

Medical Policy



Title: Ketogenic Diet as a Treatment of Refractory Epilepsy

Professional

Original Effective Date: January 12, 2009

Revision Date(s): March 8, 2010;

February 28, 2014

Current Effective Date: January 12, 2009

Institutional

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DESCRIPTION

The ketogenic diet is a high-fat, low-carbohydrate, low-protein diet that has been used to treat epilepsy. The composition of the diet induces ketosis, a physiologic state in which circulating ketone bodies are used as the primary fuel source in the absence of simple sugars. Ketosis may inhibit seizures through an unknown mechanism. The diet was developed in the 1920s, but used infrequently subsequent to the introduction of anti-epileptic drugs. Over the last several decades, the ketogenic diet has gained attention as a treatment option in patients with epilepsy that is refractory to medications.

The ketogenic diet is quite restrictive, requiring the cooperation of the patient, family, and an appropriately trained dietitian. The ratio of fat to carbohydrates must be strictly maintained, meaning that the precise contents of each food item must be known, and exactly measured. Given the restrictions, compliance with the diet can be problematic,

especially in children over 10 years of age who have well-established dietary habits and preferences.

As currently practiced, the diet is initiated in the hospital setting. Children are admitted to the hospital and fasted for 1 to 2 days. The diet is then instituted gradually over a number of days. A full ketogenic diet is attained by approximately day 5 in most children, at which time the patient is discharged home and followed up as an outpatient. The main reason for hospitalization is the period of fasting. Fasting potentially exposes children to dehydration and metabolic derangements that could become life threatening if not properly monitored and treated.

POLICY

Hospitalization for initiation of a ketogenic diet may be considered **medically necessary** in the treatment of children with refractory epilepsy.

RATIONALE

This policy is based on a 1998 TEC Assessment (1) that offered the following conclusions:

- While the published data regarding ketogenic diets consists of uncontrolled case series, the data are consistent in showing that some children benefit from the ketogenic diet, as demonstrated by a significant reduction in seizure frequency; i.e., complete cessation of seizures in 16% of children, greater than 90% reduction in 32%, and a greater than 50% reduction in 56%.
- These results exceed any expected placebo effect or spontaneous remission of seizures.

As currently practiced, the ketogenic diet is typically initiated in an inpatient setting, principally to monitor the patient during the initial fasting period, but also presumably to provide the intense education required to maintain a ketogenic diet once discharged. There are currently no data that focus on initiating the diet in the outpatient environment. However, the published studies do not explicitly delineate the adverse effects that occurred during the inpatient stay and whether their management required hospitalization. Another possibility is the gradual initiation of the diet such that fasting (and hospitalization) would not be required. This approach should, in principle, achieve the same end point of ketosis, although over a longer time period. However, it is also possible that the fast itself is responsible for some degree of response seen in the published studies.

As part of the TEC Assessment, 14 programs offering ketogenic diets were surveyed. Thirteen of the 14 programs reported that they always or virtually always instituted the diet in the inpatient setting. Four programs reported that they would rarely institute the diet in the outpatient setting under special circumstances, such as when it was being restarted after a period off the diet. One program reported that they routinely initiate the diet in the outpatient setting and that they feel that their results were comparable to other centers that followed the inpatient protocol. This program reported that they have neither published any data on their outcomes nor formally presented outcome data in any scientific forums. While these data do not represent a comprehensive catalogue of practice patterns, it is clear that the most common approach is initiation of the diet in the inpatient setting, but that there are instances in which the diet has been successfully initiated in the outpatient setting.

A literature search was performed for the period of 1998 through November 2004 with a focus on outpatient initiation of therapy. At the time of the TEC assessment, there were no studies that examined the safety and efficacy of initiating the diet in an outpatient setting. However, in 2004, Vaisleib and colleagues reported on a case series of 37 patients who underwent outpatient induction of the ketogenic diet, whose outcomes were compared retrospectively to those who underwent inpatient dietary induction. (2) The mean age of the patients was 6.6 years, with a range of 1.8 to 14 years. The authors reported that there was no evidence that inpatient initiation of the ketogenic diet was superior to outpatient initiation. Additional studies identified in the literature search focused on the long-term effects of the ketogenic diet on the growth and development of children (3-5), and whether or not the less restrictive Atkins diet, which also produces a mild ketosis, is option to the ketogenic diet. (6)

CODING

The following codes for treatment and procedures applicable to this policy are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

CPT/HCPCS

No Specific Code

ICD-9 Diagnoses

345.01	Generalized nonconvulsive epilepsy, with intractable epilepsy
345.11	Generalized convulsive epilepsy, with intractable epilepsy
345.41	Localization-related (focal) (partial) epilepsy and epileptic syndromes with complex partial seizures, with intractable epilepsy
345.51	Localization-related (focal) (partial) epilepsy and epileptic syndromes with simple partial seizures, with intractable epilepsy
345.61	Infantile spasms, with intractable epilepsy
345.71	Epilepsia partialis continua, with intractable epilepsy
345.81	Other forms of epilepsy and recurrent seizures, with intractable epilepsy
345.91	Epilepsy, unspecified, with intractable epilepsy

ICD-10 Diagnoses (Effective October 1, 2014)

G40.311	Generalized idiopathic epilepsy and epileptic syndromes, intractable, with status epilepticus
G40.319	Generalized idiopathic epilepsy and epileptic syndromes, intractable, without status epilepticus
G40.211	Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, intractable, with status epilepticus
G40.219	Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, intractable, without status epilepticus
G40.011	Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, intractable, with status epilepticus
G40.019	Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, intractable, without status epilepticus

G40.111	Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, intractable, with status epilepticus
G40.119	Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, intractable, without status epilepticus
G40.411	Other generalized epilepsy and epileptic syndromes, intractable, with status epilepticus
G40.419	Other generalized epilepsy and epileptic syndromes, intractable, without status epilepticus
G40.803	Other epilepsy, intractable, with status epilepticus
G40.804	Other epilepsy, intractable, without status epilepticus
G40.823	Epileptic spasms, intractable, with status epilepticus
G40.824	Epileptic spasms, intractable, without status epilepticus
G40.89	Other seizures
G40.A11	Absence epileptic syndrome, intractable, with status epilepticus
G40.A19	Absence epileptic syndrome, intractable, without status epilepticus
G40.B11	Juvenile myoclonic epilepsy, intractable, with status epilepticus
G40.B19	Juvenile myoclonic epilepsy, intractable, without status epilepticus
G40.911	Epilepsy, unspecified, intractable, with status epilepticus
G40.919	Epilepsy, unspecified, intractable, without status epilepticus

REVISIONS

03-08-2010	<p>In the policy section: The policy wording was changed From: "A ketogenic diet may be considered medically necessary in children with refractory epilepsy when initiated during an inpatient hospital stay." To: "Hospitalization for initiation of a ketogenic diet may be considered medically necessary in the treatment of children with refractory epilepsy." <ul style="list-style-type: none"> ▪ The policy intent was not changed, the wording was changed to more clearly reflect that hospitalization to initiate a ketogenic diet for children with refractory epilepsy is considered medically necessary. </p>
02-28-2014	<p>Medical Policy Description, Rationale, and References reviewed with no changes.</p> <p>In Coding section: <ul style="list-style-type: none"> ▪ Removed ICD-9 Diagnoses Codes: 345.00, 345.10, 345.2, 345.3, 345.40, 345.50, 345.60, 345.70, 345.80, 345.90 ▪ ICD-10 Diagnoses Codes added. </p>

REFERENCES

1. TEC Assessment, 1998; Tab 20
2. Vaisleib II, Buchhalter JR, Zupanc ML. Ketogenic diet: Outpatient initiation, without fluid, or caloric restrictions. *Pediatr Neurol* 2004; 31:198-2002
3. Kwiterovich PO, Vning EP, Pyzik P. Effect of a high-fat ketogenic diet on plasma levels of lipids, lipoproteins, and apolipoproteins in children. *JAMA* 2003; 290:912-20.
4. Liu YM, Williams S, Basualdo-Hammond C et al. A prospective study; growth and nutritional status of children treated with the ketogenic diet. *J Am Diet Assoc* 2003;103:707-12.
5. Vining EP, Pyzik P, McGrogan J et al. Growth of children on the ketogenic diet. *Dev Med Child Neurol* 2002; 44:796-802.
6. Kossoff EH, Krauss GL, McGrogan JR, Freeman JM. Efficacy of the Atkins diet as therapy for intractable epilepsy. *Neurology* 2003; 61:1789-91.