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ANAESTHETIC MANAGEMENT FOR FULL MOUTH REHABILITATION IN A CHILD WITH MULTIPLE CARBOXYLASE ENZYME DEFICIENCY

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SUMMARY
MCD Multiple carboxylase deficiency, is a rare autosomal recessive metabolic disorder, that results in impaired biotin-dependent enzyme activity, causing extensive metabolic, neuromuscular and respiratory abnormalities that can be fatal during anesthesia. Here we discuss the clinical and anaesthetic management of a 9 year old child with MCD, the relevant Preoperative investigations and preparations done including metabolic optimisation and systemic optimisation. In this case general Anaesthesia with intravenous induction and maintenance with volatile anaesthetics and non-depolarizing muscle relaxant ensured stable intraoperative conditions. Close monitoring of metabolic parameters and meticulous postoperative care facilitated a smooth recovery, highlighting the importance of tailored anaesthesia in managing paediatric patients with complex metabolic disorders.

BACKGROUND
Multiple carboxylase deficiency was first described in 1982, it is a rare inherited autosomal recessive metabolic disorder characterized by deficiencies in biotin-dependent enzymes, which play crucial roles in various metabolic pathways. These deficiencies include neurological abnormalities, dermatological, neuromuscular, metabolic, respiratory, immunodeficiency.)
In this presented case, A 9-year-old child pre diagnosed with MCD was referred from pedodontics department for extensive full mouth rehabilitation under general anesthesia as child was refusing to take oral feeds due to painful dentition and infected periapical abscess exhibited, patient was previously admitted to PICU with severe symptoms of MCD including seizures.
The article aims to provide insights into the specialized perioperative management required for paediatric patients with complex metabolic disorders like MCD, [1]

CASE PRESENTATION
We report a case of a 9-year-old child diagnosed with MCD, presenting for full mouth rehabilitation under general anesthesia. The patient weighed 29 kg and exhibited signs including poor feeding, mental and developmental disability, severe oral drooling, various stages of severe dental decay including periapical abscess in the oral region. She was on biotitin supplements and other drugs .

Anaesthetic Considerations:
These Laboratory investigations included complete blood count, Renal function test, Liver function test Electrolytes ECG ECHO CHEST XRAY, ABG , BUN , Serum Ammonia, Serum lactate and Urine ketones was done and was within normal limits .
Given the metabolic instability associated with MCD, The following factors should be taken into consideration

1. Primarily, Dyselectrolytemia was ruled out as serum electrolytes was normal .
2. ABG was within normal limits no metabolic acidosis no lactic acidosis
3. Important to avoid ketoacidosis and Maintain euglycemia and preoperative CBG was done .
4. preoperative normothermia was maintained
5. preoperative monitoring of liver enzymes including Serum Ammonia . Febrile seizures was ruled out .
6. succinyl-choline was avoided as it can cause hyperthermia

7. To monitor temperature before induction to r/o febrile seizures
8. pre operative monitoring to ensure no acute fluid loss such as AGE or vomiting .
9. it was checked I the child has taken pre operative biotin supplementation before the surgery .

ANAESTHETIC MANAGEMENT: The child received premedication with Syrup Phenergan to reduce oral secretions for 3 days as anti-sialagogue, midazolam (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg) to minimize anxiety and reduce secretions. Dexamethasone (0.1 mg/kg) was administered to prevent inflammation, and fentanyl (2 mcg/kg) for analgesia. Induction of anesthesia was achieved with propofol (2 mg/kg) and neuromuscular blockade with atracurium (0.5 mg/kg) and sevoflurane was used as the volatile agent.

During anesthesia induction and maintenance, careful monitoring of metabolic parameters including blood gases, electrolytes, and ammonia levels was conducted to promptly address any deviations. Nasotracheal intubation was performed with a size 5 tube to secure the airway. Throughout the procedure, meticulous fluid management and maintenance of euglycemia were maintained to prevent metabolic decompensation. normothermia was maintained with the hlp of forced air warmer.

RECOVERY: The intraoperative course was uneventful, with stable hemodynamic and respiratory parameters. Postoperatively, the child was successfully reversed from neuromuscular blockade using neostigmine (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg). Extubation was smooth, with adequate suctioning and postoperative monitoring revealing no significant complications.post operstive period was monitored for normothermia and normovolemia and electrolytes were monitored daily.

DISCUSSION

The discussion of the article on anaesthetic management for full mouth rehabilitation in a child with Multiple Carboxylase Deficiency (MCD) underlines several critical points and considerations in managing such complex cases.

1. **Metabolic Challenges and Anaesthetic Considerations:** Children with MCD face significant metabolic challenges, including potential for ketoacidosis, hyperammonaemia, and metabolic acidosis. These metabolic disturbances can be exacerbated by the stress of surgery and anesthesia. Therefore, meticulous preoperative evaluation and optimization are crucial.[5] The choice of anaesthetic agents, such as avoiding those that can further impair metabolic function or trigger acidosis, is paramount. The case highlights the importance of selecting anesthesia techniques and medications that minimize metabolic stress and maintain stability throughout the procedure [6]
2. **Interdisciplinary Collaboration:** The case underscores the necessity of interdisciplinary collaboration among various medical specialties, including paediatricians, metabolic disease specialists, anaesthesiologists, and dental surgeons. Each specialist contributes expertise in managing different aspects of the patient's condition, ensuring comprehensive care from preoperative assessment to postoperative recovery. This collaboration ensures that the treatment plan is tailored to the unique needs and challenges posed by MCD.
3. **Outcome and Long-term Management:** The successful management of the case, with no significant perioperative complications reported, demonstrates the effectiveness of a well-planned approach to anesthesia and surgical procedures in children with metabolic disorders. Long-term follow-up and monitoring are essential to assess growth, development, and metabolic stability. Genetic counselling may also be necessary to guide family planning and provide support.[8]
4. **Research and Future Directions:** Given the rarity of MCD and similar metabolic disorders, further research is needed to refine anesthesia protocols, understand the long-term outcomes of surgical interventions, and explore new treatment modalities. Case reports and studies like this contribute valuable insights into optimizing care for children with complex metabolic conditions undergoing surgical procedures.

DIFFERENTIAL DIAGNOSIS

Biotinidase Deficiency

 Other Inborn Errors of Metabolism (IEM):

Propionic Acidemia: Deficiency in propionyl-CoA carboxylase enzyme.
Methylmalonic Acidemia: Deficiency in methyl malonyl-CoA mutase enzyme.
Glutaric Aciduria Type 1: Deficiency in glutaryl-CoA dehydrogenase enzyme.

 Urea Cycle Disorders (UCD) , **Organic Acidemias and Mitochondrial Disorders**

TREATMENT OF MCD

The treatment of a child with Multiple Carboxylase Deficiency (MCD) undergoing full mouth rehabilitation under general anaesthesia involves a multidisciplinary approach aimed at managing metabolic disturbances, ensuring safe anaesthesia, and optimizing perioperative care. Here's a comprehensive outline of the treatment considerations:

- **Medical Management:**
  - **Biotin Supplementation:** Since MCD is characterized by deficiencies in biotin-dependent enzymes, high-dose biotin supplementation is essential to bypass enzyme deficiencies and support metabolic functions.
  - **Metabolic Support:** Continuous monitoring and management of metabolic acidosis, hyperammonaemia, and other biochemical abnormalities are crucial. This may involve specific interventions such as intravenous fluids, electrolyte correction, and medications to stabilize metabolic parameters.
  - **Nutritional Support:** Ensure adequate nutrition with a focus on biotin-rich foods or supplements to support metabolic needs.
- **Anaesthetic Management:**
  - **Preoperative Evaluation:** Comprehensive assessment of metabolic status, including electrolytes, blood gases, ammonia levels, and liver function tests.
  - **Anaesthetic Agents:** Selection of anesthesia agents that minimize metabolic stress and avoid triggering metabolic decompensation. For instance, avoiding agents that can impair mitochondrial function or exacerbate acidosis.
  - **Monitoring:** Continuous monitoring of vital signs, electrolytes, blood gases, and acid-base status throughout the procedure.
  - **Perioperative Care:** Meticulous perioperative management to maintain metabolic stability, including careful fluid management, temperature control, and avoiding prolonged fasting to prevent catabolism.
- **Surgical Management:**
  - **Procedure Planning:** Coordination with dental and surgical teams to plan a comprehensive rehabilitation procedure that minimizes stress and optimizes outcomes.
  - **Intraoperative Considerations:** Ensuring smooth induction and maintenance of anesthesia, meticulous airway management, and monitoring for signs of metabolic disturbances.
  - **Postoperative Care:** Immediate recovery in a monitored setting with continued assessment of metabolic parameters, prompt treatment of any abnormalities, and ensuring adequate pain management.

OUTCOME AND FOLLOW-UP

Long-term Follow-up:

- **Monitoring and Surveillance:** Regular follow-up visits to monitor growth, development, and metabolic status.
- **Genetic Counselling:** Providing genetic counselling and support to the family regarding the inheritance pattern and potential risks for future pregnancies or family members.

- **Multidisciplinary Collaboration:**
  - Involvement of specialists such as paediatricians, metabolic disease experts, anaesthesiologist's, nutritionists, and dental specialists to ensure coordinated care and management.
- **Education and Support:**
  - Providing education to caregivers and family members about the condition, signs of metabolic decompensation, and the importance of adherence to treatment and follow-up.

CONCLUSION

This case highlights the importance of tailored anesthesia management in paediatric patients with multiple carboxylase deficiency undergoing surgical procedures. By understanding the underlying metabolic disturbances and employing meticulous perioperative strategies, including careful selection of anaesthetic agents and monitoring parameters, optimal outcomes can be achieved in this complex patient [2][4]

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