Dental Management of Patient with Aase Syndrome Under General Anesthesia: Case Report

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Introduction

Aase Syndrome (AS) is a rare genetic disorder mostly characterized by extremity and joint anomalies, as well as hypoplastic anemia. AS has a prevalence of 1 in 1,000,000 worldwide. Understanding the physical and health disabilities in these patients is important to provide multidisciplinary dental care, especially when the patient is suffering from severe early childhood caries with a high caries risk. The most common signs and symptoms of this syndrome are: growth delay (third percentile), radial hypoplasia, pale skin, delayed closure of fontanelles, narrow shoulders, triple joint thumb, deformed ears, and droopy eyelids/ptosis. This syndrome is considered of autosomal recessive inheritance. Oral manifestation may include cleft /lip palate, micrognathia, and constricted maxillary arch. Although uncommon, the dental community should be familiarized with this syndrome in order to provide a comprehensive dental care.

Case Report

Case of 5-year-old female patient with past medical history of Aase Syndrome, cleft palate, growth delay, epilepsy, microtia/hearing impairment, bilateral triphalangeal thumb, microcephaly, micrognathia, and multiple dental caries. At the time, patient was taking 1.0mg of Keppra daily. Patient was referred to our special needs pediatric dentistry clinic to asses severe early childhood caries. Clinical and radiographic examination revealed deep dental caries in all teeth except #M, #N, and #Q. Dental history revealed past dental trauma of tooth #E when the patient was 1-year-old. Intrusion and yellow discoloration was observed. No treatment was provided at the moment of the trauma and no further symptoms developed prior. Patient also presented mild generalized gingivitis. Past surgical history disclosed tracheotomy placement, G-tube placement, pyloric stenosis correction, cleft palate correction, and left cochlear implant placement with no reported complications.

Clinical Examination

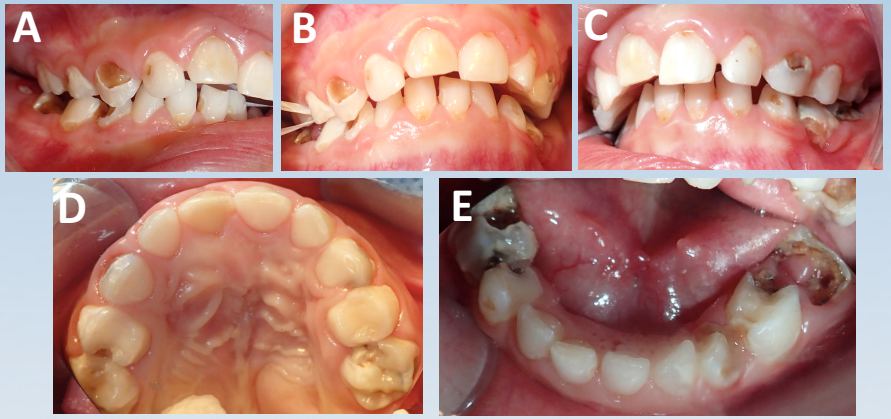

Figure 1. Intraoral Pictures. A) Right Lateral View. B) Frontal View. C) Left Lateral View. D) Maxillary Occlusal View. E) Mandibular Occlusal View

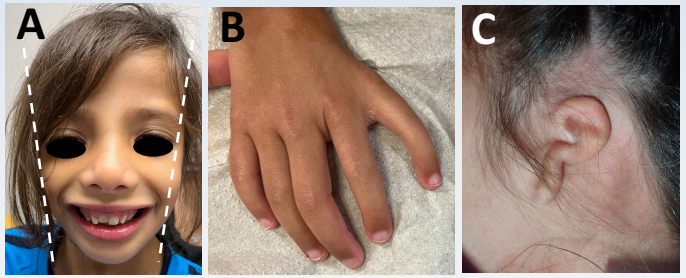

Figure 2. Characteristic Signs and Symptoms of Aase Syndrome Exibited in the Patient. A) Ptosis and Delayed Closure of Fontanelles. B) Triphalangeal Thumb. C) Microtia.

Figure 3. C-MAC Videolaryngoscope. Intubation assistance device used to aid in the intubation process of patients with craniofacial deformities i.e. cleft palate.

Management

- **Preparation for General Anesthesia (GETA)**
  - Consultation Request for Medical Clearance: Pediatrician, Geneticist and Neurologist
  - Case Presentation to Anesthesiology Team to Asses General Anesthesia Induction and Intubation techniques
  - Preoperative Lab Analysis - CBC, CMP, PT, PTT, INR and Urinalysis
- **Oral Rehabilitation Under GETA - Treatment Plan**

Preventive
-Prophylaxis
-Fluoride Varnish
-Oral Hygiene Itrusctions

Surgical
Simple and Surgical Extractions

Orthodontics
Right Side Distal Shoe

Restorative
-Amalgam Resroration
-Composite restoration
-SSC cementation

Behavior Management
GETA

Conclusion

-General Anaesthesia Considerations: Anemia appraisal must be done preoperatively to prevent hematological emergencies during procedure. Nasal intubation techniques should be assessed since cleft palate and craniofacial abnormalities may present challenges during intubation. Laryngoscopy technology like fibber optic and C-MAC Videolatyngoscope may be helpful during nasal intubation process. Also, peripheral vascular access may be difficult due to radial hypoplasia.

-Multidisciplinary Approach: Integrating all health care providers in the patient's treatment will ensure comprehensive and patient cantered care.

-Dental Treatment: Thorough clinical and radiographic examination, caries risk assessment and the discussion of anticipatory guidance's with caregivers are key factors in providing a comprehensive treatment. A preventive plan must be discussed prior and after restorative care.

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References

References upon request