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Prosthodontic Rehabilitation for a Patient with Ellis-Van Creveld Syndrome: A Case Report

Keywords

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ABSTRACT

Patients suffering from Ellis-Van-Creveld syndrome are a challenge for dental management. Aesthetics are a major concern with limited manual dexterity, making choice of treatment critical. A 38 year old female diagnosed with Ellis-Van-Creveld syndrome presented with stained teeth and un-aesthetic smile and related that to her low self-esteem and depression. Intra-oral examination revealed mal-aligned megadont central incisors in the maxillary arch, fused mandibular canines with laterals and missing central incisors with space discrepancy and pronounced reverse over jet and overbite. Treatment involved non-surgical periodontal management, fabrication of veneers and dentine bonded crowns for maxillary anteriors and fixed-fixed resin retained bridge for mandibular arch.

INTRODUCTION

Ellis-van Creveld (Chondroectodermal dysplasia) is a rare autosomal recessive disorder resulting from a mutation in genes EVC1 and EVC2 located in chromosome 4p16 affecting all races. The first full description of the syndrome was given by Richard Ellis and Simon Van Creveld in 1940.[1] The prevalence of this condition has been estimated to be 7 per 1,000,000. Familial consanguinity has been shown in 30% of cases.[9]

The four principal characteristics of this syndrome are chondrodysplasia, polydactyly, ectodermal dysplasia and congenital heart defects (60% cases).[8] The patients are generally of small stature, short limbs with hypoplastic finger nails and fine sparse hair. Most patients have normal intelligence but occasional nervous system involvement has been noted. Oral findings are variable and include both soft and hard tissue abnormalities. Fusion of the upper lip to the gingival margin, multiple frena, micro or macrodont teeth with abnormal shapes and hypodontia are commonly encountered.[5] Hypodontia involving the maxillary and mandibular arches is a constant finding. In the mandibular incisor region, multiple notches may be present on the crest of the thin alveolar ridge.[8]

The clinical report describes the prosthetic management for Ellis-van Creveld syndrome patient with conservative restorations.

CASE REPORT:

A 38 year old female was referred by her General Dental Practitioner to the Restorative Department of the Royal London Dental Hospital. The patient had a confirmed diagnosis of Ellis-Van Creveld syndrome. From childhood she was being managed at a local dental hospital. Composite restorations on the maxillary anterior teeth were provided 8 years ago after which she was discharged stating that her appearance could not be improved further. On presentation, the patient was taking a number of anti-depressants and reported smoking 10 cigarettes per day, had a cardiac defect which was repaired when she was 5 years old and had endometriosis. Patient was really concerned about the appearance of her teeth as it was affecting her confidence and life style. She had some of the typical features of Ellis-Van Creveld syndrome, including bilateral post axial polydactyly, short stature with hypoplastic finger nails of the hands and bimanual hexadactyly was noted on the ulnar side. Extra-oral examination revealed a skeletal class III base with maxillary hypoplasia, prognathic mandible and increased lower facial height. She had a medium smile line though her general smile was guarded *(Figure 1*). She had poor oral hygiene with generalised plaque and calculus deposits and immediate bleeding on probing. Intra-oral examination revealed mild hypodontia with congenitally missing mandibular central incisors and maxillary right canine, macrodont maxillary central incisors, fused 42 with 43 and 32 with 33. Mandibular teeth were lingually inclined and buccal incomplete cross bites were noted bilaterally with reverse overjet and over bite anteriorly *(Figure 2, 3 & 4*). Her static occlusion was class III with group function on both sides and protrusive excursions on second molar teeth only. All teeth were responsive to sensibility testing checked with Ethyl chloride spray and Electric Pulp tester. Radiographically, fusion of 32/33 and 42/43 were evident. Macrodont 11 and 21 had wide pulp chambers without clearly discernible apex *(Figure 5*).

Figure 1: Pre-treatment photograph upon smiling.

Figure 2: Pre-treatment intraoral photograph in intercuspal position.

Figure 3: Maxillary occlusal view.

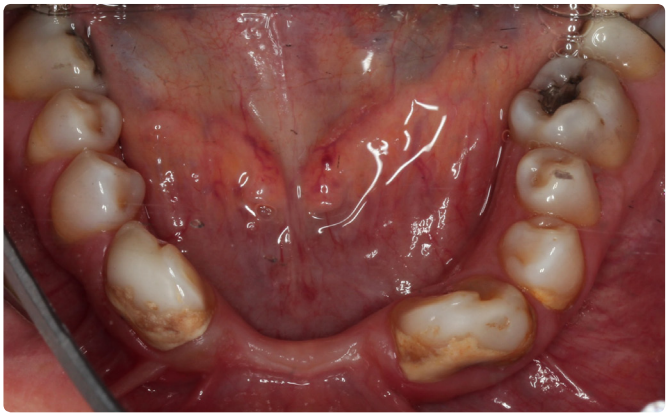

Figure 4: Mandibular occlusal view.

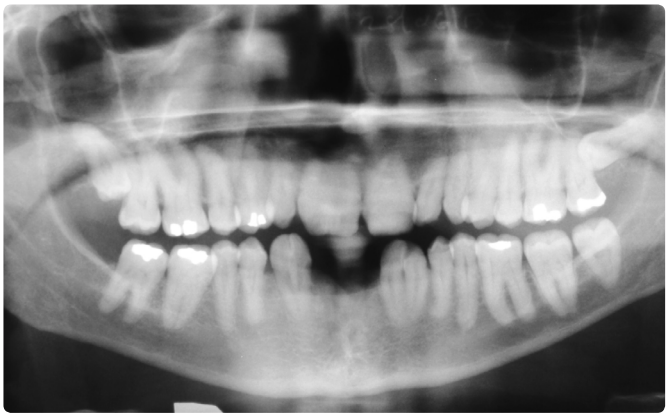

Figure 5: Dental panoramic tomograph before rehabilitation.

TREATMENT PLANNING:

Study casts were made and split cast mounted in a semi adjustable articulator to devise treatment options. The following options were presented to the patient:

1. Alignment of teeth with orthodontics with or without orthognathic surgery to correct malocclusion. Accept the size of upper central incisors and placement of 2 endosseous implants in the anterior mandible to support a fixed bridge after ridge augmentation (with block graft).
2. Extraction of maxillary central incisors and provision of implant supported restorations in the maxilla and mandible after ridge augmentation.
3. Minimally invasive fixed-fixed resin retained bridge for the mandibular anterior teeth and ceramic veneers to manage tooth proportions in maxillary arch.

In consultation with the patient, an initial treatment plan was developed which included non-surgical periodontal treatment for the remaining lower anterior teeth, provision of fixed-fixed resin retained bridge for the mandibular anterior teeth and ceramic veneers and dentine bonded crowns to manage tooth proportions in maxillary arch. Smoking cessation advice was provided in the dental hospital with a further referral to a cessation service. The patient had expressed her inability to clean thoroughly because of her inability to hold the brush properly; she also expressed displeasure with her dental aesthetics and the belief that a favourable outcome could not be achieved as being major reasons to her lack of motivation.

For assessment of proposed definitive result and to motivate the patient for efficient oral hygiene maintenance vacuum formed thermoplastic vacuum formed retainer was used to transfer the diagnostic wax-up to patient's mouth *(Figure 6 & 7)*.

After thorough scaling and polishing, oral hygiene instructions were given. After 2 months, hygiene maintenance was reviewed and a significant improvement was noted. Recorded plaque scores decreased from 90% at initial presentation to 30% at review.

Veneer preparations for 11 and 21 and dentine bonded crown preparations for 12, 13 and 22 were performed, polyvinylsiloxane impressions made and provisionalised according to the proposed wax-up. Try-in and cementation of ceramic veneers and dentine bonded crowns (IPS e.max) was done with adhesive cement.

Veneer preparations were then performed on the fused 32/33 and 42/43, impressions made and temporised. Try-in and cementation of all ceramic fixed-fixed resin retained bridge (Milled Zirconia framework over-layed with porcelain) was done with adhesive resin cement *(Figure 8-11)*.

On the 1st post treatment review, oral hygiene was re-assessed. No evidence of bleeding was noted although some localised plaque deposits were visible upon use of a disclosing tablet. Oral hygiene instructions were reinforced. Patient demonstrated use of super floss around the fixed-fixed resin retained bridge and tape floss around the maxillary veneers and dentine bonded crowns apart from interdental brushes in localised areas. Occlusion (static and dynamic) were checked, with protrusive guidance on the upper second molar teeth only and group function in lateral excursions.

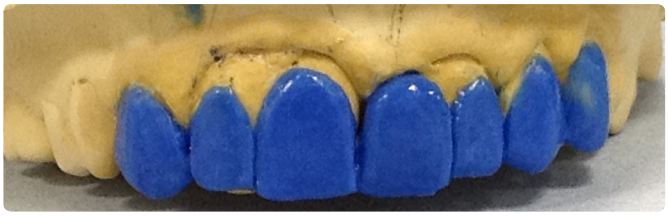

Figure 6: Wax-up of proposed restorations.

Figure 7: Trial of proposed restorations by means of vacuum formed retainer.

Figure 8: Post-treatment photograph on smiling.

Figure 9: Frontal view of restorations after rehabilitation.

Figure 10: Maxillary occlusal view of restorations.

Figure 11: Mandibular occlusal view of restorations.

DISCUSSION:

The Ellis-Van Creveld syndrome can be diagnosed during the prenatal period by ultrasonography or by clinical examination after birth. Although all embryonic layers are involved; ectodermal involvement is usually limited to nails, gums, teeth and eyes. Mesodermal involvement presents as bones, heart and kidney abnormalities and although rare, endodermal involvement presents as lung and liver abnormalities.[8,10]

Treating patients presenting with dental anomalies is always a challenge and are best managed with a multi-disciplinary approach. Likewise, this case was challenging with respect to patient management and technical demands.

Upon introduction to electric brush with a wider handle, patient demonstrated better cleaning. The use of thermoformed retainer to demonstrate the proposed plan served to motivate for improved oral hygiene and compliance which was reflected in her use of super floss and interdental brushes which may be challenging for certain group of patients.

In this case, Orthodontics could have assisted in the restorative management as it would have improved the alignment of 11 and 21, eliminated need for excess bulk of material and over contour. In the mandibular arch patient declined dental implants under sedation as it necessitated block grafting.

The angulation of the maxillary anterior teeth was a significant limitation to providing ideal tooth proportions whilst maintaining cleansibility. The veneers and dentine bonded preparations were conservative with a light chamfer at gingival margin level without compromising on strength of material and minimising over contour.[3,11] The margins were evaluated visually and using a sharp probe to ascertain any horizontal discrepancy defects as described by Holmes *et al* (1989).[6] Hayashi *et al*.[7] showed that the tip of explorers available generally measure 70-80µm. The probe did not catch and the margins were continuous with the anatomic form of the teeth. Owing to the reverse over jet and overbite, retainer wings for mandibular all-ceramic fixed-fixed bridge had to be placed buccally. Ceramic laminate veneers offer a predictable and successful treatment modality having a survival probability of 91% over 10 years.[4,2]

CONCLUSION:

This case report illustrates the successful dental rehabilitation of patient suffering from Ellis-Van Creveld Syndrome using veneers, dentine bonded crowns and resin retained fixed bridge. Owing to the complexity of the case, patient would continue being reviewed for maintenance.

REFERENCES:

1. Alves-Pereira D, Berini-Aytes L, Gay-Escoda C. Ellis –Van Creveld Syndrome. Case report and literature review. *Med Oral Patol Oral Cir Bucal.* 2009;**14**(7):E340-3.
2. Beier U.S, Kapferer I, Burtscher D, Dumfahrt H. 2012. Clinical performances of porcelain laminate veneers for up to 20 years. *Int J Prosthodont.* 2012;**25**(1): 79-85.
3. Burke F.J.T, Qualtrough A.J.E, Hale R.W. Dentine-bonded all-ceramic crowns: current status. *J Am Dent Assoc.* 1998;**129**: 455-460.
4. Dumfahrt H, Schaffer H. Porcelain laminate veneers. A retrospective evaluation after 1 year to 10 years of service: Part II-Clinical results. *Int J Prosthodont.* 2000;**13**(1): 9-17.
5. Hegde K, Puthran R.M, Nair G, Nair P.P. Ellis van Creveld syndrome–a report of two siblings. *BMJ Case Reports* 2011.
6. Holmes JR, Bayne SC, Holland GA, Sulik WD. Considerations in measurement of marginal fit. *J Prosthet Dent.* 1989 Oct;**62**(4):405-8.
7. Hayashi, M., Wilson, N., Ebisu, S., & Watts, D. Influence of explorer tip diameter in identifying restoration margin discrepancies. *J Dent.* 2005;**33**(8), 669-674.
8. Jenkins S, Morrell D.S. Ellis-Van Creveld Syndrome: Case report and Review of the literature. *Cutis* 2009;**83**:303-305.
9. Kalaskar R, Kalaskar A.R. Oral Manifestations of Ellis-van Creveld Syndrome. *Contemp Clin Dent.* 2012;**3**: S55-S59.
10. Varela M, Ramos C. Chondroectodermal dysplasia (Ellis-van Creveld syndrome): a case report. *Eur J Orthod.* 1996;**18**:313.
11. Walls A.W.G, Steele J.G, Wassell R.W. Crowns and extra-coronal restorations: porcelain laminate veneers. *Br Dent J.* 2002;**193**(2): 73-82.

Manufacturers' Details:

- Virtual® hydrophilic vinyl polysiloxane impression material, Type 3 and Type 2, Ivoclar Vivadent, Schaan, Liechtenstein.
- Rely X Ultimate® dual cure adhesive resin cement, 3M ESPE.
- MultiLink® Automix, Ivoclar Vivadent, Schaan, Liechtenstein.
- Quicktemp®, Temporary crown material, Schottlander, UK.