

INTRODUCTION

Understanding the medical management and dental manifestations of rare childhood diseases in patients has been a challenge for the pediatric dentist.

Among the current literature, only one study exists that describes dental manifestations of patients with methylmalonic acidemia. Furthermore, no current literature exists regarding the anesthetic management of dental patients with methylmalonic acidemia. This case report presents details about the clinical examination, diagnoses, imaging, radiographs, treatments, and follow-ups related to a pediatric dental patient with methylmalonic acidemia.

The patient's treatment required advanced behavior guidance, namely sedation with general anesthesia. The goal of this report is to expand literature regarding the clinical implications and anesthetic management of pediatric dental patients with methylmalonic acidemia.

METABOLIC PATHWAY OF MMA

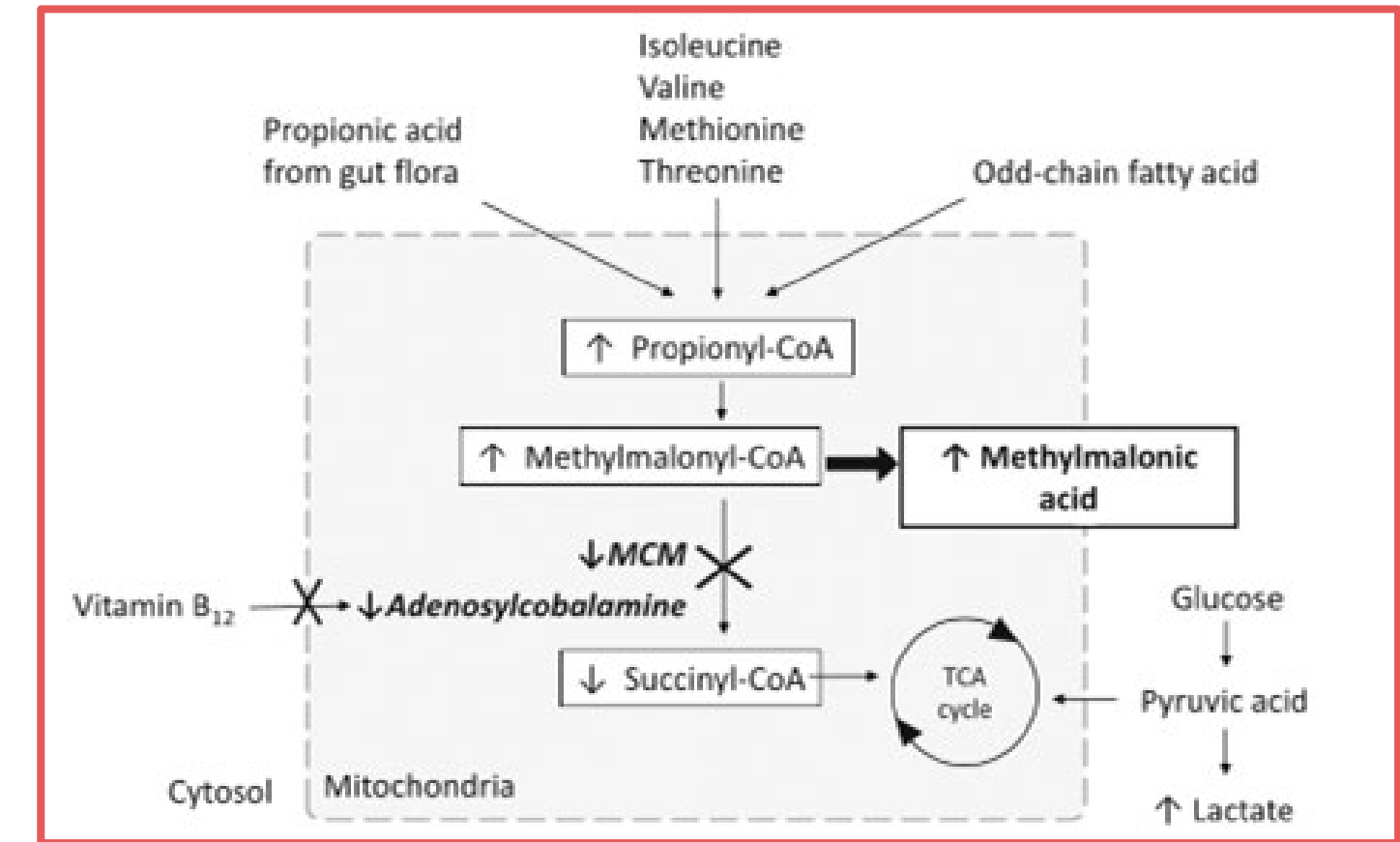


Figure 3. Metabolic Pathway in MMA (4). As the chart depicts, due to a deficiency of enzyme MCM, an excess of methylmalonic acid is present

MMA BACKGROUND

- The collective incidence of isolated methylmalonic acidemia (MMA) is still inconclusive, but is most likely between 1 in 50,000 to 100,000 births (3)
- MMA is a metabolic disorder that results from a deficiency in the enzyme Methylmalonyl-CoA mutase (MCM). (1)
- MCM is present in the metabolic pathway of four types of amino acids (valine, isoleucine, methionine, and threonine). See Figure 3. (4)
- It results in an accumulation of methylmalonic acids due to the inactivation of MCM
- Patients who have this disorder have increased protein catabolism which can result in severe metabolic acidosis, hyperammonemia, and ketosis.
- Patients with MMA can have an ATP deficiency since the MMA succinyl CoA deficiency causes the operation of the TCA cycle to mainly depend on glucose. (2)
- Severity of the deficiency of the enzyme can vary, and affect mortality outcomes accordingly; those with complete deficiency, for example, usually progress to coma and death within the first few days to weeks of life. (5)
- Given its rare incidence, there is limited medical literature surrounding MMA and by extension, even more limited literature describing dental manifestations in patients with MMA.
- Only one such study exists, "Enamel Defects and Salivary Methylmalonate in Methylmalonic Acidemia," which depicts that there is a possible association between metabolic disorders and enamel developmental pathology.

CLINICAL PHOTOS

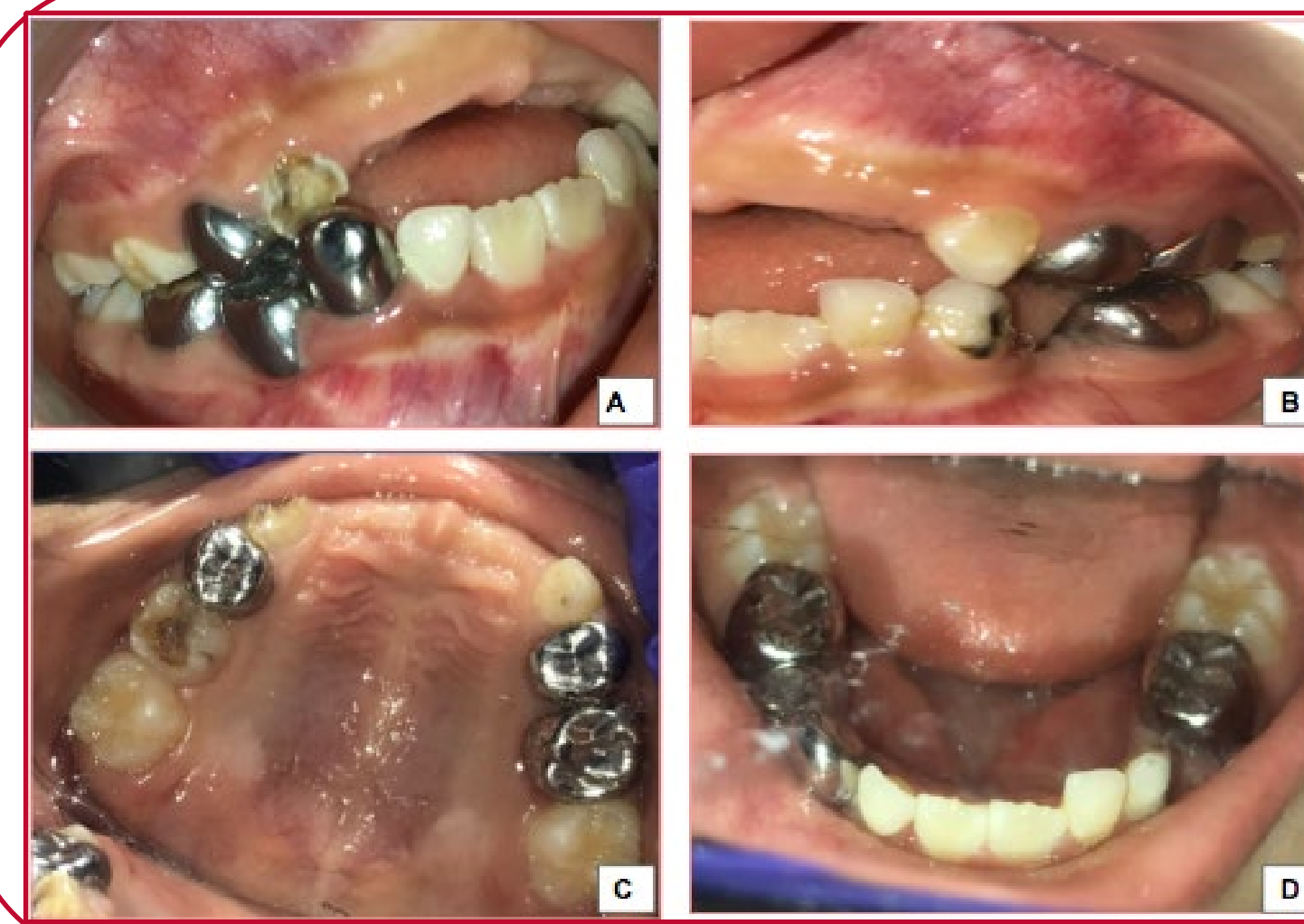
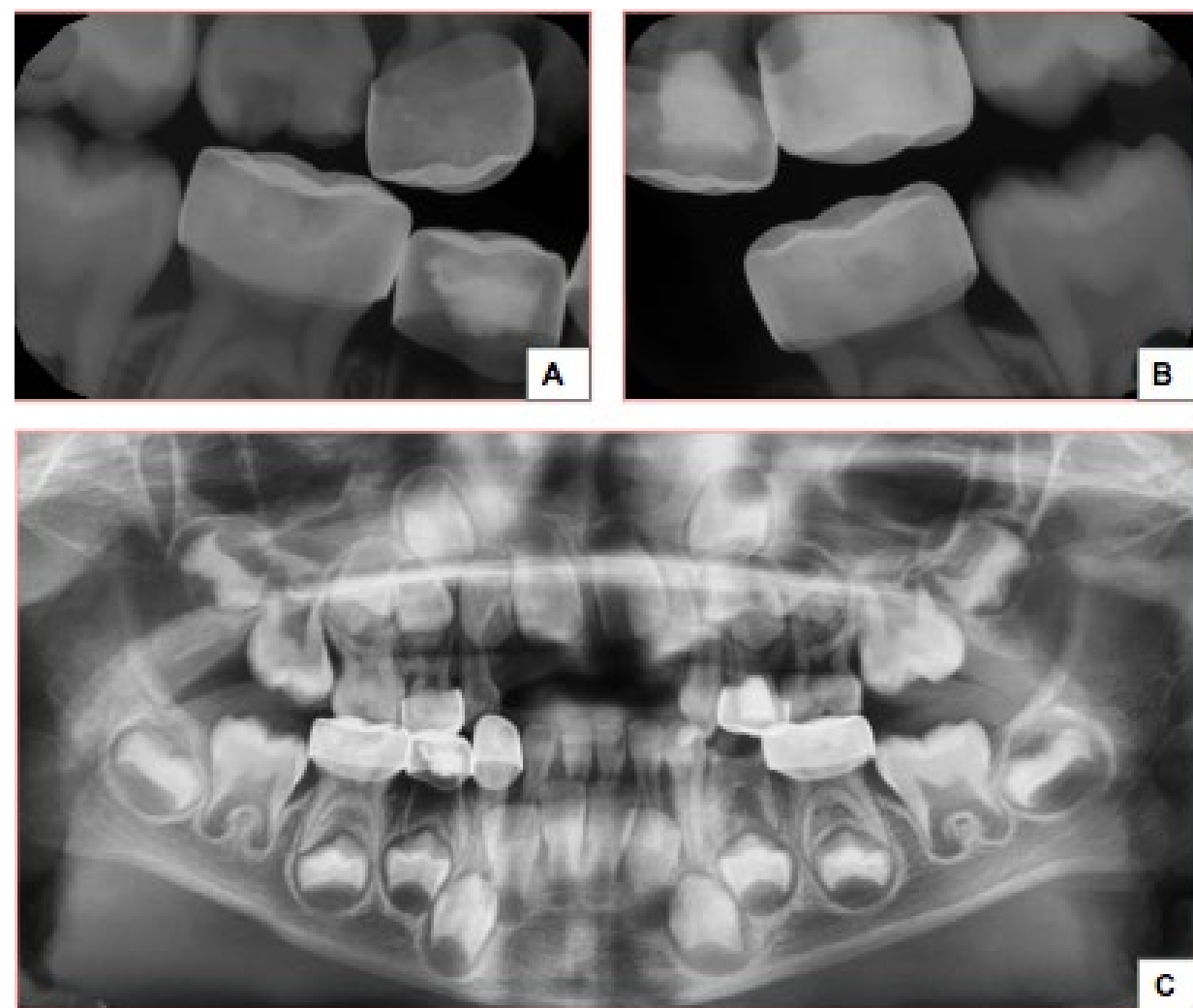


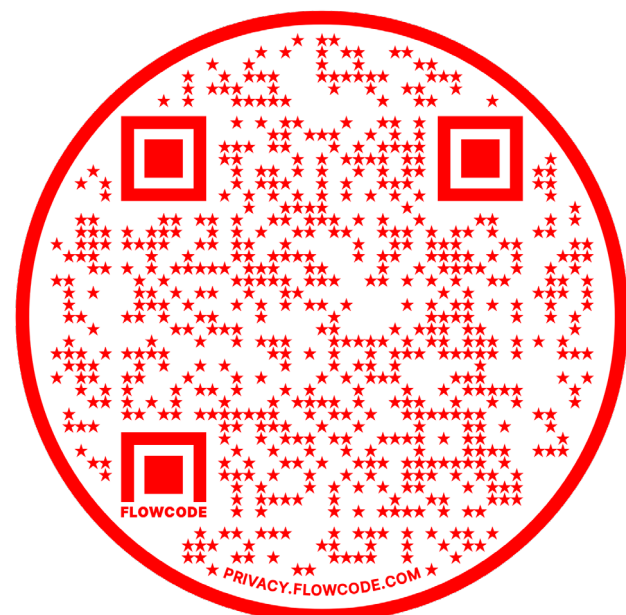
Figure 1. Patient's intraoral photographs taken on 9/8/2021: (A) right side biting, (B) left side biting, (C) lower occlusal, and (D) upper occlusal photographs are pictured.

RADIOGRAPHS

Figure 2. Patient's radiographs obtained on 9/8/2021: (A) right side bitewing - #A: occlusal caries; #S - PARL, #L - PARL (B) left side bitewing, (C) panoramic - mixed dentition, all permanent teeth present. No pathology noted.



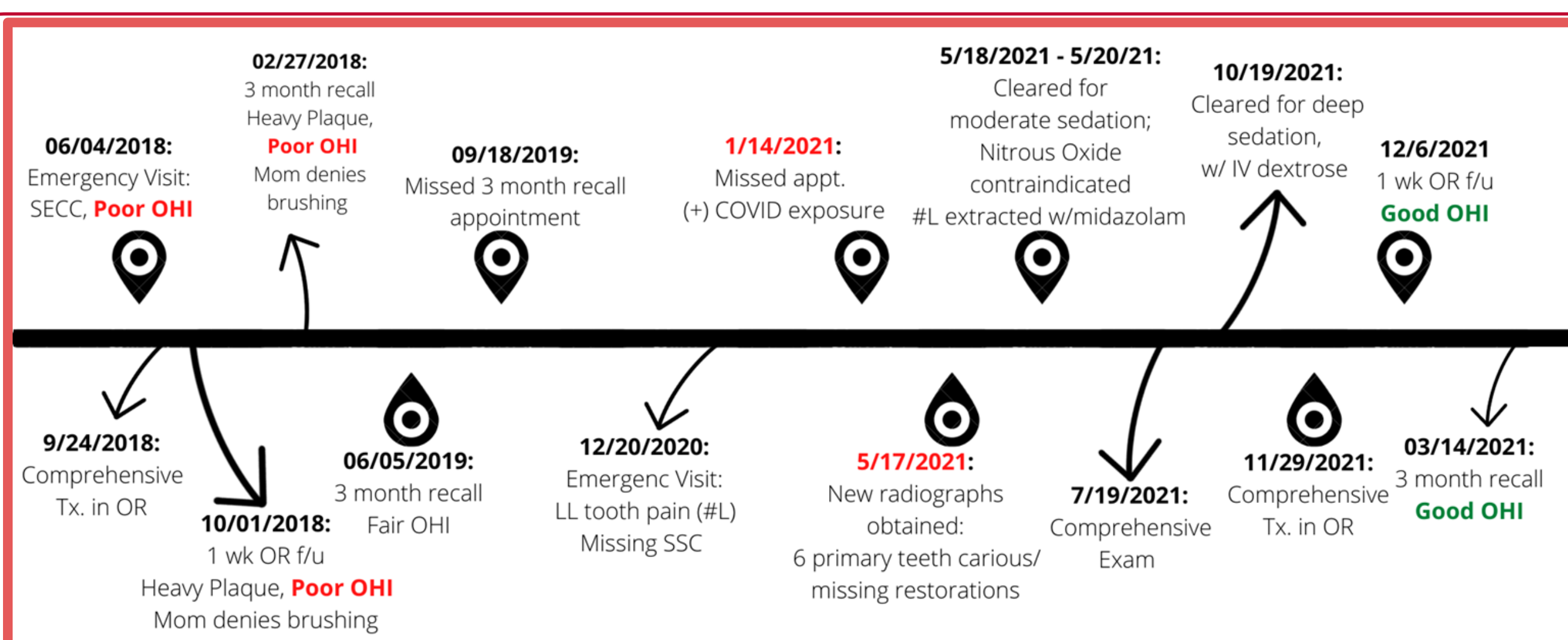
REFERENCES



DISCUSSION

- This discussion will illustrate different studies detailing the anesthetic management techniques of patients with MMA including appropriate drug selection, administration of glucose to prevent hypoglycemia, and adjustments regarding preoperative fasting.
- One study depicted a patient undergoing a liver transplant, an optimal fasting period is noted as 6 hours and the patient was given continuous IV glucose, vitamin B12 and L-carnitine infusion on the day of the procedure. The patient's general anesthesia was maintained with sevoflurane, fentanyl, remifentanyl, and rocuronium. Furthermore, due to a possible risk of metabolic acidosis, the team had prepared a catheter for CHDF (continuous hemodiafiltration) but it was not used. This study **avoided nitrous oxide and propofol during the surgery.** (2)
- Another study also noted that nitrous oxide should be avoided due to interference with adenosyl cobalamin, which is a cofactor of MCM and plays a role in the metabolism of vitamin B12; this essentially results in MMA accumulation. The study also noted that propofol should not be administered due to its association with a deterioration of metabolic failure during general anesthesia of a liver transplantation. (6)
- In a separate study looking at the anesthetic management of a patient with MMA, it is emphasized that anesthetic management is crucial to prevent acute metabolic failure. Due to the **risk of hypoglycemia** in these patients, it is also recommended that glucose loading be avoided and that preoperative fasting be as short as possible, and it be administered during surgery if the patient has decreased blood sugar levels. (6)
- Similarly, in a lens surgery procedure, it was noted that **IV dextrose should be administered one day before the surgery.** (5)
- Although a majority of sources cited avoiding propofol, one study did conclude that it is safe to administer to these patients. (5)
- In order to contribute to the above literature, it is important to note that during our patient's surgery, the anesthesia team did not use propofol nor did they use nitrous oxide during the procedure. Our team also gave the patient continuous IV dextrose throughout the procedure. The patient recovered with no complications. Additionally, our patient had a high caries risk.

PAST DENTAL & MEDICAL HISTORY



A 6-year-old Hispanic girl was seen for comprehensive dental treatment under general anesthesia; **Medical History:** MMA, Weight = 21 kg, Height = 43.5 inches, BMI = 15.6; **Medications:** L-carnitine; **Dental History:** Brookdale Hospital 2018 - 2022: After initially presenting to our clinic as an emergency visit, the patient was seen in the operating room due to severe early childhood caries in 2018. She presented for follow-up visits with poor oral hygiene and heavy plaque, and mom was encouraged to start supervised brushing. The patient then missed several recall appointments, after which she presented again as an emergency with several carious lesions resulting in a second OR visit. After this OR visit, the patient had consistent recalls and presented with good oral hygiene. As a result, the timeline of events depicts that regular recall visits and repetitive oral hygiene instruction are essential for managing this patient's oral health.