

PROTOCOL FOR FEVER OR WHEN INFECTION SUSPECTED

In the Primary Care Physician's Office –

Assessment:

1. Assess fever based on patient's baseline.
2. Determine source of fever.
3. Assess hydration.
4. Assess for worsening of the patient's "typical" symptoms.
5. Assess for gut motility and bladder function.
6. Does the patient have the ability to consistently eat or at least drink, with extra fluids given for higher temperature?
7. Anti-pyretic management.

Management:

1. Treat infections aggressively.
2. When cultures obtained, initiate antibiotic therapy until results become available.
3. Anti-pyretic management.
4. Promote calorie intake, either from food or fluids. Patient may tolerate better small amounts or volumes frequently rather than boluses or meals.
5. When there is a history of fasting intolerance, hypoglycemia, or secondary fatty acid oxidation dysfunction, the patient should take in foods or fluids high in carbohydrate and low in fat on a regular basis. If the patient cannot tolerate this regimen, is vomiting, or refuses (e.g., because of pain), IV fluids are required (see ***In the Emergency Room***, below):
6. When there is a history of gut dysmotility, the patient should take in foods or fluids high in carbohydrate and low in fat since fat can slow gastric emptying ().
7. Monitor carefully.

PROTOCOL FOR FEVER OR WHEN INFECTION SUSPECTED

In the Emergency Room –

Assessment:

1. Draw labs to assess for infection and hydration.
2. If the patient has a history of hypoglycemia, draw a blood glucose level.
3. If the patient has a history of abnormal lab tests when unstable (including blood gases, electrolytes and bicarbonate, lactate and pyruvate, ammonia, or liver functions) or if this is not known, these should be drawn.
4. If the patient looks ill or is at risk for sepsis, cultures should be drawn.

Management:

1. Start IV with balanced solution of dextrose (glucose) and saline. No Ringer's lactate.
2. For patients with a history of fasting intolerance, hypoglycemia, or secondary fatty acid oxidation dysfunction:
 - a. If bolus required – 5% dextrose with normal saline.
 - b. For maintenance fluids – 10% dextrose with appropriate electrolytes running at least at 1.25x maintenance. Providing such an infusion provides supplemental calories and dampens catabolism including fatty acid oxidation which is defective in some mitochondrial patients.
3. For patients without this history:
 - a. If bolus required – normal saline or 5% dextrose with normal saline.
 - b. For maintenance fluids – 5% dextrose with appropriate electrolytes.
4. For patients with pyruvate dehydrogenase complex deficiency, or for patients on a ketogenic diet:
 - a. If bolus required – normal saline.
 - b. For maintenance solution – no glucose or use a glucose-poor solution.
 - c. Consult the managing physician/dietitian for additional recommendations.
5. If patient cannot tolerate oral medications, L-carnitine can be administered IV. Consult a metabolic and/or mitochondrial disease specialist to calculate an appropriate dose (50-100 mg/kg/day divided 8-hourly).
6. Many patients have gut dysmotility. Infectious illnesses can further impair motility resulting in a worsening of gastroesophageal reflux, vomiting, distention, abdominal pain, and/or constipation. It may best to allow this organ to rest by stopping feeds temporarily. IV fluids should be provided and continued until the patient demonstrates an ability to at least maintain their own hydration status. Enteral fluids and feeding should be started gradually.
7. A treatable infection that goes without treatment can severely impact a mitochondrial patient. Antibiotic coverage until culture results are available should be a strong consideration.

8. Treat fever aggressively.
9. For respiratory conditions -
 - a. If there is lower respiratory involvement, patients with a history of muscle fatigue or weakness are at risk for pneumonia and respiratory failure. Monitor carefully and promote aggressive pulmonary toileting.
 - b. Watch for signs of aspiration risk (choking, gagging, drooling)
10. When urinary retention occurs –
 - a. Having the patient try and urinate on a schedule (e.g., every two hours) may help reduce the incidence of infection (as well as other symptoms such as urgency, frequency, dribbling and incontinence). Prophylactic antibiotics may be a consideration.
 - b. In more advanced cases, intermittent catheterization may be indicated.
11. If the patient has a central line and develops fever –
 - a. Evaluate for a focus of infection.
 - b. If the cause of the infection is not clear, or if the patient's status is concerning, blood cultures (peripheral and from the line) should be drawn and antibiotics begun.
 - c. Medical centers that facilitate the surgical placement of central lines generally have management protocols for febrile patients who have indwelling lines.