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A case of Idiopathic systemic capillary leak syndrome (ISCLS) in a toddler

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Case Study: Introduction
Idiopathic systemic capillary leak syndrome (ISCLS) also known as *Clarkson's disease* is a rare disorder that typically begins in midlife. But there are also cases in young children. It is characterized by episodes of severe hypotension, hypoalbuminemia without albuminuria, and hemoconcentration. Treatment for this disease during an episode is mainly supportive, aiming to stabilize symptoms and prevent severe complications.

Case
A 13 month-old girl with chief complains of irritability and sweating was referred with abnormal laboratory tests. Initial vital signs were as follows: BP 80/40, HR 128, RR 27, BT 35.9. Initial laboratory findings were as follows: WBC 31.43 10E3/uL, RBC 5.86 10E6/uL, Hb 15.2g/dL, Hct 43.2%, Plt 337K, BUN 27.1 mg/dL, Na 123mmol/L, albumin 2.9 g/dL, uric acid 7.3, creatinine 0.41. We could not find the exact cause of hypoalbuminemia (no proteinuria, normal stool alpha -1 antitrypsin l). During treatment with albumin and furosemide, pulmonary edema developed, but with massive diuretic therapy, she improved completely. Second episode occurred 2 years later with following symptoms: general edema, vomiting, abdominal pain, drowsy mentality, low BP, weight gain from 16kg to 17.4kg, low urination and at the same time she had URI symptoms. Laboratory tests were as follows: WBC 27.4 10E3/uL, Hb 15.3g/dL, Hct 43.2%, Plt 266K, BUN 27.1 g/dL, creatinine 0.36, Na 126mmol/L, albumin 2.3g/dL. Urine analysis showed mild proteinuria transiently. There were similar symptoms several times between of two episodes mainly related to the cold. We suspected ISCLS considering recurrent hypoalbuminemia and shock without definite cause. Last laboratory findings were done after 6 month and were normal. During 6 month after second episode there was no relapse.

Conclusion
ISCLS is rare disorder, especially in children. Nevertheless, we should consider ISCLS when we meet patients having recurrent hypoalbuminemia and shock without definite cause.