

Letter to the Editor

An effective medical replacement therapy: ketogenic diet for intractable childhood epilepsy

Sir,

Ketogenic diet (KD) a high fat, adequate protein and low carbohydrate restrictive diet has a long history of its use in intractable epilepsy of childhood. The diet produces biochemical changes mimicking that of starvation. The high levels of ketone bodies produced by KD act as a major source of energy for brain replacing the usual glucose.¹ Comprising the ratio of 4:1 (fat:carbohydrate and protein) by weight, the diet produces state of ketonemia or ketosis that leads to reduction in frequency of epileptic seizures by its unique mode of action. To increase the palatability medium chain triglycerides (as coconut oil) in ratio of 3:1 is used which is more efficiently absorbed and have lesser gastro intestinal side effects as compared to traditional 4:1 ratio diet with long chain triglycerides like PUFA.²

The diet is indicated as an adjuvant therapy in children (2-10 year) with intractable epilepsy and is generally effective in all type of seizure semiology especially atypical absence, myoclonic abd atonic. The difficult to treat epilepsy syndrome mainly tuberous sclerosis, infantile spasm and Lennox-Gastaut Syndrome may respond well to KD if instituted early in course of disease. The diet is considered to be lifesaving in children with glucose transporter GLUT-1 deficiency and in pyruvate dehydrogenase deficiency.³ KD is contra indicated in mitochondrial disorders, fatty acid oxidation defects, pyruvate carboxylase and carnitine deficiency thus justifying the detailed biochemical assessment before institution of KD.

KD induces its antiepileptic response by changing the basic physio-biochemistry of central nervous system by activation of ATP dependent potassium pump, adenosine upregulation, inhibition of excitatory glutamatergic transmission, synaptic inhibition of cortical motor pathways etc. The low calories intake & excess ketone bodies further accentuate the response of KD.⁴

Due to its unpleasant and bland taste and strict dietary protocol, the institution of KD is a challenge in young children. KD is not a supplementary diet so no carbohydrate rich food like cold drink, biscuit & chocolates is permitted to the recipient children. Under supervision (preferably in a hospital after attaining a 24-72 hrs state of fasting) the KD is initiated gradually from 1:1 to 4:1 ratio.

As KD is not a balanced diet so essential vitamins & minerals are supplemented to maintain adequate growth. The clinical response is generally seen within 5-7 days but it takes around 3 months to produce its optimal anticonvulsant effect. The commonly encountered side effects in initial period includes nausea, vomiting, constipation, diarrhoea and hypoglycemia.

The occurrence of hypoglycemia early in course of institution of KD pose a real therapeutic challenge as correction of hypoglycemia by intravenous glucose leads to immediate loss of ketosis with resultant breakthrough seizure. During the maintenance phase of KD weight loss, growth retardation, hyperlipidemia, hypoproteinemia, pancreatitis are seen more often as complication.⁵

The antiepileptic drug in use should be continued with KD with aim to gradually reduce or even withdraw over a period of 2 years or until the patient is seizure free off medications for a year. Amongst all antiepileptics sodium valproate is more effective with KD and has synergistic effect.⁶ The clinical efficacy of KD is variable and depends upon number of factors like compliance, seizure semiology, time of institution etc. Around 30% children show more than 90% reduction in seizure frequency, 20% show complete seizure control while remaining 50% show substantial decrease in seizure frequency.⁷ The associated improvement in behaviour, cognition, sleep pattern etc. further add to the benefits of KD as against multiple antiepileptics.

The unique mode of action of KD expand its horizon as future implication in various diseases like parkinson's disease, alzheimer disease, motor neuron disease, diabetes mellitus apart from childhood epilepsy.

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