

Conservative Management of a Child Patient with CHARGE SYNDROME

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INTRODUCTION

CHARGE syndrome (CS) is a rare, complex genetic syndrome caused by a mutation in the CHD7 gene resulting in multiple organ abnormalities². The acronym CHARGE represents the common features of the condition, including **C**oloboma of the eye, **H**eart defects, choanal **A**tresia, **R**estriction of growth and development, **G**enital abnormalities, and **E**ar abnormalities and/or deafness¹. Prevalence of CS is approximately 1 in 10,000-15,000 births. Oral manifestations include congenital dental anomalies, bruxism, periodontal disease, difficulty with tongue coordination and speech, and breathing problems. Findings such as cleft lip, cleft palate, micrognathia, and facial palsy are less frequently seen, but still observed^{2,3}. This is a case report of a pediatric patient with CS who was treated conservatively in the One Brooklyn Health Brookdale Hospital pediatric dental clinic.





Case Study

Dental Findings

Dental History:

- An 18- year old female presented to the Brookdale Pediatric Dental Clinic for her periodic recall examination
- Patient was initially seen in 2016 and was subsequently seen on a 3-month recall interval, with some gaps in care including during the pandemic
- At the time of most recent medical consult in 2021, antibiotic prophylaxis was recommended

Medical History/Allergies/Medications:

- HEENT: Coloboma, atresia chonae, Hx of cerebral abscess
- Growth retardation; Scoliosis
- Malnutrition, Chronic constipation, Pulmonary: mild persistent asthma

• Patient was found to have moderate- severe calculus

- Patient allowed hand scaling to be done; clinical calculus was removed, and teeth were examined
- No clinical caries were noted
- Periodontal risk: HIGH
- Caries risk assessment: Low (G-J tube dependent)

Medical History/Allergies/Medications

ions Oral Health Management

- Genital abnormalities, Ear abnormalities, Hydrocephalus
- Nocturnal Hypoxia
- G-J tube dependence; Global developmental delay, Optic Nerve Neuropathy
- Medications: Aspirin, Albuterol, Diazepam, Erythromycin ophthalmic ointment, Levalbuterol, Levetiracetam, Losartan, Menthol- Zinc Oxide, Mupirocin, Keppra, starch-malto dextrin, nutritional supplements
- Allergies: peaches, cefazolin, seasonal

- Three- month recalls- prophy done with hands scalers and in segments
- Non-foaming toothpaste was recommended, and fluoride varnish was applied
- Surgical History: Cardiac surgeries (ventriculoperitoneal shunt, repair of ventriculoperitoneal shunt, bidirectional glen shunt), Gastrojejunostomy, Tracheostomy

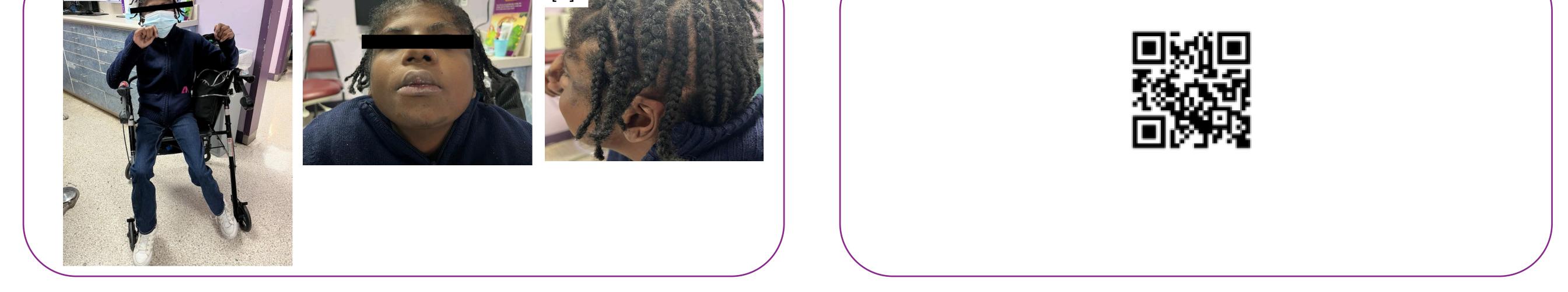
Presentation of CHARGE Syndrome

[A} Patient is wheelchair bound, Growth retardation- height= 4'2'; weight= 24.5 kg; puts patient in <1%) [B] Coloboma- patient has no view in right eye, large area of coloboma in left eye [C] Ear abnormalities- pointed at top, smaller [D] Dental findings: poor oral hygiene, gingival inflammation, hyposalivation, and no clinical caries noted [E) Moderate- severe calculus noted upon intraoral examination

Discussion

- Patients with CHARGE syndrome can have one or several congenital dental anomalies such as taurodontism, hypodontia, ectopic eruption, submergence of primary molars, agenesis and supernumerary teeth³.
- Periodontal disease is often seen due the poor oral hygiene and bruxism which can cause chronic trauma to periodontal structures
- Our patient was cooperative; therefore, prophylaxis was able to be performed effectively chairside with hand scaling and toothbrush prophylaxis
- Patient could not tolerate radiographic exam
- Some patients with CS may benefit from treatment under general anesthesia, however in this case, it was determined that the benefit of general anesthesia did not outweigh the risks due to patients' low risk of caries and her medical complexities

References



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