The European Clarkson’s syndrome registry

**Synonyms**

Clarkson’s syndrome

Monoclonal Gammopathy associated Systemic Capillary Leak Syndrome (MG-SCLS)

Systemic Capillary Leak Syndrome (SCLS)

Idiopathic Capillary Leak Syndrome (ICLS)

Idiopathic Systemic Capillary Leak Syndrome (ISCLS)

**What is Clarkson’s syndrome?**

The Monoclonal Gammopathy associated Systemic Capillary Leak Syndrome (MG-SCLS) also known as “Clarkson’s syndrome” is a rare condition characterized by unexplained recurrent attacks of systemic capillary hyperpermeability in the presence of monoclonal gammopathy (MG). Since the initial description of the disease in 1960 by Clarkson et al., fewer than 250 cases have been reported worldwide. During acute episodes, the leak of fluid and proteins from the intravascular compartment to the interstitium results in clinical signs of both acute hypovolemia (brutal and intense fatigue, thirst, faintness, profound hypotension, oliguria...) and interstitial edema (myalgia, nausea and vomiting, abdominal pain, paresthesia, segmental or generalized edema...). Biological profile is pathognomonic with marked hemoconcentration (elevated hemoglobin and hematocrit) and paradoxal hypoproteinemia/hypoalbuminemia. Compartment syndrome is a frequent complication during severe attacks. Diagnosis of MG-SCLS is based on recurring typical attacks in the presence of MG after exclusion of any other
cause of secondary capillary leak syndrome or hypoproteinemia. There is no validated treatment during acute attacks. A growing body of evidence supports the efficacy of monthly perfusion of intravenous immunoglobulins to prevent MG-SCLS attacks.

**What is EurêClark registry?**

EurêClark registry is an international study group, which gather observations of MG-SCLC and prospectively monitor attacks, preventive treatments, complications and outcome of patients. Since the EurêClark registry coordinating center has been established in the Service de Médecine Interne 2 of La Pitié-Salpêtrière Hospital, Paris, France, 49 medical centers in 8 countries (France, Italy, Israel, Switzerland, Lebanon, Spain, Canada and Turkey) have agreed to participate. The registry now contains information on more than 70 patients, which constitute the world’s largest cohort. We regularly publish up-to-date data that allow to better understand and manage MG-SCLS patients. Furthermore, we help physicians to manage acute and chronic manifestations of MG-SCLS.

**How to participate?**

If you wish to include a patient, please contact the EurêClark coordinating center (see below).

All participating physicians are associated in the EurêClark study group publications.

**Ethics**

The registry was approved by our local review board and by the Commission Nationale de l’Informatique et des Libertés in France. Every patient, or their next of kin, agreed to be included in the registry.
**How to contact EurêClark registry?**

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