

Acute kidney injury in an infant with severe combined immunodeficiency: Questions

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Case summary

A renal consult was requested for a five-month-old female infant with an acute rise in serum creatinine. She was hospitalized for management of an Interleukin 7 Receptor deficient severe combined immunodeficiency (SCID). She had initially presented at the age of 2 months with protracted bronchiolitis, persistent mucocutaneous candidiasis, diarrhea, failure to thrive, lymphocytopenia with absent T cells and severe neutropenia. Polymerase chain reaction (PCR) in bronchoalveolar lavage sample was positive for *Pneumocystis jirovecii*.

Five days prior, the infant had been commenced on daily intravenous pentamidine (4 mg/kg/day). Pentamidine was chosen over co-trimoxazole in the treatment of *Pneumocystis* in view of the severe neutropenia. She was also receiving meropenem, amikacin and liposomal amphotericin 3 mg/kg/day for new-onset febrile neutropenia. Amikacin was administered for the previous 5 days on a full dose of 20mg/kg once daily followed by an adjustment for the current renal dysfunction at 10mg/kg based on trough levels. In view of increased work of breathing related to persistent bronchiolitis, the patient was on a high caloric feed with a strict fluid restriction of 90 ml/kg/day. Watery bowel movements were ongoing with up to 8 episodes of diarrhea daily.

On examination, she had cool extremities with prolonged capillary refill time of 4 sec, good peripheral pulse volume, heart rate of 126 bpm and systolic blood pressure of 110 mmHg. Upon reviewing fluid balance, the infant had lost about 2% of the baseline body weight and she was polyuric (urine output 3.6ml/kg/h). Sequential blood results are shown in Table 1. Plasma creatinine was increasing, urine osmolality was 267mOsm/kg, fractional excretion of sodium (FENa) was 12% and transtubular potassium gradient (TTKG) was 3.7. Urine microscopy was unremarkable and urinary tract ultrasound demonstrated enlarged, echo-bright kidneys.

Table 1. Sequential laboratory parameters at the time of initial consultation

	Day -2	Day -1	Day 0	Day 0
			sample 1	sample 2
Serum Creatinine ($\mu\text{mol/L}$)	20	46	102	130
Serum Urea (mmol/L)	7.5	12.9	18.6	22.3
Serum Na (mmol/L)	139	136	132	130
Serum K (mmol/L)	4.5	5.7	6.3	6.1
Serum Albumin (g/l)	34	34	33	31
Plasma Osmolality (mOsm/kg)	ND	ND	ND	295
Venous pH/ Bicarbonate (mEq/L)/ BE	ND	ND	ND	7.31/ 21/ -3

Abbreviations**BE** Base Excess**ND** Not done*Questions*

1. What is the diagnosis?
2. What are the risk factors for renal dysfunction in this infant with immunodeficiency?
3. What should be the management and the expected outcome in this patient?