

# Calciphylaxis and Martorell Hypertensive Ischemic Leg Ulcer: Same Pattern – One Pathophysiology

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## Key Words

Calciphylaxis · Martorell hypertensive ischemic leg ulcer · Eutrophication · Arteriosclerosis · Medial calcification · Fetuin A · Sodium thiosulfate · Debridement · Skin graft

## Abstract

This review presents a closer look at four diseases which are probably closely related to one another pathophysiologicaly: (a) calciphylaxis (distal pattern); (b) calciphylaxis (proximal pattern); (c) Martorell hypertensive ischemic leg ulcer; (d) calciphylaxis with normal renal and parathyroid function (synonym: eutrophication). The four diseases have largely the same risk factors: (1) arterial hypertension, (2) diabetes mellitus (types 1 and 2), (3) secondary or tertiary hyperparathyroidism (in end-stage kidney disease) and (4) oral anticoagulation with vitamin K antagonists. They share the same clinical patterns: necrotizing livedo, skin infarctions at typical locations and acral gangrene in calciphylaxis. They also share the same histopathology: ischemic subcutaneous arteriosclerosis and small-artery disease and 'miniaturizing' Mönckeberg medial calcinosis. The treatment concept for the acute phase of the diseases is also broadly similar. In addition to an optimized control of the cardiovascular risk factors, a proactive wound approach (necrosectomy, negative pressure wound treatment with vacuum dressings, and early skin grafts supported by systemic antibiotic therapy) leads

most rapidly and effectively to a reduction of the initially severe wound pain, and finally to complete healing of the wound. Oral anticoagulation with vitamin K antagonists should be stopped. In extensive cases, the use of intravenous sodium thiosulfate is recommended. All four diagnoses are little known in the medical schools of most countries. The need to improve familiarity with these four closely related disorders is therefore great. In particular, the risk of confusion with pyoderma gangrenosum is a major diagnostic problem which can lead to false and even damaging treatment.

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## Introduction

Martorell hypertensive ischemic leg ulcer (HYTILU) and calciphylaxis (synonym: calcific uremic arteriopathy) share a common clinical appearance and pathophysiology: skin infarction as a result of subcutaneous stenotic arteriosclerosis, accompanied by medial calcinosis. The characteristic vascular pathology which unifies the two entities can be detected on skin biopsies if the appropriate sampling technique is used. The following review summarizes the characteristic clinical and histopathological features, the pathophysiological understanding and a therapeutic concept for these two closely related diseases.