

CASE STUDY 2: ASSAN

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Background and referral information

Assan was diagnosed with myoclonic astatic epilepsy (Doose syndrome) as a toddler and had trialled and failed six anti-seizure medications. Assan had severe learning disabilities and relied on his family and carers for around the clock care. During childhood, his swallow had deteriorated, resulting in weight loss, and he was fed via a nasogastric tube, followed 18 months later by a gastrostomy tube. Since Assan attended a school for special educational needs, he would be under the paediatric epilepsy service until he turned 19 years old.

Assan is an 18-year-old man who commenced a ketogenic diet (KD) just before his seventeenth birthday. When referred for assessment for KD, he was having 70-80 seizures daily presenting as clusters of absence seizures characterized by head drops and myoclonic upper limb jerks. In addition, he was having 3-5 tonic seizures each day.

His consultant neurologist referred him to the keto team for assessment. KD therapy had been discussed a few times in the past, but Assan's parents had been reluctant to trial it. There was no local adult keto service that Assan could transition to so it was felt that this would be his last opportunity to trial KD. His parents agreed to meet with the keto team to discuss their concerns.

Assan had struggled with poor feed tolerance as a child, which had been a very difficult, stressful and worrying time for the family. Although he was very settled on a standard whole protein feed and tolerating it well, these early experiences made them reluctant to make significant changes. He was also very hard to bleed, and his parents were worried about the need for regular blood tests on KD. They were concerned that so much fat would cause him to gain weight which would mean his wheelchair would need resizing and replacing. We had a long discussion about the potential for KD to help manage Assan's seizures and that the response rate in those with Doose syndrome was as high as 60-70%[1-2]. This together with the reassurance that the KD prescription would match Assan's current energy intake and be phased in slowly, helped reassure Assan's parents and they agreed to a 3 month trial of classical KD. Baseline KD bloods and urinalysis were undertaken[3,4]. Assan's calcium creatinine ratio was significantly raised which prompted a renal ultrasound. This was normal so KD commenced, and potassium citrate was started to alkalise the urine and reduce the risk of a renal calculi.

Clinical assessment

Weight: 72.6 kg (stable)

Feed regimen: 1400 mls Nutrison per day providing 1400 kcals and 56g protein.
280 ml x 5 pump assisted boluses per day and additional fluid flushes with feeds and medications.

Expectations of KD: Reduction in seizure frequency, ideally 50% or more. Parents keen to wean 1 anti-seizure medication. Assan sleeps a lot during the day and poorly at night, waking and seizing often, so keen to monitor for any change in sleeping patterns too.

Dietetic intervention

Assan had lower energy requirements than is typical for a man his age so this would limit the ratio of classical KD we could achieve. It is not uncommon for energy and protein needs to be our ratio limiting step with adults on a classical KD regime. Here we could achieve a maximum of a 2.2:1 classical KD. Assan's weight was stable, so we used the minimum pro/kg/day of 0.75g/kg/day. Table 1 outlines the calculation of the classical prescription using the dietary unit method and table 2, the feed recipe using KetoCal 4:1 LQ as a base.

Table 1. Classical KD calculations

2.2:1 Classical KD

2.2:1 diet unit = 23.8 kcals

1400/23.8 = 59 diet units/day

Fat = 2.2 x 59= 129.8 g fat

Pro = 54.5 g

CHO = 59-54.5 = 4.5 g

Total energy = 1404 kcals

Table 2. Classical KD feed recipe

Product	Fat (g)	Pro (g)	CHO (g)
750ml KetoCal 4:1 LQ	111	23.2	4.6
36g Protifar	-	31.4	-
40ml Calogen Neutral	20	-	-
Total (g)	131	54.6	4.6
Energy (kcals)	1179	218.4	18.4
1416kcals 2.2:1 ratio			

Ingredients were combined and water added up to 1400mls to mimic his existing regimen and feed volumes. The recipe met at least the lower reference nutrient intake for all vitamins and minerals, except potassium which was slightly lower, however the potassium citrate addressed this shortfall. Classical KD was phased in slowly as per table 3. Typically, we would increase the percentage contribution of the ketogenic feed every 1-3 days, however we increased more slowly owing to Assan's parents' concerns of poor tolerance.

Table 3. Phased introduction of 2.2:1 classical KD

Phase 1	Duration	Existing feed	Ketogenic feed
1	5 days	75% of energy	25% of energy
2	5 days	50% of energy	50% of energy
3	5 days	25% of energy	75% of energy
4	Day 16 onwards		100% of energy

Assan tolerated the introduction of KetoCal LQ4:1 (+ additions) very well with no adverse effects. He became ketotic on day 13 and therafter produced good ketones despite the low ratio classical KD. Morning serum ketones averaged 1.5-2mmol/l and evening 2-3mmol/l. The ratio could not be increased further (owing to low energy requirements), however his starter regimen was fine-tuned by replacing the Calogen Neutral with Liquigen. This provided 12% of total energy (8.3kcal/g fat = 166 kcals) as MCT fat, which can boost ketosis. Assan's respite centre were very keen to support his KD. The centre had not had a patient on KD for many years, so a training session was arranged with parents and the respite team to ensure all were comfortable with the feed regimen, ketone testing and management of hyperketosis should it occur.

Clinical Outcome

Outcomes were assessed at 3 months on KD, daily seizures had reduced by approximately 70%. Parents and carers reported Assan to be sleeping less during the day and he was brighter and more alert when awake. His personality and cheekiness were coming through! Assan's parents felt the greatest impact was Assan's ability to attend school more regularly and be more engaged while doing so. Overall, they felt his quality of life was improved, and by default theirs too, now that Assan's epilepsy was better controlled. The positive outcomes continued and at 6 months on KD, clobazam was slowly weaned.

Learning Points

Arguably Assan should have been referred earlier for KD but there were many factors influencing his parents' understandable reluctance to trial KD. Initially managed by a community home enteral feeding team, his care was transferred to the keto team, but the community team remained aware of his progress. It was an excellent opportunity to share the potential benefits of KD for adult patients and prompt discussion regarding other patients who may benefit. It would be ideal, in the long run, for patients such as Assan to be transferred back to the enteral feeding team once stable on KD, with support as required from the keto team. Shared care may be one approach for improving access to KD for adult patients.

If Assan's parents had met with the keto dietitian or an experienced dietetic assistant earlier, perhaps their fears and worries would have been allayed.

Often a 2- 2.5:1 ratio is the highest that can be achieved owing to protein needs and low energy requirements: it is still worth trialing the KD at this low ratio. Once KD is established, then consider fine tuning by replacing some long chain triglycerides (LCT) with MCT fat if needed to optimise ketosis. Start at 10% and increase in 5% increments as required.

Ensure the limitations of protein, energy and hence maximum ratio of classical KD are discussed with the family to ensure they have realistic expectations of KD .

References

1. Oguni H, Tanaka T, Hayashi K, et al. Treatment and long-term prognosis of myoclonic-astatic epilepsy of early childhood. Neuropediatrics. 2002;33:122–132.

2. Caraballo RH, Cersosimo RO, Sakr D, et al. KD in patients with myoclonic-astatic epilepsy. Epileptic Disord. 2006;8:151–155.

3. Kossoff EH, et al. Optimal clinical management of children receiving dietary therapies for epilepsy: Updated recommendations of the International Ketogenic Diet Study Group. Epilepsia Open. 2018. 21;3(2):175-192.

4. Cervenka MC, Wood S, Bagary M, Balabanov A, Bercovici E, Brown M-G, et al. International Recommendations for the Management of Adults Treated With KD Therapies. Neurol Clin Pract. 2021;11(5):385–97.