Consensus Conference

“NOMENCLATURE AND CLASSIFICATION CRITERIA FOR LOCALIZED SCLERODERMA”

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1. Summary of the Project

Background

Juvenile Scleroderma Syndromes are rare conditions that result in various degree of skin and internal organ fibrosis in children and adolescents below the age of 16 years.

Following a consensus-based methodology, we have recently defined the Classification Criteria for Systemic Sclerosis in children and these criteria have been endorsed by both EULAR and ACR (Arthritis Rheum 2007;57:202-213).

Various classifications of Morphoea (Localized Scleroderma) have been proposed, each one reflecting various aspects of the disease and the dermatologic or rheumatologic point of view. None of these have been validated in adults or in children. This issue should be addressed before multicenter epidemiologic and therapeutic studies can be planned in JLS.

Aim of the present project is to develop criteria for classification of patients with Localized Scleroderma on the basis of their clinical features, laboratory parameters and, if necessary, histological characteristics and following the same consensus-based methodology.

Phase 1 COLLECTION OF DATA (January 2002-December 2003)

A large, multicenter, multinational study, was conducted by collecting information on demographics, clinical and laboratory features. 727 patients with JLS from 70 centres, from Europe, North and South America, Asia and Africa, were enrolled into the study. F:M ratio was 2.4:1. The mean age at onset was 7.3 years and the disease duration at diagnosis was 18 months. Linear scleroderma (LS) was the most frequent subtype (66%), followed by plaque morphoea (PM) (25%), generalized morphoea (GM) (7%) and deep morphoea (DM) (2%). As many as 16% of patients had a “mixed subtype” in which linear and plaque-like lesions coexisted. ANA was positive in 43 % of the patients, Scl-70 in 3% and anticentromere in 2%.

Phase 2 PRE-CONSENSUS PHASE (January – May 2004)

On the basis of data collected in phase 1 and of the existing classifications, a Delphi questionnaires were developed and mailed to 19 investigators (Dermatologists (5), Pediatricians (10) and Rheumatologists(4)) selected on the basis of their clinical experience with JLS and their contribution to the previous phase of the study.

Aims of this phase were:
1) To discuss and propose changes on the **existing DEFINITIONS** of the various clinical subtypes of Localised Scleroderma as available in the literature

2) To discuss and propose changes on the **existing CLASSIFICATIONS** in order to establish the PROs and CONs of each one of them

3) To discuss and propose changes on a proposal for a **NEW CLASSIFICATION** ("Proposal 2004") that should be discussed in a new final Consensus Conference in 2007.

**Phase 3 CONSENSUS CONFERENCE**

The Consensus Conference will convene in Padua (possibly on October 4-6, 2007) to discuss the proposal for a new classification of JLS.

The participants will be the same who participated to the pre-consensus conference phase. They will be Pediatric Rheumatologists, Adult Rheumatologists and Dermatologists

Objectives to be obtained will be:

1. To discuss the nomenclature of specific subgroup of patients and the description of each subgroup on the basis of the proposed classification as resulted from the pre-consensus conference phase.

2. To classify real patients as having or not having JLS on the basis of the appearance of their sclerodermatous skin lesion and on the classification criteria previously established

The methodology to be used will be the Nominal Group Technique (NGT). The participants, in a plenary section, will discuss on the various issues merged from the pre-consensus conference phase. In particular, they will try to reach a consensus on a new and more comprehensive classification. Soon after this classification will be validated by assessing around 100 real patients whose pictures with a brief clinical description will be presented in a portable computer available for each participant. The participants will be asked to give their judgment on each patient by assigning a code corresponding to a particular subgroup. The specific cases in which a 80% agreement on classification won’t be reached, will be discussed again in a second plenary section. The few cases in which the Consensus won’t be reached after this section, will be gathered into a special subset named “Miscellanea”. 
### Classification of Juvenile Localized Scleroderma (Proposal “2004”)

<table>
<thead>
<tr>
<th>MAIN GROUP</th>
<th>SUBTYPE</th>
<th>DESCRIPTION</th>
</tr>
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<tbody>
<tr>
<td><strong>1) Linear Scleroderma</strong></td>
<td>a. Affecting trunk/limbs</td>
<td>Linear induration involving dermis, subcutaneous tissue and, sometimes, muscle and underlying bone and affecting the limbs and/or the trunk.</td>
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<tr>
<td></td>
<td>b. Affecting head</td>
<td>En coup de sabre (ECDs). Linear induration that affects the face and/or the scalp and sometimes involves muscle and underlying bone.</td>
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<tr>
<td><strong>2) Parry Romberg or Progressive Hemifacial Atrophy (PHA)</strong></td>
<td></td>
<td>It results in loss of tissue on one side of the face and may involve the dermis, subcutaneous tissue, muscle and bone. The skin is mobile.</td>
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<tr>
<td></td>
<td>a. Superficial</td>
<td>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.</td>
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<tr>
<td></td>
<td>b. Deep (morphoea profunda)</td>
<td>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.</td>
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<tr>
<td><strong>3) Circumscribed Morphea</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>a. Superficial</td>
<td></td>
</tr>
<tr>
<td></td>
<td>b. Deep</td>
<td></td>
</tr>
<tr>
<td><strong>4) Generalized Morphea</strong></td>
<td></td>
<td>Induration of the skin starting as individual plaques (4 or more and larger than 3 cm) that become confluent and involve at least 2 anatomic sites.</td>
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<td><strong>5) Panclerotic Morphea</strong></td>
<td></td>
<td>Circumferential involvement of limb(s) affecting the skin, subcutaneous tissue, muscle and bone. The lesion may also involve other areas of the body.</td>
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<tr>
<td><strong>6) Mixed Juvenile Localised Scleroderma</strong></td>
<td>a. Linear-Circumscribed morphoea</td>
<td>Combination of linear subtype and circumscribed (plaque) morphea.</td>
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<td></td>
<td>b. Linear ECD and Parry Romberg</td>
<td>Combination of linear ECD and Parry Romberg.</td>
</tr>
<tr>
<td><strong>7) Associated Conditions</strong></td>
<td>a. Lichen Sclerosus et Atrophicus (LSA)</td>
<td>Combination of one of the previous subtypes with Lichen Sclerosus et Atrophicus.</td>
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<td></td>
<td>b) Atrophoderma of Pasini and Pierini (APP)</td>
<td>Combination of one of the previous subtypes with Atrophoderma of Pasini-Pierini.</td>
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