

Introduction

Prader-Willi syndrome (PWS) is a rare neurodevelopmental disorder caused by abnormal expression of the 15q11.2–q13 region on chromosome 15, with a prevalence estimated at approximately one in 10,000 to 30,000 individuals. A hallmark feature of PWS is hyperphagia-induced obesity resulting from abnormal eating behaviors. Other systemic manifestations include hypotonia, delayed motor development, hypothalamic hypogonadism, growth hormone deficiency, intellectual disability, and behavioral problems such as temper outbursts, obsessive-compulsive behaviors, and emotional lability. Characteristic craniofacial features include a narrow forehead and downturned corners of the mouth. These systemic and endocrine abnormalities can negatively affect oral health in patients with PWS. Common oral findings include multiple dental caries, enamel hypoplasia, delayed tooth eruption, tooth wear, poor oral hygiene, gingivitis, and mouth breathing. Altered salivary characteristics—such as high viscosity, elevated ion and protein concentrations, and decreased flow rate—further elevate the risk of dental caries. Due to challenges related to airway management, risk of aspiration, and behavioral difficulties, dental treatment often necessitates sedation or general anesthesia. This case report presents two pediatric patients with PWS who underwent dental treatment under sedation and general anesthesia, highlighting the clinical considerations and management challenges associated with their care.

Case 1

Patient Information

Age / Sex: 8 years 3 months / Male
 PMH: Prader-Willi syndrome
 Chief Complaint: Regular follow-up, dental caries
 Present Illness: Dental caries involving all four quadrants of the dentition
 Congenital missing of bilateral mandibular second premolar

Pre-operation



Fig 1. pre-op panorama

Fig 2. pre-op PA view



Fig 3. pre-op intraoral photo

Operation

◆ All second primary molars were restored with stainless steel crowns to ensure full coronal coverage. In particular, for the bilateral mandibular second primary molars, where the permanent successors are congenitally missing, restoration with stainless steel crowns was selected to preserve the teeth for long-term function. All first permanent molars were restored with composite resin restorations. Dental caries on the bilateral maxillary primary canines and the maxillary left first primary molar were planned for observation, considering the extent of caries and the anticipated timing of exfoliation.

Case 2

Patient Information

Age / Sex: 2 years 8 months / Male
 PMH: Prader-Willi syndrome, type 2 Diabetes Mellitus
 Chief Complaint: Dental caries
 Present Illness: Dental caries involving all primary teeth

Pre-operation

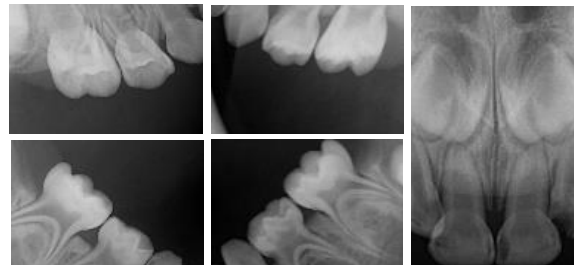


Fig 4. pre-op PA view

◆ Extensive dental caries were noted on all maxillary primary incisors and both mandibular primary central incisors. Carious lesions were also present on all first primary molars and the bilateral mandibular second primary molars.

Operation

◆ Pulpectomy followed by celluloid crown restoration was performed on the bilateral maxillary lateral primary incisors. Composite resin restorations were placed on both maxillary first primary molars and the mandibular second primary molars. The mandibular left first primary molar underwent pulpectomy and was subsequently restored with a stainless steel crown. The mandibular right first primary molar was restored with a stainless steel crown without pulpal treatment. Both mandibular central incisors were restored using glass ionomer cement. Professional topical fluoride was applied.



Fig 5. pre-op PA view

Additional operation

◆ Approximately 10 months later, the patient returned with a chief complaint of gingival bleeding in the anterior region. Pulpectomy was performed on the maxillary left primary central incisor, followed by capping with a calcium hydroxide material (Ultra-Blend®).



Fig 6. 2.5 year post-op panorama

Follow-up

◆ Following comprehensive caries treatment under sedation, the patient was placed on a three-month recall schedule for caries management and oral hygiene reinforcement. At five years of age (2.5 years post-treatment), the first panoramic radiograph was obtained to assess the development of permanent tooth buds, evaluate the status of previously restored teeth, and monitor for new caries.

Summary

Considering the altered salivary composition and increased viscosity in patients with PWS, along with difficulties in maintaining oral hygiene due to limited cooperation, enamel hypoplasia and dental caries have been reported in approximately 40% of cases. However, outpatient dental treatment is often challenging because of the risk of aspiration, behavioral difficulties, and obesity. Physical restraint is frequently not feasible, and sedation or general anesthesia is often required, as demonstrated in this case report. Therefore, prevention of obesity and dental caries is essential in patients with PWS through growth hormone therapy, appropriate weight control, and dietary modification to reduce sugar intake. Close caregiver supervision, regular dental check-ups, and preventive care are important to improve patient cooperation and facilitate outpatient treatment whenever possible. Continuous oral health management and more frequent dental visits are recommended to reduce the need for general anesthesia during dental procedures.