Sick Day & Emergency Protocols: Helpful and Potentially Harmful

Mark Korson, MD
VMP Genetics

WARNING!

- Some physicians are intimidated by patients / parents who know more about the condition than they do
- If you think things are not proceeding as they should in the ED setting:
  - Have the ED doc call your doc
  - Have your doc call the ED doc
- Have your doc run interference and call the ED *before* you get there

THE ATTRIBUTES OF A GOOD PROTOCOL?

Not every protocol letter is a practical or effective letter

Too specific

Overwhelming

Not updated

Too little information

Not specific enough

A BAD PROTOCOL…

- Gets ignored
- Introduces confusion
- Makes the reader (MD) nervous
- Causes the reader to withdraw or lose confidence that s/he can handle the case
- Can be obstructive to care

THE ATTRIBUTES OF A GOOD PROTOCOL

- Assumes the reader is not familiar with the disease
- Describes the patient's particular/unique issues
- Explains the issues clearly, in understandable language
- Breaks down the issue into parts
- Offers practical, common-sense advice, open-ended but not vague, addresses possible scenarios that can happen
- Should end with a name, a signature, a contact number, and a date. Printed on letterhead paper

EMERGENCY PROTOCOLS

OBJECTIVES OF AN EMERGENCY PROTOCOL

- Provides essential information to emergency providers in order to:
  - Teach
  - Direct
  - (And scare?)
- Provides the clinician with contacts for expert back-up

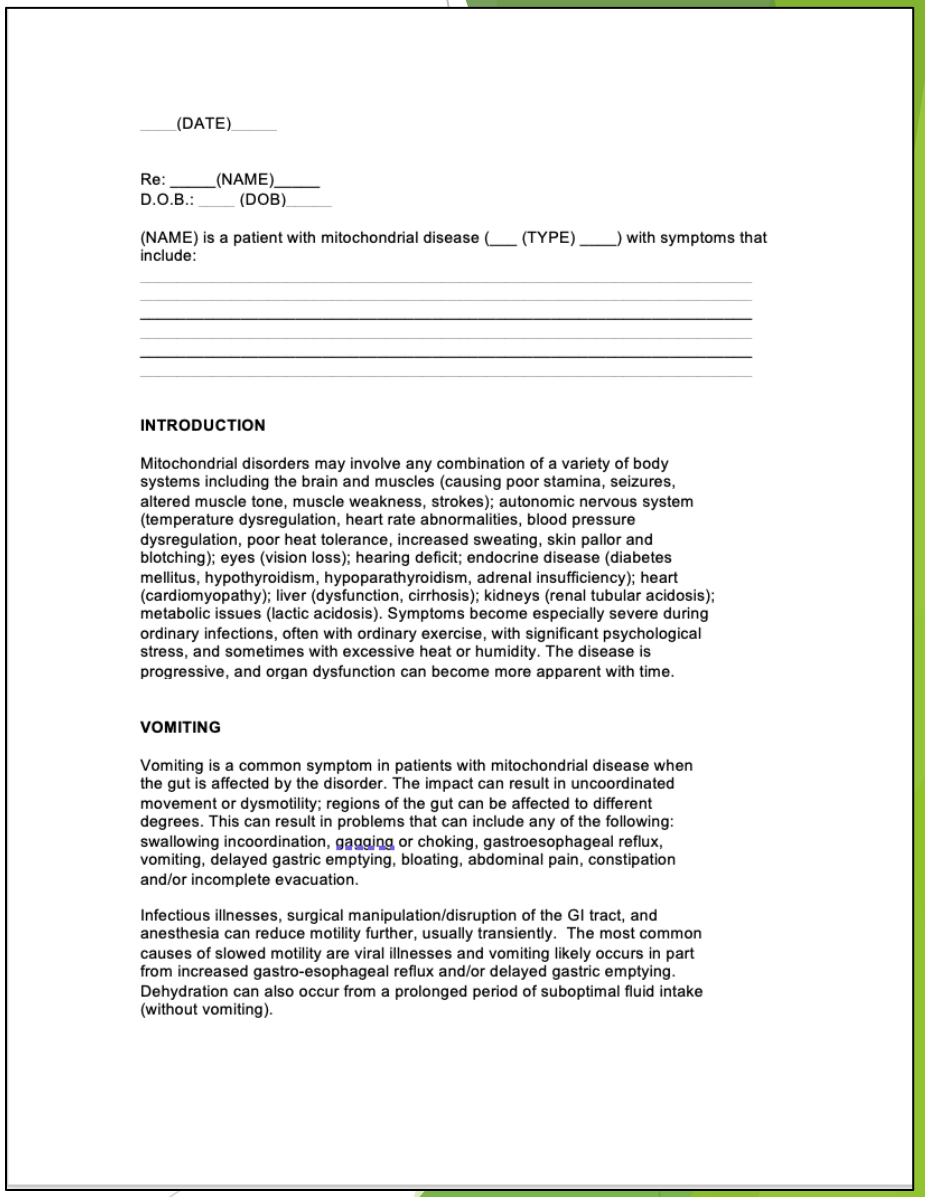
CONTENTS OF AN EMERGENCY PROTOCOL

- Information about:
  - Disease (general)
  - Disease (patient)
  - The problem(s) (include physiology)
  - Assessment
  - Considerations for treatment of the problem(s)
- Contact information

BEST PROTOCOLS ARE PATIENT + PROBLEM-SPECIFIC

TWO SAMPLE MITO PROTOCOLS

- ED – vomiting
- Anesthesia/surgery (GI surgery)



_____(DATE)_____

Re: _____(NAME)_____
D.O.B.: _____ (DOB)_____

(NAME) is a patient with mitochondrial disease (___ (TYPE) ____) with symptoms that include:

INTRODUCTION

Mitochondrial disorders may involve any combination of a variety of body systems including the brain and muscles (causing poor stamina, seizures, altered muscle tone, muscle weakness, strokes); autonomic nervous system (temperature dysregulation, heart rate abnormalities, blood pressure dysregulation, poor heat tolerance, increased sweating, skin pallor and blotching); eyes (vision loss); hearing deficit; endocrine disease (diabetes mellitus, hypothyroidism, hypoparathyroidism, adrenal insufficiency); heart (cardiomyopathy); liver (dysfunction, cirrhosis); kidneys (renal tubular acidosis); metabolic issues (lactic acidosis). Symptoms become especially severe during ordinary infections, often with ordinary exercise, with significant psychological stress, and sometimes with excessive heat or humidity. The disease is progressive, and organ dysfunction can become more apparent with time.

VOMITING

Vomiting is a common symptom in patients with mitochondrial disease when the gut is affected by the disorder. The impact can result in uncoordinated movement or dysmotility; regions of the gut can be affected to different degrees. This can result in problems that can include any of the following: swallowing incoordination, gagging or choking, gastroesophageal reflux, vomiting, delayed gastric emptying, bloating, abdominal pain, constipation and/or incomplete evacuation.

Infectious illnesses, surgical manipulation/disruption of the GI tract, and anesthesia can reduce motility further, usually transiently. The most common causes of slowed motility are viral illnesses and vomiting likely occurs in part from increased gastro-esophageal reflux and/or delayed gastric emptying. Dehydration can also occur from a prolonged period of suboptimal fluid intake (without vomiting).

VOMITING PROTOCOL – DATED

- _____(DATE)_____

  Re: _____(NAME)_____
  D.O.B.: _____ (DOB)_____

  (NAME) is a patient with mitochondrial disease (___ (TYPE) ____) with symptoms that include:

  _____
  _____
  _____
  _____
  _____
  _____

VOMITING PROTOCOL – PATIENT-SPECIFIC

_____(DATE)_____

Re: _____(NAME)_____
D.O.B.: _____ (DOB)_____

(NAME) is a patient with mitochondrial disease (___ (TYPE) ____) with symptoms that include:

_____
_____
_____
_____
_____
_____

MITO IS A PATIENT-SPECIFIC DISEASE

_____(DATE)_____

Re: _____(NAME)_____
D.O.B.: _____ (DOB)_____

(NAME) is a patient with mitochondrial disease (___ (TYPE) ____) with symptoms that include:

_____
_____
_____
_____
_____
_____

TEACHING ABOUT MITO

INTRODUCTION

Mitochondrial disorders may involve any combination of a variety of body systems including the brain and muscles (causing poor stamina, seizures, altered muscle tone, muscle weakness, strokes); autonomic nervous system (temperature dysregulation, heart rate abnormalities, blood pressure dysregulation, poor heat tolerance, increased sweating, skin pallor and blotching); eyes (vision loss); hearing deficit; endocrine disease (diabetes mellitus, hypothyroidism, hypoparathyroidism, adrenal insufficiency); heart (cardiomyopathy); liver (dysfunction, cirrhosis); kidneys (renal tubular acidosis); metabolic issues (lactic acidosis). Symptoms become especially severe during ordinary infections, often with ordinary exercise, with significant psychological stress, and sometimes with excessive heat or humidity. The disease is progressive, and organ dysfunction can become more apparent with time.

TEACHING ABOUT THE SYMPTOM

VOMITING

Vomiting is a common symptom in patients with mitochondrial disease when the gut is affected by the disorder. The impact can result in uncoordinated movement or dysmotility; regions of the gut can be affected to different degrees. This can result in problems that can include any of the following: swallowing incoordination, gagging or choking, gastroesophageal reflux, vomiting, delayed gastric emptying, bloating, abdominal pain, constipation and/or incomplete evacuation.

Infectious illnesses, surgical manipulation/disruption of the GI tract, and anesthesia can reduce motility further, usually transiently. The most common causes of slowed motility are viral illnesses and vomiting likely occurs in part from increased gastro-esophageal reflux and/or delayed gastric emptying. Dehydration can also occur from a prolonged period of suboptimal fluid intake (without vomiting).

TEACHING ABOUT THE SYMPTOM

Patients with autonomic dysregulation are also at risk for vascular dysautonomia in which they experience orthostatic changes in blood pressure and heart rate. An inadequate intake of fluids can result in dizziness or lightheadedness, syncope, and significant fatigue, and may be associated with chronic nausea and vomiting, as well as an increase in migraine frequency and severity.

WHAT TO DO?

ASSESSMENT

--Assess mental status.
--Assess cardio-respiratory status.
--Assess hydration status. Check last time tolerated fluids/feedings.
--Assess heart rate, blood pressure, look for orthostatic changes
--Look for fever, infection, or other physical stressor as a trigger for current symptom(s).
--Assess for changes compared to usual state/symptoms. More severe? Changes? New symptoms?
--Assess for acute biochemical abnormalities – electrolyte disturbances, hypoglycemia, blood gases/metabolic acidosis, hyperammonemia, liver dysfunction, CK.

TYPE OF IV FLUIDS

MANAGEMENT IN THE ED

The following guidelines are recommended for patients who are vomiting or who cannot take in adequate fluids PO/enterally:

1. An intravenous line should be placed and IV fluids provided;
2. IV fluids should contain ____ dextrose and electrolytes;
3. For patients with a history of fasting intolerance and/or documented hypoglycemia, or if there is secondary disturbance in fatty acid oxidation, IV fluids should contain 10% dextrose with electrolytes to run at 1.25x maintenance or higher. The higher glucose solution is necessary to minimize catabolism and flux through an impaired fatty acid oxidation pathway. 10% dextrose is more effective than 5% dextrose in accomplishing this goal;
4. The patient might require admission until s/he is able to tolerate consistently fluids by mouth/enterally.

LACTATED RINGER'S OK!

REFERENCE MATERIAL, TO TEACH

This reference provides comprehensive information about mitochondrial disease and its various systemic and organ-based issues: **Parikh S, et al. Patient care standards for primary mitochondrial disease: a consensus statement from the Mitochondrial Medicine Society. Genetics in Medicine 2017;19(12)**

WHOM TO CALL WITH QUESTIONS OR FOR HELP?

Please call with any questions.

_____, M.D.
Pager # _____

GI SURGERY LETTER - TEACHING

SURGICAL ISSUES

Patients with mitochondrial disease can tolerate surgery and anesthesia safely. However, procedures that impact the gut (at any level) can potentially destabilize these patients.

Mitochondrial disease is often associated with some degree of abnormal gut motility associated with uncoordinated movement or dysmotility; regions of the gut can be affected to different degrees. This can result in problems that can include any of the following: swallowing incoordination, gagging or choking, gastroesophageal reflux, vomiting, delayed gastric emptying, bloating, abdominal pain, constipation and/or incomplete evacuation. Infectious illnesses, surgical manipulation/disruption of the GI tract, and anesthesia can reduce motility further, usually transiently.

WHAT TO WATCH OUT FOR?

In the post-operative period, the oral intake of food and liquid diminishes and these patients are at risk for becoming dehydrated. This can occur for several reasons – pain at the surgical site (e.g., as with tonsillectomy), fatigue from the surgical procedure and/or anesthesia, or worsening gut motility with increased gastro-esophageal reflux and/or delayed gastric emptying. Refusal to eat or drink and poor calorie intake can exacerbate fatigue and result in a prolonged recovery and occasionally readmission.

WHAT TO CONSIDER?

MANAGEMENT

1. A pre-op appointment with Anesthesia should be scheduled in advance of the surgical date to give the team adequate time to review past courses of anesthetics, their efficacy and safety, and to make an appropriate plan for the upcoming procedure/surgery.
2. Elective procedures should be postponed if the patient develops any signs of infectious illness around the time of the procedure date;
3. Minimize the time necessary for fasting. The patient should be encouraged to take some fluids (orally or enterally) just before becoming NPO;
4. An intravenous line should be placed pre-operatively and fluids provided until the patient is eating/drinking well, or able to tolerate fluids through a g-tube if present;

FLUIDS

5. IV fluids should contain _____ dextrose and electrolytes;
6. For patients with a history of fasting intolerance and/or documented hypoglycemia, or if there is secondary disturbance in fatty acid oxidation, IV fluids should contain 10% dextrose with electrolytes to run at 1.25x maintenance or higher. The higher glucose solution is necessary to minimize catabolism and flux through an impaired fatty acid oxidation pathway. 10% dextrose is more effective than 5% dextrose in accomplishing this goal;
7. IV fluids should be continued until the patient is able to tolerate adequate volumes of fluids/food PO/enterally without vomiting;

SPECIAL IFs

8. If this patient demonstrates a refusal to eat or drink after 24 hours post-surgery, consideration should be given to providing 1-2 days of TPN which can improve calorie intake, reduce fatigue and related symptoms, and shorten the stay in hospital;
9. If the patient takes any vitamins as part of his/her mitochondrial management, these can be provided once PO/enteral fluids are tolerated;
10. For patients with respiratory insufficiency due to trunk muscle weakness, the use of a cough-assist to expand their lung capacity has been found to be beneficial.

REFERENCES… TO TEACH

If there are any questions regarding the safety of induction agents or other anesthetic medications, or other anesthesia concerns, please consult:

--Hsieh VC, Krane EJ, Morgan PG. Mitochondrial Disease and Anesthesia. J Inborn Errors of Metabolism & Screening 2017;5:1-5.

--Parikh S, et al. Patient care standards for primary mitochondrial disease: a consensus statement from the Mitochondrial Medicine Society. Genetics in Medicine 2017;19(12)

YOUR CHALLENGE – AN LBSL PROTOCOL

- Information about:
  - Disease (general)
  - Disease (patient)
  - The problem(s) (include physiology)
  - Assessment
  - Considerations for treatment of the problem(s)
- Contact information

BEST PROTOCOLS ARE PATIENT + PROBLEM-SPECIFIC

SICK-DAY PROTOCOLS

OBJECTIVES OF A SICK-DAY (HOME) PROTOCOL

- Keep the patient stable
- Keep the patient out of the hospital
- Start and change treatment plans more quickly

WHEN TO PROVIDE A HOME PROTOCOL?

- The patient's disease crises are not severe
- The course of the patient's disease crises are known and predictable
- The patient / parents:
  - Understand how to assess the patient's status
  - Understand the protocol – benefits, limitations
  - Are good communicators and reliable

MEDIUM CHAIN ACYL CoA DEHYDROGENASE (MCAD) DEFICIENCY HOME PROTOCOL

INTRODUCTION -

Your child has a metabolic disorder that is associated with acute episodes or crises when s/he can become acutely ill. If you are ever concerned about your child, call the Metabolism Clinic or page the Metabolism Physician-on-call. The team can help you decide whether your child can be treated at home or needs to be evaluated in the Emergency Department.

METABOLISM CLINIC NURSE – (111)-222-3333

METABOLISM PHYSICIAN-ON-CALL – (222)-333-4444, PAGER 123

You know your child better than anyone else. You know his/her typical routine and when s/he may be out of sorts. It is important that you pay close attention when s/he is not acting normally and convey these concerns to the Metabolism Clinic team.

You and the Metabolism Clinic staff function together as a team to best assess your child and determine a plan that is right for his/her wellbeing.

How has your child been eating?

- Has there been a change in the amount s/he is eating?
- Is s/he eating regularly during the day?
- How long is he going at night without any food or drink?
- Does s/he get a snack at bed-time or during the night? Is s/he still talking it?
- When was the last time s/he ate or drank and kept it down?

Is your child taking her/his medications?

- When was the last time s/he took them and kept it down?

TREATMENT -

This treatment plan is customized for your child, and should only be implemented after you discuss your child's status with the Metabolism Clinic team:

_____ _____

_____ _____

Continue to assess your child (as above) and have these answers ready to discuss with you team. Plan to talk with the Metabolism Team regularly.

Any questions about the patient or this protocol:

- **METABOLISM CLINIC NURSE – (111)-222-3333**
- **METABOLISM PHYSICIAN-ON-CALL – (222)-333-4444, PAGER 123**

_____ _____

Date Dr. Metabolism Service

PROBLEMS WITH HOME PROTOCOLS

- Protocols are not kept up to date
- Patients / parents implement the protocol or make changes without calling the doctor/team
- The treatment team becomes uncoordinated
  - If the child gets worse, the team does not know what's going on

VMP, LLC

expanding genetic horizons...

Thank you for participating today!

Questions?

mkorson@vmpgenetics.com

www.vmpgenetics.com