

# Acute treatment of Idiopathic Systemic Capillary Leak Syndrome: a referral center's experience

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**Background** Idiopathic systemic capillary leak syndrome (ISCLS) presents with recurrent potentially life-threatening episodes of hemoconcentration and hypovolemic shock. Due to the rarity of the disease and to misdiagnosis, evidence-based approaches as well as validated protocols are still lacking.

**Aim** To report experience on timely diagnosis and treatment of acute shock in ISCLS-patients.

**Methods** Analysis of records from 12 ISCLS patients (9 men), from a cohort of 22, admitted once or several times to hospital for hypovolemic shock.

**Results** Mean age at symptoms' onset was 51.5 years. ISCLS crises had variable frequency, timing and severity. Mean follow-up was 6 years.

*Prodromal symptoms* reported by frequency were: arterial hypotension, fatigue, oliguria, worsening edema, weight gain, presyncopal/syncopal episodes, abdominal pain, nausea, vomiting, diarrhea, arthromyalgia, sore throat, dysphonia, cough, dizziness, high temperature, thirst/polydipsia, diaphoresis, dyspnea, altered consciousness, headache, livedo reticularis.

*The acute phase* could develop rapidly, with marked hypotension, tachycardia, oligoanuria, "stone-like" edema with paresthesia/pain. Blood tests revealed high hemoglobin (highest recorded value 25.8 g/dl) and hematocrit (up to 72%), hypoproteinemia (minimum serum albumin 9 g/L). IgG monoclonal band was present in all patients.

*Acute treatment* was sparing crystalloids and amines; infusing repeated 200 ml boluses of high molecular weight plasma expanders when SAP falls below 70 mmHg. Steroids, albumin, diuretics, methylene blue, iv Ig did not seem to have significant efficacy.

4/12 patients underwent CVVH, 2/12 mechanical ventilation, 1/12 fasciotomy, 1/12 extracorporeal circulatory support with venous-arterial ECMO.

*Complications* listed by frequency were acute renal failure, compartment syndrome and neuropathy, rhabdomyolysis, myocardial edema, pericardial effusion, pleural effusion-abdominal free fluid, cerebral involvement, acute pulmonary edema, deep vein thrombosis.

**Conclusion** ISCLS may induce severe shock associated with hemocentration and

hypoproteinemia. Judicious use of fluids and amines and knowledge of the strategies to prevent/minimize complications can reduce mortality and morbidity.