

## Dental Management Consideration of Severe Early Childhood Caries in Down Syndrome with Autistic Features: A Case Report

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### Abstract

Behavioral problems are reported to cooccur with Down syndrome (DS). Some children need support in oral care due to their intellectual disability. In addition, medical problems are present in DS. All these conditions make it challenging to perform dental care because communication is burdensome, and patients are prone to high-risk infection during dental treatment.

A 3-year-old girl with DS and autistic features came to the clinic because she was unable to eat. The echocardiogram showed ASV (-), VSD (-), and PDA (1.5 mm). The intraoral examination revealed severe early childhood caries. The dental treatment was conducted under general anesthesia, which included filling, pulp therapy, and extractions.

This case report emphasized the need for a multidisciplinary approach for planning dental treatment for DS children and enhancing the importance of improving oral health to prevent infections caused by oral microbial flora in special needs children.

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### Introduction

Down syndrome (DS) is the most well-known genetic condition caused by chromosomal aberrations resulting in the presence of an additional third chromosome 21, which is known as trisomy 21. Chromosomal abnormalities are responsible for physical features, intellectual disability, and developmental delay, which affects the typical characteristics of DS people.<sup>1</sup>

Genetic conditions contribute to dental and oral manifestations. Delayed tooth formation and eruption can be observed in children with DS, 2-3 years behind the normal eruption in healthy children. Hypodontia, congenitally missing teeth, and malformations of the teeth can also be found. Generally, children with DS have more severe periodontal disease due to local risk and general risk factors, including immunodeficiency.<sup>1</sup> Several reports have shown a lower incidence of dental caries in DS children. In contrast, another study reported that there was no difference

between DS and non-DS children.<sup>1-3</sup>

Several clinical manifestations relating to oral conditions and medical problems with oral health consequences make the DS children need to receive support from dental health care professionals.<sup>4</sup> However, there are some challenges for DS children to receive dental treatment. Children with DS have problems with neurological and behavioral alterations. Intellectual disability, the most frequent clinical manifestation in DS children, impacts the ability of children with DS to perform personal or oral care. In addition, some children with DS cooccur with autism, attention deficit hyperactivity disorder (ADHD), depression, or obsessive-compulsive disorder.<sup>1,5,6</sup>

Some individuals may need more support than others. Moreover, DS children have a high risk of medical conditions. Congenital heart disease is present in up to 50% of patients with DS.<sup>7</sup> There is susceptibility to complications during dental treatments for children with DS. Here, we reported dental management considerations in children with DS.

### Case Report

A 3-year-old girl accompanied by her mother and father visited the Dental Clinic of

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Universitas Indonesia Hospital. The parents reported that the child refused to eat and was unable to chew solid food in the past 3 days. The child used a bottle to drink milk at bedtime regularly. Therefore, she only drank milk instead of eating food for the past 3 days. This time was her first visit for a dental appointment. The parents suspected that the child's unusual eating behavior was caused by her teeth condition.

The child was diagnosed with DS and showed autistic features. She was unable to make eye contact, could not speak, had speech delay, was hyperactive and aggressive, and had unusual behaviors and temperaments. The parents declined to indicate whether the child had an allergic condition and were receiving routine prescribed medication. The parents admitted that they had difficulty maintaining oral care daily for the child at home due to her hyperactive behavior. She attended speech therapy and occupational therapy classes regularly.

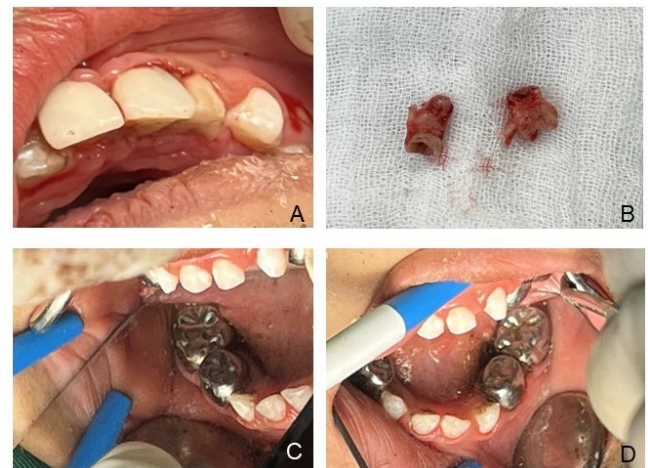
The initial dental evaluation was performed with protective stabilization with her parent's consent. The intraoral examination showed a "V"-shaped palate, missing mandibular lateral primary incisors, poor oral hygiene, and caries lesions on all primary molars and maxillary incisors (Figure 1).



**Figure 1.** Preoperative Intraoral Conditions.

We decided to perform dental treatment under general anesthesia as an inpatient procedure. After consultation with a pediatrician and anesthesiologist, the child was referred to a cardiologist for echocardiography. There were no

arterial septal defect (ASD) and ventricular septal defect (VSD), the size of the patent ductus arteriosus (PDA) was 1.5 mm, the left ventricular ejection fraction (LVEF) was 68%, and there were no leaky heart valves. The dental treatment under general anesthesia was approved. Following MTA pulpotomy procedures, the mandibular primary molars were restored with stainless steel crown. The maxillary first primary molars were extracted. The first maxillary left incisor was treated with a one-visit endodontic, followed by a composite crown. The first maxillary right incisor was restored with a composite crown. The second maxillary molars and incisors were treated with glass ionomer cement. (Figure 2)



**Figure 2.** Postoperative intraoral conditions A. Composite crown for maxillary central primary incisors. B. The remaining maxillary first primary molars (left and right) after extraction. C–D. Stainless steel crown for mandibular primary molars.

Cefixime and ibuprofen were prescribed posttreatment. The child's condition was monitored for the next 24 hours posttreatment with extra caution during the first hour after treatment. The intravenous fluid therapy was stopped after 9 hours posttreatment. The child was discharged from the hospital one day after treatment when she was asymptomatic and was able to ingest soft textured food.

The child came to the dental clinic as an outpatient after 7 days posttreatment. Her mother reported that the child was able to eat. The intraoral examination was performed with the aid of protective stabilization with the consent of her

parents. Parents were taught and motivated to brush their child's teeth at this time.

The patient is currently undergoing follow-up care. She has good oral hygiene.

### Discussion

Down syndrome patients present typical phenotypic, cognitive, and behavioral characteristics. Down syndrome is the most common genetic condition leading to mental retardation (MR) at mild (IQ 50-70), moderate (IQ 35-50), or severe (IQ 20-35) levels. People with MR usually have emotional, behavioral, and psychiatric problems. Although DS children are known to be friendly, good-tempered, affectionate, and sympathetic, they also exhibit externalizing behaviors, including hyperactivity, inattention, tantrums, agitation, impulsivity, repetitive movement, sensory dysregulation, and speech problems.<sup>5</sup> Children with DS reported co-occurring diagnosed with attention-deficit/hyperactivity disorder (ADHD), autism spectrum disorder (ASD), or behavioral or oppositional disorders.<sup>5,6</sup> These behavioral symptoms may manifest as early as 2 or 3 years of age.<sup>8</sup> The dentist should understand the child's cognitive function, liking and disliking, and any physical limitations in the initial assessment.<sup>9</sup> This information will help to build rapport between the dentist and the patient. In the present case, the MR and behavioral conditions led to a communication challenge between the DS child and the dental and medical team. During the first visit at the in-office appointment, protective stabilization was provided to deliver immediate examination/ treatment while protecting the safety of the patient, parents, and dental team. Children who lack maturity and development (emotional and cognitive) are indicated to have stabilization when they are uncooperative but require immediate diagnosis or urgent treatment.<sup>10</sup>

Based on the intraoral examination, the child exhibited several characteristics of DS oral features, including a "V"-shaped palate and hypodontia on mandibular lateral primary incisors. DS children have phenotypic oral manifestations. The typical oral feature in DS children is a narrowing "V"-shaped palate with a high arch and anterior open bite. The oral mucosa characteristics of DS children are fissured tongue, geographic tongue, cheilitis, oral candidiasis, and macroglossia. Other oral anomalies in DS

patients include periodontal diseases, enamel hypocalcifications, microdontia, diastema, agenesis, and delayed tooth eruption.<sup>1,11</sup> These conditions could lead to malocclusion. Approximately 85% of DS people have malocclusion. However, although the demand for orthodontic treatment is high, the result is often unsatisfactory because of poor oral hygiene and lack of compliance.<sup>12</sup> Therefore, in this study, we did not have plans for orthodontic treatment due to the age, compliance, or oral hygiene habits of the patient.

The dental caries rate of this patient was considered high due to the involvement of all primary molars and upper primary anterior teeth, which led to the diagnosis of severe early childhood caries. The presence of caries lesions, both cavitated and non-cavitated, indicates the presence of risk factors for the disease.<sup>13</sup> Although studies have reported that DS children have a lower prevalence of caries or are equivalent to individuals in the control group, dental caries may occur due to several risk factors (difficulty obtaining dental care access, maintaining good oral hygiene, reducing manual dexterity, or parental neglect).<sup>2,3,11</sup>

The patient had extensive caries lesions at a young age, and her mental condition made her unable to cooperate in the dental office. Therefore, the patient was recommended to receive dental treatment under general anesthesia. When working with people with special health care needs, clinicians prefer to perform comprehensive dental treatment under general anesthesia for safety and efficacy reasons.<sup>14</sup> Features of manifest DS affect almost every organ system. Therefore, a comprehensive multidisciplinary assessment is needed.

Children with DS have a higher risk of medical problems than do the general population. CHD is a comorbidity in DS, and half of DS children have CHD. Single or multiple malformations, including structural or functional cardiac defects, present at birth or can even be discovered later in life. ASD and VSD are the most common CHD associated with DS. Meanwhile, PDA is the least minor CHD in DS.<sup>7</sup> Children with CHD are at risk for infective endocarditis (IE), an infection at the endocardial surface of the heart. IE may occur when bacteremia derived from the oral cavity attaches to one of the abnormal valves.<sup>15,16</sup> People with

DS and CHD should attend regular dental care to minimize IE.<sup>7</sup>

Antibiotic prophylaxis for dental procedures has been recommended to prevent IE in CHD patients with a high risk for IE, although the proven benefits have still been debated. The dental procedures in which antibiotic prophylaxis is suggested for patients at high risk for IE involve manipulation of gingival tissue or the periapical region of teeth with perforation of the oral mucosa.<sup>15,16</sup>

In the present case, the echocardiogram showed no signs of ASD or VSD, the predisposing factors to IE. Although echocardiography revealed a small size PDA in this patient, the risk of IE was lower in these patients than in those with VSD.<sup>17</sup> We did not administer antibiotic prophylaxis before treatment since there was no evidence of a high risk for IE. However, we administered antibiotics as an adjunct therapy after extraction.

There are no specific guidelines for pulpal involvement in primary teeth in DS patients with CHD. Several authors have suggested that pulp therapy is unsafe, and extraction is recommended for CHD patients. Other studies reported that half of the respondents chose to perform pulp therapy on vital primary teeth and preferred to perform extraction of irreversible primary teeth in CHD children. Pulpotomy of reversible primary teeth has a high success rate. Moreover, multiple extractions of primary teeth in young children may cause adverse outcomes, such as mastication, growth, speech, and psychological development.<sup>18</sup>

The initial intraoral examination on the first visit revealed poor oral hygiene. The caries risk assessment for this child was considered high risk because the child still had a nursing bottle habit, the parent was unable to maintain the child's oral home care, and the child had special health care needs. In addition to being immature, the patient showed a learning disability due to neurological and behavioral alterations, which are part of the clinical manifestations of DS. Oral home care is essential for these patients. However, it was very challenging for both parents to build tooth brushing habits in the presence of behavioral problems related to the procedures and non-procedures. The child might dislike the feeling of the toothbrush in the oral cavity or the taste of the toothpaste. It is common for parents of autistic children to feel worried about their

child's aggressive behavior during tooth brushing.<sup>19</sup> To build good habits for oral home care for this child with DS with autism, cooperation among parents, dentists, and occupational therapists is necessary.

## Conclusions

In conclusion, dental treatment planning for Down syndrome children requires a multidisciplinary approach because maintaining good oral health to prevent infections caused by oral microbial flora in children with special health care needs is essential.

## Declaration of Interest

The authors report no conflict of interest.

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