Case Report: Green Dyschromia of Teeth Due to Alagille Syndrome

Mathes LP, Vergotine RJ, Boynton JR
University of Michigan, Ann Arbor, MI, C.S. Mott Children's Hospital

INTRODUCTION

Alagille Syndrome

Alagille syndrome (ALGS) is a highly variable genetic disorder characterized primarily by a paucity or complete absence of bile ducts within the liver. The most common hepatic manifestations include chronic cholestasis, jaundice and cirrhosis.

This case report details diagnosis, clinical manifestations and dental rehabilitation of a patient with Alagille syndrome. Highlighted are the challenges of preparation and restoration of teeth demonstrating a green dyschromia.

CLINICAL MANIFESTATIONS

Hepatic
- Cholestasis secondary to bule duct paucity
- Conjugated hyperbilirubinemia
- Pruritis
- Xanthomas
- Cirrhosis – leading to end-stage liver disease in 15%

Renal
- Renal dysplasia

Ophthalmologic
- Posterior embryotoxon

Skeletal
- Butterfly vertebrae

Cardiac
- Peripheral pulmonary stenosis (67%)
- Tetralogy of Fallot (16%)
- VSD, ASD
- HLHS
- Aortic stenosis
- Coarctation of the aorta

Facial
- Prominent, broad forehead
- Deep-set eyes with hypertelorism
- Prominent ears
- Broad nasal bridge

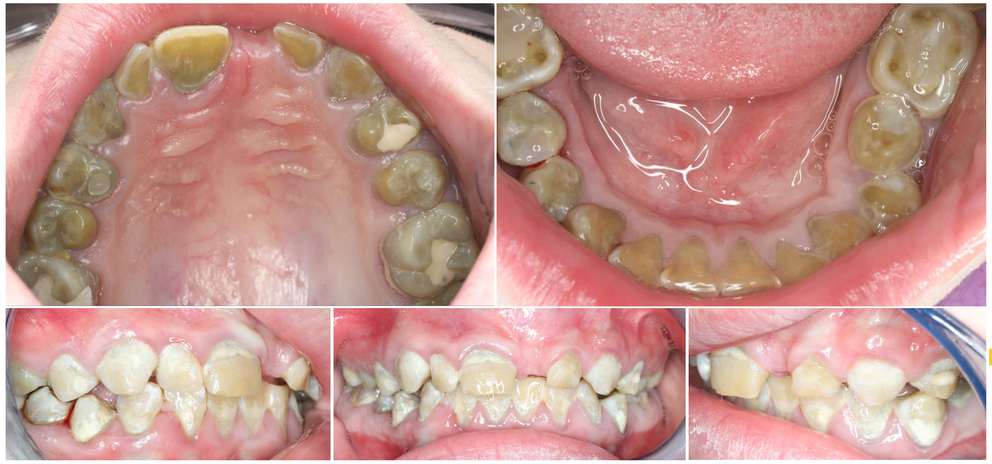
Vascular
- Aneurysm
- Moyamoya syndrome
- Cerebral artery anomalies
- Reno-vascular anomalies

MEDICAL CONSIDERATIONS

Liver transplant
- 15-47% of ALGS patients undergo liver transplant, with median age for transplant being 4-6.5 years
- Dental infections risk damage to the transplanted liver
- Ideally, complete dental treatment before transplantation
- Rigorous prophylaxis schedule

Immunosuppression
- Leukopenia, thrombocytopenia, anemia
- Cyclosporine – gingival hypertrophy

Cardiac defects
- May require antibiotic prophylaxis
- Consider dental rehabilitation under general anesthesia if cardiac sequelae are severe



DENTAL MANIFESTATIONS

Dental manifestations are not direct features, rather a consequence of cholestasis and hyperbilirubinemia
- When serum bilirubin surpasses 30mg/dL, biliverdin accumulates in the dental tissues, resulting in a green-brown dyschromia
- Timing of the hyperbilirubinemia will determine which teeth/dentitions are affected

Other associated dental anomalies
- Talon cusps, macrodontia, taurodontism (primary dentition), hypodontia
- Extensive decalcification of dentin invariably occurs

DENTAL CONSIDERATIONS

- Decalcification/hypomineralization may result in bonding difficulty and decreased bond strength
- Dyschromia may be present in both enamel and dentin, which may make distinguishing between carious and healthy dentin challenging during preparation
- Consider use of caries disclosing agents, and rely on tactile sensation for caries detection
- Orthodontia may be indicated in order to establish ideal alignment for potential esthetic restorative care in adulthood

CASE REPORT

- This patient, 13 years old at the time of these photos, was referred to the UM Pediatric Dentistry Clinic for dental restorations
- She had previously undergone dental rehabilitation under general anesthesia at UM at age 3, as well as restorations completed later at the UM Hospital Dentistry Clinic
- Her health history includes ALGS, drug-induced immunosuppression secondary to liver transplantation, CKD stage 1, LVH, and patent foramen ovale. No SBE prophylaxis is indicated per her cardiologist
- She presents with generalized green-brown dyschromia affecting both enamel and dentin, and requires multiple restorations, as well as orthodontic treatment
- She has since received multiple composite restorations, and has completed an orthodontic consultation at the UM Orthodontics Clinic

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