Title: Propionic Acidemia *GeneReview* Table 3a
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Updated: October 2016

Table 3a. Findings During the Initial Metabolic Crisis

Symptoms	Prevalence [1]	Source
Feeding difficulties	59%-87%	Grünert et al [2012]
Vomiting	45%-70%	Kölker et al [2015a]
Hypotonia	47%-61%	Grünert et al [2012], Kölker et al [2015a]
Somnolence	35%-55%	Grünert et al [2012]
Coma	12%-26%	Grünert et al [2012]
Seizures	12%-26%	Grünert et al [2012], Kölker et al [2015a]
Respiratory problems	57%	Lehnert et al [1994]
Hyperammonemia [2]	93%-97%	Grünert et al [2012]
Metabolic acidosis	67%	Grünert et al [2012]
Anemia	8%-41%	Grünert et al [2012]
Leukopenia	25%-52%	Grünert et al [2012]
Thrombocytopenia	8%-21%	Grünert et al [2012]

1. The differences in the reported prevalence of findings may reflect variable sizes of the cohorts, age of the last evaluation, length of follow-up, differences in the therapeutic approaches, availability, turnaround time and sensitivity of the newborn screening, screening method (newborn screen versus selective metabolic screen), overlap of patients in the reported cohorts, ascertainment and recall biases.

2. The mean plasma ammonia levels in patients with early onset PA can vary between 207 and 697 µmol/L [Grünert et al 2012, Kölker et al 2015a]. Reported plasma ammonia levels fall between 86 and 3377 µmol/L [Kölker et al 2015a]. In contrast to urea cycle disorders, hyperammonemia in PA is usually accompanied by normal level of glutamine [Al-Hassnan et al 2003, Filipowicz et al 2006, Kölker et al 2015a].

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