

Teaching Points

- Advocate for additional investigation into the dental implications of Xq21 Microdeletion Syndrome for the advancement of scientific understanding and treatment planning
- Explore non-pharmacological behavior management techniques for treating children with special needs, emphasizing alternatives to pharmacological interventions

Introduction/Background

- Drawing from select case reports, Xq21 Microdeletion Syndrome has been associated with choroideremia, cleft palate, intellectual disability (IQ <70), dysmorphic facial features, autism, schizophrenia (0.2%-0.6%)[4], and hearing impairment, with a 0.015% occurrence rate [1][4]
- Xq21 Microdeletion Syndrome is rare X-linked condition, with few cases in literature
- About 75% of all children exhibit hypotonia, 16% of children have seizures that typically begin during infancy [2], and 30% of those diagnosed may have generalized mild learning difficulties [4]
- Challenges in accessing dental care for patients with special needs exist due to a shortage of dentists knowledgeable about the necessary techniques to effectively manage their behaviors

Case Presentation

A 12 year old male presented to the dental clinic at Jamaica Hospital for dental examination. Parents had difficulty finding suitable dental care for their child and were told that he would need to be seen in the operating room for treatment.

Medical history:

- Xq21 Microdeletion Syndrome
- Autism spectrum disorder with intellectual disability and attention deficit hyperactivity disorder
- Hearing loss: diagnosed by audiology in 2012; patient is nonverbal
- Hyperactive and has a lot of behavioral issues; gets frustrated easily; hit his head when upset
- Cleft lip/palate: surgical correction in the past (date unknown)

Dentition: Mixed Dentition

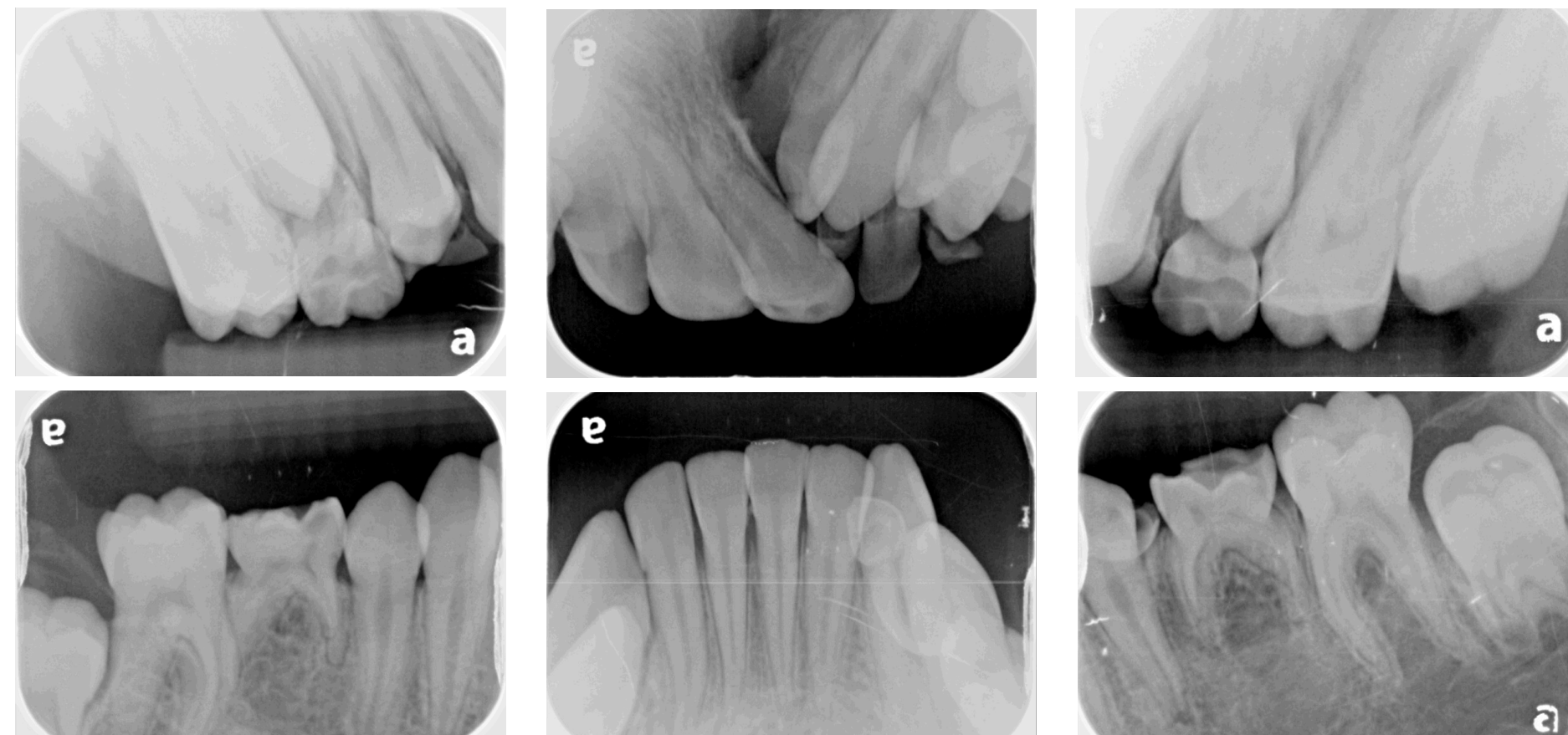
Existing restorations: None

Findings:

- Generalized moderate plaque
- No decay noted
- Over-retained primary root remnants #B, #C, #F, #G, #H, #I, #J, #M, #L
- Congenitally missing mandibular second premolars #20 and #29
- Occlusal wear noted on #A, #J, #K, #T

Sensory-adaptive techniques:

- Use of passive protective stabilization and weighted blankets



Discussion/Treatment

- This case underscores the need for further research in the Xq21 Microdeletion Syndrome. A deeper understanding of this genetic condition will enhance our ability to predict dental needs, plan treatments more effectively, and implement efficient behavior management strategies.
- Challenges arose for the parents in finding suitable dental care for their child, as some practitioners recommended sedation or treatment in a hospital's 'surgical suite' (operating room). After addressing concerns about general anesthesia and sedation, the mom agreed to conservative care with advanced behavioral management techniques.
- Leveraging passive protective stabilization and weighted blankets for deep pressure input, comprehensive care was rendered and all the patient's over-retained primary teeth were removed.
- This case highlights the potential of sensory-adapted dental environments as an effective alternative to pharmacological interventions. Creating awareness within the dental community about the benefits of sensory-adapted environments can contribute to a more inclusive approach in various dental offices and improve access to timely dental care for patients with special needs.
- This case highlights the use of behavior management choices in the clinic for a special needs patient.

References

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3. Iossa S, Costa V, Corvino V, Auletta G, Barruffo L, Cappelli S, Ceglia C, Cennamo G, D'Adamo AP, D'Amico A, Di Paolo N, Forte R, Gasparini P, Laria C, Lombardo B, Malesci R, Vitale A, Marciano E, Franzè A. Phenotypic and genetic characterization of a family carrying two Xq21.1-21.3 interstitial deletions associated with syndromic hearing loss. Mol Cytogenet. 2015 Mar 20;8:18. Doi: 10.1186/s13039-015-0120-0. PMID: 25821518; PMCID: PMC4376344.
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