

# :: Maple syrup urine disease

## Synonyms:

- ▶ MSUD
- ▶ Branched-chain ketoacid dehydrogenase deficiency
- ▶ BCKD deficiency
- ▶ Branched-chain ketoaciduria
- ▶ Leucinosi

## Forms:

- ▶ Classic maple syrup urine disease
- ▶ Intermediate maple syrup urine disease
- ▶ Intermittent maple syrup urine disease
- ▶ Thiamin-responsive maple syrup urine disease



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 an **Acute decompensation in children and adults with maple syrup urine disease:**

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

**Further Information:** see the [Orphanet Abstract](#).