Database Establishment of Ketogenic Diet Clinic in Ain Shams University Children's Hospital

Thesis

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Wafaa Mostafa Abdelwahab Shokair

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Abb.	Full term
AA	*Arachidonic acid*
ADEAF	*Autosomal dominant epilepsy with auditory features*
ADNFLE	*Autosomal-dominant nocturnal frontal lobe epilepsy*
AE	*Adverse events*
AED	*Antiepileptic drug*
BDH1	*BHB dehydrogenase*
BECTS	*Benign epilepsy with centrotemporal spikes*
BFNE	*Benign familial neonatal epilepsy*
BHB	*Beta – hydroxy- Butarate*
BMI	*Body mass index*
BNS	*Benign neonatal seizures*
CAE	*Childhood absence epilepsy*
CPT	*Carnitine palmitoyl ltransferase*
CSWS	*Continuous spike-and-wave during sleep*
DHA	*Docosahexanoic acid*
EEG	*Electroencephalogram*
EME	*Early myoclonic encephalopathy*
ESES	*Electrical Status Epilepticus in Sleep*
FIRES	*Febrile infection–related epilepsy syndrome*
FS	*Febrile seizures*
FS+	*Febrile seizures plus*
GA	*Gestational age*
GABA	*Gamma-amino butyric acid*
GI	*Glycemic index*
GLUT-1	*Glucose Transporter Protein 1*

List of Abbreviations cont...

Abb.	Full term
Glut1DS	*Glucose transporter protein 1 (Glut-1) deficiency syndrome*
GSH	*Glutathione*
HMG-CoA	*3-hydroxy-3-methylglutaryl CoA*
ICIDH-2	*International Classification of Functioning, Disability and Health*
ILAE	*International League against Epilepsy*
JAE	*Juvenile absence epilepsy*
JME	*Juvenile myoclonic epilepsy*
KD	*Ketogenic Diet*
LCAD	*Long-chain acyl dehydrogenase deficiency*
LGIT	*Low glycemic index treatment*
LKS	*Landau-Kleffner syndrome*
LMIC	*Low and middle-income countries*
MAD	*Modified Atkins diet*
MCAD	*Medium-chain acyl dehydrogenase deficiency*
MCT	*Medium-chain triglyceride*
MEG	*Magneto-encephalography*
MEI	*Myoclonic epilepsy in infancy*
MRI	*Magnetic resonance image*
MTLE with HS	*Mesial temporal lobe epilepsy with hippocampal sclerosis*
NAD^+	*Nicotinamide adenine dinucleotide*
NE	*Norepinephrine*
NPY	*Neuropeptide-Y*
NS	*Non significant*
PDH	*Pyruvate dehydrogenase*

List of Abbreviations cont...

Abb.	Full term
PDHD	*Pyruvate dehydrogenase deficiency*
PME	*Progressive myoclonus epilepsies*
PUFAs	*Polyunsaturated fatty acids*
ROS	*Reactive oxygen species*
S	*Significant*
SANAD	*Standard versus New Antiepileptic Drugs*
SCAD	*Short-chain acyl dehydrogenase deficiency*
SRS	*Stereotactic radio-surgery*
SSPE	*Subacute sclerosing panencephalitis*
UCPs	*Uncoupling proteins*
VNS	*Vagus nerve stimulation*

INTRODUCTION

Epilepsy is a disorder that occur in about 1% of population and the onset of epilepsy in 60% of cases start in childhood ***(Armeno et al., 2014).***

Epidemiological data indicate that 20-30% of patient will become refractory to therapy. Refractory epilepsy is defined as seizures that cannot be controlled with at least two first line antiepileptic drugs in adequate doses, as single or combined drug therapy ***(Freeman et al., 2007).***

The ketogenic diet, a non-drug treatment had proven its effectiveness in treatment of epilepsy in children in the past decade especially in management of refractory epilepsy. The ketogenic diet is highly effective and reduce the incidence of seizures by 50%in a half of patient, and 90% in one third of patients ***(Lee and Kossoff, 2011).***

The ketogenic diet is a high fat, low carbohydrate, adequate protein diet that cause ketosis and leads to metabolic state that resemble the fasting state ***(Neal et al., 2008).*** It works through multiple mechanisms that target a specific biochemical pathways linked to cell substrate (e.g, ion channel) and mediators responsible for neuronal hyperexcitability. It is also thought that the ketone bodies have direct anticonvulsant effect ***(Rho and Neuroscil, 2015).***

The classical ketogenic diet is considered the treatment of choice for patient with a glucose transporter protein type 1 (GLUT1) deficiency or a pyruvate dehydrogenase (PDH) deficiency ***(Nangia, 2012).***

Its use in Egypt has been started since 2011and full publication on 2013 ***(El-Rashidy et al., 2013).***

It is also important to exclude clinical condition for which the ketogenic diet is contraindicated (e.g, disorder of fatty acid oxidation, disorder of fatty acid transport, pyruvate carboxylase deficiency and porphyria) and assess risk factors that may complicate the use of ketogenic diet (e.g, gastroesophageal reflux ***(Kossoff et al., 2009).***

The more common complications are metabolic acidosis and gastrointestinal manifestations, such as abdominal pain, nausea and vomiting with a risk of dehydration and hypoglycemia especially in patients who remain fasting for an extended period of time. Less common, but very important, effects because of their difficult management include eating disorders, such as loss of appetite, fluid rejection, and self-induced vomiting ***(Ballaban et al., 1998).***

The ketogenic diet is a meal plan with an unbalanced intake of micro- and macronutrients; it may result in energy, protein, mineral and vitamin deficiency and excessive lipid intake, with a risk of unwanted side effects. However, its