# Protocol: Hypoglycemia in CMD

## What is hypoglycemia?

Hypoglycemia may occur when the blood glucose levels are too low. This can be dangerous because glucose is the main source of energy for the body, especially the brain. Blood glucose levels normally range from 70-140 mg/dl (milligrams of glucose per deciliter of blood). Our bodies closely regulate glucose levels; however, in susceptible individuals (e.g. those with CMD), this regulation can be stressed beyond its limits and result in dangerously low blood sugar levels.

## Signs and Symptoms

Signs and symptoms of hypoglycemia may vary depending on the affected individual. Most common symptoms experienced are neurologic symptoms (i.e. fatigue, headache, decreased responsiveness), difficulty concentrating, loss of coordination, but may include sweating, irritability, and sudden behavior changes as well. If hypoglycemia reaches critical levels and remains uncorrected, seizures and strokes may also occur.

#### **First Steps**

If hypoglycemia is suspected at home, first steps include: immediate sugar intake (e.g. orange juice, milk, hard candy), initiating a feed via G-tube (if available), or oral glucose. If symptoms persist, then the affected individual should be taken to an emergency room for proper medical intervention. The affected individual should be promptly checked for low blood glucose levels by first responders to direct appropriate fluid management. The course of treatment would include restoring and maintaining a safe blood glucose level.

#### **Proactive Measures**

To avoid future episodes of hypoglycemia in children and adults with CMD, caregivers should ensure adequate food intake and close attention during illness when food intake may not keep up with body demands for glucose. Caregivers should also be more attentive when the affected individual is required to fast for long periods of time in preparation for blood tests or anesthesia. For children with recurring episodes of severe hypoglycemia, additional abortive interventions such as a glucagon shot may be helpful. These interventions should be discussed with your healthcare team.

# **Preparation for Surgery/Anesthesia**

Preparation for surgery and/or anesthesia requires a communicative team of doctors and caregivers who should be aware of the potential for hypoglycemia in CMD affected individuals. Fasting times should be minimized when possible and a careful monitoring system should be put in place to maintain safe blood glucose levels.

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