Case Report

Oral Health Management of a Child with Hirschsprung's Disease: An Uncommon Clinical Entity

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Abstract

This article presents a case review of a three years eight months old master with a prediagnosed rare medical condition involving the gastrointestinal tract (GIT), namely Hirschsprung's disease. This medical condition involving GIT ailment and its usual treatment can influence the mouth and the oral environment. This article attempts to present a review of the prediagnosed medical condition and its possible influence on child's overall oral symptoms and signs if any and the management of the same. It is of utmost importance for every dentist seeing children in their operatory to be well-aware of the rare conditions which possibly may have influence on the oral health, its treatment approach, and thereby the outcome of the same in case they encounter such pediatric cases at their chair-side dental practices.

Keywords: Hirschsprung's disease, medication caries, oro-motor exercises, tube feeding

Introduction

Hirschsprung disease (pronounced as HIRSH-sproongz), a gastrointestinal (GI) disease, named after Harald Hirschsprung, a pathologist from Copenhagen, Denmark, described two cases of children with megacolon in 1887.[1] Tittel in 1901 noted the absence of ganglion cells in the distal colon of a child with Hirschsprung disease, and hence, it is also called as congenital aganglionic megacolon[2] [Figure 1b and c].

It is a developmental disorder or a neurocristopathy (disease arising from maldevelopment of neural crest) caused by genetic mutations in the DNA sequence characterized by the absence of the enteric ganglia along a variable length of the intestine occurring in about 1/5000 live-born babies and is more common in boys than girls at the ratio of 4:1 as documented by Spouge and Baird in 1985.[3] Children with Down syndrome have a higher risk as well.[3,4]

Nutrition is the fundamental basis for good health and development during the early years of life till puberty. If children do not receive the right amounts of macronutrients and micronutrients, it can lead to delayed mental and motor development that can have enduring adverse effects beyond childhood.[5] Any GI disease such as Hirschsprung disease thus can cause the failure of the child to meet the expected growth targets as predicted from a simple height and weight growth chart [Figure 1a and d]. The prediction chart is concerned primarily with the intake and digestion of food and the absorption of nutrients. The mouth is a specialized part of the GI tract (GIT) and can be affected by many of the diseases encountered in other GIT regions.[6] Dental development can also be affected with delayed eruption of teeth and altered timing of the growth of the face and jaw. The quality of the dental hard tissues may be affected during the period of malnutrition with higher caries reported as a consequence.

The following manuscript aims to present few of the oral findings, the dental practitioners should anticipate in a child with known diagnosis of Hirschsprung's disease along with its management protocols. The oral health measures emphasized to the parents and the child along with few preventive measures are also explained with a brief background of the GI disease itself as an attempt to create awareness of the same.

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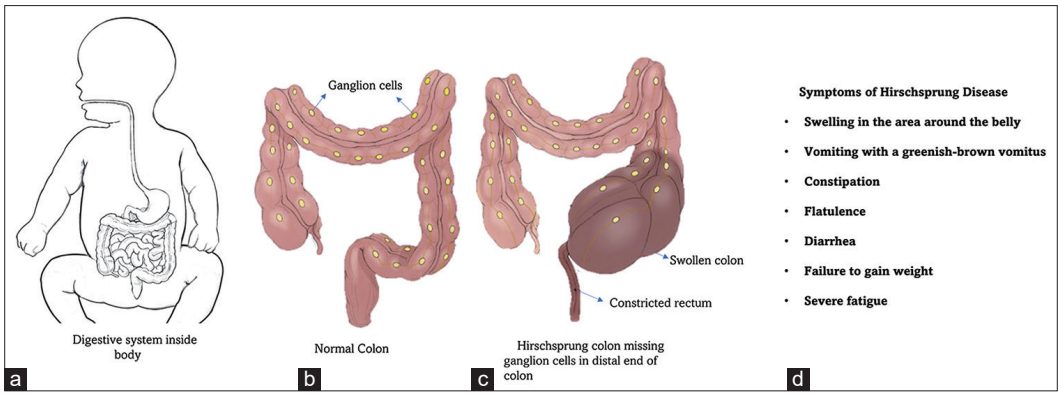

Figure 1: (a) showing normal digestive system inside the body (b) showing normal colon with ganglion cells throughout the colon (c) showing Hirschsprung disease with swollen colon and constricted rectum and missing ganglion cells in the distal end of colon (d) showing the symptoms of Hirschsprung disease.

Case Report

A three years eight months old master along with his father reported to our dental unit for generalized upper front and back teeth erosion. Upon interviewing the parents, no relevant prenatal or postnatal history was noted, there was no family history of occurrence of the disease and it was further inferred that the child was an isolated case of prediagnosed Hirschsprung's disease in the family with parents being unaffected. The child was the firstborn with a normal sibling born after 4 years. After normal term gestational birth weight of 3860gms, he developed constipation, abdominal distension, and fever. Ultimately, Hirschsprung's disease was diagnosed by barium enema and treated surgically through a procedure called pull-through surgery as reported by the attending parent.

General physical examination revealed, the child being undernourished, weak, and underweight. The oral hygiene measures were seemingly compromised with a lack of regularity due to the systemic health priorities. Clinically, the child presented with primary dentition and intraoral findings included severely demineralized and broken maxillary front teeth 51, 52, 61, 62 and decalcification of 53, 63 [Figure 2] and grossly destroyed 75 with root stump remnants.

The child was cautious but positively cooperative as per Frankel's Rating Scale no. 3 as he was familiar with the doctor approaches and hospital environment. Not much of behavioral support strategies were required apart from familiarization, tell-show-do techniques, and social reinforcers.

Treatment plan formulated was to perform thorough oral prophylaxis, restoration of 51, 52, 61, 62 with anterior strip crowns [Figure 3], following which the child was sent to seek an opinion from speech pathologist regarding the therapy sessions if required, as the child presented difficulty with few word pronunciation due to the loss of major portion of upper anterior teeth in the earlier years of life.

Extraction of grossly destructed 75 root remnants and placement of a modified distal shoe space maintainer – Band loop with distal extension bar [Figure 4a-c] as the permanent

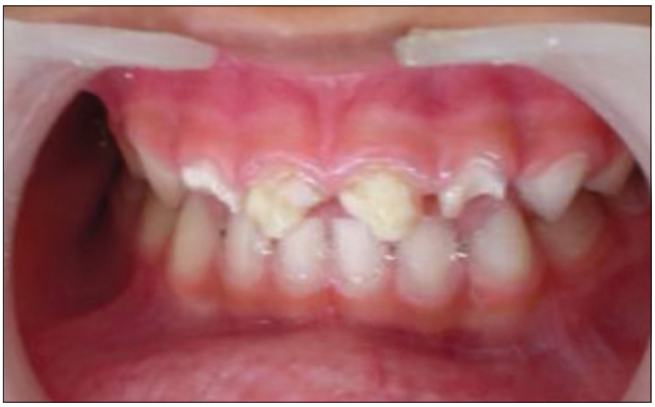

Figure 2: Showing severely demineralized maxillary anterior teeth.

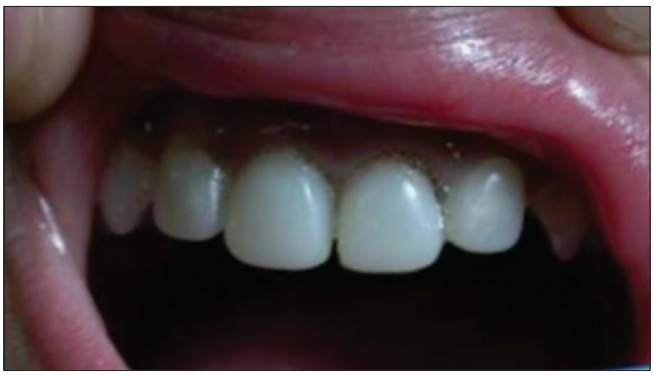

Figure 3: Showing restoration of upper anterior teeth with strip crowns.

molar 36 had not yet erupted to achieve a regular aligned teeth avoiding malocclusion was implemented. The advantage of the band loop with distal extension bar space maintainer was to avoid additional visits for impression and fabrication of another appliance but rather to cutoff the distal extension bar once the permanent molar starts erupting. This implied a better compliance from the parents and the child during extraction of the remnant root stump and impression-taking appointment.

In the follow-up visits, regular fluoride regimens, sealant application for 55, 65, 85 with diet counseling, and

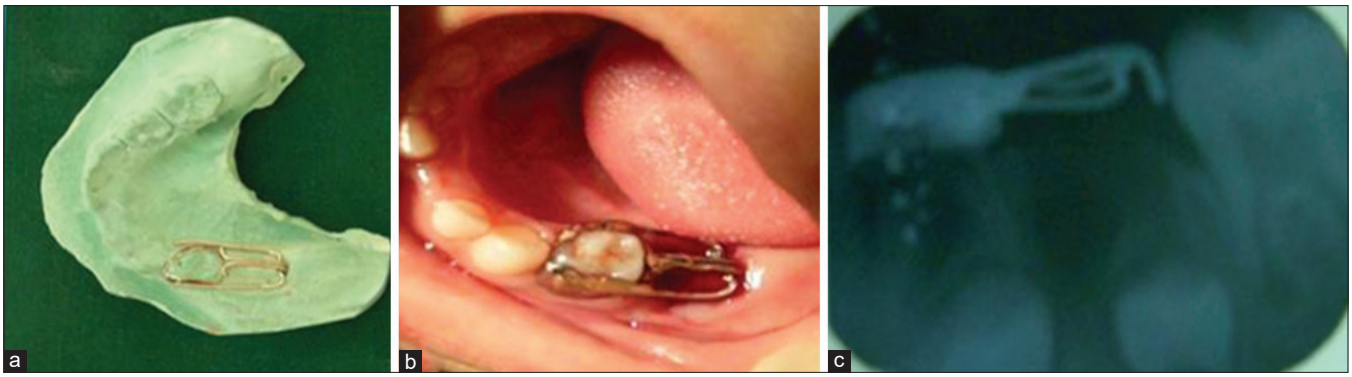

Figure 4: Showing (a) mandibular cast with band and loop with distal extension bar (b) intra oral positioning of the band and loop over 74 (c) intra oral periapical radiograph showing the position of the distal extension bar mesial to the crown of 36.

maintenance of diet chart diary were encouraged to trace the sugar intakes and snack intake frequency in-between meals. Fone's technique/circular method of brushing was demonstrated and taught to the patient with the use of remineralizing toothpaste. The authors further on, teamed up with occupational therapists specialized in oro-motor exercises (OME) and an expertise personnel on feeding skills and swallowing ability of the child by targeting the sensory orofacial regions and OMEs. These were demonstrated with modified fluid and foods, special feeding equipment, and strategies for general improvisations in the health of the child to catch up regular development and growth milestones. The progressive changes in the child's follow-up visits exhibited a marked nutritional gain and simultaneous alertness of oral health awareness.

Discussion

Hirschsprung's disease is thought to originate embryologically by maldevelopment of the neural crest involving neurologic, cardiovascular, urologic, and GI abnormalities. Conditions that have been linked to Hirschsprung's disease include congenital deafness, hydrocephalus, diverticulum of the bladder, Meckel's diverticulum, imperforate anus, ventricular septal defect, and renal agenesis.[1,4] Feeding and swallowing disorders could lead to unexplained respiratory problems and put the affected children at the highest risk for aspiration pneumonia.[7]

The oral region anomalies in children afflicted with Hirschsprung's disease as per the literature which is scarce the geographic tongue feature is documented as the most frequent find.[6] Torsiversion, an uncommon dental anomaly consisting of rotation of a tooth on its long axis, initially thought to arise from both environmental and genetic reasons has been reported with the bilateral molar involvement in few cases of Aarskog's syndrome with Hirschsprung's disease.[8] Green-colored deciduous teeth causing parental anxiety seeking cosmetic treatment for children with Hirschprung's disease have been reported in a single case report so far.[9]

Before the surgical management in Hirschsprung's disease, the children are fed by tube who usually have abundant calculus and low caries activity with increased potential for dental erosion related to gastroesophageal reflux and oral hypersensitivity. Low caries activity is attributed to the fact that reduced oral feeding decreases the amount of fermentable carbohydrates in the mouth in children fed by tube even if a "caries-producing diet" is fed through the tube it did not result in tooth decay, despite the penalty of desalivation.[10,11] Furthermore, it is important for regular oral health-care checks, to reduce the risk of aspiration pneumonia in children with Hirschsprung's disease than children who are fed orally.[7]

Apart from the above concerns, it is imperative to address few other inevitable conditions associated with long-term hospital stay, postanesthetic care, pediatric medications with cariogenic potential (sugar addition to mask the unpleasant taste of some active constituents, allowing for better acceptance by patients) leading to "Medication Caries," etc.[6,10,11] A long-term follow-up with psychological counseling, medication, and toilet training is necessary along with optimization of oral health checks, prioritizing oral care allowing for the implementation of appropriate feeding skills, and swallowing ability enhancement with supplemental OMEs for the overall well-being of the child, thereby improving the quality of life during the important growing years to achieve appropriate milestones for the development.

Conclusion

The oral health practitioner's treating children must be familiar with a range of medical problems which can affect the mouth or general health of children. The features such as weak body stature, retarded growth, or impairment in any routine milestones while monitoring the oral health should always prompt the dentist to make inquiries about the possible comorbidities associated with physical or systemic or mental conditions if any. Oral health clinicians are ideally placed to

help with the detection of a range of GI issues at times and should know when to refer to the pediatric specialist for advice. This article presents a case report of rare GI condition that can affect children, review the condition, its usual presentation and treatment, and how it can influence the mouth and the oral environment. In an attempt to create further awareness among the dental fraternity as comprehensive health care providers, we should consider both the systemic and oral health of the child.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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