Ketogenic dietary therapy for Lennox Gastaut syndrome

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First described in 1966 (1), Lennox-Gastaut syndrome (LGS) is an epileptic encephalopathy which typically presents before eight years of age and accounts for up to 10% of all childhood epilepsies (2). It is characterized by multiple seizure types (most commonly tonic or atonic drop attacks) and abnormal electroencephalographic (EEG) features; with progressive intellectual disability and behavioural problems that may both present later (3). Anti-epileptic drug (AED) therapy is the usual first-line treatment (4) however LGS seizures are difficult to control with medication and failure of a first drug will significantly reduce the likelihood of subsequent seizure freedom (5). Intractable seizures and adverse effects on cognitive and social functioning (6) will have a major impact on the health-related quality of life of those affected (7) with considerable NHS and social cost implications for supporting medical and educational needs. Lifelong persistence of LGS is usual (8) with poor outcomes continuing into adulthood and long-term dependence on caregivers for daily living abilities (9).

In view of these challenges it is important that all therapy options for the LGS child are explored. Non-pharmacologic treatments include vagal nerve stimulation and dietary therapy (4, 10). The ketogenic diet (KD) is a high fat, restricted carbohydrate dietary regime that has been used as a treatment for epilepsy since the 1920s, newer modified variants include the modified Atkins diet (MAD). Efficacy of these diets has been demonstrated in many prospective studies and randomised trials (11, 12, 13).

Retrospective reviews show KD to be efficacious for treatment of LGS. One study of 71 LGS patients from USA reported 51% had over 50% seizure reduction after 6 months on KD with 23% experiencing over 90% reduction and similar results after 12 months (14). Another study of 47 LGS patients from China reported over 50% seizure reduction in 49% after 3 months on KD with four becoming seizure free after 6 months; response to diet was associated with positive EEG changes (15). A review of 25 LGS children treated with MAD in India reported a similar responder rate: 48% with over 50% seizure reduction after 3 months and 44% after 6 months (16). In a prospective study of 20 Argentinian LGS children on KD, seizures were reduced by over 50% in eight (40%) after 18 months with three seizure free (17). A literature review of 18 different studies with outcome data on 189 LGS children reported 88 (47%) with over 50% seizure reduction after 3-36 months on KD (14). One Korean study has assessed long term outcomes of 68 LGS patients over a mean follow up period of 19 years: of the 19 who tried diet therapy, 5 maintained seizure freedom on classical KD and one on MAD (18). The same group reported diet therapy to be efficacious and feasible in a recent review of 20 LGS patients with mitochondrial dysfunction (19).

International consensus recommendations suggest that the KD should be strongly considered in a child with epilepsy which has failed to respond to two or three AEDs (20). Guidelines (NICE) on management of the epilepsies in the UK also suggest that children and young people with epilepsy whose seizures have not responded to appropriate AEDs are referred to a tertiary paediatric epilepsy specialist for consideration of the KD (CG137, nice.org.uk). In view of these recommendations, we would propose that all children with LGS who have failed
appropriate AED therapy are funded for an initial assessment of suitability for dietary therapy. Children who start KD will require a minimum of three months on treatment to allow adequate assessment of benefit and appropriate fine-tuning of the diet prescription to individual needs; it is recommended that successful treatment is continued for at least two years during which the potential for weaning AEDs will be regularly reviewed.

References: