

Dietary management of a 10-year old boy with Arginosuccinic Aciduria (ASA) using basecal®.

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Patient Details & Medical History

Age:

10

Gender:



Anthropometry:

Weight: 33.4kg, 50th-75th centile Height: 123.7cm, 0.4th -2nd centile

Initial presentation:

Our case is the second child of unrelated Caucasian-origin parents. At 5 days of age he was admitted at a local hospital with respiratory symptoms and presumed sepsis. On follow-up he had neurological investigations for developmental delay. Following sedation for an MRI scan, he became encephalopathic with ammonia elevated at 350µmol/L. He was started on 10% IV dextrose and ammonia concentrations reduced to 70µmol/L. Further analysis of urine and plasma showed an elevated level of arginosuccinic acid (urine) and low arginine (plasma) - the patient was diagnosed with ASA at 4 months of age. A low protein diet and oral medications (sodium benzoate and arginine) commenced at diagnosis. He was then transferred to an inherited metabolic disorders (IMD) team for ongoing care.

Medical and dietary management:

ASA, a urea cycle disorder (UCD), is a complex condition requiring a multi-faceted dietary and medicinal management regimen. In addition to the IMD team, the patient is reviewed by several other medical disciplines (e.g. liver). His prescription medications include the "nitrogen scavenging" medications sodium benzoate and sodium phenylbutyrate. He is also on:

L-Arginine: 750mg TDS
Levetiracetam: 750mg BD
Gaviscon liquid: 5mls after feeds

Omeprazole: 20mg OD
Clonazepam: 600mcg OD
Potassium Chloride: 10mmols BD

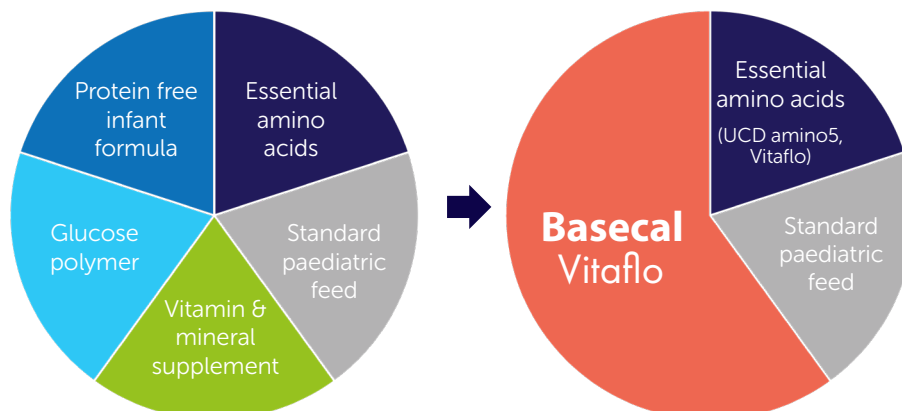
His dietary management involves following a low protein diet to meet WHO/FAO/UNU 2007 safe levels of protein intake¹ which is achieved through oral intake and a low protein gastrostomy feed given day and night. Adequate energy intake is essential to achieve good metabolic control. In times when the oral intake is minimal, all his nutritional intake comes from enteral feeds. Emergency regimens, when required, are glucose polymer feeds (S.O.S20™, Vitaflo).



Aim and Plan of Management

Prior to the introduction of basecal®, the patient was on a complex modular feed, comprised of multiple ingredients which required regular adjustments.

With the introduction of basecal, a protein free feed containing carbohydrate, fat and micronutrients, there was an opportunity to simplify this aspect of his management.



Outcome

Although the new and old feed recipes were isocaloric, the introduction of basecal simplified the patient's feeding regimen. The patient tolerated the new tube feed well - there was no feed intolerance. His ammonia levels were stable and his blood essential amino acids levels were mainly within reference range.

Recent Developments

The feeding plan has been followed well and has proved easily adaptable to changes in the patient's appetite, growth needs, nutritional requirements and metabolic stability. The patient's main carer is a single, working mother and this increased ease of preparation has proved very valuable. Although the patient's behaviour remains challenging, periods of metabolic instability and inpatient admissions became less frequent. Furthermore, his appetite has improved, reducing his overall dependence on tube-feeding and eliminating the need for any supplementary daytime gastrostomy feeds which previously had sometimes been necessary.

Discussion

This case study highlights several familiar challenges for those involved in the dietary management of UCD;

- Difficulties and delays in diagnosis
- A significant and chronic disease burden
- Complexity of dietetic and medical management

With regard to this last point, dietitians can play a pivotal role in reducing dietary management complexity. In patients requiring bespoke modular feeds, research has indicated that a regimen with fewer ingredients is more accurate and easier for caregivers to prepare^{2,3,4}.

In this case, basecal helped reduce the number of ingredients in the patient's feed. It was well accepted by the patient orally, so it boosted oral intake which, in combination with his improvements in appetite, helped reduce reliance on enteral feeds. With metabolic patients at risk of metabolic decompensation, it is noteworthy that these practical benefits were accompanied by good gastro-intestinal tolerance and improved metabolic stability.

References

- 1 Häberle J, Boddart N, Burlina A, Chakrapani A, Dixon M, Huemer M et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet J Rare Dis.* 2017; 7(1):32.
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