

A CASE REPORT

Prosthodontic rehabilitation of a patient with Sturge Weber Syndrome using modified Cast Partial Denture

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ABSTRACT

Background: Dentists must possess a thorough understanding of the clinical characteristics and treatment options associated with SWS to address its oral manifestations effectively. Collaborative consultations with diverse medical experts are essential for optimal treatment which aids both the patient and their family in navigating challenges and enhancing prognosis. Regular systemic and oral assessments are advisable to mitigate the risk of cranial and oral complications.

Case detail: A 46-year old female patient reported to the dental OP with the complaint of inability to chew foods. The patient presented with unilateral portwine stain of the face, glaucoma of the right eye, and frequent headaches. Intraoral examination revealed hypertrophied right maxilla, and multiple missing teeth with collapsed bite. The patient was diagnosed with SWS and a simple and conservative prosthetic rehabilitation with a cast partial denture having a unique major connector design was planned for the patient to fulfill both masticatory and esthetic needs.

Conclusion : When rehabilitating the oral structures, it is crucial to consider the patient's overall health rather than solely focusing on achieving function and aesthetics through surgical procedures. We must carefully weigh the benefits and risks to ensure the best outcome.

Key words: Sturge Weber Syndrome, prosthodontic rehabilitation, major connector, non-invasive approach

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INTRODUCTION

Sturge-Weber Syndrome (SWS) is an uncommon congenital neurocutaneous disorder marked by a triad of cutaneous, neurological, and ocular anomalies.¹ Most commonly observed features include a unilateral port wine facial stain, leptomeningeal angioma, and elevated ocular pressure, predisposing individuals to glaucoma.² Oral management in Sturge-Weber Syndrome (SWS) encompasses more than just controlling bleeding from hemangiomas; it also involves addressing issues like poor oral hygiene, malocclusion, and gingival enlargement.³

It's crucial to ensure that patients don't experience anxiety, as heightened blood pressure could potentially exacerbate lesions in the brain and eyes.⁴ Additionally, patients with SWS may be on multiple medications for epilepsy, hemorrhage control, and behavior management, which can further complicate treatment outcomes. Therefore, a comprehensive approach to oral care must consider these factors to optimize patient health and well-being.

Prosthetic rehabilitation in long-term edentulous patients with a collapsed bite presents numerous

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challenges, encompassing limited vertical dimension, aesthetic concerns, and Temporomandibular joint (TMJ) disorders. Muscular adaptations further complicate establishing a new occlusal relationship, often leading to discomfort and muscle fatigue. Additionally, bone resorption and sinus pneumatization impede implant placement, sometimes rendering it impractical. The presence of SWS and associated complications restrict the treatment options drastically. In a patient with Sturge-Weber syndrome, where minimal tissue alteration is desirable, a cast partial denture would be beneficial.

Given the rarity and the necessity for improved understanding regarding the dental management of SWS, a case report has been detailed in the article. The patient was diagnosed with SWS and a simple and a conservative prosthetic rehabilitation with a cast partial denture having a unique major connector design was planned for the patient to fulfil both masticatory and esthetic needs.

CASE REPORT

A 46-year-old female, a homemaker, presented to the outpatient department with a complaint of difficulty chewing food persisting for the past three years. She underwent extraction of her upper and lower posterior teeth due to caries four years ago, followed by extraction of her lower anterior teeth two months ago. Upon extraoral examination, a port-wine birthmark was noted on the sclera of her right eye, right cheek, nose, and lip, which has been present since birth. Additionally, the patient reported experiencing blurred vision in her right eye accompanied by frequent headaches. There was no pertinent family history. The patient was diagnosed with glaucoma of right eye 10 years back.

Upon examination of the temporomandibular joint, no signs of clicking or tenderness were observed. Furthermore, there was no deviation of the mandible during opening and closing, and mouth opening was found to be adequate. The patient exhibited competent lips with normal length and tonicity for her age. Notably, unilateral macrochelia was present on the right side due to the birthmark. Additionally, the patient's face appeared asymmetrical and ovoid, with a convex facial profile. A medium smile line was noted. [Figure 1]



Figure 1: Extraoral profile of patient

During intraoral examination, poor oral hygiene was evident. Thick layers of calculus deposition was seen on the lingual surfaces of the teeth. The gingival biotype appeared flat and thick and there was hemihypertrophy of alveolar and buccal mucosa with a reddish pigmentation depicting a concentration of blood vessels in the right maxillary region. The tongue was also excessively large, flattened and broadened. Furthermore, there was evidence of loss of vertical dimension, accompanied by limited interocclusal space. Examination of the hard tissue revealed root stumps in 15,14,13,23 and 25 and dental caries in 18,21,26,27 and 33. There was supraeruption in 26 and 35 and attrition of 41, 42 and 43. The patient also had missing 17,16,28,38,37,36,34,32,45,46,47 and 48. [Figure 2]



Figure 2: Intraoral profile of patient

Radiological examinations, including orthopantomogram and CBCT, revealed no abnormalities in the underlying bone structure but indicated an inadequate amount of hard tissue in both the maxilla and mandible, rendering them unsuitable for endosteal implant placement. Prior to the dental appointment, the patient had undergone several radiographical investigations of the head to rule out abnormalities. Blood investigations were conducted to ascertain the presence of iron deficiency anemia. Histopathological examination of tissue samples from the right maxillary alveolar mucosa revealed findings suggestive of inflammatory fibroepithelial hyperplasia with vascular proliferation. The culmination of clinical, radiological, and histopathological examinations

led to the diagnosis of class II type Sturge-Weber syndrome.

Before initiating treatment, the patient was thoroughly informed about various treatment options, including cast partial dentures (CPDs), removable complete dentures, overdentures, and cusil dentures. Written consent was obtained from the patient, along with medical clearance from the general physician. The treatment plan agreed upon included CPD for teeth 13, 14, 15, 16, 17, and 27 in the maxilla, with surveyed crowns in 18, and 26, as well as a fixed partial denture (FPD) for teeth 11, 12, 22, 24, and 26. In the mandible, CPD was planned for teeth 31, 32, 33, 34, 36, 37, 45, 46, and 47, with FPD in 35, 41, 42, 43, and 44, along with precision attachments. Crown lengthening procedures were avoided in the maxilla to mitigate the risk of haemorrhage due to the underlying condition. Implants were not advised due to insufficient bone quantity and quality, as well as to prevent bleeding complications. A consent was obtained from the patient to proceed with the treatment and to publish the same with relevant photographs.

The initial phase of treatment involved oral prophylaxis followed by instruction on maintenance using 0.2% chlorhexidine mouthwash (Hexidine, ICPA health products) for 2 weeks, brushing, and flossing twice daily. Subsequent phases included the removal of root stumps and root canal treatment for the remaining teeth where the bleeding was controlled using topical hemostatic agents like collagen sponges and adrenaline.

Procedure

During the restorative phase, the focus was on rehabilitating the edentulous areas and restoring the vertical dimension of the patient's dentition.

1. Shade selection: Shade selection was performed with manual shade guide (VITA 3D Master, Vita Zahnfabrik, Germany) to achieve optimal aesthetics.
2. Diagnostic impressions and temporary partial denture: Two sets of primary impressions were taken using alginate, and models were fabricated accordingly. The first set of impressions was utilized for the fabrication of a temporary partial denture, which served to assess the patient's vertical and centric relation.

The patient was instructed to wear this denture for a duration of four weeks. Meanwhile, the second set of impressions was employed for diagnostic mounting, alongside a face bow transfer in a semiadjustable articulator (Stratos 300, Ivoclar Vivadent Inc., Germany). Notably, a discrepancy in interocclusal space between the right and left sides was observed, attributed to the hemangiomatic growth present in the right maxillary arch.

3. Tooth preparation: Following a thorough assessment of interocclusal space adequacy, oral musculature adaptation, and patient comfort with the temporary denture, tooth preparation was performed. In the maxilla, tooth preparation for a fixed partial denture (FPD), surveyed crown in 26 and 18 was carried out. In the mandible, tooth preparation for FPD involving teeth 35, 41, 42, 43, and 44, equipped with precision attachments, was undertaken. Shoulder finish lines with equigingival margins were meticulously established. Subsequently, single-stage wash impressions using addition silicone (Aquasil, Dentsply, USA) were obtained, and casts were poured using die stone (Gemstone, Bharath Dental Supplies, Hyderabad). Bite registration was recorded using upper and lower occlusal rims to ensure precise articulation.
4. Articulation: Articulation in a semiadjustable articulator along (Hanau wide view, Whip mix, USA) with facebow transfer was made. [Figure 3]



Figure 3: Fabrication of fixed and removable components

5. Design of CPD: Metal copings with rest seats in 26 and 18 and precision attachments in 34 and 43 for the fixed prosthesis and metal frame work for CPD was done. The major connector is a modified closed horseshoe design to accommodate multiple missing teeth and avoid the deep, curved palate. It was also designed to minimize the gag reflex, food accumulation and improve speech. This unique modification

includes thin, lightweight bars, with the anterior part reinforced by an additional bar to prevent undue stress on the abutments and reduce flexion of the thin connector. The direct retainers were of the suprabulge type with both mesial and distal rests. Since cingulum bars are given indirect retainers were no required for the patient. The additional rests not only provide vertical support and distribute occlusal loads but also prevents settling of the denture and impingement of soft tissue.

6. Altered cast: Functional impressions were taken with the framework and the altered casts were rearticulated.
7. Denture trial: At the next appointment, the focus was on evaluating the esthetics and adaptation of the fixed partial denture (FPD) and crowns, as well as conducting the wax trial of the cast partial denture (CPD). [Figure 4] Bilateral balanced occlusion was achieved for posterior teeth to ensure distribution of forces.



Figure 4: Postoperative occlusal views

8. Insertion : During the insertion stage, the FPD and crowns were cemented in place using type I glass ionomer cement (GC Gold label, GC International), followed by the insertion of the CPD. [Figure 5] Comprehensive assessments were made regarding the esthetics, fit, occlusion, and retention of the prostheses, ensuring patient satisfaction. It was found that the lip support and smile improved drastically after the treatment. Post-insertion instructions were provided to the patient.



Figure 5: Postoperative extraoral view

8. Follow up: The patient adhered to scheduled follow-up appointments at one week, one month, one year and two year post-insertion. During these visits, any necessary adjustments were made, ensuring the continued functionality and comfort of the prostheses.

DISCUSSION

Differential diagnosis of Sturge-Weber Syndrome (SWS) includes nevus flammeus, Rendu–Osler–Weber syndrome, Von Hippel–Lindau disease, Wyburg–Mason syndrome, and Maffucci syndrome.⁶ Diagnosis relies accurately on clinical and imaging features. However, there is still insufficient data on the inheritance and progression of the syndrome over time.⁷ A multidisciplinary approach is advocated for both diagnosis and treatment planning.

An important precaution is to have a suitable arsenal available in case of any complication. Regarding oral hygiene, the findings of this case align with those of other authors who report an increase in biofilm or plaque leading to rampant dental caries and periodontal issues.^{8,9} Therefore, a prosthesis with a simple design, aimed at preventing biofilm accumulation and easy maintenance, was provided to the patient. It is believed that the design of the cast partial denture discussed is well suited in SWS as it reduces the chance of undue stress on the highly vascularised tissues apart from avoiding gag reflex, and aiding in speech.

Notably, there are no case reports on prosthodontic rehabilitation of Sturge-Weber Syndrome patients. Hence, the prosthodontic considerations and precautions have been elaborately discussed in the article. The rehabilitation proved to be challenging due to the conservative non-invasive approach. The rehabilitation focused more on the function than on the esthetics. Enlarged gingival tissues and the tongue also posed difficulties during impression procedures and denture seating. Additionally, mucosal changes could occur along the hemangioma following treatment completion, underscoring the importance of regular checkups to enable adjustment of the denture base and prevent soreness.¹⁰ Doh et al. in 2015 similarly encountered difficulties in denture fabrication due to insufficient supporting tissue, vestiges of hemangioma, and enlarged soft tissue in their case report.¹¹

CONCLUSION

An innovative major connector design was advocated for the patient to avoid injury to both the vascularised tissue and avoid stress on the abutments. Dentists must possess a thorough understanding of the clinical characteristics and treatment options associated with SWS to address its oral manifestations effectively. Collaborative consultations with diverse medical experts are essential for optimal treatment, complemented by professional counseling, which aids both the patient and their family in navigating challenges and enhancing prognosis. Regular systemic and oral assessments are advisable to mitigate the risk of cranial and oral complications.

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