Management of Oral Manifestations of Child with Heimler Syndrome-2
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ABSTRACT

Heimler Syndrome 2 is a rare, autosomal recessive mild form of a peroxisomal biogenesis disorder. Though knowledge regarding Heimler Syndrome 2 is limited, emerging research has shown sensorineural hearing loss, amelogenesis imperfecta, enamel hypoplasia of the permanent dentition, occasional or late onset retinal pigmentation, and nail abnormalities. An eight-year-old male presented to Geisinger Medical Center with a chief complaint of lack of enamel on multiple teeth. Patient’s medical history was significant for patent ductus arteriosus, bilateral sensorineural hearing loss, and biallelic mutation of the PEX6 gene. The patient’s medications included a probiotic capsule taken daily. While no abnormalities were noted, there were reports of the patient having retinal pigmentation abnormalities. An intraoral examination revealed the following clinical findings:

- Hypoplastic enamel of permanent dentition
- Dental overcrowding
- Hypoplasia or hypocalcification of permanent mandibular molars
- Hypoplasia or hypocalcification of permanent maxillary molars
- Hypoplasia or hypocalcification of #23,26 (mandibular anteriors)
- No caries present on hypoplastic/hypominalized molars.

An 8-year-old male presented to Geisinger Medical Center (GMC) Pediatric Dentistry Clinic for a new patient comprehensive examination with the chief complaint of “no enamel on his back teeth and lower front tooth.” The patient’s medical history reveals that he had a patent ductus arteriosus, bilateral sensorineural hearing loss, and a biallelic mutation of PEX6 gene. The patient’s medications included a probiotic capsule taken daily. While no abnormalities were noted, there were reports of the patient having retinal pigmentation abnormalities. An intraoral examination revealed the following clinical findings:

- Hypoplastic enamel of permanent dentition
- Dental overcrowding
- Hypoplasia or hypocalcification of #23,26 (mandibular anteriors)
- Hypoplasia or hypocalcification of permanent mandibular molars
- Hypoplasia or hypocalcification of permanent maxillary molars
- No caries present on hypoplastic/hypominalized molars.

COMPREHENSIVE DENTAL TREATMENT PLAN

Treatment options for Hypoplastic/Hypominalized Molars (#3,14,19,30):

1. No treatment. Management can include strong oral home care, fluoride varnish, and prescription toothpaste (Sodium Fluoride 1.1% Dental Cream) utilized before bed.
2. Glass ionomer Sealants. This temporary measure can help prevent caries and decrease sensitivity, in conjunction with management listed above.
3. Silver Diamine Fluoride (SDF). The benefit of SDF is that it is desensitizing and caries-arresting. However, application of this interim therapeutic medication can cause discoloration resembling brown-grey-black color on affected teeth.
4. Full coverage stainless steel crowns. This treatment option should be utilized, as tooth declines and patient’s teeth begin to clinically deteriorate. Advantages include preservation of remaining tooth structure so that definitive restorative treatment, such as crowns, can be completed, when patient is older.
5. Extraction and substitution with the second molars. This treatment is indicated when current molars cannot be maintained and continue to deteriorate. However, the second molars, which have not fully erupted into patient’s oral cavity, may have a similar appearance/condition as the first molars.

Treatment options for Mandibular Anterior Teeth (#23,26):

1. No treatment. This option would not address patient’s chief complaint of the sharp feeling with these teeth and the esthetic concerns brought forward by patient and mother.
2. Temporary white fillings with glass ionomer. This interim therapeutic measure can cause discoloration resembling brown-grey-black color on affected teeth.
3. Silver Diamine Fluoride (SDF) or glass ionomer. This interim therapeutic measure can cause discoloration resembling brown-grey-black color on affected teeth.
4. Full coverage stainless steel crowns. This treatment option should be utilized, as tooth declines and patient’s teeth begin to clinically deteriorate. Advantages include preservation of remaining tooth structure so that definitive restorative treatment, such as crowns, can be completed, when patient is older.
5. Extraction and substitution with the second molars. This treatment is indicated when current molars cannot be maintained and continue to deteriorate. However, the second molars, which have not fully erupted into patient’s oral cavity, may have a similar appearance/condition as the first molars.

REFERENCES

4. Black GC, Vernon H, O’Neill M. Emerging evidence has shown that definitive restorative treatment, such as crowns, can be completed, when patient is older.
5. Full coverage stainless steel crowns. This treatment option should be utilized, as tooth declines and patient’s teeth begin to clinically deteriorate. Advantages include preservation of remaining tooth structure so that definitive restorative treatment, such as crowns, can be completed, when patient is older.
6. Extraction and substitution with the second molars. This treatment is indicated when current molars cannot be maintained and continue to deteriorate. However, the second molars, which have not fully erupted into patient’s oral cavity, may have a similar appearance/condition as the first molars.