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). Seizures are severe and may range in type but LGS is characterised by tonic ‘drop attacks’ (3) which can cause frequent injuries. Anti-epileptic drug (AED) therapy is usually commenced as a treatment for epilepsy after presentation of two or more unprovoked seizures; however LGS is difficult to control with medication and failure of a first drug will significantly reduce the likelihood of subsequent seizure freedom (4). This will have considerable cost implications for NHS services due to a child’s need for regular clinical review, ongoing medications (both routine and emergency), support of other therapies and hospital admissions. LGS is usually accompanied by learning disability which will require educational assessment and support. The intractable seizures and associated cognitive and behavioural problems will have a major impact on the health-related quality of life of the affected children as well as their caregivers (5).

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 (6). The ketogenic diet is a high fat, restricted carbohydrate dietary regime that has been used as a treatment for epilepsy since the 1920s; efficacy has been demonstrated in many prospective studies and a randomised controlled trial (7). Modified variants include the modified Atkins diet (MAD). Retrospective reviews have shown the ketogenic diet (8, 9, 10) and MAD (11) to be efficacious for the treatment of LGS. In a prospective study of twenty LGS children on the ketogenic diet, seizures were reduced by over 50% in eight (40%) after 18 months (12). A literature review of 189 LGS children reported 88 (47%) with greater than 50% seizure reduction after 3-36 months on the ketogenic diet (8). One 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 the traditional ketogenic diet and one on MAD (13).

International consensus recommendations suggest that the ketogenic diet should be strongly considered in a child with epilepsy which has failed to respond to two or three AEDs (14). 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 use of a ketogenic diet (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 ketogenic dietary therapy. Children who start a diet will require a minimum of three months on treatment to allow adequate assessment of benefit and appropriate fine-tuning of the 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: