Mortality from systemic sclerosis in Portugal: results from the Rheumatic Diseases

Portuguese Register (Reuma.pt)

Proponents: Gonçalo Boleto – Rheumatology Department, Unidade Local de Saúde de

Santa Maria, Centro Académico de Medicina de Lisboa.

Research team: Gonçalo Boleto, Daniela Russell, Mariana Pereira Silva, Catarina Resende, João Eurico

Fonseca (Rheumatology Department, Unidade Local de Saúde Santa Maria, Centro Académico

de Medicina de Lisboa (CAML), Faculdade de Medicina, Universidade de Lisboa, CAML).

Abstract

Systemic sclerosis (SSc) is a severe autoimmune disease characterized by skin and organ fibrosis due

to immune dysfunction and microvascular damage. Mortality is significantly higher than in the general

population, mainly from complications such as interstitial lung disease, pulmonary hypertension,

myocardial damage, and scleroderma renal crisis. Advances in immunosuppressive therapies,

antifibrotic drugs, vasodilators, and stem cell transplants have improved survival. Several studies

highlight variations in mortality and risk factors across populations, emphasizing the need for localized

data to refine clinical practices and therapeutic guidelines, particularly considering genetic,

environmental, and healthcare access differences in specific populations like Portugal.

Therefore, our main goal is to identify the causes of death and risk factors associated with SSc in

Portugal.

This study will utilize the Rheumatic Diseases Portuguese Register (Reuma.pt). Patients meeting the

2013 SSc criteria will be analyzed. Individuals lost to follow-up over a 10-year period (2013-2023) will

be cross-checked with death certificates to determine any deaths and their causes. We will then

identify the risk factors associated with mortality in this population.