

## Congenital nephrotic syndrome with acute renal failure: questions

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### Case presentation

We report a case of congenital nephrotic syndrome with acute renal failure associated with multiple organ injury requiring peritoneal dialysis. Maternal serology, including hepatitis B, HIV, and *Treponema pallidum* hemagglutination (TPHA) and the Venereal Disease Research Laboratory (VDRL) tests were all negative at 10 weeks of gestational age (GA). At 34 weeks of gestation, the ultrasound examination showed hydrops fetalis. The umbilical cord puncture found anemia, with hemoglobin at 8.4 g/dl, thrombocytopenia at 43,000/mm<sup>3</sup>, cytomegalovirus (CMV)-polymerase chain reaction (PCR) and parvovirus-PCR were negative. The child was born at 35 weeks of gestational age and immediately transferred to an intensive care unit because of septic shock. The clinical findings were sclerema neonatorum with blisters, hepatomegaly, splenomegaly, and macroscopic hematuria. The

initial blood analysis showed hemoglobin 11 g/dl, thrombocytopenia 22,000/mm<sup>3</sup> (despite intrauterine blood and platelets transfusion), reticulocytes >150,000/mm<sup>3</sup>, C-reactive protein (CRP) 233 mg/L, urea 4 mmol/L, serum creatinine 72 μmol/L, proteins 56 g/L, serum glutamic oxaloacetic transaminase (SGOT) 482 UI/L, serum glutamic pyruvic transaminase (SGPT) 224 UI/L, haptoglobin <200 ng/ml, total bilirubin 108 μmol/L, and lactate dehydrogenase (LDH) 1,042 UI/L. Treatment was immediately initiated by amoxicillin, cefotaxime, and gentamicin. All bacteriological findings were initially negative (blood culture, spinal puncture, peripheral samplings). At the 7th day of life, serum creatinine decreased to 39 μmol/L.

Since birth, the patient had a macroscopic hematuria. The ultrasound found both kidneys increased in size, hyperechogenic, but without Doppler flow abnormality. A nephrotic syndrome was diagnosed at 4 days of life (proteinuria 60 g/L and albuminemia 20 g/L). Hematuria was 10 × 10<sup>6</sup>/ml and hemoglobinuria >600 μg/dl. Symptomatic treatment was initiated by albumin, anticoagulation, and recombinant erythropoietin. At the 9th day of life, CRP was 51 ng/ml, and hemoculture showed *Staphylococcus epidermidis*, motivating vancomycin treatment. Serum creatinine increased concomitantly to 262 μmol/L.

The answer to this question can be found at <http://dx.doi.org/10.1007/s00467-011-1845-6>

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### Questions

1. Which investigations should be performed (repeated) rapidly?
2. Which diagnosis is the most probable to explain the clinical presentation?