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# The hand in systemic diseases other than rheumatoid arthritis

## *La main dans les maladies systémiques autres que la polyarthrite rhumatoïde*

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**Abstract**

This review outlines the skin, vascular and musculoskeletal symptoms affecting the hand during systemic inflammatory diseases other than rheumatoid arthritis. Skin lesions are diagnosed clinically and their symptomatology is documented through an extensive series of photographs. These conditions may require specific care before a surgical procedure can be performed. Vascular lesions are also diagnosed clinically and their symptomatology is described in detail. It is important to recognize that acrocyanosis is always benign. The surgeon should be able to distinguish between primary, but benign Raynaud's disease and secondary Raynaud's syndrome, which has a high risk of finger necrosis. Current preventative and curative treatments for finger necrosis are described. The clinical, radiological, progressive and therapeutic features of musculoskeletal lesions are reviewed, namely those associated with psoriatic arthritis, systemic sclerosis and lupus.


*Keywords:* Wrist; Hand; Scleroderma; Lupus; Dermatomyositis; Polymyositis; Raynaud; Ischemia; Necrosis; Arthritis; Surgery

**Résumé**

Cet article dresse un tableau des manifestations cutanées, vasculaires et ostéoarticulaires des maladies inflammatoires autres que la polyarthrite rhumatoïde. Les lésions cutanées sont diagnostiquées cliniquement et leur sémiologie est rappelée ; elles peuvent nécessiter un traitement spécifique avant une intervention chirurgicale. Les lésions vasculaires sont elles aussi de diagnostic clinique et leur sémiologie est rappelée ; il importe de connaître le caractère toujours bénin de l'acrocyanose et de savoir distinguer des syndromes de Raynaud primitifs et bénins, d'une part, des syndromes de Raynaud secondaires et à fort potentiel de nécrose digitale ; le traitement actuel, préventif et curatif, des nécroses digitales est rappelé. Les lésions ostéoarticulaires présentent des caractéristiques cliniques, radiologiques, évolutives et thérapeutiques qui sont rappelées, en particulier celles du rhumatisme psoriasique, de la sclérodermie et du lupus. L'article est enrichi d'une abondante iconographie.


*Mots clés :* Poignet ; Main ; Sclérodermie ; Lupus ; Psoriasis ; Dermatomyosite ; Polymyosite ; Raynaud ; Ischémie ; Nécrose ; Arthrite ; Chirurgie

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## 1. Introduction

Systemic inflammatory diseases are fairly rare. They have a varied clinical expression and typically affect multiple tissues and organs. There are three progressive stages:

- acute: vascularitis;
- chronic: systemic sclerosis and Gougerot-Sjögren syndrome;
- subacute with attacks: systemic lupus erythematosus (SLE), dermatomyositis, polymyositis.

Although the hand surgeon may be familiar with joint and tendon aspects of rheumatoid arthritis (RA), he may be perplexed by a hand condition occurring in the context of rarer systemic diseases that he knows less about, and by their extra-articular signs, which are nevertheless typical and have the potential to cause post-operative complications.

The goal of this review is to paint a non-exhaustive picture of hand involvement in systemic diseases other than RA, by presenting their tissue-specific symptoms and providing extensive imagery.

The pathophysiology and background therapy of these conditions will not be discussed here, as they differ from one disease to the other. The surgeon must remember that biological therapies have to be discontinued before surgery (except in emergency cases) for a period of time equal to 4–5× the product's half-life, as in RA [1] (Table 1).

The following topics will be reviewed:

- skin symptoms;
- vascular symptoms;
- musculoskeletal symptoms: hand surgeons will be more familiar with these because they can mimic RA lesions. However, their specific features led us to describe the clinical symptoms, imaging modalities and care of the most common conditions in greater detail, namely psoriatic arthritis, lupus-related arthritis and systemic sclerosis.

## 2. Skin symptoms

Upon performing a clinical hand exam, the hand surgeon may bring up the possibility of an inflammatory disease. Skin lesions can increase the risk of complications during a surgical procedure, especially in terms of infection and wound healing (Section 2.6).

### 2.1. Lupus

The term lupus encompasses a continuous spectrum of conditions, ranging from isolated skin lesions to systemic disease, which can affect several organs in the context of systemic lupus erythematosus (SLE). Cutaneous and mucosal symptoms make a significant contribution to the diagnosis, and sometimes even the prognosis.

In terms of disease classification, there are three skin-related forms of lupus:

- acute cutaneous lupus erythematosus (ACLE) which is mainly found in women (9:1 female to male ratio);
- subacute cutaneous lupus erythematosus (SCLE), which affects women after 50 years of age (70% of cases);
- chronic cutaneous lupus erythematosus, which is not as strongly associated with women (3:2 or 3:1 female to male ratio) and combines several clinical conditions including chilblain lupus erythematosus.

These various types of cutaneous lupus can sometime appear simultaneously in a patient.

The diagnosis of lupus skin lesions is based on a range of clinical, histological and immunological evidence. Dermatological features that are considered specific to lupus are mainly located in sun-exposed areas such as the back side of the hands along with the face.

In ACLE, involvement of the back of the hands is characterized by erythematous papules, which may or may not show signs of edema, or squamous lesions, mainly located between the joints (Fig. 1). The face may also have a rash, especially the nose and cheeks, with characteristic wolf-like or bat-like appearance (Fig. 2). A diffuse, measles-like, eczema-like or blister-like rash is possible, predominantly in sun-exposed areas. The surgeon must also look for mucous membrane involvement, notably erosion of the buccal mucosa.

Chilblain lupus erythematosus is the second type of lupus that can lead to typical hand involvement. The condition is localized to the tips of the fingers and toes. Chilblain lupus lesions can also affect the ears, nose, calves, heels, elbows and

Table 1
Half-life of the most commonly used biological therapies (based on Perdriger [1]).

| Drug name | Half-life (days) | Recommended number of days to stop before surgery | | | |
|---|---|---|---|---|---|
| | | Low risk | Medium risk | High risk | Very high risk |
| | | 2× half-life (days) | 3× half-life (days) | 4× half-life (days) | 5× half-life (days) |
| Etanecerpt (Enbrel®) | 3 | 10 | 15 | 20 | 25 |
| Infliximab (Remicade®) | 10 | 20 | 30 | 40 | 50 |
| Adalimumab (Humira®) | | | | | |
| Certolizumab (Cimzia®) | 15 | 30 | 45 | 60 | 75 |
| Golimumab (Simponi®) | | | | | |
| Rituximab (Mabthera®) | 20 | 40 | 60 | 80 | 100 |