CASE STUDY OF A CHILD WITH EPIDERMOLYSIS BULLOSA (E.B.)

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This case study relates to a boy who was diagnosed at ten days with Recessive Dystrophic Epidermolysis Bullosa.

Initial feeding history:

Prescribed 150% of the Reference Nutrient Intake (RNI) for protein and calories, fed via a Silk nasogastric tube. This was changed to a cup and spoon/syringe, due to the constant re-passing of the tube. Feed of choice was concentrated infant formula with additional fat, carbohydrate, minerals and vitamins. Spoon feeding was commenced at four months of age.

Weight progressed normally for the first year of life and the only nutritional problem noted at 11 months was constipation. By two years and five months, the boy had settled into a weight centile of above and parallel to the third centile on Irish Clinical Growth Charts (1). His height centile followed just above and parallel to the tenth centile. A progression to sip feed Paediasure™ was attempted but refused and he returned to concentrated infant formula. At two years and ten months, he was receiving concentrated infant formula and a fibre rich sip feed, Enrich™. Difficulties with lumpy food and the presence of oesophageal strictures were reported. At this stage, the possibility of a gastrostomy for poor weight gain was mentioned to parents. His weight and height continued on the centiles until the age of six and a half when the possibility of artificial feeding was broached once more. At six years and ten months, his weight had fallen below the third centile and the insertion of a skin level device was performed.

POST BUTTON

- DAY 1 Commenced enteral feed. Nutrini Fibre (Nutricia). Small volumes increasing as tolerated.
- DAY 2 Tolerating – plan to (increase) feed o/n. 100ml x 10H.
- DAY 4 Discharged home on feed as above.
- DAY 6 Mother contacted. B O ++ overnight. Alternate feed with Nutrini d/c Duphalac.
- Regrade as tolerated to Nutrini Fibre.
- DAY 25 Father phoned. PU++ at night. Advised (decrease) feed o/n. Add 2 bolus.
- Cubby™ dislodged. Admitted for resiting.
At three months post button insertion, he was tolerating 1000 ml of a multifibre polymeric feed overnight, with small amounts of food during the day. Eleven months post button, his weight had increased by 4kg from the time of insertion and he had achieved the tenth centile for weight. At eighteen months post button, he was having a weight plateau. He was occasionally not having his feed overnight. A specialised wound repair feed Cubitan™ was recommended as a sip feed, but refused. Recommendation to change the enteral feed to an energy dense feed with fibre. Nutrison Energy Multifibre was recommended. This was introduced gradually and he is tolerating same.

<table>
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<th>FEED/</th>
<th>NUTRINI</th>
<th>NUTRINI</th>
<th>MULTIFIBRE</th>
<th>ENERGY</th>
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<tr>
<td>100ml</td>
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<td>MULTIFIBRE</td>
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Reference:

(1) Irish Clinical Growth Standards Boys: Birth to 19 years, Hoey, Tanner and Cox (1986), Castlemead Publications.