

Overnight survival tips for working with patients who have inborn errors of metabolism:

If the relative of a patient phones you and says the patient is not well, believe them. Relatives have been right many times when the doctors are wrong.

Blood ammonia levels above 100 $\mu\text{mol/L}$ are an emergency and the patient needs to have treatment started ASAP. Levels below 100 $\mu\text{mol/L}$ should be treated with calories and repeated in 4 hours. If the levels do not improve, the patient may need to be transferred to a hospital with dialysis capability or to VGH where ammonia scavenging therapies are available.

Here are some “tips” for the peripheral hospital:

1. How to start calories in the periphery:
 - Have local hospital start IV D10W at 100 ml/hr
 - Have local hospital start 20% lipids (via peripheral vein) at 10 ml/hr if available
 - Have NG tube inserted locally – start DuoCal (protein free, calorie dense enteral formula) via NG at 50 ml/hr.
 - Repeat NH₃ in 4 hours. If not improving, patient may require transfer.

If the patient is at VGH:

2. Make sure that everything is done rapidly – delays can cause brain damage.
3. DuoCal is available (frozen) in the Pharmacy 24 hours/day
4. All medications used on the preprinted orders are in the Pharmacy
5. If an NG tube is inserted, IT DOES NOT NEED TO BE X-RAYED. This is why the orders say a Levine NG tube and not an enteroflex. This avoids a delay in waiting for the x-ray.
6. Look for precipitants of the episode – noncompliance with diet, sepsis, vascular event etc.
7. Watch the K⁺ (- the phenylacetate/benzoate mixture leaches potassium)
8. When calculating calories, if in doubt, overestimate their requirements. It is better to provide too many calories than too few.
9. Make sure the lab is drawing the NH₃ specimens correctly – they must be drawn with tourniquet OFF and on ice (just like ionized calcium). If drawn incorrectly, the results will be falsely elevated.
10. Consider hemodialysis if NH₃ does not decrease within 6 hours of therapy onset or if patient has deteriorating level of consciousness.
11. If vomiting occurs, try the following:
Slow the rate of the DuoCal– but you need to make up the calories elsewhere
Add an additional line of D10W (340 kcal/L) and can help you make up calories
Add a prokinetic agent as gastric stasis can occur with high ammonia
12. Cerebral edema is the feared complication of high NH₃ levels. However, it is impossible to restrict fluids and still get adequate calories into the patient to prevent catabolism. Calories are more important than fluid in this case as failure to provide calories will cause rapid deterioration. (due to catabolism.)

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