A Case Report on Morquio Syndrome

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Introduction

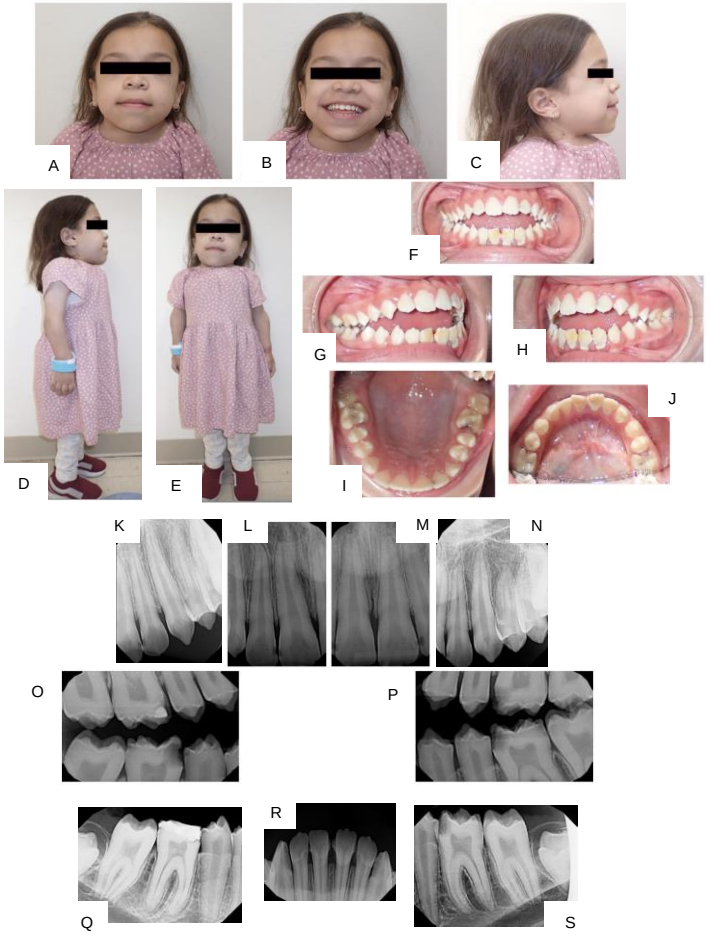
Morquio Syndrome, also known as mucopolysaccharidosis type IV, is a rare autosomal recessive lysosomal storage disorder. It is caused by a deficiency in the enzyme *N-acetylgalactosamine-6-sulfatase*, resulting in an excess of glycosaminoglycans in the bones, cartilage and ligaments. As a result, patients with Morquio Syndrome develop multiple musculoskeletal abnormalities. These include short trunk stature, cervical spinal cord compression, kyphoscoliosis, pectus carinatum (outward protrusion of ribs and sternum), genu valgum (inward tilting knees), hypermobility of joints and abnormal gait leading to increased fall risk. Other characteristics of Morquio Syndrome include cardiac abnormalities, respiratory failure, vision problems (corneal clouding), hearing difficulties, hepatomegaly and coarse facial features. Despite the numerous phenotypical changes, patients with Morquio Syndrome do not have any intellectual disabilities.

Case Report

A 14-year-old female presented with a chief compliant of dental pain resulting in tolerating only soft foods. Medical history revealed that the patient was diagnosed with Morquio Syndrome (4 years of age), eustachian tube dysfunction, sensorineural hearing loss, gastric reflux, kyphoscoliosis, mitral valve prolapse and aortic root dilation. The patient is wheelchair dependent. Past surgical history includes spinal fusion surgery, knee stabilization, hip fusion and tube myringotomy. The patient currently receives weekly Vimizim enzyme replacement therapy infusions and takes Ibuprofen as needed for generalized body pain. The patient has no reported drug allergies. The patient moved from Pakistan to the United States in 2021 to seek care and receive enzyme replacement therapy with Vimizim. The patient had multiple surgeries in Delaware and now lives in New York to continue receiving medical care.

Findings

- **Extraoral exam** (figures A-E): short stature, short neck with limited mobility, genu valgum, pectus carinatum, altered gait, scoliosis, coarse facial features and a broad nasal bridge.
- **Intraoral exam** (figures F-J): recurrent caries on upper right first molar; caries on upper left first molar, upper left lateral incisor, lower left first molar, lower left second premolar and lower right first molar; generalized calculus accumulation; sharp cusp tips; prominent mamelons on lower incisors; macroglossia; anterior open bite; tongue thrust habit.
- **Radiographic findings** (figures K-S): extensive caries in all four quadrants, taurodontism and generalized enamel hypoplasia consistent with amelogenesis imperfecta.



Treatment

- Treatment was attempted in the dental chair with nitrous oxide and local anesthesia. Due to intraoperative hypersensitivity, all remaining treatment was completed in the operating room under general anesthesia.
- Final treatment: stainless steel crowns (SSC) placed as a preventative measure on all first and second molars; #14 and #19 partial pulpotomy with SSC due to extensive caries; composite restorations #7, #8, #9, #10, #12, #29; sealant on #4, #5, #13, #20, #21, #28 as a preventative measure.

Discussion

The biggest obstacle to treatment was hypersensitivity. Thin enamel seen radiographically may be partially responsible for the morphologic changes, such as sharp cusp edges, yellow discoloration of dentition and narrow crowns. These findings may also be attributed to the hypersensitivity that this patient experienced which prevented her from being able to comfortably receive restorative treatment in the dental chair. Patients with hypoplastic dentition, such as molar-incisor hypomineralization (MIH), tend to have difficulty in achieving profound anesthesia for operative treatment due to chronic subclinical pulpal inflammation resulting in altered nerve potential. In such situations, it is critical to consider alternative local anesthesia techniques. In patients with MIH and hypersensitivity, it was found that intraosseous (IO) injections are more successful than infiltration injections. Onset of analgesia was significantly faster and pain experienced during the injection was found to be statistically lower[4].

Conclusion

Morquio Syndrome, also known as mucopolysaccharidosis type IV, presents with multiple systemic and dental manifestations that all dental professionals should be familiar with. Being aware of the findings discussed in this case report will allow providers to alter their treatment to be able to deliver quality care for those with Morquio Syndrome, and other special healthcare needs. Specifically, alter anesthetic techniques to be able to complete treatment without the potential need for general anesthesia and/or sedation.

References