The Need of Interdisciplinary Approach for the Treatment of Children with Down Syndrome with Severe Caries Unintentionally Facilitated by Hypotonia Therapy

Enrique Bimstein*/ Camilla Miskovich**

This manuscript describes a case of a 4-year-old child with Down syndrome involving hypotonia in which the use of sweets to induce oral muscular function contributed to the need to extract all the primary teeth due to extensive caries. The need for interdisciplinary education and practice targeted to the treatment of children with special health care needs is emphasized.

Key words: Caries, hypotonia, special needs.

INTRODUCTION

own syndrome (DS) is usually diagnosed on the first day of life, mostly identified by the presence of slanted palpebral fissures, epicanthal folds and hypotonia.¹⁻³ Hypotonia is ubiquitous in DS,⁴ one study reporting a prevalence of 76.3%.⁵ Most patients with Down syndrome have anterior open bite, anterior and posterior crossbite, anterior proclination and underdevelopment of the maxilla and midface, hypodontia, peg shaped teeth, enamel hypocalcifications, and motor speech disorders.^{2,3,6}

While hypotonia cannot be reversed, it can be compensated for by strengthening the muscles, including oral motor exercises involving the mouth muscles.^{7,8} Therefore, the treatment of children with DS may include language intervention, and oral motor and sensory stimulation utilizing speech therapy and palatal plates.⁷⁻¹⁰ The oral stimulation may be accomplished with an electric tooth brush, a chewy tube bitten up to 10 seconds, and by motivating the use of the oral muscles with sweets.^{7,8}

This manuscript reports a case of a 4 year old DS patient who was treated for oral hypotonia with muscle stimulating objects covered with ice cream and sweets, unintentionally leading to carious destruction all the primary teeth. This case strongly emphasizes the need for interprofessional education and practice are essential to achieve adequate comprehensive treatment for children with special health care needs.

Send all correspondence to: Enrique Bimstein Levona 70, Jerusalem, Israel, 93843 Phone: +972546267280 Email: bimsteine@gmail.com

Case Report

A 4 year old child (AH) was referred to a university health center pediatric dentistry clinic for dental treatment under general anesthesia (GA). The child had DS involving muscular hypotonia, asthma, developmental delay and "unclear spinal issues", received Albuterol for the treatment of asthma, Fluticasone spray for the treatment of nasal allergy symptoms,¹² and Montelukast to prevent asthma attacks.¹³ AH was born without chewing instinct and for the treatment of hypotonia from birth to age 18 months speech therapists used candy and a chewing tube dipped in ice cream to induce chewing and oral muscle movements. It was unclear if AH ever had oral pain related to dental caries due to his inability to communicate.

The oral examination revealed extremely severe early childhood caries affecting all the primary teeth, to the point that several teeth had pulp polyps (Figure 1). With AH under GA, radiographic and clinical examinations were followed by an unsuccessful attempt to restore his primary teeth, since the vast carious lesions did not allow to provide successful dental restorations (Figure 1). Therefore, all the primary teeth were extracted followed by 3.0 chromic gut sutures for hemorrhage control. The teeth appeared to be smaller than normal, and concrescence of the distal root of a mandibular right first primary molar with the mesial root of the mandibular right second primary molar was evident (Figure 1). Two days after treatment, clinical evaluation revealed that the patient did not develop any significant post-op discomfort or fever, and the extractions sites were healing well. A phone conversation with AH's mother 3 weeks after the treatment indicated that the patient had gradually gotten used to eat with no teeth, returning to eat "whatever he wants" and actually gaining some weight.

^{*}Enrique Bimstein, Professor Emeritus, University of Kentucky Levona 70, Jerusalem Israel

^{**}Camilla Miskovich, Pediatric Dentist, Private practice.



Figure 1. Clinical picture of primary teeth extracted due to extensive caries lesions. Note the pulp polips in teeth 1 and 2, and the concrescence of the roots in tooth # 3.

DISCUSSION

The treatment of hypotonia by speech therapist is most significant since it targets the improvement of basic oral functions such as chewing, swallowing and speaking that are fundamental for the child's function and development. The present case is an example of unawareness of the etiology of dental caries while treating developmental or systemic diseases in individuals with special needs: the basic concept that the coexistence of susceptible teeth, bacteria and carbohydrates may eventually lead to caries, especially in the absence of adequate oral hygiene. In the present case, it may be argued that the development of caries was also facilitated by enamel defects that may be found in children with Down syndrome,² however, it is likely that the main culprits for the extreme severity of dental destruction in the present case was the frequent use of sweets for oral stimulation and a lack of adequate oral hygiene during and after the oral hypotonia therapy.

A hypotonic child is floppy and may have difficulty in maintaining head support or a straight back while sitting.¹⁵ Involvement of the face, tongue, palate and extra ocular muscles provides an important distinction in the differential diagnosis of neuromuscular disorders.¹⁶ In fact, low muscle tone or hypotonia is a major contributor to the typical differences between movements performed by persons with and without DS, due to laxity of ligaments, more compliant tendons, and shorter limb segments that provide less than half of the inertial resistance to motion in persons with DS.¹⁷ Hypotonia is highly prevalent in infants with DS, it is defined as decreased resistance to passive muscle stretching and therefore, individuals with DS have inherent joint laxity resulting in reduced gait stability, increased energetic costs for physical exertion and low muscle tone in the facial muscles resulting in an open mouth posture.^{3,4}

When a child presents a feeding problem a comprehensive assessment needs to evaluate what oral motor pattern the child is using, and also what factors (medical, motor or learned patterns of behavior) are contributing to maintain the child's use of that pattern.¹⁸ Children with hypotonia, like in the case of children with Down syndrome,^{3, 10} may require oral stimulation as a preparation

for eating, with oral-motor practice followed by oral stimulation to practice food manipulation, transport and swallow.¹⁸ Oral stimulation should provide the child with the necessary sensory and movement input to adequately prepare the child for controlled practice with food, the use of varying foods is the key to increasing the frequency of positive practice and thus increasing the acceptance of variety and volume of food consumed in children exhibiting oral-motor-difficulties.¹⁸

The treatment of hypotonia is complicated, patients with low oral muscle tone have a limited understanding of what is happening in their mouths and therefore, providing stimulation will help awaken their sense, and the treatment of oral hypotonia may include "fun exercises" such as using a z-bive (oral stimulator for speech and feeding), an electric tooth brush to help regulate the oral sensitivity, biting hard on a chewy tube, making funny faces, blowing bubbles, whistling, using straws to drink, mouth games, and with a lollipop either by using a preferred taste, encouraging the patient to stick the tongue out to try and reach for the lollipop, and after a small taste start moving the lollipop with their tongue as giving them a small reward consisting of occasional taste and reminder of what they are working for, or licking suckers in various positions.^{7,8}

While the treatment of hypotonia is most relevant, it is crucial that the treatment be based on an interprofessional collaborative model of healthcare, that optimizes the use of multiple professional skills set to provide well-coordinated collaborative high quality patient centered care.^{19, 20} In general, health care of children with special health care needs should be based on a team approach to include the patient's pediatrician, pediatric dentist, specialist on the different medical disciplines such as neurologist, physical therapist, neurologist, etc. based on the individual patient characteristics. This, in addition to parental education and involvement in the child's management, and compliance with suggested preventive practices as early as possible.¹¹

Many individuals with Down syndrome have oral, anatomical, and structural differences that predispose them to potential feeding problems, detailed feeding assessment have shown both delays and aberrant oral-motor function with abnormal -tongue /jaw function and difficulty initiating and maintaining the feeding sequence; commonly reported feeding concerns of parents were suck and lip closure, difficulty chewing, and uncoordinated swallow leading to chocking and gagging ²¹ Despite that Down syndrome children have been typically reported to have lower or similar caries experience than children without Down syndrome,²² health providers most take in consideration that he dental biofilm of children with Down syndrome may have a higher cariogenic potential than that of children without this condition, oral hygiene in children with Down syndrome is worse than in children with no Down syndrome, and self-cleaning in children with hypotonia may be reduced resulting in more food remaining on the teeth after eating.3, 23, 24 Risk assessment is a key element of contemporary care of infants, children and adolescents, and persons with special health care needs.²⁵ Furthermore, dental caries-risk assessment, based on the child's age, biological factors, protective factors, and clinical findings should be a routine component of new and periodic examinations by oral health and medical providers.26

The finding that our patient teeth were found to be smaller than normal is consistent with the report that 13% of children with Down syndrome may show microdontia in the permanent dentition;^{27, 28} the present case reporting microdontia in the primary dentition in a child with Down syndrome. Concrescence is a twining anomaly, consisting of a form of fusion that occurs after root formation, the teeth being united by cementum only, with individual root canals and root formation; it may be related to trauma or crowding with resorption of the interdental bone so that the roots approximate and fuse by the deposition of cement between them, usually involving only two teeth.^{29, 30} Our search of the literature revealed that concrescence in the primary dentition is infrequent and usually affecting the anterior teeth.³¹ It appears therefore, that this is the first case in which concrescence has been reported affecting primary molars.

CONCLUSIONS

- 1. Hypotonia in infants is a most significant problem that requires immediate effective treatment since its success is crucial for the child's function, development and growth.
- 2. Health providers who treat hypotonia in children must be aware of the possibility of dental carious destruction that may be induced by the frequent presence of sweets in the oral cavity, and that dental pain, infections and extractions significantly reduce the child's quality of life.
- 3. Unambiguously, the adequate treatment of hypotonia in children requires interprofessional collaboration which will lead to a balance of adequate oral stimulation by non-cariogenic means and diet and proper oral hygiene to prevent or at least minimize the occurrence of oral diseases.
- 4. Interprofessional practice emphasizes teamwork, understanding other professions' roles and responsibilities, decreasing the possibilities of medical errors, with the ultimate goal to improve patients' safety and health outcomes while addressing the changing needs of the patient ³².

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