

# :: Hyperammonaemia due to transport defects

## Diseases:

- ▶ Hyperornithinemia-hyperammonemia-homocitrullinuria syndrome
  - Synonyms: HHH syndrome, Triple H syndrome
- ▶ Lysinuric protein intolerance
  - Synonyms: Hyperdibasic aminoaciduria type 2, LPI



The British Inherited Metabolic Disease Group (BIMDG) has published on its website guidelines for the emergency management of patients with inherited metabolic disorders.

Here are the ones for the management of **hyperammonaemia due to a HHH syndrome or a lysinuric protein intolerance in children.**

- ▶ See [children BIMDG guidelines](#) (last reviewed in Feb 2012)

**Further Information:** see the Orphanet Abstracts for the [HHH syndrome](#) and the [lysinuric protein intolerance](#).