Non-ketotic hyperglycinaemia (NKH) is a rare inborn error of glycine metabolism characterised by accumulation of glycine in body fluids and tissues, resulting in neurometabolic symptoms of variable severity.

Sodium benzoate is the sodium salt of benzoic acid that conjugates with nitrogen containing glycine to form the molecule hippurate. Hippurate can be excreted by the kidneys, reducing plasma glycine levels. N-methyl-D-aspartate receptor antagonists may ameliorate neurological symptoms although it remains to be established whether they improve long term outcome. Lack of authorised presentations for treatment of rare diseases is an important obstacle that is usually resolved by hospital pharmacy formulations, especially in children.

Aim and objectives

The aim was to provide an adequate, stable and well accepted oral sodium benzoate formulation for a patient with NKH, to improve her general status

Material and methods

A 4-year-old girl with severe NKH needed oral treatment with sodium benzoate, although there is no standard oral formulation for children. To find an optimal and suitable solution, a literature search was carried out in the National Library of Medicine’s (MEDLINE) database, including terms ‘sodium benzoate/chemistry’, and ‘administration, oral’ with no other filter. Our national and regional formulation databases were also checked.

Results

The patient was initially treated with 16 mL, three times a day, sodium benzoate syrup 112.5 mg/mL, but the volume needed was impossible to swallow by the patient due to her clinical status.

Subsequently, 2 g sodium benzoate sachets were given with meals (four times a day) but they were not well tolerated.

We then dispensed a 250 mg/mL suspension in Ora-Sweet with a stability of 90 days. Despite it being a new formulation for our pharmacy service, glycine levels were reduced from 900–1000 m/L to 500 m/L over 2 months. Currently, her clinical situation is stable, and the patient receives 8 g/24 hours of sodium benzoate which is well tolerated.

Conclusion and relevance

Sodium benzoate oral suspension dispensed with Ora-Sweet seemed to be an adequate solution to NKH treatment in our patient. Although the formulation is a basic operation for hospital pharmacy services, it is essential, especially in children with rare diseases that need orphan drugs.