



FAOD Management Strategies at Home and in the Hospital: *Keeping your child safe*

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- **Scenario:**

- Joey is a 10 month old child with a fatty acid oxidation defect
- MCAD deficiency

- **Diagnosis by newborn screening:**

- Primary screening - significant biochemical abnormality
- Back-up DNA testing – common MCAD mutation, known to cause symptoms

- **Management:**

- Occurs in an established metabolic clinic with a seasoned physician and dietitian
- Regular feeding schedule during the day:
 - Eats food and snacks about 5-6 times during the day
 - Also drinks regularly
- An 8-hour fast is permitted at night
- “Heart-healthy diet” – dietary fat is limited, but not restricted
- Carnitine was measured and found to be borderline low, so L-carnitine supplement provided

Home - Viral illness!

- Joey develops a fever (up to 102F) and a runny nose
- Fever responds to ibuprofen
- Older brother had the same illness; cleared within a week
- Joey is still feeding and drinking without a problem

What is your plan?

What is your plan?

- Diet - if feeding well, then the FAO defect is likely being kept under control because calories are maintained
- Fasting at night – extra feeding?
- Medications – L-carnitine
- Triggers - treat fever
- Monitoring - watch carefully
- Action - call if there are concerns!

Home - Day 2

- Fever continues to rise but still responds to ibuprofen
- Joey is slower today but still fairly active; did not sleep well the night before (interrupted by coughing)
- Today he is eating about half of what he usually eats, but drinking well
- He is taking his L-carnitine

What is your plan?

What is your plan?

- Diet – calories problematic from food, but if drinking well, can switch calories to liquids, as long as he is drinking regularly. Avoid high fat foods now.
- Fasting at night – should be fed every 4 hours around the clock
- Medications – L-carnitine
- Triggers – treat fever
- Monitoring – watch carefully
- Action – call if not meeting his calorie requirements, or if there are concerns!

Home - Day 3

- Fever continues to rise but still responds to ibuprofen
- Joey is clearly tired
- Joey vomits following a small meal, and is given juice, then vomits again
- Wants to go to sleep after vomiting

What is your plan?

What is your plan?

- Diet – Oral calorie intake is no longer dependable for fluids or solids. Encourage high-sugar fluids on the way to the Emergency Department
- Medications – L-carnitine
- Action – go the Emergency Department for evaluation and management

Emergency Department - assessment

- Joey is assessed in a community hospital, and is found to be febrile (102.5F) and mildly dehydrated
- Given ibuprofen
- Joey is tired but rousable and not lethargic
- Has only sips of juice and then vomits
- Diagnosed with a viral illness associated with vomiting
- Blood glucose is measured in the ED – 70 mg/dL (not low)

Emergency Department - plan

- The ED doctor comes in with a treatment plan
- “Good news – Joey is not hypoglycemic!”
- Proposes a bolus of normal saline and if Joey eats a popsicle, he can be discharged
- You ask about including dextrose in the IV fluid but the ED doctor says there is no need and reiterates “Joey is not hypoglycemic!”

What is your plan?

What is your plan?

- Emergency Letter
- Have the ED doctor call the Metabolism specialist
- If the ED doctor won't call the Metabolism MD, or if management isn't happening as it should, you call the Metabolism MD with the name of the ED doctor
- Consider calling the Metabolism MD in advance of going to the ED in order to prepare them

Emergency Department - management

- Joey is given a bolus of IV fluid (5% dextrose and normal saline) in the ED, followed by IV 10% dextrose with $\frac{1}{4}$ normal saline at 1-1/2 times maintenance
- Following his bolus of fluid - more alert and interactive
- Vomits again in the ED
- Fever is managed with ibuprofen
- The ED doctor recommends giving the L-carnitine IV and says the Ward MD will order it (but forgets to document that recommendation)

Hospital Ward

- The management plan continues on the Ward
- The Ward MD writes the order for oral L-carnitine
- You mention the ED MD's recommendation for IV L-carnitine
- The Ward MD does not want to change the order, and when you go back to the room, you learn that Joey has just thrown up twice more

What is your plan?

What is your plan?

- Get the Ward MD to call the ED MD
- Approach the ED MD personally and ask her to talk with the Ward MD
- Ask the Metabolism Clinic MD for an opinion and intervention
- Ask the Ward MD for a team meeting
- Ask the Patient Liaison to intervene

More Serious Encounters

- When health professionals:
 - Question your motives for making suggestions or advocating
 - Refuse to involve you in your child's medical management
 - Suggest that you are contributing to your child's illness

Maintaining records at home

- Keep information catalogued by physician and ordered by date
- When symptoms are unclear or unexplained, keep a portfolio of verification by independent observers

Passing on information

- Avoid passing on information from one doctor to the next, and try not to get in the middle of a debate
- Consider a bound clinic journal that you carry from appointment to appointment
- During hospitalizations, where many services are involved or when major decisions are being made, consider asking for a team meeting (can also occur in the outpatient setting)

Behavior

- Be an advocate
- Be assertive
- Don't be aggressive or abusive

Reflection

- If your interactions keep leading to confrontation...
- If you are firing a lot of physicians or they are firing you...
- Consider reaching out to someone (social worker?) to discuss how to make communication skills more effective

If tension is building...

- Call a team meeting
- Don't make accusations, just say how you feel
- If the situation worsens and you think it is unfair:
 - Speak to the Patient Liaison
 - Ask for an Ethics Consult



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