Clinical commentary

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The ketogenic diet in two paediatric patients with refractory myoclonic status epilepticus

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The term “myoclonic status epilepticus” is used for a wide range of electroclinical presentations with different prognostic and treatment implications. Myoclonic status epilepticus has been described in generalized epilepsy syndromes, neurodegenerative disease, infectious or inflammatory neurological disease, toxic-metabolic states, and following anoxic brain injury.

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In this study, we evaluated the efficacy and tolerability of the KD in two patients who met the diagnostic criteria for pharmacoresistant myoclonic SE.

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Case report 1

- The patient was a 23-month-old boy, born after a full-term pregnancy with a normal weight for gestational age. His perinatal history was unremarkable. Both parents had a history of febrile seizures.

- Developmental milestones and neurological examination were according to age, up to one year of life.

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- Based on the electroclinical features, the patient was considered to have myoclonic epilepsy of unknown aetiology.

- At two years and five months of age, the patient was admitted with a repetitive myoclonic SE characterized by erratic and generalized jerks. The patient lost independent gait, however, during the period without myoclonic status epilepticus, his motor development improved significantly. The ictal video-EEG showed continuous diffuse and asymmetric spike-and-polyspike waves associated with massive myoclonias

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-The patient was refractory to different AEDs, such as midazolam, levetiracetam, ethosuximide, clobazam, topiramate, as well as corticosteroids which were tried for two weeks.

-Therefore, at two years and six months of age, the KD was started at a 4:1 ratio and glucose administration was discontinued, including that by intravenous infusion. After a 24-hour fast, the patient was administered a commercial preparation (KetoCal [KetoCal, SHS]) via a nasogastric tube. Urine ketosis was monitored daily with Labstix. The urine ketone strip measured 4 (++++) throughout the dietary treatment.

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- During the first week on the KD, the patient slowly improved, achieving a 75-90% seizure reduction, with less EEG abnormalities. Since then, his neuropsychological performance, but mainly his motor development, has significantly improved. The child has recovered independent gait.

- After one year on the diet, the patient has isolated myoclonias. He is successfully attending kindergarten. The last control EEG showed occasional generalized spike-and-polyspike waves.

- The number of AEDs has been reduced to one (valproic acid). The diet was switched to the classic oral diet at three years of age.

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Case report 2

- The patient was a 17-month-old boy. He was born after an uneventful, full-term pregnancy with a normal weight for gestational age. His family history was unremarkable.

- On the first day of life, he started with recurrent refractory status epilepticus requiring mechanical ventilation. He received valproic acid, vigabatrin, and levetiracetam, with poor response.

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-At one year of age, the patient was referred to our centre due to progressive encephalopathy and myoclonic epilepsy.

- On neurological examination, slow growth of head circumference, generalized hypotonia, and pyramidal signs were observed. Cognitive development was impaired. Fundoscopy was normal.

- EEG recordings showed multifocal spikes and diffuse fast spike-and-polyspike waves that increased during sleep. Intermittent photic stimulation was negative. The background activity slowly became impaired.

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- At that time, brain MRI showed bilateral hypointensity in the white matter on T1-weighted and hyperintensity on T2-weighted images with mild ventricular asymmetry, mega cisterna magna extending to both cerebellar hemispheres, and mild cerebellar hypoplasia.

- Neurometabolic investigations, including blood, urine, and CSF analysis, as well as karyotyping, were normal.

-The myoclonic episodes were refractory to classic and new oral AEDs. The video-EEG showed continuous diffuse spike-and-wave paroxysms associated with subintrant multifocal and erratic myoclonias.

- The boy was admitted to the ICU because of refractory myoclonias, and intravenous levatiracetam, valproic acid, benzodiazepines, and corticosteroids were tried without response over three weeks before starting the KD.

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-The KD was begun at a 4:1 ratio and glucose administration was discontinued, including that by intravenous infusion. After 24 hours of fasting, the child was given a commercial preparation (KetoCal [KetoCal, SHS]) via a nasogastric tube. Urine ketosis was monitored daily with Labstix. The patient maintained a level of urine ketones of 3 (+++) or 4 (++++) throughout the treatment. Plasma ketone bodies were not measured.

- One week after KD initiation, the patient slowly improved achieving a 50% seizure reduction.

- He was discharged from the ICU and two weeks later from the hospital. He had become more alert and his eye contact improved.

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-He had become more alert and his eye contact improved.

- After a six-month follow-up, in spite of the progressive encephalopathy, his quality of life improved and the AEDs were reduced to levetiracetam and topiramate.

- The patient continues to receive the same formula orally.

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Conclusion

-The KD is a promising therapy for myoclonic SE. It should be considered as an alternative option in the management of this type of SE, regardless of the aetiology.

- In patients with refractory myoclonic SE, the KD should be tried earlier in the course of the treatment.

- When good seizure control is achieved, the use of the diet may avoid cognitive deterioration and behavioural disturbances.

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