Complete Oral Rehabilitation of an Alagille Syndrome Patient Case Report

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Background and purpose:

Alagille syndrome is a rare genetic disorder that can affect multiple organ systems of the body including the liver, heart, skeleton, eyes and kidneys. The mutations that cause Alagille Syndrome can be inherited in an autosomal dominant pattern, but in about 50% of cases, the mutation occurs as a new change de novo" in the individual and was not inherited from a parent. The syndrome affects females and males in equal numbers. Common symptoms of the syndrome which often develop during the first three months of life, include Cholestasis, which is blockage of the flow of bile from the liver. Jaundice which is yellowing of the skin and mucous membranes, poor weight gain and growth.

Additional symptoms include heart murmurs, congenital heart defects like aorta coarctation, skeletal differences, thickening of the ring that normally lines the cornea in the eye and distinctive facial features. The distinctive facial features include deep set and hypertelorism of the eyes, broad forehead and a pointed chin. In these patients chin may appear larger and more prominent (prognathism).

Oral manifestations are not a primary feature of the syndrome, but they invariably occur as a complication of the long-lasting cholestasis and are linked to hyperbilirubinemia. As a consequence of cholestasis during odontogenesis, hypomineralization, enamel opacities, and hypoplasia of tooth enamel can appear. Our purpose is to present a case of an Alagille Syndrome patient with multiple health conditions and multiple surgical interventions and hospitalizations with high carbohydrate intake including prolonged bottle feeding and poor oral hygiene posed an adverse effect on his oral health.

Case Description:

- Age: 4 y/o
- Sex: Male
- Previous Medical History: Alagille Syndrome, Aorta Coarctation, Astigmatism, Myopia, born premature at 29 weeks and in rule out of Autism diagnosis at the time
- Allergies: NKDA/NKFA Meds: Urzodiol
- Previous Surgical History: Thoracotomy, 3 Angioplasty surgeries
- Patient presented Severe Early Childhood Caries and uncooperative behaviour

Clinical & Radiographic Examination

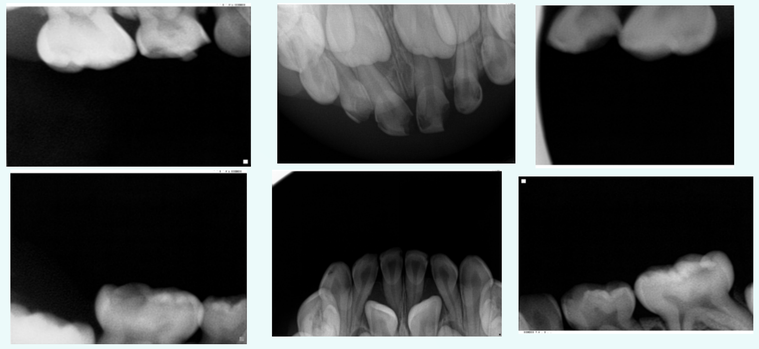
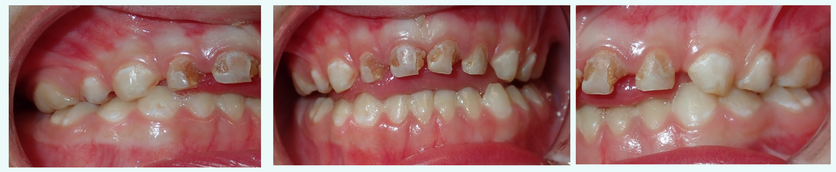


Figure 1: Xrays showing Severe Early Childhood Caries on 4y/o patient with Alagille Syndrome



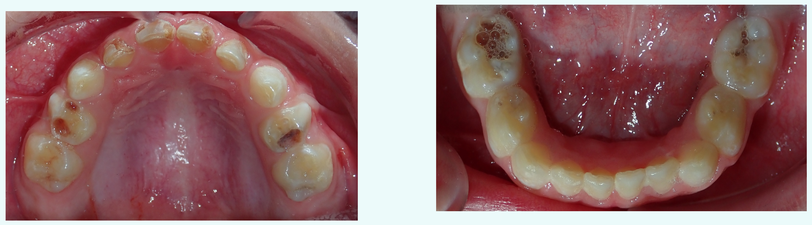


Figure 2: Clinical photos of Severe Early Childhood Caries on 4y/o patient with Alagille Syndrome

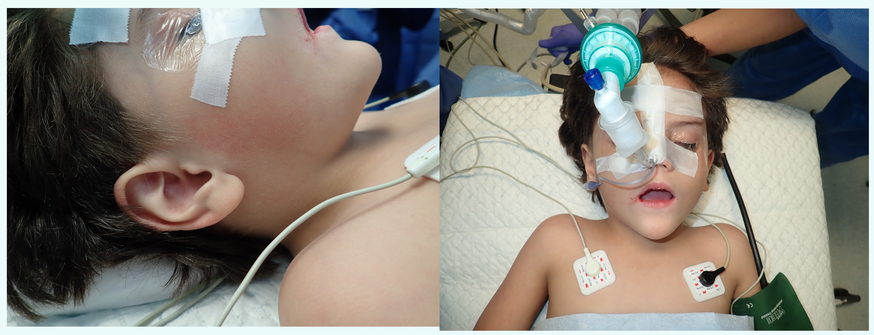


Figure 3: In this photo we can observe patients pointed chin, broad forehead which is a typical facial feature of Alagille Syndrome

Discussion and Concluding remarks:

The patient was evaluated clinically and radiographically and proper medical history was taken. It was determined that the best scenario for dental treatment was a complete oral rehabilitation under General Anesthesia. Due to patients medical and surgical history and uncooperative behavior. Proper consultations to the patients multidisciplinary care team that included pediatrician, gastroenterologist and cardiologist were done. Their recommendations were followed. Prior to procedure date, anesthesiologist were presented the case and type and screen was done. This patients oral health was affected by prolonged bottle usage but also by prolonged cholestasis as mentioned before.

The patient received dental treatment in the OR, that included extractions, Stainless steel crowns and amalgam restorations. The patient has continued to receive follow up in the University Pediatric Hospital Dental Clinic as a dental home. It is important for Pediatric Dentist and General dentist to recognize medically compromised patients because oral health problems pose increased risk to their systemic health. Alagille Syndrome Prolonged cholestasis during odontogenesis predisposes patient to an increased caries risk. The appropriate treatment setting and management as a multidisciplinary team should be done. An improved oral health will enhance the quality of life of the patient.

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References

Alagille syndrome. NORD (National Organization for Rare Disorders). (2020, August 3). Retrieved January 18, 2022, from https://rarediseases.org/rare-diseases/alagille-syndrome/

Berniczei-Royko, A., Chałas, R., Mitura, I., Nagy, K., & Prussak, E. (2014). Medical and dental management of Alagille syndrome: A review. *Medical science monitor: international medical journal of experimental and clinical research*, *20*, 476.

Consent was received for photos for dental proffesional education purposes