

Modified Rodnan skin score

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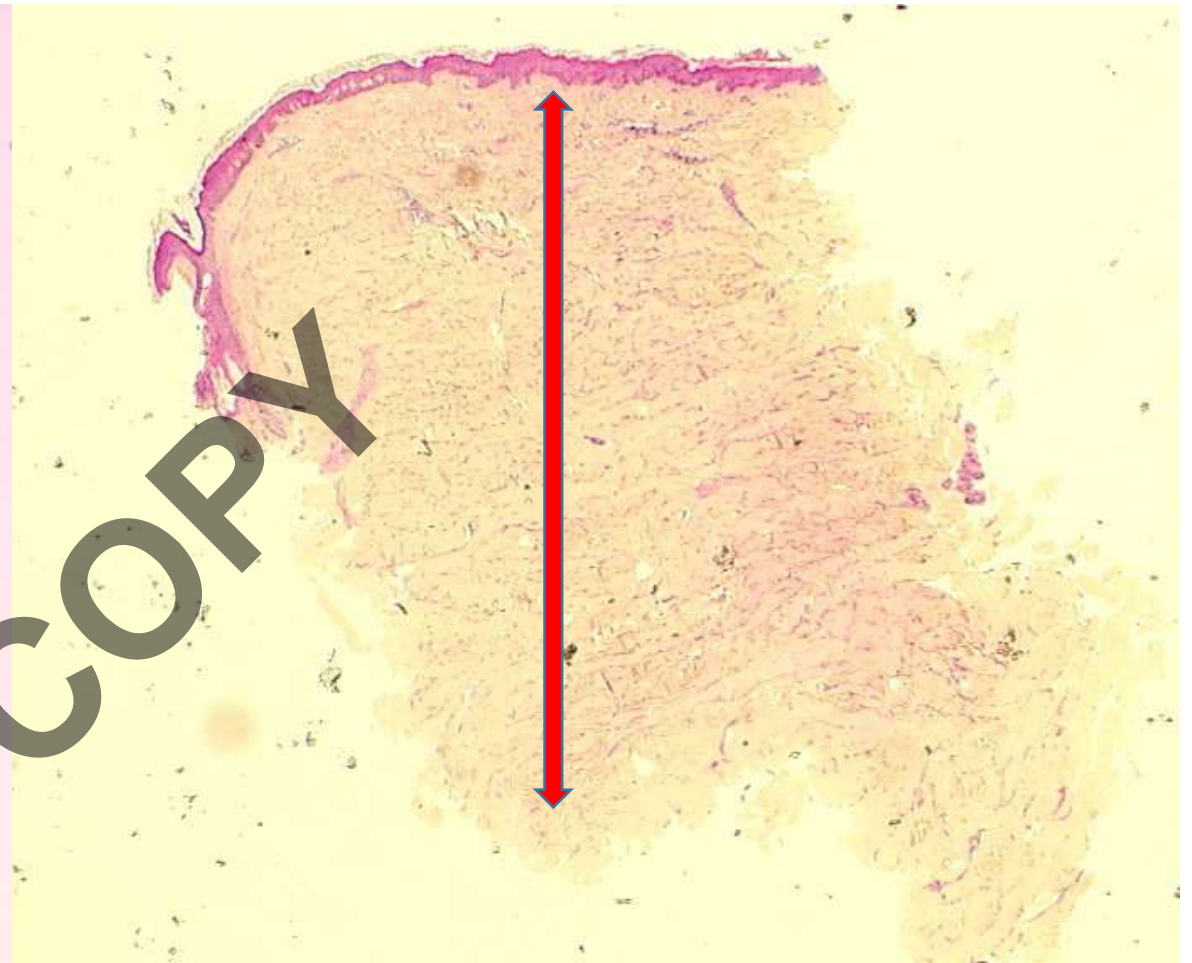
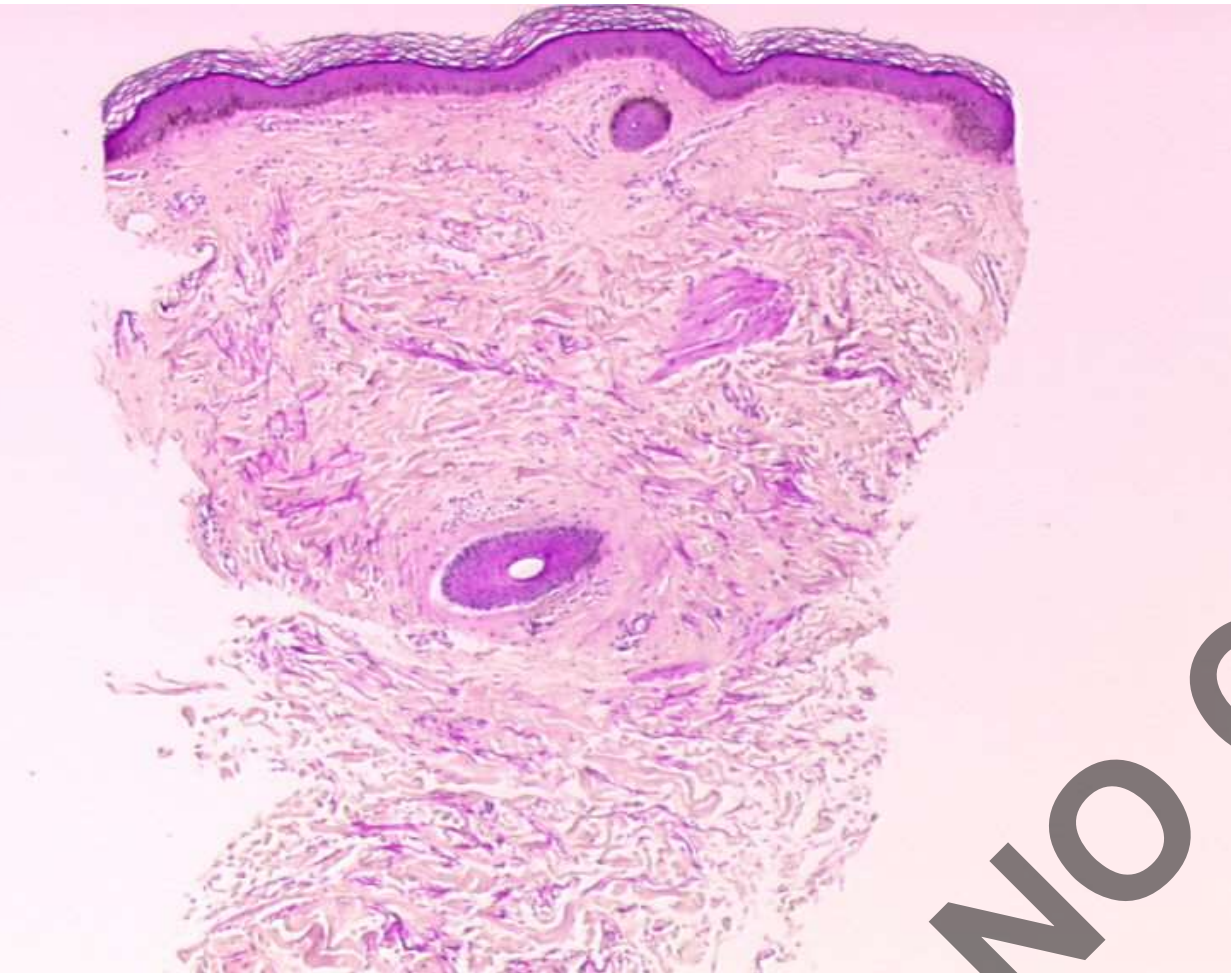
ACR/EULAR 2013 CRITERIA

Table 1 The American College of Rheumatology/European League Against Rheumatism criteria for the classification of systemic sclerosis*

Item	Sub-item(s)	Weight/ score†
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (<i>sufficient criterion</i>)	–	9
Skin thickening of the fingers (<i>only count the higher score</i>)	Puffy fingers	2
	Sclerodactyly of the fingers (distal to the metacarpophalangeal joints but proximal to the proximal interphalangeal joints)	4
Fingertip lesions (<i>only count the higher score</i>)	Digital tip ulcers	2
	Fingertip pitting scars	3
Telangiectasia	–	2
Abnormal nailfold capillaries	–	2
Pulmonary arterial hypertension and/or interstitial lung disease (<i>maximum score is 2</i>)	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) (<i>maximum score is 3</i>)	Anticentromere	3
	Anti–topoisomerase I	
	Anti–RNA polymerase III	

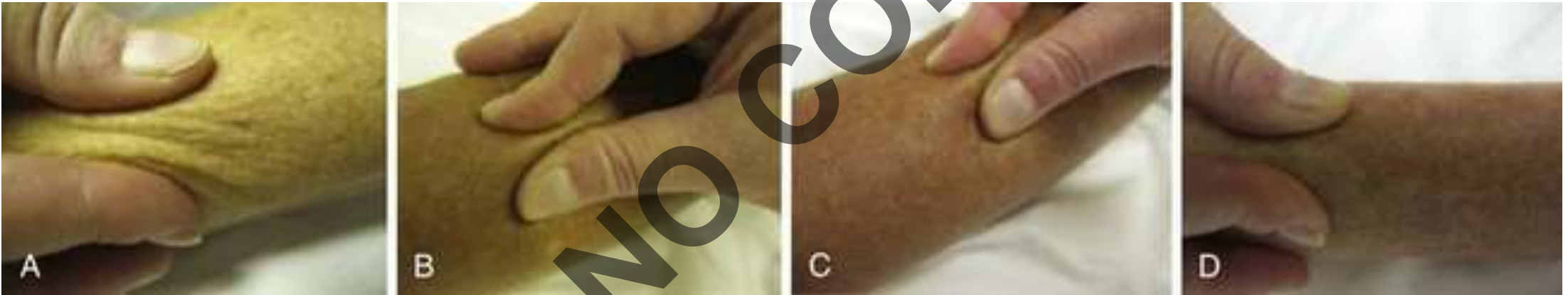
*These criteria are applicable to any patient considered for inclusion in a systemic sclerosis study. The criteria are not applicable to patients with skin thickening sparing the fingers or to patients who have a scleroderma-like disorder that better explains their manifestations (eg, nephrogenic sclerosing fibrosis, generalised 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 systemic sclerosis. SSc, systemic sclerosis.



- Normal epidermis
- Thickened dermis with disorganised collagen
 - Decrease in adnexal structures
 - Reduced surrounding adipose tissue
 - Perivascular lymphocytic infiltrate
 - In red, the dermal thickness

Measurement of skin thickness



The OMERACT Filter for Outcome Measure Validation

Face Validity : does it make sense?

Content Validity : is it comprehensive?

Construct validity : agree with other measures or with a “gold standard”? – ability to discriminate subsets?

Accuracy - criterion validity

Discrimination. Does the measure discriminate between situations of interest?

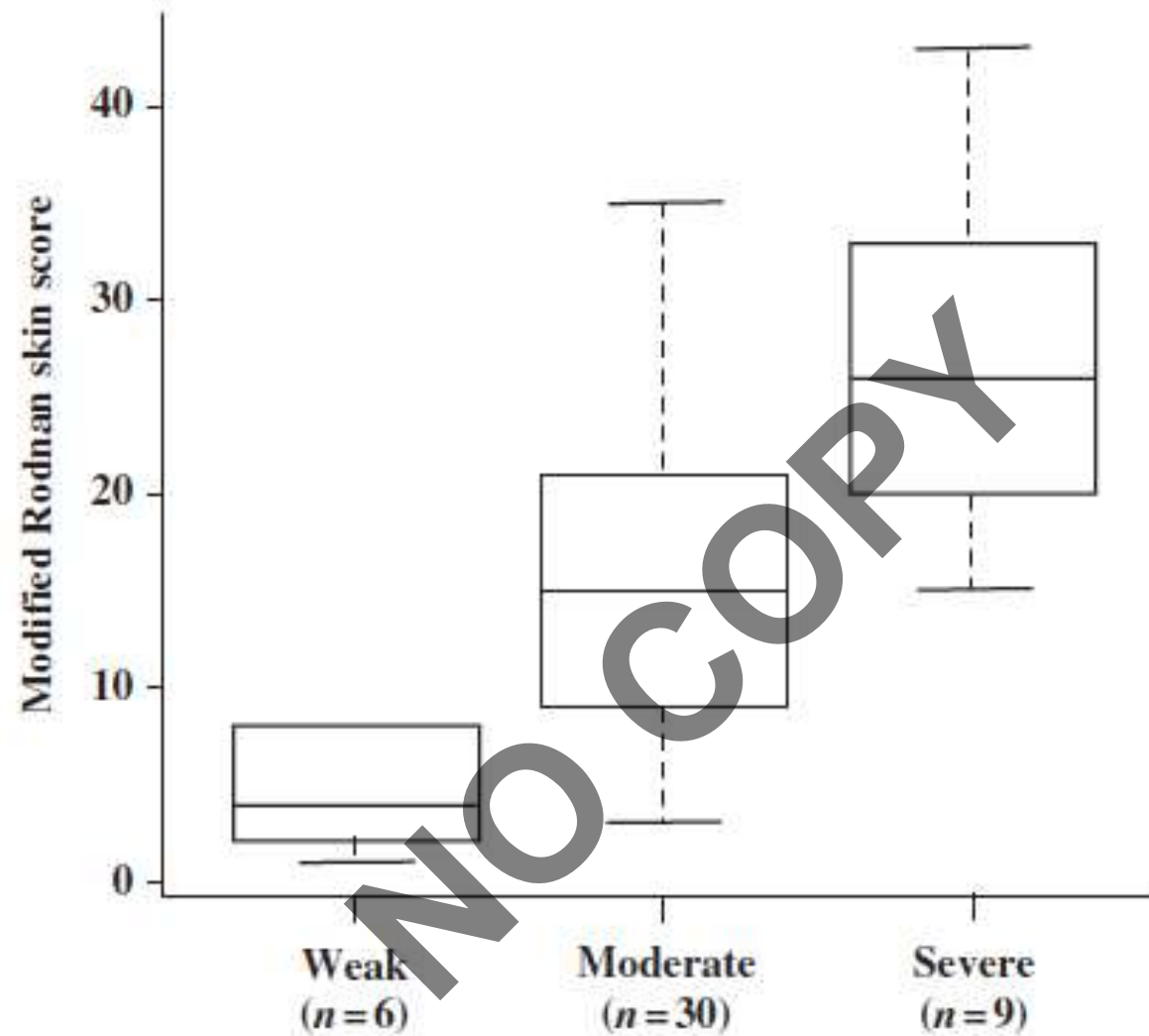
Reproducibility

Sensitivity to change

Feasibility: Can the measure be applied easily, given constraints of time, money, and interpretability?

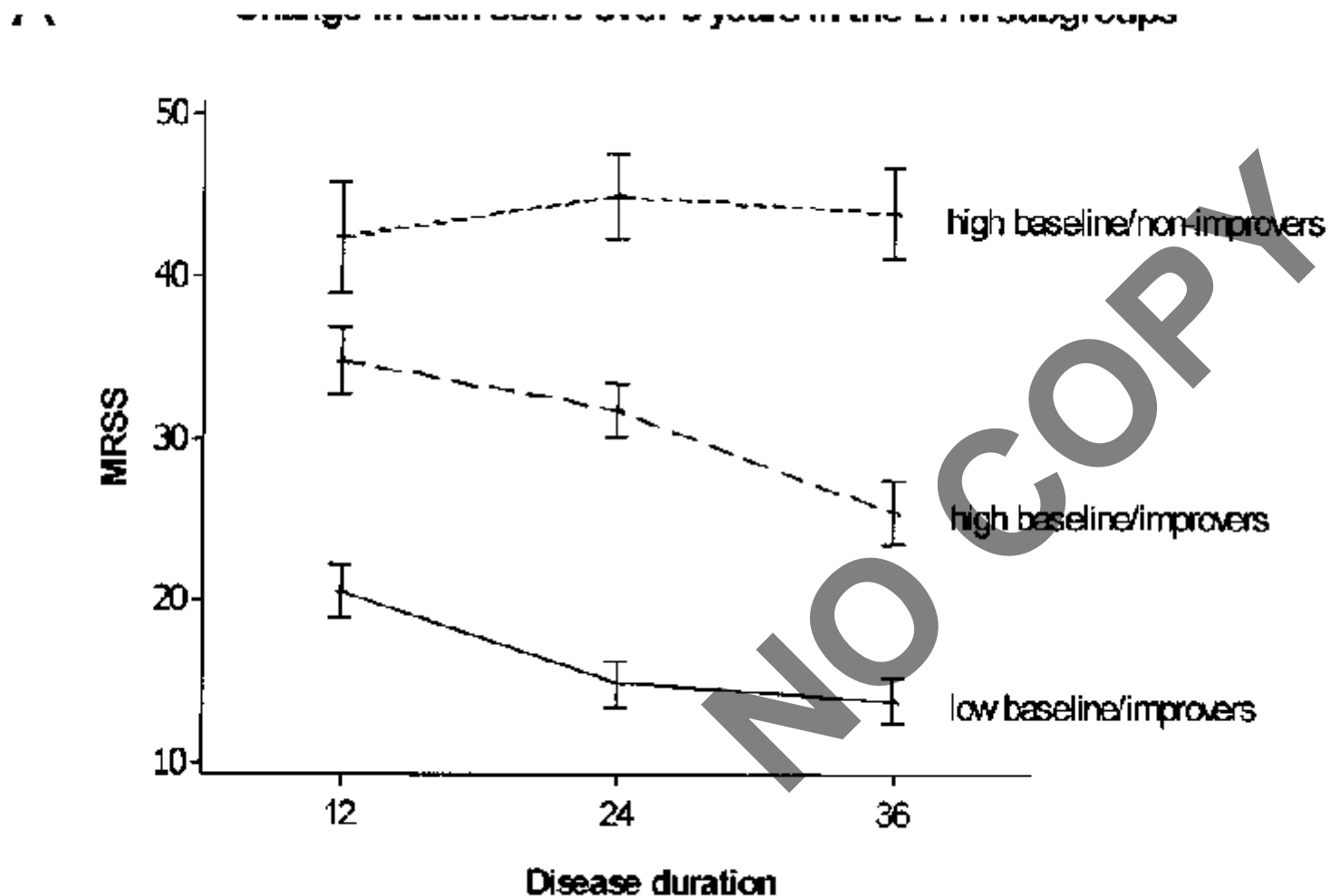
mRSS : a valid outcome measure

- **Accuracy:** Inter-observer variability 5 units (20-25%)
- **Reproducibility:** Intra-observer variability 3 units (10-15%)
- **Accessibility:** Clinical examination technique
- **Construct validity:** good correlation with skin biopsies (weight and histology)
- **Sensitivity to Change:** Characteristic



Correlation between 3 histological sub-groups and the mRSS

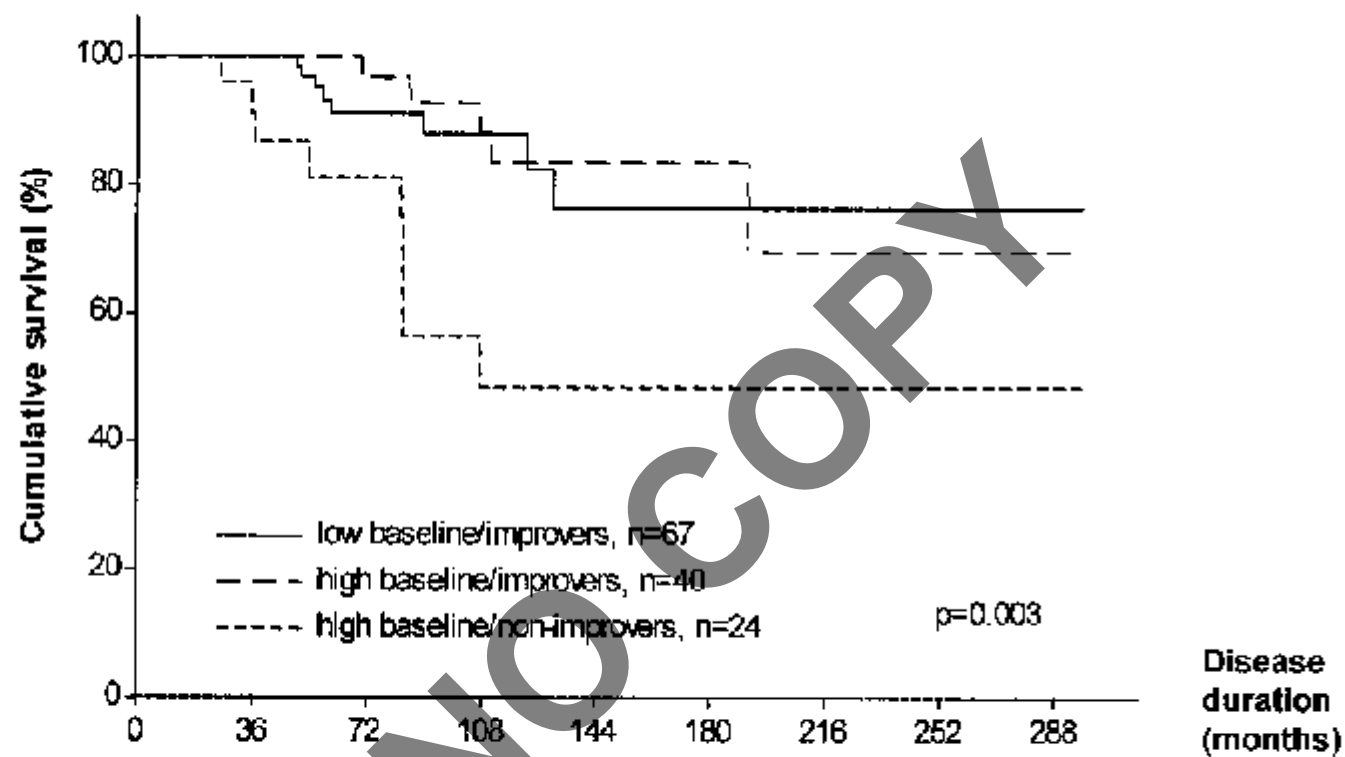
SSc subsets according to cutaneous changes



SSc subsets according to cutaneous changes

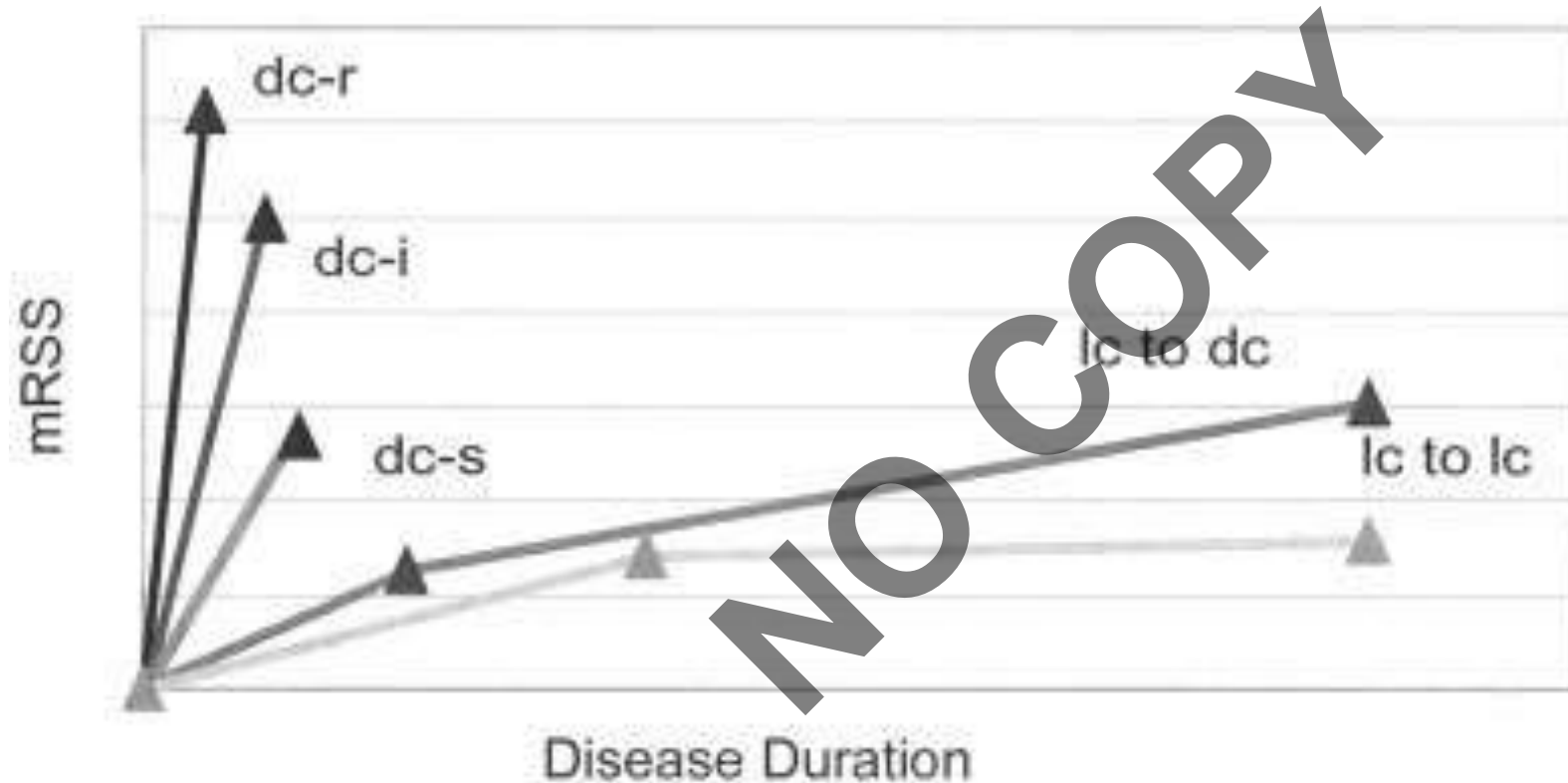
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Survival in the LTM subgroups

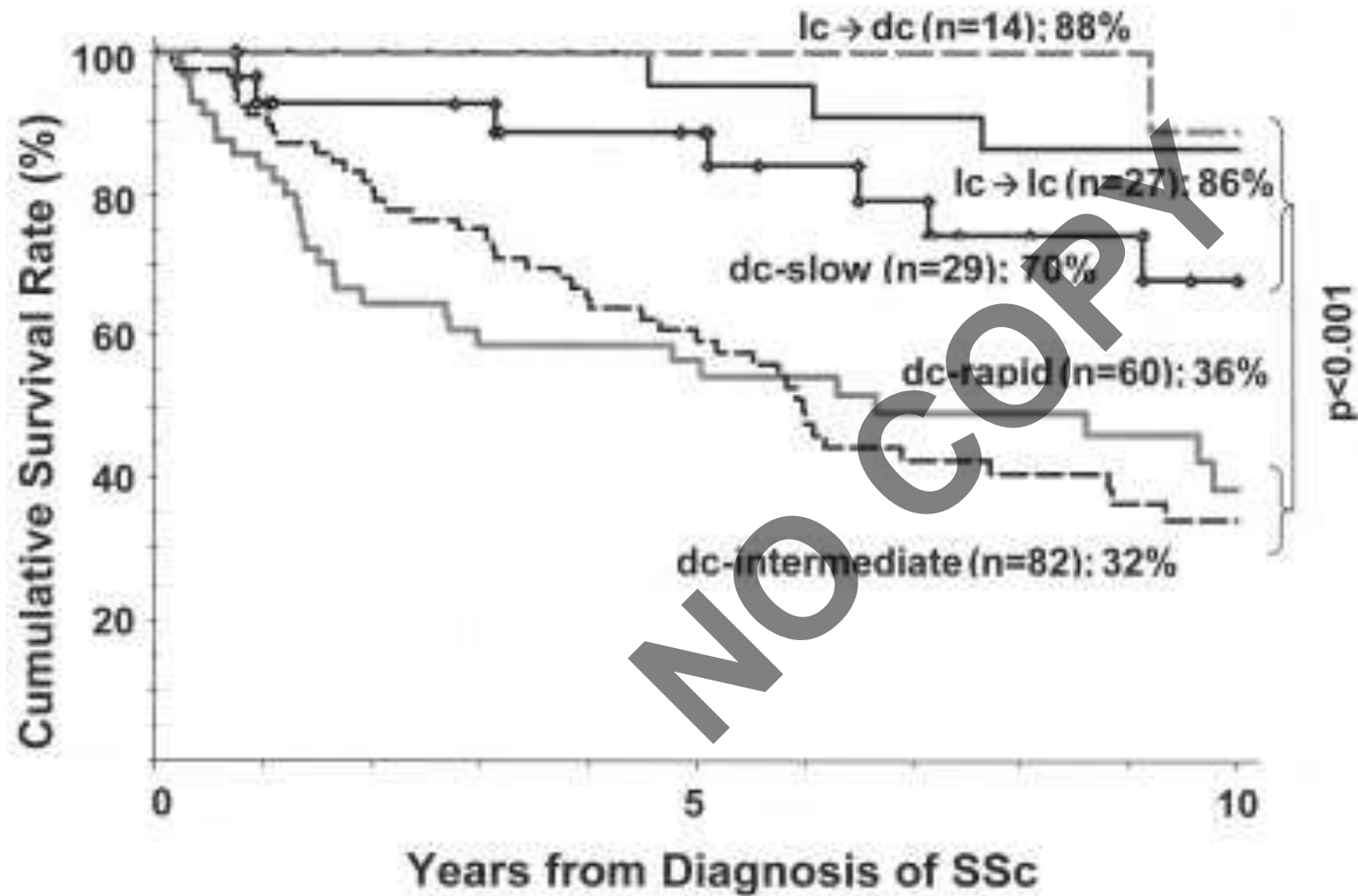


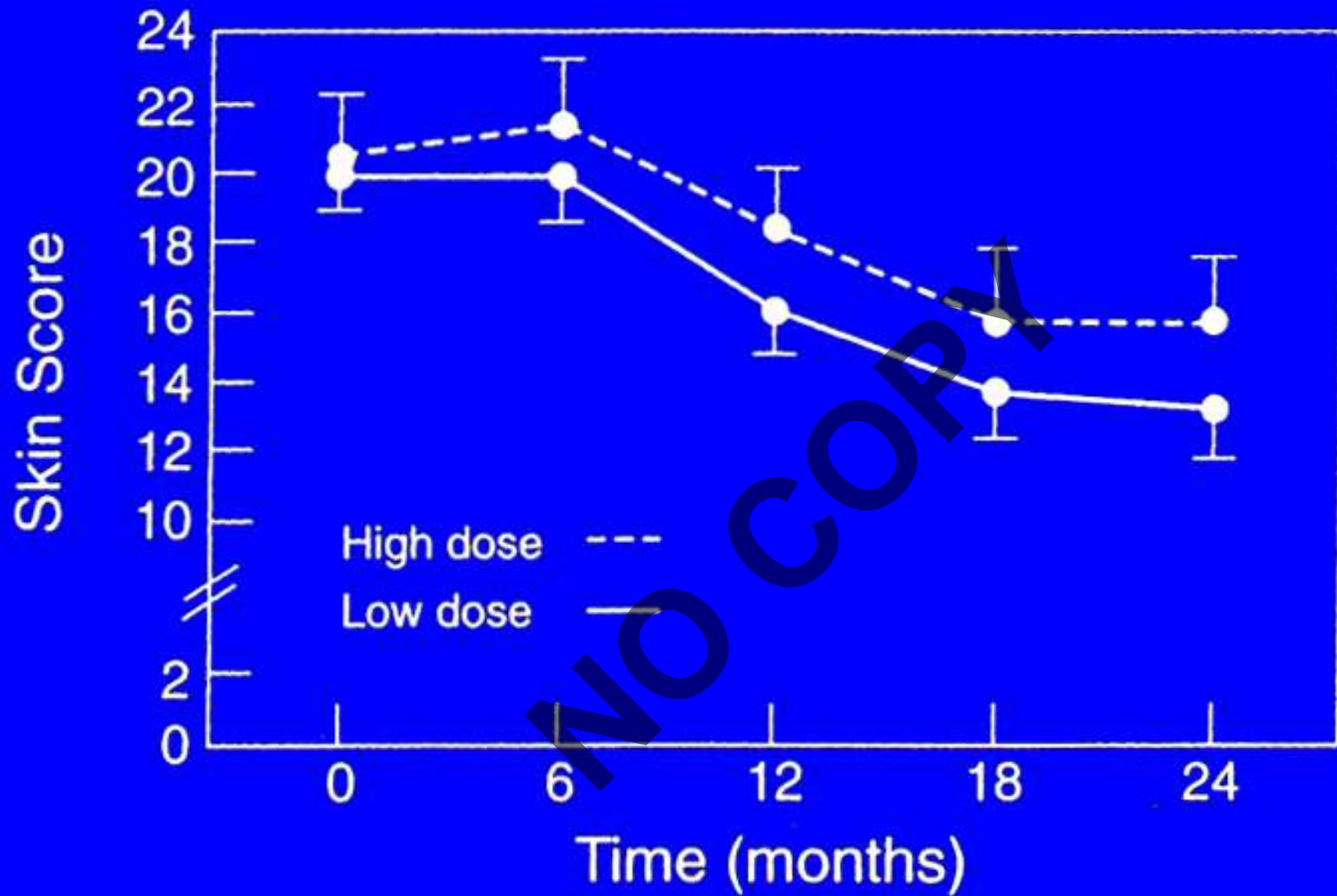
low baseline/improvers	67	67	39	17	9	5	3	2	1	Numbers at risk
high baseline/improvers	40	39	30	20	10	7	3	0	0	
high baseline/non-improvers	24	21	13	7	4	3	1	0	0	

SSc subsets according to cutaneous changes



SSc subsets according to cutaneous changes

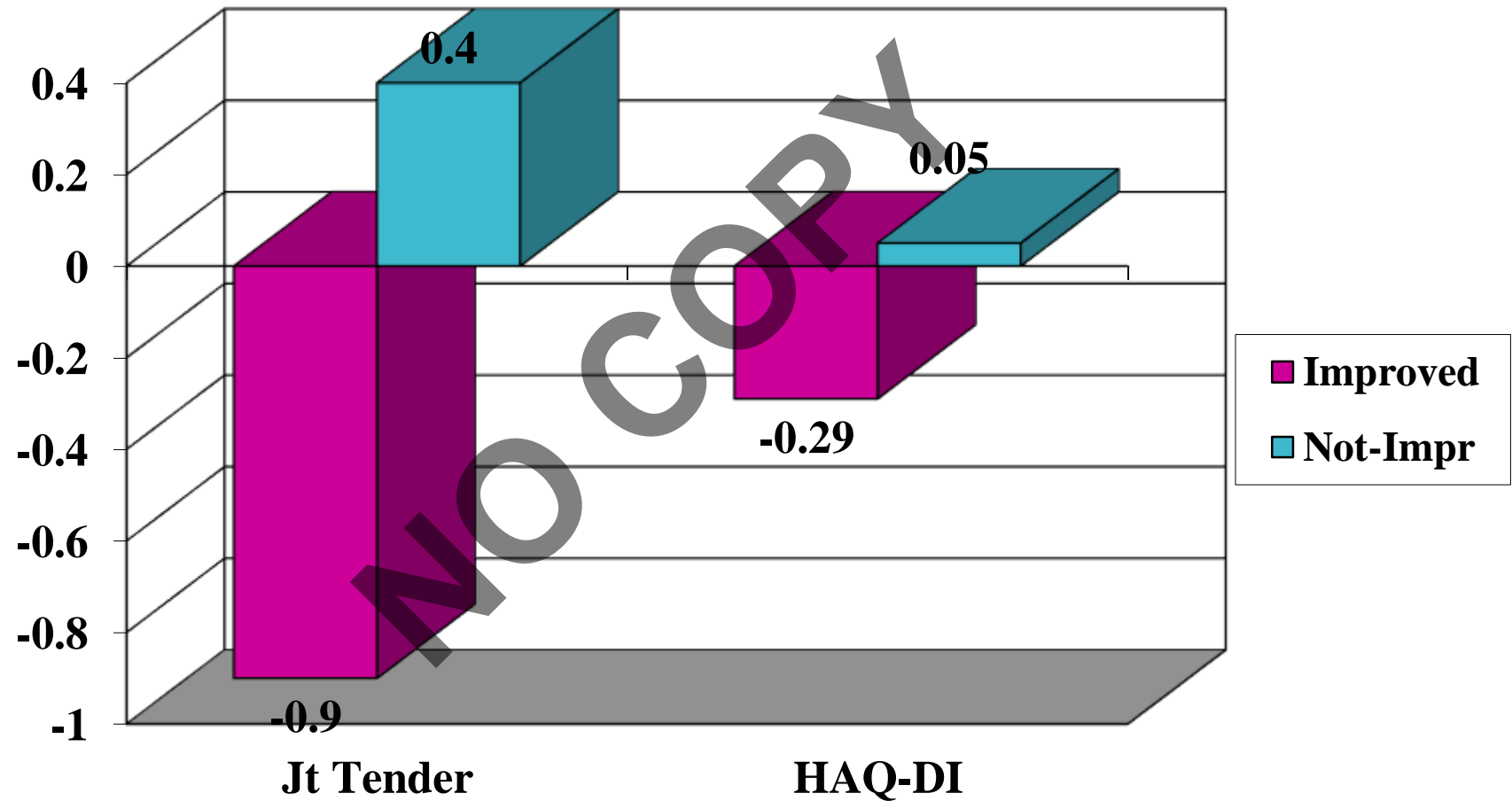




Prediction of renal crisis in D-Pen trial (n=18/133)

Variable	OR	95% CI	p-value
Skin Score >20	10.00	2.213-45.907	0.003
Large Joint Contracture	16.12	2.075-25.254	0.008
Prednisone Use	3.63	1.304-10.051	0.014
Heart involvement	2.93	1.010-8.482	0.048

mRSS and disability: D-Pen study



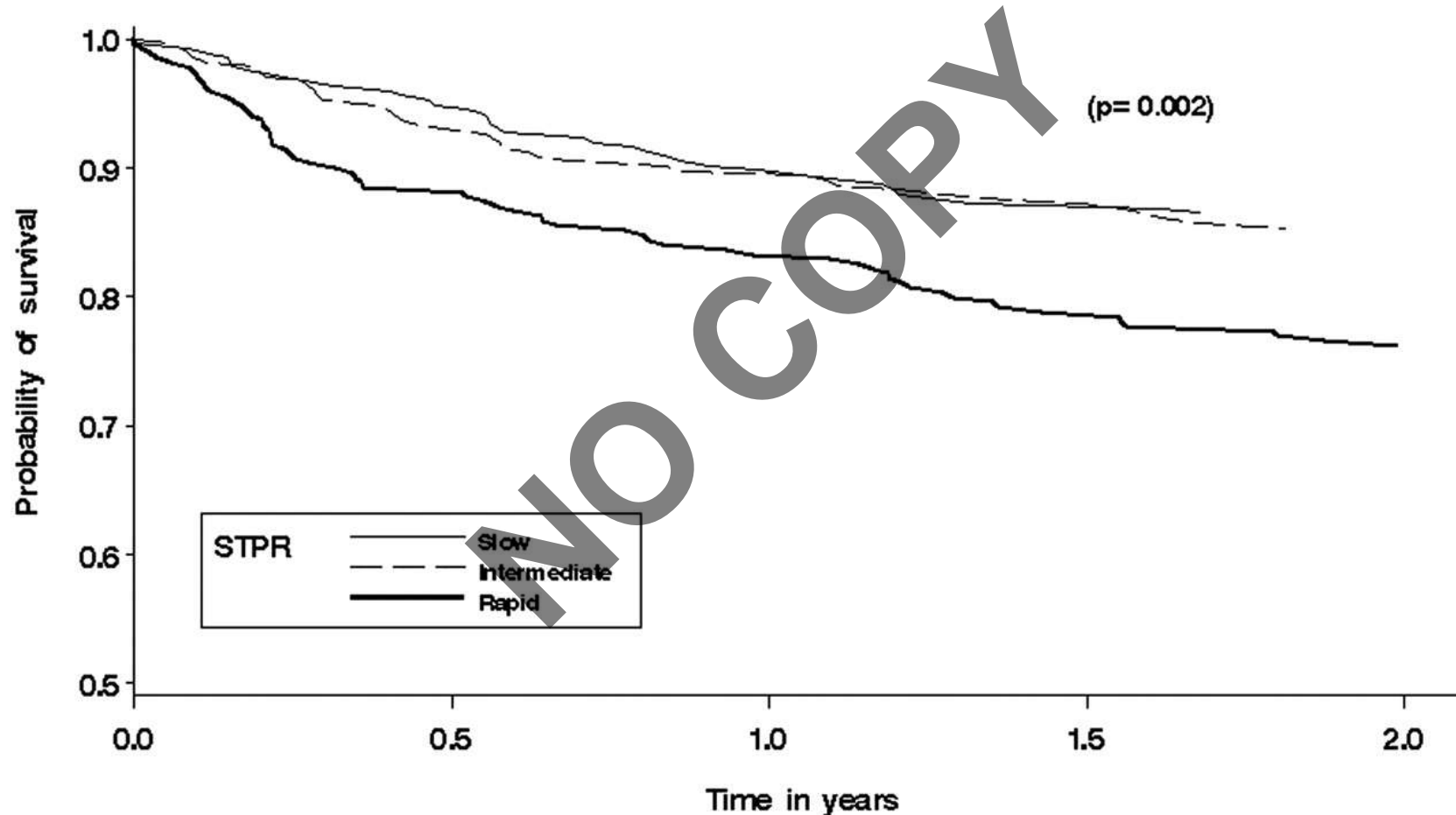
mRSS : a valid outcome measure

- Increase in mRSS predicts worsening of SSc
- Stability in mRSS predicts reduced new internal involvement
- Improvement in mRSS predicts improved survival
- High scores (>20) predict renal crisis
- High scores (>20) predict mortality
- Improved mRSS correlates with joint involvement, hand function, QOL (HAQ-DI)

*Clements PJ, et al. Arthritis Rheum. 1990 and 2000.
Steen and Medsger. Arthritis Rheum 2001.*

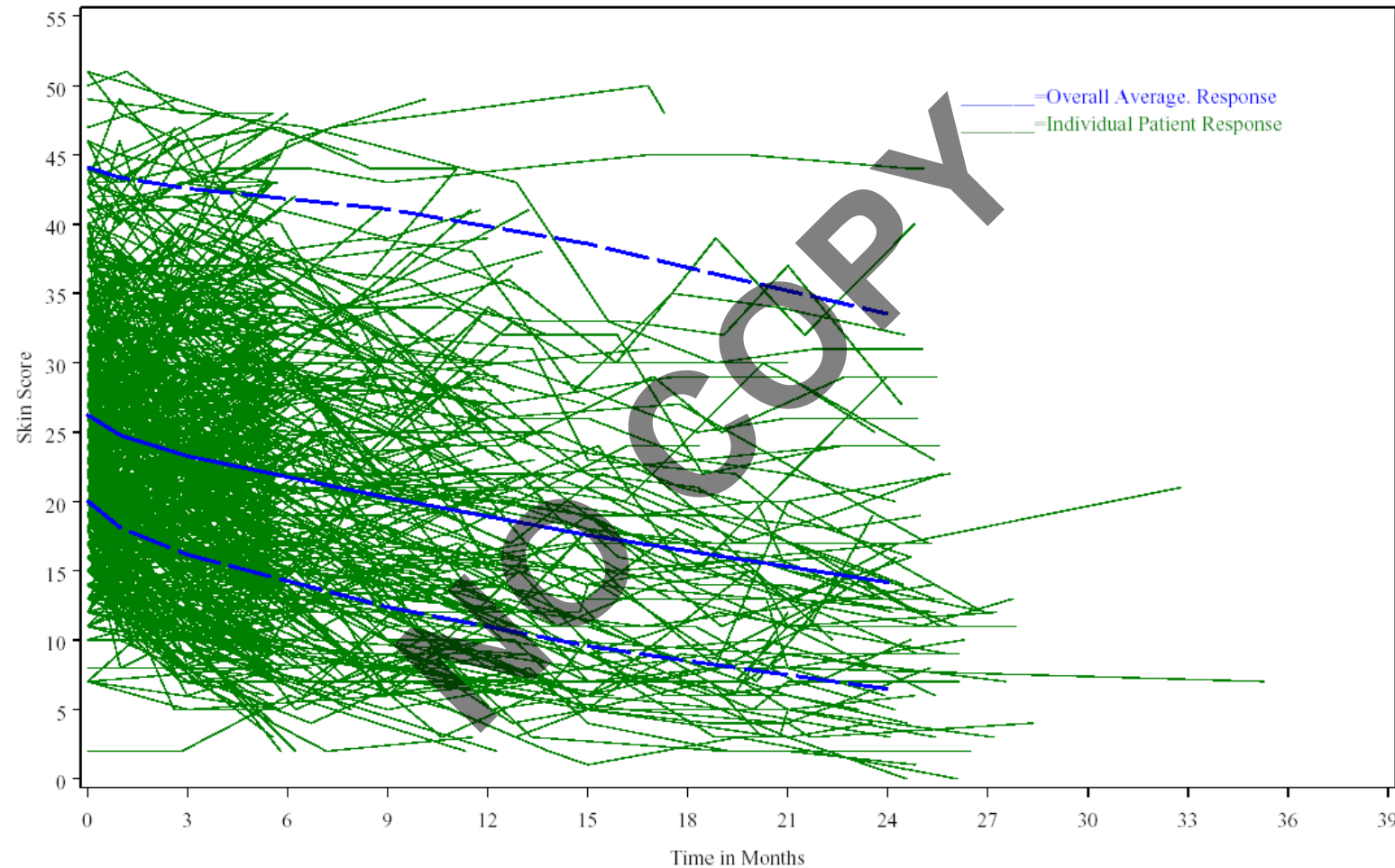
Skin thickness progression rate: a predictor of mortality and early internal organ involvement in DcSSc

STPR = mRSS at the 1st visit divided by the duration of skin thickening (in years)



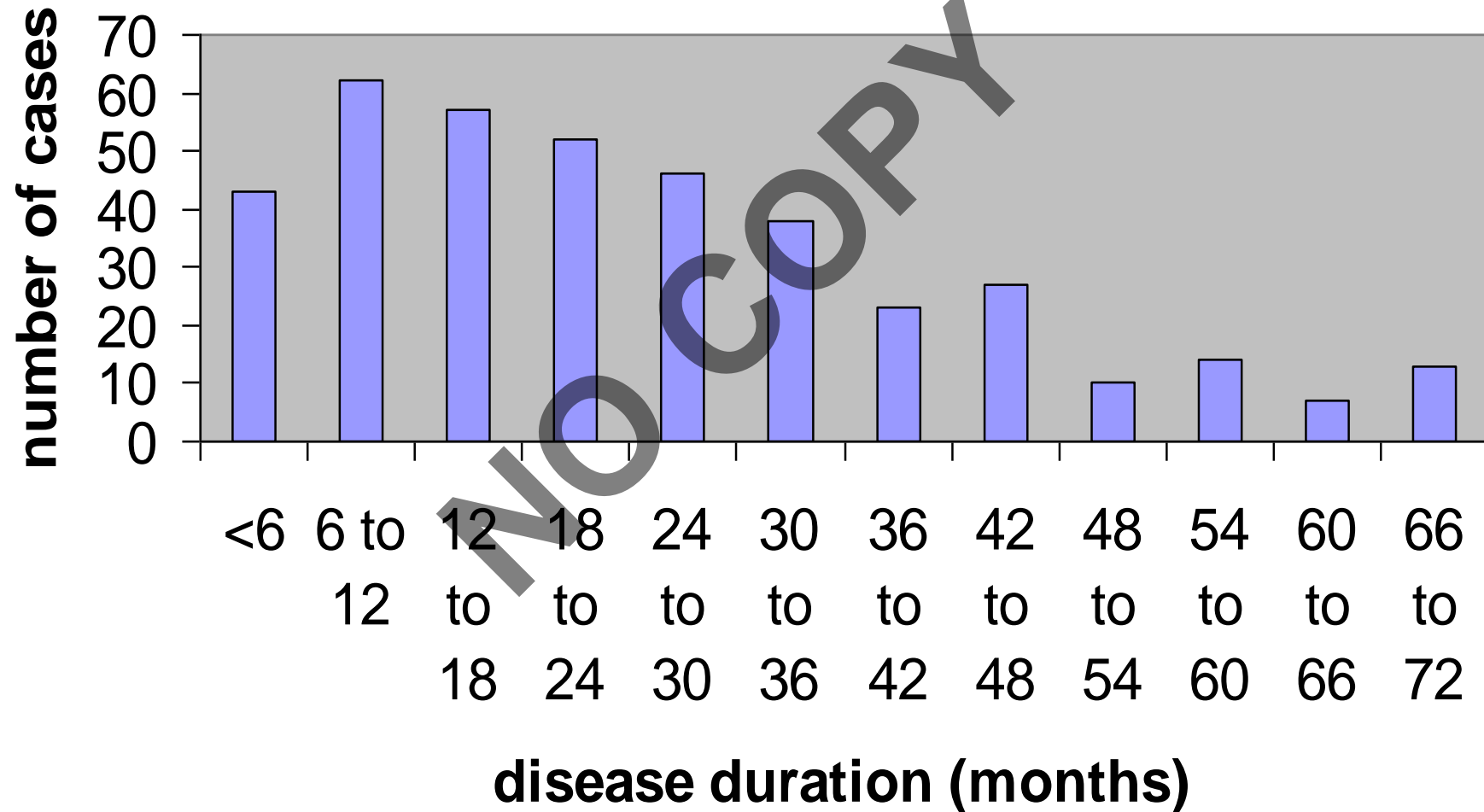
Plot of Individual Patient mRSS over Time

FIGURE 2.1-2
PLOT OF SKIN SCORE OVER TIME, ALL PATIENTS, WITH OVERALL AVERAGE RESPONSE

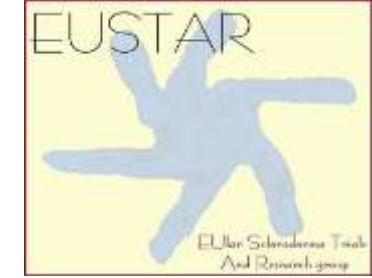


Note: The average line is produced by calculating the average value at each timepoint, and smoothing this curve using a Lowess Function

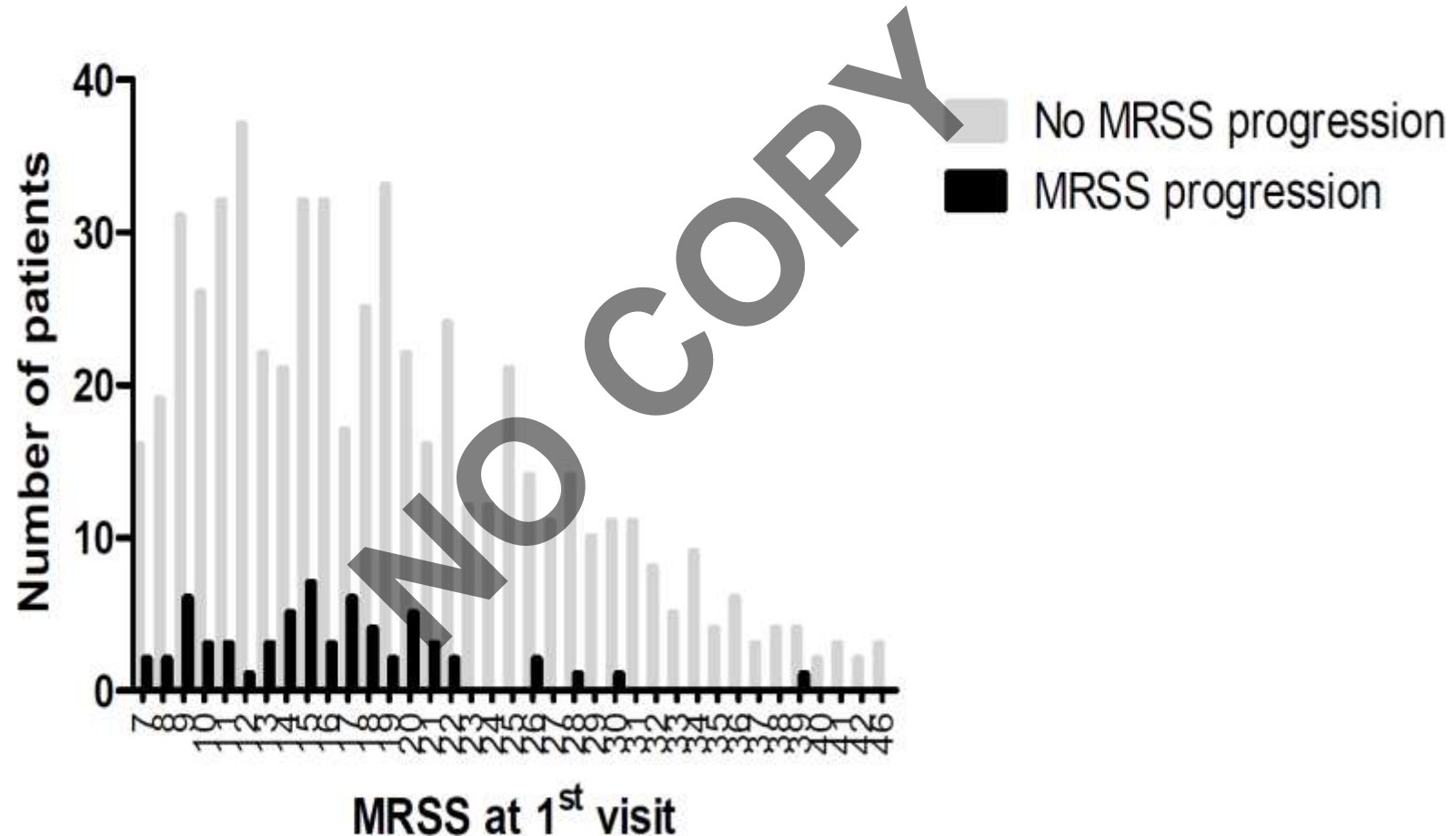
Duration of disease in dcSSc at peak skin score



Cohort enrichment: skin



637 dcSSc patients with longitudinal mRSS data



mRSS as an outcome measure

- Good performance metrics and validated in early DcSSc
- Surrogate for internal organs and it can predict outcomes
- But not linear regarding the natural history and some patient dependance (distinct subsets and role of auto-antibodies)
- High inter-individual variability for scoring
- Various targeted population according to the goal of the trials: regression versus prevention of progression