



# Prader-Willi Syndrome: Periodontal-prosthodontic rehabilitation in an adult patient

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## ABSTRACT

Prader-Willi syndrome (PWS) is a rare genetic multisystemic disease that is the most common inherited cause of severe childhood obesity. PWS patients are prone to significant oral and systemic health issues that detrimentally affect quality of life and decrease longevity. This report documents full-mouth pre-prosthetic surgical and restorative care in an adult PWS patient.

## INTRODUCTION

Prader-Willi syndrome (PWS) is a rare genetic disorder resulting from partial or complete chromosome 15 deletions that manipulate gene expression. The occurrence frequency of PWS has been reported to be from 1:10 000 to 1:30 000 births.<sup>1</sup> Major symptoms early in life include hypotonia (decreased muscle tone) leading to feeding problems, and conversely, later in childhood, hyperphagia which can progress to morbid obesity and a shorter life expectancy. In a 40-year mortality survey by Butler et al. it was reported that respiratory failure was the most common cause of death (31%), and a 99% mortality rate was reached by the age of 60. Short stature with increased fat composition, and low levels of growth and sex hormones are regularly observed, along with mild to moderate intellectual deficiency. The impaired cognition may lead to significant behavioral and social issues. Recombinant human growth hormone (rhGH) until adulthood is a mainstay of treatment, along with diet, exercise, and behavioral/mental therapy. Once the PWS patient reaches adulthood, however, the focus usually becomes one of treating multiple chronic problems resultant from this condition. Oral health conditions commonly seen in PWS children and adults include rampant caries, minimal thick salivary flow, enamel hypoplasia leading to severe dental erosion and attrition, candidiasis, delayed dental eruption with possible malocclusion and periodontal disease. High refined carbohydrate intake combined with inadequate oral hygiene can further advance the complications seen in this patient population, as well as higher rates of diabetes, respiratory problems, and cardiovascular issues. Continual support and maintenance by family/caregivers is imperative for the patient's well-being, and to increase the likelihood of achieving longevity. Due to the rareness of PWS, documentation of successfully treated adult cases in the literature is uncommon. There is only one previously published PWS case in periodontology journals, and that was 32 years ago in a pediatric patient. This case report documents full-mouth periodontal surgical and prosthodontic treatment with follow-up in a PWS adult patient.



Figure 1: Pre-op Smile



Figure 2: Pre-op maxillary arch



Figure 3: Pre-op mandibular arch



Figure 4: Post-op smile



Figure 5: Post-op maxillary arch



Figure 6: Post-op mandibular arch

## Case Report

Preliminary impressions were made using an irreversible hydrocolloid impression material and poured into a type IV gypsum product. A facebow transfer was used to mount the maxillary cast on a D5A Denar articulator. A centric relation record was made using an anterior deprogramming jig and a rigid polyvinyl siloxane registration material (Extrude). The centric relation (CR) record was verified three times intraorally. After assessment of the VDO, the patient's existing VDO was adequate and maintained. Diagnostic casts were duplicated and cross-mounted using the CR record. A diagnostic wax-up was completed to plan the anticipated occlusion and the potential esthetic outcome. Periodontal *full mouth crown* surgery was completed with a prefabricated surgical guide created from the diagnostic wax-up. Under intravenous sedation, mucoperiosteal flaps were reflected and clinical crown lengthening was completed in two appointments, one arch at a time.

After a 4 week healing period, existing composite restorations were removed, and all teeth were inspected for secondary caries and evaluated for restorability. Mechanically retained composite resin was used to restore and block out any undercut. VDO was re-established with good function and esthetics. Provisionals were fabricated from diagnostic wax-up utilizing laboratory and milled PMMA. The patient wore the completed provisional restorations for 8 weeks. The patient was happy with the esthetics, phonetics, occlusion, and comfort of the provisionals. Impressions were made of the provisional restorations with irreversible hydrocolloids and poured into type IV gypsum stone. Custom impression trays were fabricated. Teeth preparations were refined for final impressions. Gingival retraction was done using a double-cord technique.

The final restorations were inserted. Marginal fit was confirmed. The occlusion and interproximal contacts were checked with shimstock and adjusted as needed and polished thoroughly. Self-adhesive dual cure resin cement was used for sequential cementation of all ceramic restorations. Closure of all margins and interproximal contacts were confirmed after cementation with radiographs. A mutually protected occlusion was achieved. An occlusal device with an anterior ramp was fabricated for the patient.

Completion of treatment was successful, and the patient was placed on a lifelong recall regimen every 3-month periodontal maintenance interval, with professional and at-home maintenance regimen for tooth-born restorations to provide biological and mechanical maintenance of the health of the hard and soft tissues. The patient's attitude, motivation, and the outcome of his treatment support an overall good long-term prognosis.

## Treatment Plan

- 1. Oral prophylaxis and oral hygiene instructions.
- 2. Prescription for fluoridated toothpaste and Chlorohexidine Gluconate oral rinse.
- 3. Determine incisal edge position.
- 4. Analog wax up.
- 5. Full mouth crown lengthening.
- 6. Removal of existing restorations to assess restorability of the existing dentition and restoring the coronal structure with new foundation material.
- 7. Provisionalization of all teeth after a 4-month healing period.
- 8. PMMA (polymethyl methacrylate) provisional phase to test the occlusion design, existing VDO (vertical dimension of occlusion), patient comfort, function, and esthetics.
- 9. Root canal treatment if needed due to severe tooth sensitive.
- 10. Cement-retained full mouth lithium disilicate ceramic restoration.
- 11. An occlusal device at the completion of treatment.
- 12. Periodontal and prosthodontic maintenance every 3 months (CAMBA Protocol).

## Discussion

There are only two cases of periodontal patients with PWS in the literature, and only one of these was in an adult. This report is the first to document full mouth surgical and prosthetic reconstruction in a PWS patient. There is no doubt that the severe malocclusal discrepancies in this patient were strong secondary etiological factors leading to difficult plaque control in an already compromised rehabilitate with PWS. A lot of time was spent with patients and guardians on disease etiology and needed oral hygiene and long-term maintenance. There are previous reports in the literature stressing the importance of family involvement in the motivation and maintenance of oral health and hygiene habits. It is essential that family and/or caregivers be involved early on with PWS patients' daily hygiene needs. Only with continued follow-up will such involved treatment reach lasting success. This report highlights how an adult PWS patient can successfully manage later life challenges with much-needed special care and therapy when available. Although this patient was usually very cooperative and in adulthood, due to the long, multiple periodontal-prosthodontic appointments needed from start to finish, these treatments were made much easier for practitioner and patient utilizing IV conscious sedation. This maximized efficiency for all involved, to include the patient. Roman-Torres et al. recently described a pediatric PWS case successfully treated in a hospital environment with general anesthesia, underscoring how sedation should be considered versus conventional treatment in this category of patients. It is the authors' conclusion that sedation, in this case, was imperative and that complete oral rehabilitation would have been extremely difficult to complete without it. Special care patients should be afforded the accessibility to all regimens that enable needed care to be delivered. Disorders such as PWS can present practitioners with unique challenges, but with a multidisciplinary team approach these patients can get needed support and live a normal lifespan.