skin

**Generalized morphea**

Widespread skin involvement with multiple indurated plaques, hyperpigmentation and frequent muscle atrophy

(Asano 2018)

## Peterson et al.classification 1995

**Plaque morphea**
- Plaque morphea
- Guttate morphea
- Atrophoderma of Pasini and Pierini
- Keloid morphea (nodular morphea)

(Lichen sclerosus et atrophicus)

**Generalized morphea**

**Bullous morphea**

**Linear morphea**
- Linear morphea (linear scleroderma)
- Morphea en coup de sabre
- Progressive facial hemiatrophy

**Deep morphea**
- Morphea profunda
- Subcutaneous morphea
- Eosinophilic fasciitis
- Panscleroticmorphea of

## Padua Consensus classification 2006

**Circumscribed morphea**
(i) Superficial
(ii) DeepLinear

**Scleroderma**
(i) Trunk/limbs
(ii) Head

**Generalized Morphea**

**Pansclerotic morphea**

**Mixed morphea**

# Learning Objectives

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**Sclerosis of skin that can result in significant cosmetic and functional impairment**

- Often well-circumscribed
- Firm with loss of normal elasticity
- Often with a central pale or pigmented area surrounded by reddening at the margins

**Two distinct phases;**

1. Inflammatory active stage with erythema and enlargement
2. Inactive, sclerotic, atrophic stage of damage

**Can have a varied clinical presentation**

PAEDIATRIC DERMATOLOGY DOI 10.1111/j.1365-2133.2008.08647.x

# Linear morphoea follows Blaschko's lines

L. Weibel and J.I. Harper

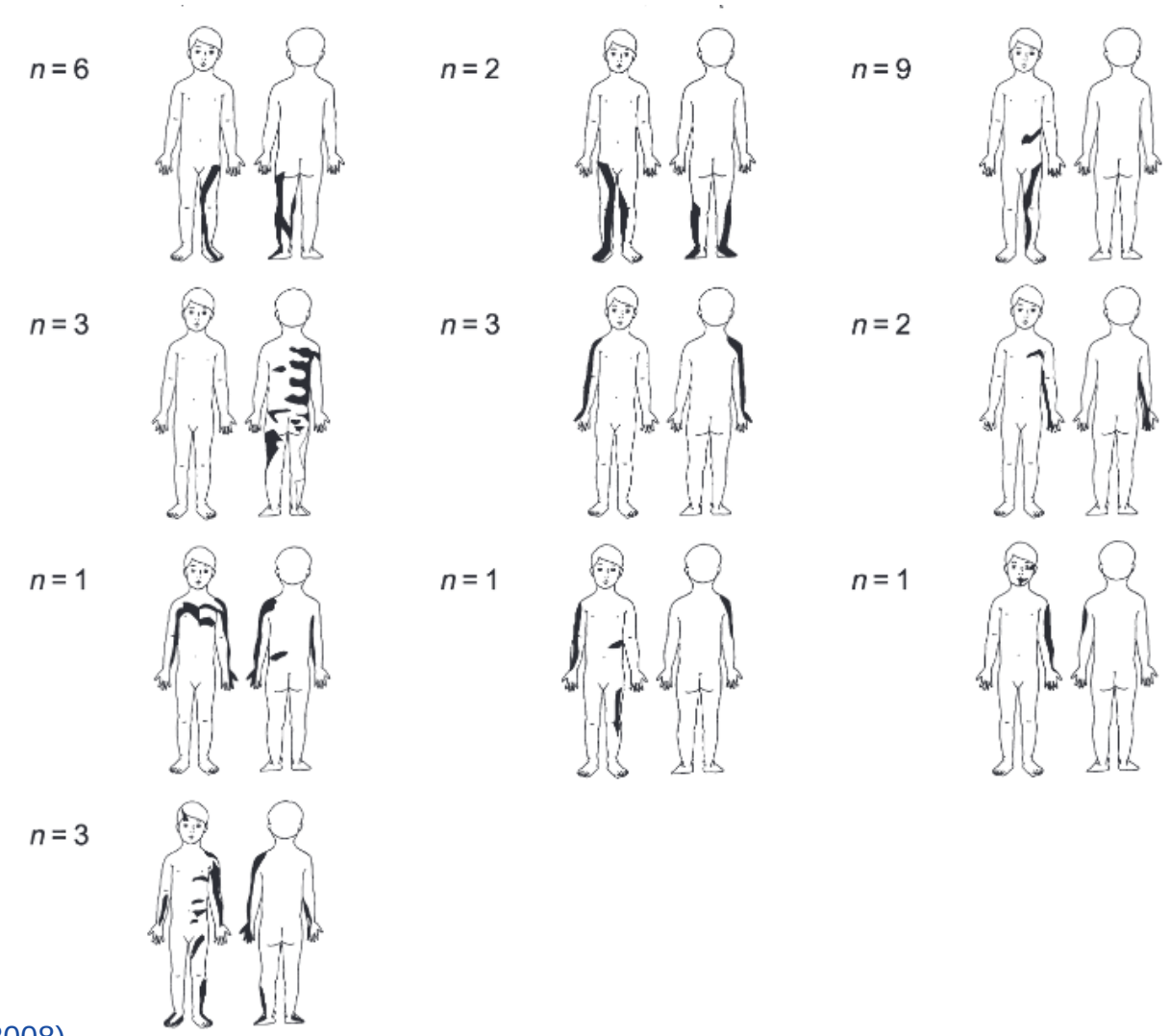
Department of Paediatric Dermatology, Great Ormond Street Hospital for Children, London WC1N 3JH, U.K.

A retrospective chart review of 65 children with linear morphoea

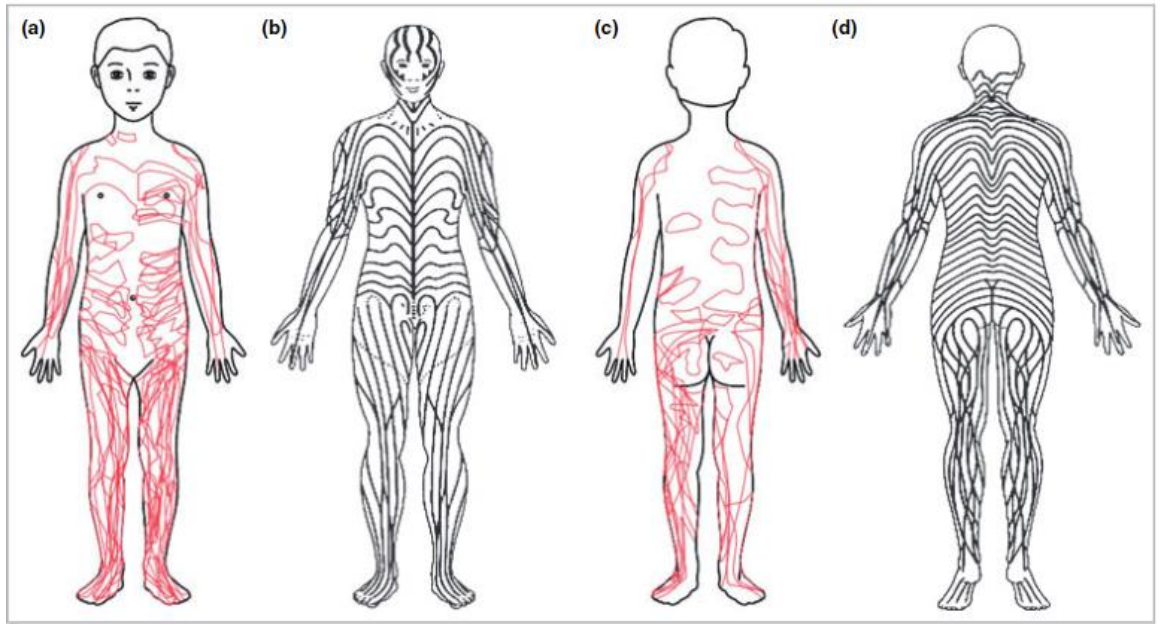
- 53% had the en coup de sabre subtype,
- 41% linear morphoea on the trunk and/or limbs
- 6% a combination
- 85% were confined to one side of the body

Data indicated linear morphoea follows the lines of Blaschko

Authors hypothesize that in these patients' susceptible cells are present in a mosaic state, exposure to a trigger may result in the condition




(Weibel 2008)

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# Learning Objectives

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MEDICAL DERMATOLOGY

BJD
British Journal of Dermatology

# Using the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) to classify morphoea by severity and identify clinically significant change

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**Linked Comment:** Torok. *Br J Dermatol* 2020; **182**:272–273.

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**Conflicts of interest**
None to declare.

DOI 10.1111/bjd.18097

## Summary

Background Validated scoring measures in morphoea can facilitate clinical trials.
Objectives To ascertain the clinical significance of scores on the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) and identify the change in scores correlated with clinically meaningful change.
Methods A prospective study of 120 participants from the Morphea in Adults and Children (MAC) cohort was undertaken. Physician's subjective assessments of severity and of improvement were completed at each visit. Receiver operating characteristic analysis determined LoSCAT scores corresponding with mild, moderate and severe disease, and absolute and percentage changes in scores corresponding with improved or worsened disease activity or damage.
Results Mild, moderate and severe activity corresponded with LoSCAT activity index (LoSAI) scores of 0–4, 5–12 and 13 and over, and with Physician's Global Assessment of activity (PGA-A) scores of 0–10, 11–30 and 31 and over. Mild, moderate and severe damage corresponded with LoSCAT damage index (LoSDI) scores of 0–10, 11–15 and 16 and over, and with PGA of damage (PGA-D) scores of 0–18, 19–30 and 31 and over. Improved activity was best indicated by LoSAI decrease of at least 2 points or 27·5%, or PGA-A decrease of at least 6 points. Improved damage was best indicated by LoSDI score decrease of at least 2 points. Worsening activity was best indicated by LoSAI increase of at least 2 points or 19·5%, or PGA-A increase of at least 4 points. Worsening damage was best indicated by LoSDI increase of at least 25·5%.
Conclusions The LoSCAT can be used to classify patients with morphoea by disease severity, and identify clinically significant improvement in activity.

**What's already known about this topic?**

- The Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) is a clinical tool that separately quantifies disease activity and damage in morphoea, and prior studies have demonstrated validity and reliability.

**What does this study add?**

- The LoSCAT can be used to classify patients with morphoea by disease severity into

# LoSCAT

## Localised Scleroderma Cutaneous Assessment Tool

## LoSAI & LoSDI

Validated clinical measure
Accounts for evolution of over time

LoSAI & PGA-A - Activity
LoSDI & PGA-D - Damage

(Teske 2020)

**LoSCAT**
Localized Scleroderma Cutaneous Assessment Tool

| | | LoSAI (Localized Scleroderma Skin **Activity** Index) | | | LoSDI (Localized Scleroderma Skin **Damage** Index) | | | |
|---|---|---|---|---|---|---|---|---|
| | | New/Enlarged (past month)<br>0 = none<br>3 = N / E | Erythema<br>0 = none<br>1 = pink<br>2 = red<br>3 = dark red /violaceous | Induration (skin swelling at EDGE)<br>0 = none<br>1 = mild<br>2 = moderate<br>3 = marked | Dermal atrophy<br>0 = none<br>1 = shiny<br>2 = visible vessels<br>3 = diff drop | Sub Q / Deep atrophy<br>0 = none<br>1 = flat<br>2 = concave<br>3 = marked | Dyspigmentation (hyper or hypo)<br>0 = none<br>1 = mild<br>2 = moderate<br>3 = marked | Skin Thickness (at CENTER)<br>0 = none<br>1 = mild<br>2 = moderate<br>3 = marked |
| | Scalp/Face | | | | | | | |
| | Neck | | | | | | | |
| | Chest | | | | | | | |
| | Abdomen | | | | | | | |
| | Upper Back | | | | | | | |
| | Lower Back | | | | | | | |
| RT | Arm | | | | | | | |
| RT | Forearm | | | | | | | |
| RT | Hand | | | | | | | |
| RT | Thigh | | | | | | | |
| RT | Leg | | | | | | | |
| RT | Foot | | | | | | | |
| LT | Arm | | | | | | | |
| LT | Forearm | | | | | | | |
| LT | Hand | | | | | | | |
| LT | Thigh | | | | | | | |
| LT | Leg | | | | | | | |
| LT | Foot | | | | | | | |

LoSAI _____ LoSDI_____

**PGA-A** (Physician Global Assessment of Disease Activity)

(0=inactive) (100=markedly active)

**PGA-D** (Physician Global Assessment of Disease Damage)

(0=no damage) (100=markedly damaged)

## LoSAI – Activity index

| | |
|---|---|
| 0-4 | Mild |
| 5-12 | Moderate |
| ≥13 | Severe |

## LoSDI – Damage index

| | |
|---|---|
| 0-10 | Mild |
| 11-15 | Moderate |
| ≥16 | Severe |

Example scores over 2 years:

- LoSAI = 15; LoSDI = 9; PGA-A =80; PGA-D =70 (Dec 22)
- LoSAI = 0 LoSDI = 12 ; PGA-A =80; PGA-D =70 (June 23)
- LoSAI = 0 LoSDI = 13 ; PGA-A =0; PGA-D =70 (Sept 23)
- LoSAI = 0 LoSDI = 13 ; PGA-A =0; PGA-D =70 (Dec 23)
- LoSAI = 2 LoSDI = 13 ; PGA-A =0; PGA-D =70 (Mar 24)
- LoSAI = 2 LoSDI = 10 ; PGA-A =0; PGA-D =50 (Jun 24)
- LoSAi = 0 LoSDI = 9 ; PGA-A = 0: PGA-D =50 (today)

# Learning Objectives

- Clinical features
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Recommendation

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]

Ann Rheum Dis: first published as

**Handling editor** Josef S Smolen

► Additional material is published online only. To view please visit the journal online (http://dx.doi.org/10.1136/

ABSTRACT

In 2012, a European initiative called Single Hub an Access point for paediatric Rheumatology in Europ (SHARE) was launched to optimise and disseminat diagnostic and management regimens in Europe fo children and young adults with rheumatic diseases.

(Zulian 2019)

| | Overarching principle |
|---|---|
| | All children with suspected localised scleroderma should be referred to a specialised paediatric rheumatology centre. |
| 1 | LoSSI, which is part of LoSCAT, is a good clinical instrument to assess activity and severity in JLS lesions and is highly recommended in clinical practice. |
| 2 | LoSDI, which is part of LoSCAT, is a good clinical instrument to assess damage in JLS and is highly recommended in clinical practice. |
| 3 | Infrared thermography can be used to assess activity of the lesions in JLS, but skin atrophy can give false-positive results. |
| 4 | A specialised US imaging, using standardised assessment and colour Doppler, may be a useful tool for assessing disease activity, extent of JLS and response to treatment. |
| 5 | All patients with JLS at diagnosis and during follow-up should be carefully evaluated with a complete joint examination, including the temporomandibular joint. |
| 6 | MRI can be considered a useful tool to assess musculoskeletal involvement in JLS, especially when the lesion crosses the joint. |
| 7 | It is highly recommended that all patients with JLS involving face and head, with or without signs of neurological involvement, have an MRI of the head at the time of the diagnosis. |
| 8 | All patients with JLS involving face and head should undergo an orthodontic and maxillofacial evaluation at diagnosis and during follow-up. |
| 9 | Ophthalmological assessment, including screening for uveitis, is recommended at diagnosis for every patient with JLS, especially in those with skin lesions on the face and scalp. |
| 10 | Ophthalmological follow-up, including screening for uveitis, should be considered for every patient with JLS, especially in those with skin lesions on the face and scalp. |

# Investgations

- Clinical assessment – specialist centre
- Photos
- Assessment scoring - LoSCAT
- Examine joints +/- MRI
- Consider skin biopsy and bloods
- Consider imaging eg MRI head
- Consider thermography
- Consider ophthalmology review
- Consider maxilofacial or orthodontic review

25% may have extra cutaneous associations…

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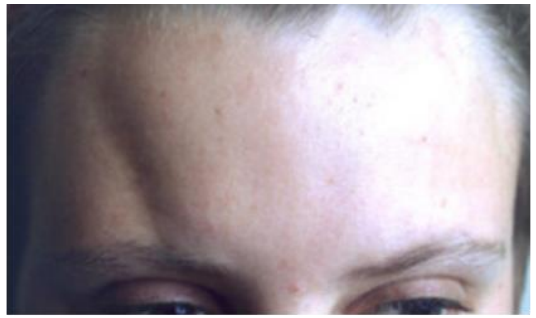
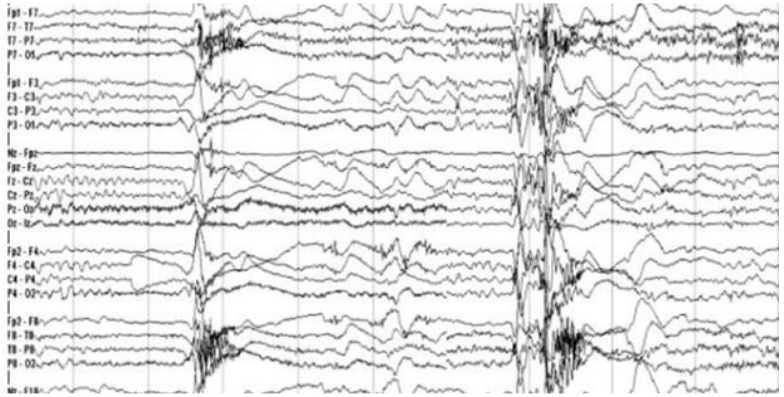
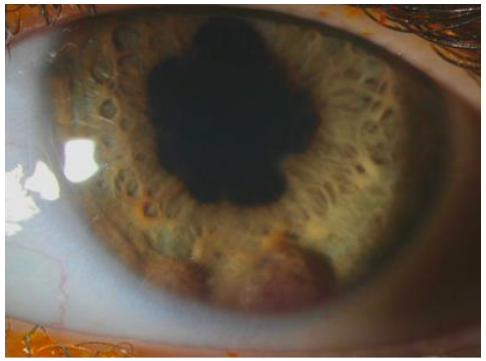


Image from Dermnetnz.org

Arthritis



Seizures



uveitis

Bloods

- Inflammatory markers – if elevated can monitor response
- Muscle enzymes – suggest muscle involvement
- Anti-nuclear antibodies (43-50%) may suggest extensive / severe disease
- Rheumatoid factor (15 – 25 %) extra-cutaneous features in LSc

(Wu 2019, Zulian 2006)

- **Deformities can occur**

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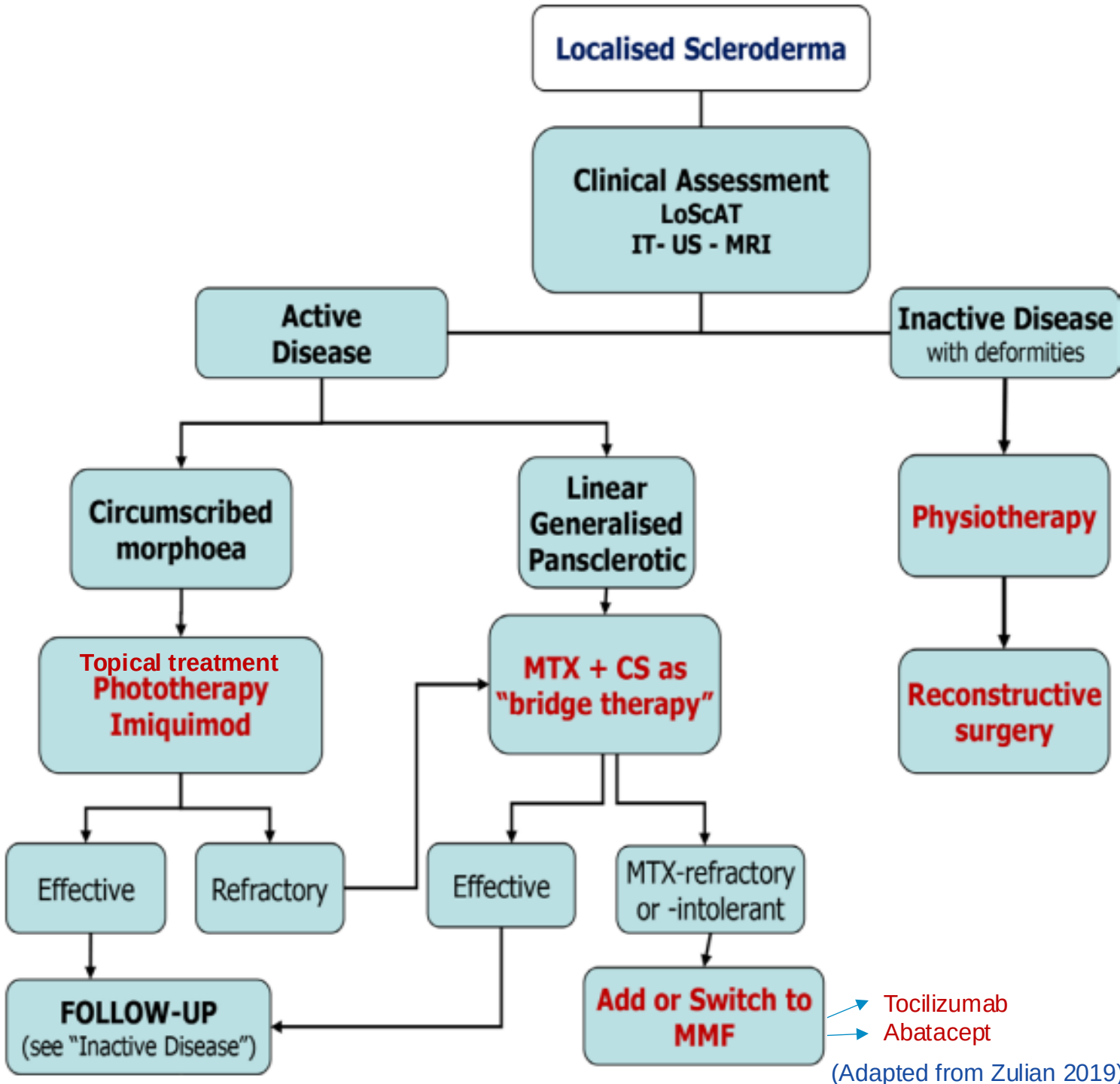
Affected foot - small, tight tendons. Affecting gait

Subcutaneous fat loss & joint contractures

# Management


Localised Scleroderma
Clinical Assessment
LoScAT
IT- US - MRI
Active Disease
Inactive Disease
with deformities
Circumscribed morphoea
Linear Generalised Pansclerotic
Physiotherapy
Topical treatment
Phototherapy
Imiquimod
MTX + CS as "bridge therapy"
Reconstructive surgery
Effective
Refractory
Effective
MTX-refractory or -intolerant
FOLLOW-UP
(see "Inactive Disease")
Add or Switch to MMF
Tocilizumab
Abatacept
(Adapted from Zulian 2019)

# Learning Objectives

- Clinical features
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- **Case examples**
- Psychosocial impact

# A localised patch

- Aged 7

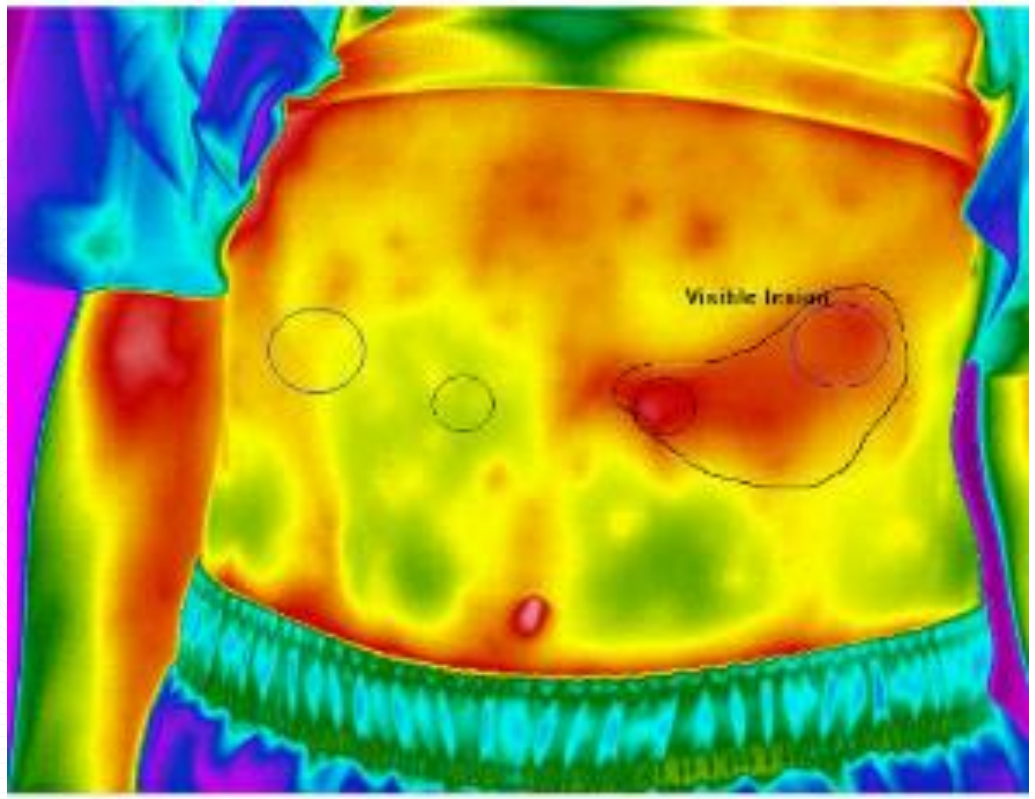
- Managed with super potent topical steroid
- Not increasing in size
- Inactive and symptomatic –topical tacrolimus

PATIENT IMAGES REMOVED

Aged 7

Aged 9

"The lesion is generally warmer than the contralateral area, the medial edge is the hottest area of the lesion. A small region drawn here measured 3.3 ° C warmer than the contralateral area . In the absence of any other explanation this does appear to indicate inflammation in this area"

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- 6 years later
- Concern over erythema
- Thermography performed
- Considering MTX
- LoSDI 6

**LoSDI**
(Localized Scleroderma Skin **Damage** Index)

| Dermal atrophy | Sub Q / Deep atrophy | Dyspigmentation (hyper or hypo) | Skin Thickness (at CENTER) |
|---|---|---|---|
| 0 = none<br>1 = shiny<br>2 = visible vessels<br>3 = cliff drop | 0 = none<br>1 = flat<br>2 = concave<br>3 = marked | 0 = none<br>1 = mild<br>2 = moderate<br>3 = marked | 0 = none<br>1 = mild<br>2 = moderate<br>3 = marked |

## En coup de sabre

3 yrs old

8-week history pale hairless patch no better with oral terbinafine

MRI head; Right frontal thinning of calvarium consistent with linear scleroderma
no brain abnormality

MTX for 2 years  stopped due to intolerance

Topical Sebco

PATIENT IMAGES REMOVED

# 18 months later

PATIENT IMAGES REMOVED

- LoSAI = 0 & LoSDI = 3 (06/22)
- LoSAI = 0 & LoSDI = 4 (09/23)
- LoSA1=0 today LosD1 = 4 today

## Extending plaque

9 yrs old

7-month history

No response to topical antifungals

FH autoimmune disease

Asthma

Hyperhidrosis

PATIENT IMAGES REMOVED

PATIENT IMAGES REMOVED

Initial IV steroid - 3 months
MTX 15 months (nausea and neutropenia)
MMF from April 2024

LoSAI 15 - 0

- LoSAI = 15; LoSDI = 9; PGA-A =80; PGA-D =70 (Dec 22)
- LoSAI = 0 LoSDI = 12 ; PGA-A =80; PGA-D =70 (June 23)
- LoSAI = 0 LoSDI = 13 ; PGA-A =0; PGA-D =70 (Sept 23)
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- LoSAi = 0 LoSDI = 9 ; PGA-A = 0: PGA-D =50 (today)

# Bilateral changes

PATIENT IMAGES REMOVED

Referred aged 10 4-year history

MRI x3 shows microhaemorrhages in brain parenchyma

Coexisting Alopecia areata

MTX for 2 years and initial IV steroid

3 months after stopping treatment - relapse
Switched to MMF for 2 years

PATIENT IMAGES REMOVED

4 years later

No activity
Minimal extension

Some new alopecia to scalp and eyelashes
No relapse after stopping MMF

# Extensive linear change

PATIENT IMAGES REMOVED

14 yr old

Noticed incidentally

Present months

Keen athlete

PATIENT IMAGES REMOVED

6 months later

20 months of MTX
Initial 5 months of IV pred

Nausea with MTX

Great anxiety about relapse

LoSAI 21 - 0
LoSDI 20 - 3

- September 2022: LoSAI 21 and LoSDI 20. PGA-A 80/100; PGA-D 80/100 (at diagnosis)
- March 2023: LoSAI 0 and LoSDI 8. PGA-A 0/100; PGA-D 50/100
- June 2023: LoSAI 0 and LoSDI 11. PGA-A 0/100; PGA-D 50/100
- September 2023: LoSAI 0 and LoSDI 10. PGA-A 0/100; PGA-D 50/100
- December 2023: LoSAI 0 and LoSDI 10. PGA-A 0/100; PGA-D 40/100
- March 2024: LoSAI 0 and LoSDI 8. PGA-A 0/100; PGA-D 30/100
- June 2024: LoSAI 0 and LoSDI 8. PGA-A 0/100; PGA-D 20/100
- Today: LoSAI 0 and LoSDI 3. PGA-A 0/100; PGA-D 20/100

# Extensive en coup de sabre

Seen aged 3 with 6 week history of increasing macule on forehead
History of congenital nystagmus

Urgent biopsy and ophthalmology review

**Admitted within days for IV steroid**

- Initial IV steroid 14 months
- MTX 18 months initially – stopped due to LFT's
  (Skin still active)
- Switched to MMF + Tocilizumab for 2 years
  with IV pred / oral pred intermittently
- Methotrexate added back in to allow reduction in steroids

- Currently in remission

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Skin biopsy scalp aged 3 – early linear scleroderma

MRI head; left frontal skull abnormality, with loss of scalp tissue and thinning of the underlying bone
.
Multiple foci calcification – progression on subsequent scans (neurology supported use of tocilizumab)

Fat transfers with plastic surgery
Considering further surgery

Camouflage clinic
Eyebrow tattooing
Ophthalmology input
Clinical psychology

PATIENT IMAGES REMOVED

Aged 7

Aged 11

# Learning Objectives

- Clinical features
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- **Psychosocial impact**

# Psychosocial Impact

## Factors

- Self-image
- Bullying
- Fear and anxiety – progression / recurrence
- Burden of treatment – needles , blood tests, medication
- Visits – school attendance

## Tools

- Health psychology
- Scores – GAD, CDLQI, PHQ
- Art therapy
- Camouflage make up clinic
- MDT team input

**PHQ-9 modified for Adolescents (PHQ-A)**

Name: _____ Clinician: _____ Date: _____

**Instructions:** How often have you been bothered by each of the following symptoms during the past **two weeks**? For each symptom put an "**X**" in the box beneath the answer that best describes how you have been feeling.

| | (0) Not at all | (1) Several days | (2) More than | (3) Nearly every |
|---|---|---|---|---|
| 1. Feeling down, depressed, irritable, or h | | | | |
| 2. Little interest or pleasure in doing thing | | | | |
| 3. Trouble falling asleep, staying asleep, ( much? | | | | |
| 4. Poor appetite, weight loss, or overeatin | | | | |
| 5. Feeling tired, or having little energy? | | | | |
| 6. Feeling bad about yourself – or feeling | | | | |

**CHILDREN'S DERMATOLOGY LIFE QUALITY INDEX**

Hospital No
Name: Diagnosis: CDLQI SCORE:
Age:
Address: Date:

**The aim of this questionnaire is to measure how much your skin problem has affected you OVER THE LAST WEEK. Please tick ✓ one box for each question.**

| | | |
|---|---|---|
| 1. | Over the last week, how **itchy**, "**scratchy**", **sore** or **painful** has your skin been? | Very much ☐<br>Quite a lot ☐<br>Only a little ☐<br>Not at all ☐ |
| 2. | Over the last week, how **embarrassed** or **self conscious**, **upset** or **sad** have you been because of your skin? | Very much ☐<br>Quite a lot ☐<br>Only a little ☐<br>Not at all ☐ |

*Hospital Anxiety and Depression (HAD) Scale*

Name: _____ Trial No: ☐☐☐☐
Hospital: _____ Date of Completion: ☐☐ ☐☐ ☐☐

Doctors are aware that emotions play an important part in most illnes
will be able to help you more.
This questionnaire is designed to help your doctor to know how you fe
opposite the reply which comes closest to how you have been feelir
Don't take too long over your replies: your immediate reaction to eac
thought-out response.

*Tick only one box in eac*

I feel tense or 'wound up': I feel
Most of the time ☐
A lot of the time

Newcastle Hospitals NHS Foundation Trust

Children and young people's health psychology - Newcastle Hospitals NHS Foundation Trust

SCLERODERMA & RAYNAUD'S UK

CHANGING FACES
Providing support and promoting respect for everyone with a visible difference

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# Morphoea in children, experience from a joint dermatology rheumatology clinic

Dr Charlotte Goodhead

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