



## CASE STUDY 4: GEORGE

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### Patient Profile

George is a 2 year old male. His birth weight was 2.319kg (below 0.4th centile). Infancy onset epilepsy occured at 3-month-old. He was diagnosed with microcephaly and congenital pontocerebellar hypoplasia.

### Background

His current medication consists of Carbamazepine (120 mg, oral, twice daily) and Levetiracetam (300 mg, oral, twice daily).

An internal referral from the epilepsy team was made due to drug resistant epilepsy.

George has tonic seizures, described by his parents as arms and leg outstretched and stiff, head turns to the left. They currently occur 2-3 times per fortnight and last less than a minute, previously they occured up to 6 times per week. They can also come in clusters lasting up to two hours requiring hospital intervention.

George has poor oral intake and associated faltering growth. He has only gained 1.4 kg over a year, his weight dropping from the 0.4th centile to between -3 and -4 SD below the 0.4th centile. His length plots along the 2nd centile. In the initial appointment maintaining his nutritional status was difficult due to his decreased intake of milk. George was not taking sufficient formula from a bottle and parents were using a syringe to give milk and fluids. He was managing three small pureed meals per day and ~ 500ml fluid daily. Meals take 45 to 90 minutes to finish. There were no overt signs of aspiration, for example coughing or choking, but he does drool. It was felt prior to ketogenic diet a speech and language assessment was appropriate to ensure safety of swallow. It was also apparent that medication adherence was impacted by George spitting out medications. Dietetic input and the need for a nasogastric tube (NGT) and the likely long term need for gastrostomy was discussed.

The MDT agreed the need for nutritional support via a tube to support nutritional and hydration status and medication adherence. Referrals were made to a local dietitian, to speech and language therapy, and a local paediatrician for urgent placement of a NGT and a possible gastrostomy.

### Clinical assessment

Weight: 7.88 kg (-3 and -4 SD below the 0.4th centile)

Weight age: 5/6 months

Height age: 14 months

### Estimated Nutritional Requirements Actual Age

Energy: 646 - 1000 kcal (82 kcal – EAR)

Likely low estimated energy requirements due to low activity, therefore consider low physical activity energy expenditure for growth and maintenance estimated at 77%: 497 – 770kcal

Protein: 7.6 - 14.5 g (0.97 g/kg, safe intake - RNI)

Could also consider weight age due to significant difference from actual age.

### Estimated Nutritional Requirements Weight Age

Energy: 567 – 756 kcal (72 – 96 kcal/ kg)

Protein: 9.8 - 12.7 g (1.24 g/kg, safe intake - RNI)

### Estimated fluid requirement: ~800 - 1000ml

### Estimated current intake:

250 m whole cows milk – 160 kcal

3 meals per day: 460 - 560 kcal

Total energy: 450 – 550 kcal

Takes healthy start vitamins (Vitamin A, C and D)

### Speech and language therapy recommendations:

- Unsafe swallow currently to both fluids and food due to risk of aspiration. Can continue tastes. Repeat assessment in 6 months.
- Therefore need to provide 100% nutrition and fluids via NGT. Poor growth on current intake therefore consider adding add additional 10% energy to current estimated intake with the aim for improved weight gain.
- Target energy requirements: 600 kcal, 14.5 g protein.

### Dietetic interventions:

1. Change macronutrient composition of diet to a classical ketogenic diet to initiate ketosis.
2. Improve growth aiming for increases towards 0.4th centile.

Upon discussion with the family they would like to initiate 4 feeds per day, ideally if tolerated via gravity bolus feeding. Parents asked about the available products discussed the need to ensure the recipe is simple and easy and meets his nutritional needs. Parents expressed a preference for liquid feeds with their main reasoning that they are expecting a new baby in the next 8 weeks and time at home is under pressure. Mum also expressed that she is concerned that having multiple powder formulas might be difficult to manage.

### Reviewing Ketocal formula feeds to determine most appropriate feed considering:

- Initial ratio and potential for ratio changes
- Patient age
- Nutritional completeness of the feed including at a ratio 4:1
- Is a multivitamin supplementation required?
- Ease of preparation e.g. number of ingredients, mixing of ingredients
- Parental preferences
- Local home enteral feeding contract company

The initial ratio target was 2:1, with a potential transition to a 4:1 ratio, if clinically indicated. Initial target regime was to reduce side effects, improve tolerance and initiation of ketogenic diet. Through considering both the current plan and potential future needs this can help reduce risk of feed changes increasing workload and waste. Ketocal 3:1, Ketocal 4:1 Powder and Ketocal 4:1 LQ meets vitamin, mineral requirements but does not meet reference nutrient intake (RNI) for protein. Although Ketocal 2.5:1 LQ meets and exceed protein RNI, it is also higher in vitamins, mineral and trace elements and not licensed in under 3s. It is important to consider safety aspects of feeding. There was also a need to consider the challenges of measuring and mixing on ingredients whether using scoops, syringes and/or scales.



### Initial feed plans

400ml Ketocal 4:1 LQ

125ml Paediatric 1 kcal/ ml fibre containing formula

Made up to 1000 ml with water

Ratio: 2:1

Provides per 100 ml: 72 kcal, 1.6 g protein.

Feeding regimen: 4 x 200ml

Initiated feeding by switch alternating feeds between Paediatric 1 kcal/ ml fibre containing formula and ketogenic formula (Ratio 2:1) within one week George was fully on the ketogenic diet at a ratio of 2:1. Ketones range between 0.9 – 1.5 mmol/l. Over the next 4 weeks the ratio was increased to 4:1. Ketones increased to 4.5 – 5.5 mmol/l. His feed was diluted Ketocal 4:1 LQ, 72 kcal and 1.5g protein per 100ml.

### Clinical Outcome

Over the first 8 weeks George tolerated the feeds well. However, weigh gain slowed and his weight centile was moving closer to -4SD below 0.4th centile. Therefore the recipe was increased by ~10% in two increments with the feed providing 79 kcal and 1.6 g protein per 100 ml which helped improve growth.

### Outcome

At 12 weeks on diet he had a clinical follow up. George had stable ketosis for 8 weeks. He had suffered with constipation; his bowels were opening every 48-72 hours and involved straining and despite dietetic management he required a laxative to regulate bowels to open daily. His weight was tracking along -3SD below the 0.4th centile.

He continued to have seizures however, he had not had any prolonged clusters of seizures since initiating the ketogenic diet and could have up to 16 days seizure free. In the last 4 weeks, he had 4 seizures which were more subtle. Therefore, it was agreed to continue ketogenic diet due to the improvements in seizure control.