

:: Glycogenosis due to glucose 6 phosphatase deficiency

Synonyms:

- ▶ G6P deficiency
- ▶ Glycogen storage disease type 1
- ▶ GSD type 1
- ▶ Type 1 glycogenosis
- ▶ Hepatorenal glycogenosis
- ▶ Von Gierke disease

Forms:

- ▶ GSD type 1a
- ▶ GSD type 1b



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 a G6P deficiency.

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

N.B.: Guidelines for management of children suffering from GSD type 1b are also available on BIMDG website.

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

Further Information: see the Orphanet Abstract for [G6P deficiency type 1](#), [G6P deficiency type 1a](#) and [G6P deficiency type 1b](#).