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Clinical Presentation

Baby L was born in August 2012 in Ireland. He was diagnosed with combined immune deficiency (CID) and a zinc transporter defect after birth and was referred to a UK specialist tertiary unit for assessment for a bone marrow transplant (BMT).

History

Baby L was born full term and his birth weight was 3.6 kg (25th centile; UK-WHO growth chart). He was initially fed on an extensively hydrolysed semi-elemental powdered formula taking 150 ml/kg/day (102 kcals/kg/day). However, his weight began to fall due to feed intolerance secondary to immune dysregulation. By two weeks of age his weight was 3.175 kg (9th centile). His feeds were increased to 175 ml/kg/day (119 kcal/kg/day) but due to lethargy he could not finish his feeds so a nasogastric feeding tube was inserted to provide top-up feeds.

He was unable to cope with the additional volume (he started vomiting), so an energy dense whole protein infant feed (1.0 kcal/ml) was started. At the same time, his feeding regimen was changed from top-up boluses to continuous feeding (22 ml/ml/hour). He continued to

show signs of feed intolerance - e.g. increased secretions, had a sore bottom, and his weight remained poor - and by one month of age he weighed 3.72 kg (9-25th centile).

Despite his continuing symptoms of poor feed tolerance, he managed to gain some weight on the high energy infant feed. As his investigations for BMT were completed he was prepared for transfer back to his local tertiary unit in Ireland. Prior to discharge to his local tertiary hospital, he was switched to Infatrini (1.0 kcal/ml) following a bolus feeding regimen and did quite well. He was now 8 weeks old but weighed only 4.15 kg (just below 2nd centile). It is important to note that Baby L had continued to have faltering growth whilst being assessed for BMT and, indeed, was sent back to his local tertiary unit with poor growth.



Management

He returned to our unit at the age of 26 weeks because of worsening bowel symptoms. He weighed 6.2 kg (0.4th-2nd centile). He was on Infatrini Peptisorb (1.0 kcal/ml extensively hydrolysed ready-to-use feed) having been changed over by his local hospital in Ireland. He was having 5 feeds a day plus some solid food. The feed was providing him with 88 kcal/kg/ day. He had loose stools and vomiting but it was also noted that he had Norovirus.

He was started on parenteral nutrition (PN) as he could no longer maintain his fluid and electrolyte balance and his enteral feeds were reduced to provide 40 kcal/kg/day as the enteral feeds were increasing his bowel losses. PN was initiated for three weeks but later stopped due to rapidly deteriorating liver function tests. At the end of his period on PN his weight was 6.98 kg (2nd-9th centile). However. he remained on electrolyte infusions whilst taking Infatrini Peptisorb given as 5 x 50 ml feeds per day. Over the next month his clinical condition deteriorated significantly though his weight remained static. He was fluid-restricted so unable to meet requirements and he could not have PN because his liver was now showing signs of failure. In addition, he could not maintain his blood sugars so feeds were given continuously.

His liver function slowly improved following cessation of PN and he was able to consume more Infatrini Peptisorb to meet his requirements, but his weight remained static. Over time his oral intake increased and he managed more oral bolus feeds. However, he had eight loose stools per day and his weight at the age of 45 weeks was 6.6 kg (0.4th centile). He was discharged home a week later and continues to be assessed as an outpatient.

His last weight at the age of 46 weeks was 6.81 kg (0.4th centile) showing a slight improvement.

Discussion

This child's pathway is fairly typical of those seen in this unit who have an immune dysregulation disorder and await a BMT. These patients are extremely difficult to manage due to their underlying conditions and they may have significant gut problems, usually presenting with symptoms such as vomiting, diarrhoea, electrolyte imbalance and significant faltering growth. Gut function can deteriorate and often complications with liver function necessitate significant fluid restrictions. Once fluid and electrolyte balance can be maintained these children can return home until a donor for BMT can be found. PN is avoided for as long as possible due to the risks, such as line infection and liver dysfunction, whilst being on immunosuppression.

Infatrini Peptisorb is now the feed of choice when managing these infants. Gut failure can impair fat absorption and the 50% medium-chain triglycerides in Infatrini Peptisorb provides an alternative energy source, being absorbed by a different pathway to long-chain triglycerides. In addition, a 1 kcal/ml product maximises energy in a smaller volume which is important for infants when they are fluid restricted.

Summary

For vulnerable infants such as baby L, Infatrini Peptisorb is the feed of choice as it is nutrient dense (optimal energy, protein and micronutrients) in a ready-to-use format and extensively hydrolysed. This enables us to avoid the risks associated with concentrating powder feeds. Infatrini Peptisorb proved invaluable for this child.

Peptisorb

