

Date: \_\_\_\_\_

Patient Name: \_\_\_\_\_

DOB: \_\_\_\_\_

Insurance Company Name: \_\_\_\_\_

Insurance ID: \_\_\_\_\_

Relevant ICD-9 Codes\*: \_\_\_\_\_

<b>Letter of Medical Necessity</b>
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To Whom It May Concern:

I am requesting insurance coverage and reimbursement for my patient, \_\_\_\_\_, who has been diagnosed with \_\_\_\_\_ and for whom I have prescribed the use of **Liquigen**<sup>®</sup> (manufactured by SHS International and distributed by Nutricia North America).

**Liquigen** is a medical food for conditions where medium chain triglycerides (MCTs) are part of dietary management, such as long-chain fatty acid oxidation disorders, conditions managed by the ketogenic diet, or fat malabsorption disorders.

- **Long-chain fatty acid oxidation disorders (FAODs)** are a group of rare inherited inborn errors of metabolism. Individuals with long-chain FAODs are unable to use dietary fat (long chain fat) for energy. The inability to break down fats for energy and the buildup of fatty acids causes serious health problems. MCTs are metabolized differently than long chain fats and can be used as alternative fat source in patients with long-chain FAODs.
- The **ketogenic diet** is used for the dietary management of intractable epilepsy, as well as for certain disorders of carbohydrate metabolism, including Glucose Transporter Type 1 Deficiency (Glut-1 Deficiency) and Pyruvate Dehydrogenase Complex Deficiency (PDCD). This very restrictive high fat, low carbohydrate diet causes the body to burn fat rather than carbohydrate for energy. The metabolism of fat results in the production of ketones, which provide an alternative energy source to the brain. Because MCTs are more “ketotic” than dietary fat, meaning that they produce more ketones, they are often used to boost ketone production for patients on a ketogenic diet.
- MCTs may be used for patients with **malabsorption disorders** who need calories but are unable to absorb or metabolize dietary fats. The unique structure of MCTs allows them to be easily absorbed in the gastrointestinal tract. Most fats are broken down in the intestine and remade into a special form that can be transported in the blood. But MCTs are absorbed intact and taken to the liver, where they are used directly for energy.
- MCTs are sometimes used for patients with **liver disease** who do not produce sufficient bile. In the absence of bile, fats become indigestible and are instead excreted in feces, a condition called steatorrhea. This can lead to deficiencies in essential fatty acids and fat-soluble vitamins.

Although some foods contain small amounts of MCTs, isolated MCT oil is not found naturally and must be provided by a special medical food. **Liquigen** is an ideal product for providing MCTs because it is a unique emulsion of 50% MCT oil and 50% water, which helps improve tolerance and palatability. Most importantly, **Liquigen** blends well with formulas and stays in suspension, which makes it ideal for patients who are tube-fed, as standard oils will separate in the feeding bag.

**Liquigen** is not a drug but is considered a “Medical Food”, which is not available over the counter and is used only under strict medical supervision. **Liquigen** has been prescribed and is medically necessary as the optimum treatment for my patient.

Sincerely,

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*Signature*

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*Name*

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*Title*

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*Provider Number*

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*Center/Hospital/Institution/Practice*

**Prescribed amount:** \_\_\_\_\_ mL per \_\_\_\_\_ (day/month).

**Product and Reimbursement Information for Liquigen**

Product Code	Packaging	Calories per Bottle	Reimbursement Code	HCPCS Code
71957 (Case)	3 x 4 x 250 mL (8.5 fl oz)	1125	49735-011957	B4155
71957-3 (Unit)	4 x 250 mL (8.5 fl oz)	1125	49735-019573	B4155

**Examples of Relevant Diagnosis and ICD-9 Codes**

Diagnosis	ICD-9 Code*
Epilepsy, unspecified, with intractable epilepsy	345.91
Fatty Acid Oxidation Disorders	277.85
Malabsorption (Fat)	579.8
Unspecified disorder of carbohydrate transport and metabolism.	271.9
Other specified disorders of carbohydrate transport and metabolism	271.8

## A Selection of Clinical References for the Usage of Medium-chain Triglycerides (MCTs) in the Nutritional Management of Various Medical Conditions

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### **Ketogenic diet for epilepsy:**

- Liu YM, Wang HS. Medium-chain triglyceride ketogenic diet, an effective treatment for drug-resistant epilepsy and a comparison with other ketogenic diets. *Biomed J*. 2013 Jan-Feb;36(1):9-15.
- Neal EG, Chaffe H, Schwartz RH, Lawson MS, Edwards N, Fitzsimmons G, Whitney A, Cross JH. A randomized trial of classical and medium-chain triglyceride ketogenic diets in the treatment of childhood epilepsy. *Epilepsia*. 2009 May;50(5):1109-17.
- Liu YM. Medium-chain triglyceride (MCT) ketogenic therapy. *Epilepsia*. 2008 Nov;49 Suppl 8:33-6.
- Neal EG, Chaffe HM, Edwards N, Lawson MS, Schwartz RH, Cross JH. Growth of children on classical and medium-chain triglyceride ketogenic diets. *Pediatrics*. 2008 Aug;122(2):e334-40.
- Liu YM, Wang HS. Medium-chain triglyceride ketogenic diet, an effective treatment for drug-resistant epilepsy and a comparison with other ketogenic diets. *Biomed J*. 2013 Jan-Feb;36(1):9-15.
- Schwarz RH, Eaton J, Bower BD, Aynsley-Green A. Ketogenic diets in the treatment of epilepsy: short-term clinical effects. *Dev Med Child Neurol* .1989;2:145-51.

### **Disorders of long-chain fat metabolism:**

- Behrend AM, Harding CO, Shoemaker JD, Matern D, Sahn DJ, Elliot DL, Gillingham MB. Substrate oxidation and cardiac performance during exercise in disorders of long chain fatty acid oxidation. *Mol Genet Metab*. 2012 Jan;105(1):110-5.
- Gillingham MB, Scott B, Elliott D, Harding CO. Metabolic control during exercise with and without medium-chain triglycerides (MCT) in children with long-chain 3-hydroxy acyl-CoA dehydrogenase (LCHAD) or trifunctional protein (TFP) deficiency. *Mol Genet Metab*. 2006 Sep-Oct;89(1-2):58-63.
- Lund MA, Dixon PV, Leonard JV, Morris AAM. What is the role of medium-chain triglycerides in the management of long chain 3-hydroxyacyl-CoA dehydrogenase deficiency? *J Inherit Metab Dis* 2003;26: 353-360.
- Morris AAM, Turnbull DM. Fatty acid oxidation defect in muscle. *Curr Opin Neurol* 1998; 11:485-90.

### **Short bowel syndrome:**

- Tandon RK, Rodgers JB Jr, Balint JA. The effects of medium chain triglycerides in the short bowel syndrome. Increased glucose and water transport. *Am J Dig Dis* 1972;17:233-8.
- Thompson JM, Brett A, Rose S. Dietary management of intestinal lymphangiectasia complicate by short gut syndrome. *Human Nutrition: Applied Nutrition* 1986; 40A:136-140.
- Winawer SJ, Broitman SA, Wolochow DA, et al. Successful management of massive small-bowel resection based on assessment of absorption defects and nutritional needs. *N Engl J Med* 1966; 274:72-8.
- Bochenek W, Rodgers JB Jr, Balint JA. Effects of change in dietary lipids on intestinal fluid loss in the short bowel syndrome. *Ann Intern Med* 1970;72:205-13.

### **Intestinal lymphangiectasia**

- Tift W and Lloyd JK. Intestinal lymphangiectasia. *Arch Dis Child* 1975;50;269-276.

Thompson JM, Brett A, Rose S. Dietary management of intestinal lymphangiectasia complicate by short gut syndrome. *Human Nutrition: Applied Nutrition* 1986; 40A:136-140.

### **Malabsorption and chronic diarrhea:**

Wanke CA, Pleskow D, Degirolami PC, Lambi BB, Merkel K, Akrabawi S. A medium chain triglyceride-based diet in patients with HIV and chronic diarrhea reduces diarrhea and malabsorption: a prospective, controlled trial. *Nutrition*. 1996 Nov-Dec;12(11-12):766-71.

Thomas AG, Stanton RHJ, Miller V. Nutritional management of chronic diarrhoea and / or malabsorption. *J Pediatr Gastroenterol Nutr* 1990;11 (1)142-143.

Smallridge RC, Burman KD, Wartofsky L. Malabsorption of thyroxine, calcium, and vitamin D in a thyroparathyroidectomized woman: efficacy of therapy with medium-chain triglyceride oil. *Mil Med* 1990; 155(4):156-8.

### **Disorders of carbohydrate metabolism**

Moore R, et al. Long-chain 3-hydroxyacyl-coenzyme A dehydrogenase deficiency—diagnosis, plasma carnitine fractions and management in a further patient. *Eur J Pediatr*. 1993 May;152(5):433-6.

### **Cholestasis and liver disease:**

Beath S, Johnson T, Willis K et al. Superior absorption of medium chain triglycerides compared with conventional dietary long chain fats in children with chronic liver disease. *Proc Nutr Soc* 1993;52:252

Protheroe SM and Kelly DA. Cholestasis and end-stage liver disease. *Bailliere's Clinical Gastroenterology*. 1998;12(4):823-841.

### **Chylothorax:**

Machado JD, Suen VM, Marchini JS. Is oral nutritional therapy effective for the treatment of chylothorax? A case report. *Nutrition*. 2008 Jun;24(6):607-9.

Chan EH, Russell JL, Williams WG, Van Arsdell GS, Coles JG, McCrindle BW. Postoperative chylothorax after cardiothoracic surgery in children. *Ann Thorac Surg*. 2005 Nov;80(5):1864-70.

### **Pancreatic insufficiency:**

Caliari S, Benini L, Sembinini C, Gregori B, Carnielli V, Vantini I. Medium-chain triglyceride absorption in patients with pancreatic insufficiency. *Scand J Gastroenterol* 1996;31(1):90-4.

### **Biliary atresia:**

Cohen MI, Gartner LM. The use of medium chain triglycerides in the management of biliary atresia. *J Pediatr* 1971;79(3):379.