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# **Case Report**

# Prosthodontic management of tuberous sclerosis complex patient with deranged occlusal plane– A case report

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# ABSTRACT

Tuberous sclerosis complex (TSC) is a syndrome that is not uncommon affecting many organs of the body. Dental manifastations are fibrous growth, enamel pits, enamel hypoplasia. Long term partial edentulous condition of such TSC patient resulted in tooth positional changes altering the occlusal curve and the esthetics. Broadrick occlusal plane analyzer was used to coorect the deranged occlusal plane and to improve the esthetics and functional occlusion.

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#### 1. Introduction

Tuberous sclerosis is a complex condition affecting multiple systems resulting in tumours and hamartomas. TSC is caused by mutations or defects on either of two genes-TSC1 gene, (discovered in 1997, is on chromosome 9 -9q34) and TSC2 gene, (discovered in 1993, is on chromosome 16 -16p13.3) which encode for the proteins hamartin and tuberin respectively. These proteins act as tumor growth suppressors, agents that regulate cell proliferation and differentiation  $1^{-4}$  is the second most common neurocutaneous syndrome after neurofibromatosis with an estimated incidence of one case per 6,000 to 10,000 live births, and has no predilection for gender or race.<sup>1-4</sup>TSC can affect many different systems of the body, causing a variety of signs and symptoms as per the system affected. The natural course of TSC varies from individual to individual, with symptoms ranging from very mild to quite severe. In addition to the benign tumors that frequently occur in TSC, other common symptoms include seizures, mental retardation, behaviour problems, and skin

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In 1998, at the Tuberous Sclerosis Complex Consensus Conference in Annapolis, Maryland, a revised criterion was formed which requires tuberous sclerosis complexassociated lesions of two or more organ systems or at least two dissimilar lesions of the same organ to confirm the diagnosis.<sup>2,3</sup> Major features include facial angiofibroma, Ungual/periungual fibroma, Hypomelanotic macules, Shagreen patch, Cortical tuber, Subependymal nodule, Subependymal giant cell astrocytoma, Multiple renal nodular hamartomas, Cardiac rhabdomyoma, Lymphangiomyomatosis, Renalangiomyolipoma. Minor features are Multiple pits in dental enamel, Hamartomatous rectal polyps, Bone cysts, Migration tracts, Gingival fibromas, Nonrenal hamartoma, Retinal achromatic patch, Retinal achromatic patch, Multiple renal cysts.<sup>3</sup> The various signs are then marked against the diagnostic criteria to produce a level of diagnostic certainty:<sup>3,4</sup>

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abnormalities. Tumors can grow in nearly any organ, but they most commonly occur in the brain, kidneys, heart, lungs, and skin. Because of its striking variability of clinical expression and severity, the diagnosis of tuberous sclerosis complex can be difficult, especially in young individuals or in those with subtle findings.

- 1. Definite Either two major features or one major feature plus two minor features.
- 2. Probable One major plus one minor feature.
- Suspect Either one major feature or two or more minor features.

# 2. Clincal Report

A male patient aged 33 years reported for the restoration of missing teeth to improve appearance and function. On clinical examination, angiofibromas are present in the face over the bridge of the nose and in the zygoma region and a skin tag in left lateral forehead (Figure 1a). Past medical history reveals an epileptic attack and has been under treatment. Intra oral examination reveals multiple nodular growth in the palate (Figure 1b), buccal mucosa and in the gingiva (Figure 1c).

Based on the diagnostic criteria of the TSC, the patient was diagnosed as a definite case of TSC as the patient exhibited major feature like angiofobroma, minor features like gingival fibroma and skin tags. The dental charting of the patient, illustrating the missing 15,21,23,34,35,36,37,38,46,47,48 and rotated 12, supraerupted 26, 28, 16 all of which affected the occlusal plane. The teeth present in the mouth were discoloured and hypoplastic in nature. An OPG was taken to assess and evaluate the condition of the teeth present

# 2.1. Treatment plan

The main aim and objective of the treatment were to replace the missing teeth, improve the esthetics by correcting hypoplastic condition and the occlusal curve. The treatment plan was formed with clinical, radiographic examination, and diagnostic mounting. Left maxillary third molar was planned for extraction based on the bone loss and level of supraeruption. The Kennedy class I partial edentulous condition in mandible was planned for restoration with attachment (Rhein-83) retained partial dentures. All the remaining maxillary teeth were planned for tooth preparation to replace the missing teeth as well as to correct the supraeruption and the unesthetic hypoplastic condition. To carry out the procedures, intentional pulp canal therapy was needed due to the supraeruption of maxillary first molars rotation of right maxillary lateral incisor. Broadrick occlusal plane analyser was used to correct the deranged occlusal plane.

# 2.2. Procedures

In Maxilla, Intentional Root Canal Treatment was done in 16, 12, and 26 and all the remaining maxillary teeth were prepared to receive Porcelain Fused to Metal restorations. An elastomeric impression was obtained and the maxillary cast was mounted on the articulator using Hanau springbow in the Hanau Wide Vue articulator. In Mandible, left lower



Fig. 1: Sowing angiofobromas, skin tags, nodular growth in palate and buccal mucosa.



Fig. 2: Showing hypoplastic teeth, deranged occlusal plane



Fig. 3: Showing the occlusal plane correction using bocclusal plane analyser



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Fig. 4: Showing the fabricated maxillary and mandibular prostheses



Fig. 5: Completed restorations in mouth.

lateral and canine and right lower premolars were prepared to receive porcelain fused to metal (PFM) crowns with male part of the attachment (fig ). The metal framework of the RPD(removable partial denture) with female part that provides housing for resilient retentive ring was tried for fit and then an occlusal rim was prepared. The mandibular cast was mounted in the semi adjustable HANAU WIDE VUE articulator using centric relation CR record. The articulator was programmed with protrusive record and right and left lateral record.

To correct and establish occlusal plane, Broadrick occlusal plane analyzer was used. As per the manual, 4 inch arc was taken to draw the long arc from the tip of the mandibular canine (ASP- Anterior Survey Point) and short arc from the point –half the height of the retromolar pad (PSP- Posterior Survey Point). From the point of intersection (OPSC - occlusal plane survey centre) an arc is drawn over the occlusal rim (Fig-). The occlusal rim is corrected to that level, a putty index was made to the corrected occlusal plane and the teeth were set.

Maxillary Anterior teeth were fabricated with guideline of 2mm over jet and over bite in the articulator and checked for esthetics and phonetics. Maxillary posterior occlusal pattern was fabricated to occlude with the corrected occlusal plane of the lower teeth. The maxillary restorations in a bisque stage, and mandibular teeth set in a wax were tried in the patient's mouth. The restorations and wax trial were sent to the laboratory for processing and finishing. The maxillary crowns and fixed partial dentures luted with type I GIC - Glass Ionomer Cement. Mandibular crowns with male part of the attachment are luted with type I GIC. The RPD with the female part of the attachment was seated in the male part with silicone o ring (yellow color). Occlusion was checked intra orally and minor corrections were done. The patient is informed about his general condition and referred to dermaotology and neuromedicine department for opinion and management as the patient had visible lesions on the face and also he had history of epileptic attack.

# 3. Discussion

The clinical manifestions of TSC are multifaceted and include neurological disorders (e.g., epilepsy, autism. and mental retardation), skin lesions, lymphangioleiomyomatosis (LAM), and the failure of affected organs. The most common oral manifestations of TSC are fibromas, gingival hyperplasia and enamel hypoplasia. Other less frequent findings in the oral cavity are a high arched palate, bifid uvula, harelip and/or cleft palate, delayed dental eruption and the presence of diastemas.<sup>4</sup> The skin lesion of TSC can be corrected by cosmetic procedures like laser and cryo therapies.<sup>5</sup> The oral features of this TSC patient hypoplastic condition, rotated, drifting and supraeruption called for shortened dental arch SDA concept. 6,7

Failure to replace the missing teeth immediately after extraction slowly produced supraeruption, tipping, rotation of the existing dentition, resulting occlusal plane anomaly.<sup>8</sup> Tooth positional changes following posterior tooth loss may alter arch forms and occlusal planes and may result in the development of occlusal interferences altered curve of spee.<sup>5</sup> The occlusal plane should be corrected lest it may produce repeated fractured restorations, muscle fatigue and temporomandibular joint problem.<sup>9</sup> This can be avoided by reconstructing the curve of Spee to pass through the mandibular condyle, which has been demonstrated to allow posterior disclusion on mandibular protrusion as the angle of condylar guidance is greater than the curve of Spee, posterior disclusion is achieved.<sup>10,11</sup>

The Broadrick flag is a useful tool in establishing the occlusal plane in prosthodontic and restorative dentistry, as it identifies the most likely position of the center of the curve of Spee.<sup>11,12</sup> Esthetics and function place a considerable demand on the design of the occlusal plane. Compromise can be achieved by altering the length of the radius of the curve.<sup>11</sup> The use of broadrick occlusal plane was supported by many clinical studies and confirmed that it may produce an accurate or approximate curve even in deranged occlusal conditions or when the teeth forming the posterior determinant is tipped.<sup>13</sup> The full mouth rehabilitation of this patient was completed with occlusal plane correction using broadrick occlusal plane analyser and SDA concept.

# 4. Conclusion

An increasing awareness about TSC will help in recognizing such patient and improve the quality of their life. Knowledge about TSC will help us in management of them. Based on the systemic and oral presentation of the patient, treatment plan was formulated, and executed successfully. Broadrick occlusal plane was a valuable tool in correcting the occlusal plane and improving the esthetics in harmony with the posterior condylar guidance. It is not the extensive prosthodontics, but the procedures done in the patient's mouth should be in harmony with the entire stomatognathic system, for longevity of the restorations and for the comfort of the patient.

#### 5. Conflict of Interest

The authors declare that there is no conflict of interest.

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None.

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