

SPA 2014 PBLD

A Child with Mitochondrial Metabolic Disorder

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Objectives:

1. - Understand the nature of mitochondrial metabolic disorders (MMD) and recognize the challenges in the anesthetic management for these patients.
2. Develop a safe anesthetic plan for MMD patients who are vulnerable to several metabolic derangements.
3. Facilitate the postoperative course, including pain management and other measures resulting in optimal outcomes.

Case history:

Our patient is a 10 years old female with a history of MMD. She is scheduled for a tonsillectomy on the following day.

Questions:

1. What are the pathophysiologic alterations in patients with mitochondrial metabolic disorders (MMD)? And what are their symptoms?
2. How common are MMDs? How are they treated?
3. What is the natural progress of MMD? What is their prognosis?
4. What would you focus on during the preoperative assessment in a patient with MMD?

Case history (continued) and Physical examination:

The patient had progressive muscular weakness with limited ability to move, along with severe restrictive lung disorder. She is BiPAP dependant at night and oxygen nasal canula at 1 L/min at daytime. She had past medical history of grand-mal seizures, dysphagia, gastroparesis, and malnutrition. Her weight is 30 Kg and she is 135 cm tall.

Her current medications were: Co-Enzyme Q10, levomefolate, and riboflavin.

She previously underwent subclavian intravenous subcutaneous port for total parenteral nutrition under sedation and monitored anesthesia care. The patient family expressed great concern about undergoing another anesthetic. They reported that she experienced a grand-mal seizure

progressing to status epilepticus, and resulting into a prolonged intubation, after her last anesthetic.

Physical examination revealed a thin female laying flat in bed with no signs of distress. Vital signs were: SpO₂ 92% on oxygen nasal canula at 1 L/min, HR 110/min (regular), and blood pressure was 90/45. Mouth opening and neck mobility were normal (Mallampati score: I). Lung examination was normal.

Questions:

5. The parents were wondering if anesthesia is safe for their daughter, and if you can discuss the risk of another seizure?
6. Should home medications be held or continued?
7. How do we handle the NPO guidelines for MMD patients?

Preoperative studies:

An EKG shows normal sinus rhythm. An earlier cardiac echocardiogram showed moderately depressed left ventricular function with an Ejection Fraction of 45% and mild pulmonary hypertension. Chest x-ray exam was clear. Pulmonary function studies showed pulmonary restrictive disease (FVC and FEV₁ at 25% of expected). Laboratory tests show normal electrolytes with the exception of low K⁺ = 2.9 and Creatinine 1.6 and two-fold increase in liver enzymes.

Questions:

8. Do you need any additional information before proceeding with the anesthetic?
9. Would you premedicate this child? If yes, how would you like to premedicate her?

Case progression:

An appropriate level of sedation was achieved, and the patient was transferred to the operating room without problems. Standard monitors were applied and pre-oxygenation was started.

Intraoperative care:

10. How would you like to induce and maintain anesthesia?
11. Do we need any special monitoring (what would you use)?
12. Which anesthetic agents are better avoided?
13. What is your plan for IV access?
14. Do you have alternative ways of taking care of this patient?
15. What airway (if any) would you use?
16. Are you planning on using a muscle relaxant?
17. Is there an association between MMD and malignant hyperthermia?
18. How would you manage the fluid balance? Which IV fluids would you use?
19. The surgeon is planning on injecting local anesthetic, what do you think?

Case progression:

The patient had an uneventful induction of anesthesia and placement of an airway. The procedure lasted 45 minutes.

Questions:

20. Are you going to attempt extubation in the operating room? Why?
21. Under what circumstances would you keep the patient intubated?

Case progression:

Upon completion of the procedure, the patient was successfully extubated. Just prior to departure from the operating room, the patient started to experience a seizure.

Questions:

22. How would you manage the patient's seizure?
23. What are the possible causes of this seizure?
24. You succeeded in treating the seizure, how do you manage the respiratory support?
25. Would you reintubate the patient? Why?

Postoperative care:

The patient arrived to the recovery unit in a stable condition. She is hemodynamically stable and receiving oxygen through a face mask but she seems to be having some pain and discomfort.

Questions:

26. What are your main concerns in the recovery period?
27. Which analgesics would be optimal for this child?
28. What kind of hospital bed does this patient need: regular floor bed, step-down bed, or ICU bed? Why?
29. Under what circumstances should MMD patients be admitted, and when can they get discharged to home?

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