

Information for Pharmacists

What is GA 1?

Glutaric Aciduria Type 1 (GA 1) is an inherited metabolic disorder affecting the metabolism of foods containing protein. People with GA 1 cannot use protein in the normal way. Protein is made up of amino acids, two of which are lysine and tryptophan. In GA-1, the enzyme which breaks down lysine and tryptophan is absent or only present in small amounts. The resulting build up of these two amino acids but in particular lysine in the blood can cause brain damage and death unless treatment is commenced. Some lysine is essential for growth.

GA 1 is treated by restricting the natural protein in the diet. This prevents the build up of excess lysine in the blood, while providing sufficient lysine for growth and development. High protein foods are replaced by a synthetic amino acid mix and the diet supplemented by a variety of low protein foods available on prescription. These low protein foods provide energy, bulk and variety in the diet. At present the recommended treatment for GA 1 is **Diet for Life**.

In addition a medication called carnitine is prescribed to aid the breakdown of lysine. The dose of this is usually doubled if the patient is unwell.

Prescriptions

Children and adults with GA 1 will require regular prescriptions for their low protein foodstuffs. These items are an integral part of the treatment of GA 1 and without these foods the dietary management will not be successful. In Ireland, most patients are entitled to a medical card which entitles them to order low protein foods and their prescribed synthetic protein/amino acid supplement without cost. In some cases it may be necessary to apply for the hardship scheme for certain foods which have not been GMS listed.

Low protein foods: There are a large variety of low protein foods available from several manufacturers. The patient will often prefer one manufacturer to another for different items. It is very important to encourage variety in the diet so that patients will not resort to seeking restricted foods and thereby upsetting their dietary management. We provide an updated list of manufactured food each year.

Amino Acid Supplements: Every patient following a low protein diet requires an amino acid supplement to replace the forbidden high protein foods. It is vital they have a constant supply of this amino acid supplement. If they do not receive this supplement then their lysine levels will rise because the patient will become catabolic. If this continues over time it will effect the patient's growth and development.

There are several types of amino acid supplement, prescribed according to age and suitability for the patient. Once the child or adult is established on one they tend not to change for several years. Supplies can therefore be ordered in advance to ensure a constant intake. There are a range of amino acid supplements for other metabolic disorders which have similar label colour, label design and names to the lysine free supplements. These are **not** suitable for the treatment of GA1.

What if you have supply issues?

Should you experience any problems obtaining any of the low protein foods or amino acid supplements it is worth contacting the manufacturer directly as they often have sufficient stocks of the products.

A metabolic dietitian can be contacted at any time during office hours to discuss any problems or concerns you may have about the prescriptions for the patients with GA1.

Trouble-Shooting

- When new products become available on prescription, they are often not easy to obtain because the wholesaler you use may not have obtained supplies. It is best to put in your order as usual and then contact the supplier and order the product direct until stocks are available locally.

- There are some low protein foods produced by small companies that may not be available from your wholesaler. If you have difficulty obtaining any of the low protein foods they should be ordered directly from the company.
- Many gluten free products are similar in appearance to the low protein foods, **but are not suitable** for the patient on a protein restricted diet as they often have protein added in the form of egg, milk or soya. This will increase the protein content of the product to a level too high for patients following a low protein diet. **Many low protein products are Gluten free, but Gluten free products are not necessarily low protein**

For more information about GA1 contact:

NCIMD

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PK Foods Email: ellis@glutenfree-foods.co.uk	All Phar	01-4041600
Juvela Freephone number: 1800 405090 Email: lowprotein@juvela.co.uk	United Drug	01-4632300 01-4598877
Promin UK number: 0044161 474 7576 Email: info@prominpk.com or katya.sheridan@firstplaydf.com	Biofact	01-612544
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