

Table 1 Summary of demographic, clinical, and laboratory findings of 41 Vietnamese patients with beta-ketothiolase deficiency.

Findings	No. of patients
Demographic	
Male/female	23/18
Positive consanguinity	0/41
Positive family history	16/41 (39%)
Kinh ethnicity	41/41 (100%)
Clinical (1st episode)*	
Fever	36/39 (92%)
Vomiting	35/39 (90%)
Diarrhea	20/39 (51%)
Cough/sneezing	15/39 (39%)
Tachypnea	38/39 (97%)
Dehydration	38/39 (97%)
Altered mental status	39/39 (100%)
Convulsions	6/39 (15%)
Hypotension	4/39 (10%)
Laboratory (1st episode)*	
Metabolic acidosis	39/39 (100%)
Ketonuria	36/36 (100%)
Hyperglycemia	11/39 (28%)
Hypoglycemia	3/39 (8%)
Hyperammonemia	7/31 (23%)
Brain imaging abnormalities	4/28 (14%)

* Two of three patients diagnosed by screening have remained asymptomatic to date; 39/41 patients have manifested episodes of ketoacidosis.