

Telescopic crowns in adult case with lip and palate cleft. Update on the etiology and management

José Félix Mañes Ferrer ⁽¹⁾, Amparo Martínez González ⁽²⁾, Begoña Oteiza Galdón ⁽³⁾, Kheira Bouazza Juanes ⁽⁴⁾, Francisco Benet Iranzo ⁽³⁾, Ana Candel Tomás ⁽⁵⁾

- (1) Profesor Responsable Prótesis I. Facultad de Ciencias Experimentales y de la Salud. Odontología. Universidad Cardenal Herrera CEU
 (2) Profesor Responsable Prótesis II. Facultad de Ciencias Experimentales y de la Salud. Odontología. Universidad Cardenal Herrera CEU
 (3) Profesor Asociado Prótesis I. Facultad de Ciencias Experimentales y de la Salud. Odontología. Universidad Cardenal Herrera CEU
 (4) Profesor Asociado Prótesis II. Facultad de Ciencias Experimentales y de la Salud. Odontología. Universidad Cardenal Herrera CEU
 (5) Licenciada en Odontología. Valencia

Correspondence:

Dr. José Félix Mañes Ferrer

Prótesis I

Facultad de Ciencias Experimentales y de la Salud. Odontología.

Universidad Cardenal Herrera CEU.

Edificio Odontología

Cl Del Pozo, s/n

46115 Alfara del Patriarca (Valencia)

E-mail: jfmanes@uch.ceu.es

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ABSTRACT

Lip and palatal clefts are among the most important congenital craniofacial malformations to be taken into account in general dental practice, due to their high incidence and important repercussions upon the oral cavity. The underlying causes are genetic and fundamentally environmental, and the disorders manifest as early as in the embryonic period. Males are predominantly affected, with a 7:3 ratio versus females. Our patient, a 20-year-old male, presented the most common association, i.e., total unilateral hare lip with palatal cleft. A description is provided of the treatment for his dental problem, together with an update on the etiology and management of adults with malformations of this kind.

Key words: *Telescopic crowns, cleft lip, palatal cleft, etiology, treatment.*

RESUMEN

La fisura labiopalatina es una de las malformaciones congénitas craneofaciales más importantes a tener en cuenta por el odontólogo general debido a su alta incidencia y a su elevada repercusión en la cavidad oral. Sus causas se deben a factores genéticos y predominantemente a factores ambientales, haciendo su aparición ya en el período embrionario. El sexo masculino se encuentra más afectado, en una relación de 7:3. La asociación más frecuente es el labio leporino unilateral total con fisura palatina, que es el caso que nos atañe. Presentamos el caso de un varón de 20 años que acude a nuestra consulta demandando una solución estética a su situación dental. El tratamiento efectuado al paciente, así como una puesta al día en cuanto a la etiología y tratamiento en adultos de esta malformación es el motivo del presente artículo.

Palabras clave: *Coronas telescópicas, labio fisurado, fisura palatina, etiología, tratamiento.*

INTRODUCTION

Lip and palatal clefts are congenital structural alterations secondary to defective coalescence among some of the embryonic facial development processes. According to the European Commission on Science, Research and Development for the European Community, the incidence of these malformations is one case out of every 600 individuals – thus underscoring the importance of these congenital malformations in the health care setting, not only because of their esthetic impact but also due to the swallowing, speech, skeletal, dental and psychological effects involved.

These malformations are also typically varied in terms of severity – the latter depending mainly on the degree of structural involvement. Thus, presentations range from unilateral clefting limited to the lip (not very serious) to extremely complex cases corresponding to clefts extending from the base of the nose to the uvula.

The classification of these disorders is based on the incisor foramen as reference. Thus, preforamen clefts are located anterior to the incisor foramen and affect the premaxilla (group I); postforamen clefts are located posterior to the foramen (group II); and transforamen clefts extend from the premaxilla to the soft palate (group III). These three groups in turn can be divided into uni- or bilateral presentations (1).

Regarding the treatment of these malformations, it is essential to integrate not only the different specialized fields in dental practice but also medicine, psychology, phoniatrics, etc. (2). As part of the management team, the dentist, or better still the different specialists, are responsible for supervision of all the dental aspects of the patient in the course of development. These individuals pose the greatest prosthodontic challenge, as reflected by the patient presented in this study.

CLINICAL CASE

A 20-year-old male consulted seeking an esthetic solution for his anterior sector. In the first 5 years of life he underwent three surgical operations to close the soft tissues of the lip and palate. Extraoral examination showed upper lip asymmetry secondary to the different operations undergone in childhood.

Intraoral examination revealed evident dental malpositioning and malocclusion, as well as different dental agenesis (affecting 15, 12, 11, 25, 34, 33 and 44) and maxillary-mandibular discrepancy. Moderate gingivitis due to poor dental plaque control was also diagnosed (Fig. 1).

The patient presented severe maxillary-mandibular dysmorphism advising orthodontic treatment before deciding any kind of prosthodontic management. The case was consulted with a specialist in orthodontics, who recommended orthognathic surgery before prosthetic treatment. The situation was explained to the patient who nevertheless refused surgery. Analyzing the habits of hygiene of our patient, with the identification of clearly deficient plaque control, we decided to apply a fixed prosthesis on telescopic crowns (removable

under professional supervision) to improve plaque control (this being essential for treatment prognosis), and ensure stenting of the arch on both sides of the palatal cleft.

Telescopic crowns as a double-crown prosthodontic system allow cross-stenting of the dental arch (this being limited to the anterior maxillary sector in our patient) – thereby facilitating tooth stabilization over the long term. The double-crown concept and the intrinsic design involved ensure maximally favorable masticatory force transmission, since the latter always takes place axial to the teeth. The fact that the patient may remove the secondary structure in turn facilitates hygiene of the dental abutments – particularly when compared with the cleaning difficulties associated with a conventional fixed bridge. This was one of the main reasons for deciding to provide treatment with telescopic crowns in our patient, since hygiene in this case was suboptimal.

From the start the patient was informed that the esthetic and functional outcome would not be ideal, and he accepted the fact. Prior to prosthodontic treatment, the root fragments of tooth 13 were removed, and definitive treatment was carried out after healing of the wound. Such treatment consisted of the preparation of dental abutments 14, 21, 22, 23 and 24, with conventional trimming for telescopic crowns. Of note is the important trimming required in the case of the crown of tooth 21, in order to achieve parallelism between abutments – this being a crucial aspect when working with telescopic crowns (Fig. 2A).

Posteriorly, imprints and intermaxillary records were obtained, and the resulting models were used in the laboratory to prepare the primary crowns. These were in turn tested in the mouth of the patient, confirming correct fit, followed by obtainment of a second imprint to position the primary crowns with respect to the rest of the oral structures of the patient. A new model was thus obtained for elaboration of the secondary crowns conforming the fixed prosthesis that could be removed by the dental professional. This secondary structure was likewise tested in the mouth of the patient, confirming both fit and occlusal relation (Fig. 2B-C).

Following selection of the color, finishing and placement was carried out. The end result (Fig. 3) was simply acceptable from our perspective, though the patient was quite pleased with the outcome – probably because he was not convinced that the original situation could be bettered.

The case has been subjected to follow-up during 5 years, and despite persistently deficient plaque control, the prosthetic abutments remain in perfect condition, and both patient comfort and esthetic performance are satisfactory. This case reflects the correct functioning over the medium and long term of treatments with telescopic crowns. The latter should always be taken into consideration as a treatment option, particularly when dealing with patients presenting a poor dental prognosis, since the biomechanical behavior of telescopic crowns offers long-term survival of the teeth.



Fig. 1. Intraoral view of the patient.

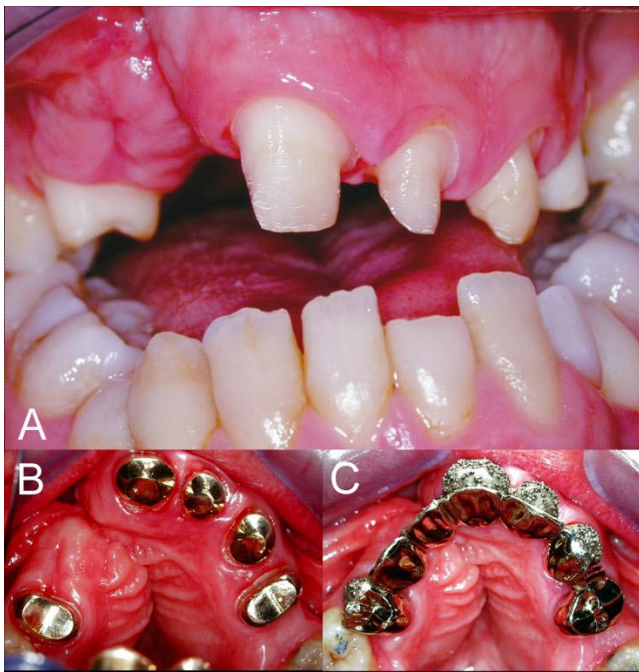


Fig. 2. A. Trimming for telescopic crowns. B. Metal testing for the primary crowns. C. Testing of the secondary structure.



Fig. 3. End result of the case with telescopic crowns.

REVIEW OF THE LITERATURE

The presentation of this case of palatal cleft allows us to review two important aspects of this pathology: (a) the causes underlying the malformation and which can help us define preventive measures destined to avoid their development; and (b) the existing therapeutic possibilities in patients such as our own, where prior corrective measures have not been adopted at the correct time.

The causes of such malformations are highly diverse, though three major groups can be considered: genetic causes, environmental factors and multifactor causes. A fourth category could also be included, corresponding to those cases in which the underlying causes are unknown (idiopathic cases).

- Genetic causes

Orofacial clefts have been classified in the scientific literature into two large groups (syndromic and non-syndromic oral clefts), according to the way in which they manifest clinically. However, this classification has been questioned by the latest advances in our knowledge of the genetic causes of these disorders. To date it was believed that some syndromes are clearly related to genetic alterations (3), such as alterations of the TBX22 (T-box transcription factor-22) gene related to cleft palate associated to chromosome X (4-5); the PVRL1 (poliovirus receptor like-1) gene, related with the so-called ectodermal dysplasia syndrome with cleft lip and palate (6); and the IRF6 (interferon regulatory factor-6) gene, which leads to Van der Woude syndrome (7). Recent research seems to relate these and other genes to clinical cleft lip and palate disorders of a non-syndromic nature. As an example, TBX22 gene alterations have been related not only to the aforementioned syndrome but also to isolated cleft palate (8) and to cleft palate and ankyloglossia (4,9). In the same way, the PVRL1 gene has been associated with sporadic forms of cleft lip and palate in areas of Venezuela (10), and authors such as Zuccherio et al. (11) consider the IRF6 gene to be the major modifier of non-syndromic clefts, and the factor responsible for a three-fold greater risk of recurrence in families that already have an affected offspring.

Other studies relate oral clefts to alterations in the MSX1 and other genes (FGFR1, TTF-2, FOXC2, SATB2 and ACOD4), underscoring that classification of these malformations into two groups is not so clear. Further research in this field is thus necessary (12).

- Environmental factors

Authors such as Fallin (13) have described the potential importance of interactions between genetic and environmental importance, suggesting that alteration of the MSX1 gene, accompanied by a maternal smoking habit, clearly predisposes to development of oral clefts.

In a recent study, Meyer et al. (14), analyzing 678 cases of cleft palate and 1175 cases of cleft lip with or without cleft palate, and their relation to tobacco smoking, came to the conclusion that a direct relationship effectively exists between smoking and the development of palatal clefts, though in contrast no such association is identified in the case of cleft lip with or without palatal cleft.

Another environmental factor related to the development of oral cleft defects is parent age. It is known that the risk of malformation increases with maternal age. Bille et al. (15) investigated the way in which maternal age affects the incidence of oral clefts. They concluded that advanced maternal as well as paternal age is associated with an increased incidence of cleft lip with or without cleft palate. However, advanced paternal but not maternal age was found to be linked to an increased risk of isolated cleft palate.

It has also been reported that the development of such malformations is influenced by phenytoin (inducing cleft lip with or without cleft palate) and 6-aminonicotinamide (inducing isolated cleft palate) (16). Pezzetti et al. (17) postulated that low enzymatic activity of methylenetetrahydrofolate (MTHFR) in pregnant women, generally related with variation in the form of c.665C>T, could be responsible for an increased risk of cleft lip in the offspring.

In a study of the association between cleft lip with or without cleft palate and myoinositol, glucose and zinc, Krapels et al. (18) concluded that children with cleft lip and their mothers present significantly lower blood zinc concentrations than the controls, and that a reduced combined concentration of zinc and myoinositol in blood increases the risk of cleft lip with or without cleft palate. These authors considered that glucose does not condition the appearance of such malformations, in contrast to the observations of Spilson et al. (19), who reported a 1352-fold greater risk of lip or palatal clefting in the offspring of diabetic mothers.

Some studies, based on the influence of external factors, postulate that the risk of orofacial clefts can be reduced by multivitamin supplements at the time around conception – though taking into account that excessive vitamin A (retinol) ingestion may not be advisable (20). Folic acid and vitamin B supplements have also been seen to be effective. In this sense, studies in animal models (mice) have shown total group B vitamin deficiency to induce a 15% increment in the incidence of palatal clefts (21). Rooij et al. (22) in turn reported that a low maternal concentration of vitamin B increases the risk of oral clefts in the newborn offspring.

Regarding the management of orofacial clefts, treatment begins at birth, with primary care of the newborn infant, and continues during the development period and into adult life. The multidiscipline team supervising patient management programs surgery, such as bone grafting in the cleft region, and plans orthodontic treatments to align the teeth and correct malocclusions. In turn, prosthodontic therapy is programmed to restore function and esthetic effect, adapted to each phase of dental development of the patient.

Thanks to improved knowledge of craniofacial growth and development, with improved orthodontic and surgical treatments, these patients increasingly receive better and sooner medical care. This in turn makes it possible to avoid the need for prosthodontic treatment of the permanent dentition, or alternatively such treatment requirements are only minimal. In any case, prosthodontics is an essential aspect of the global management of these patients (2).

Orthodontic and periodontal treatment must be coordinated with prosthetic restoration to ensure sufficient intermaxillary space and soft tissue morphology adapted to the requirements of definitive restoration (23). The periodontist plays an important role in this context, since dental crowding and malpositioning, hypertrophic gums, orthodontic apparatuses and prostheses all complicate plaque removal and contribute to perpetuate periodontal disease.

From the prosthetic point of view, a number of treatment possibilities exist. One option is a removable prosthesis, as reported in different studies (24-27), including overdentures on natural teeth (as in our case), and implant-supported prostheses.

Edentulous cleft patients pose increased difficulties, since the maxillas are affected, and the lips and palate sometimes present scar tissue. A prosthesis may prove necessary in some patients to seal a residual cleft palate or correct an inadequate pharyngeal vault that can complicate speech. Another management option is a conventional fixed prosthesis involving teeth stented on both sides of the cleft, thereby contributing to restore functional loading capacity (28).

Osseointegrated implants have been shown to be a restorative option affording good results when placed in the inserted bone tissue (29-30). In any case, and regardless of the rehabilitation approach adopted, prosthodontic maintenance is an essential component of long-term patient care, and serves to maintain adequate chewing and speech function, and facial esthetics.

CONCLUSIONS

The present study describes an adult patient and reviews the state of orofacial cleft pathology in the last 5 years. Perhaps the most important aspect of these disorders is the study of their underlying causes, with special attention focusing on both the genetic components and on the influence of environmental factors. Treatment of adult cases is increasingly rare, and emphasis in this sense may be placed on the growing importance of implant-based treatments.

REFERENCES

1. Spina V. Classificação das fissuras labio-palatinas: sugestão de modificação. *Rev Hosp Clin Fac Med* 1972;27:5-6.
2. Moore D, McCord JF. Prosthetic dentistry and the unilateral cleft lip and palate patient. The last 30 years. A review of the prosthodontic literature in respect of treatment options. *Eur J Prosthodont Restor Dent* 2004;12:70-4.
3. Wong FK, Hagg U. An update on the aetiology of orofacial clefts. *Hong Kong Med J* 2004;10:331-6.
4. Stanier P, Forbes SA, Arnason A, Bjornsson A, Sveinbjornsdottir E, Williamson R et al. The localization of a gene causing X-linked cleft palate and ankyloglossia (CPX) in an Icelandic kindred is between DXS326 and DXYS1X. *Genomics* 1993;17:549-55.
5. Braybrook C, Doudney K, Marcano AC, Arnason A, Bjornsson A, Patton MA et al. The T-box transcription factor gene TBX22 is mutated in X-linked cleft palate and ankyloglossia. *Nat Genet* 2001;29:179-83.
6. Suzuki K, Hu D, Bustos T, Zlotogora J, Richieri-Costa A, Helms JA et al. Mutations of PVRL1, encoding a cell-cell adhesion molecule/herpesvirus receptor, in cleft lip/palate-ectodermal dysplasia. *Nat Genet* 2000;25:427-30.
7. Burdick AB. Genetic epidemiology and control of genetic expression in van der Woude syndrome. *J Craniofac Genet Dev Biol Suppl* 1986;2:99-105.

8. Marçano AC, Doudney K, Braybrook C, Squires R, Patton MA, Less MM et al. TBX22 mutations are frequent cause of cleft palate. *J Med Genet* 2004;41:68-74.
9. Haenig B, Schmidt C, Kraus F, Pfordt M, Kispert A. Cloning and expression analysis of the chick ortholog of TBX22, the gene mutated in X-linked cleft palate and ankyloglossia. *Mech Dev* 2002;117:321-5.
10. Sozen MA, Suzuki K, Talavera MM, Bustos T, Fernandez Iglesias JE, Spritz RA. Mutation of PVRL1 is associated with sporadic, non-syndromic cleft lip/palate in northern Venezuela. *Nat Genet* 2001;29:141-2.
11. Zuchero T, Cooper M, Maher BS, Daack-Hirsch S, Nepomuceno B, Ribeiro L et al. Interferon regulatory factor 6 (IRF6) gene variants and the risk of isolated cleft lip or palate. *N Engl J Med* 2004;19:769-80
12. Stanier P, Moore GE. Genetics of cleft lip and palate: syndromic genes contribute to the incidence of non-syndromic clefts. *Hum Mol Genet* 2003;13:73-81.
13. Fallin MD, Hetmanski JB, Park J, Scott AF, Ingersoll R, Fuernkranz HA et al. Family-based analysis of MSX1 haplotypes for association with oral clefts. *Genet Epidemiol* 2003;25:168-75.
14. Meyer KA, Williams P, Hernandez-Diaz S, Cnattingius S. Smoking and the risk of oral clefts: exploring the impact of study designs. *Epidemiology* 2004;15:671-8.
15. Bille C, Skytthe A, Vach W, Knudsen LB, Andersen AM, Murray JC, et al. Parent's age and the risk of oral clefts. *Epidemiology* 2005;16:311-6.
16. Erickson RP, Karolyi IJ, Diehl SR. Correlation of susceptibility to 6-aminonicotinamide and hydrocortisone-induced cleft palate. *Life Sci* 2005;76:2071-8.
17. Pezzetti F, Martinelli M, Scapoli L, Carinci F, Palmieri A, Marchesini J et al. Maternal MTHFR (Methylenetetrahydrofolate) variant forms increase the risk in offspring of isolated nonsyndromic cleft lip with or without cleft palate. *Hum Mutat* 2004;24:104-5.
18. Krapels IP, Rooij IA, Wevers RA, Zielhuis GA, Spauwen PH, Brussel W. Myo-inositol, glucose and zinc status as risk factors for non-syndromic cleft lip with or without cleft palate in offspring: a case-control study. *BJOG* 2004;111:661-8.
19. Spilson SV, Kim HJ, Chung KC. Association between maternal diabetes mellitus and newborn oral cleft. *Ann Plast Surg* 2001;47:477-81.
20. Hozyasz K, Chelchowska M, Surowiec Z. Plasma vitamin A in mothers of children with orofacial clefts. *Ginekol Pol* 2004;75:139-44.
21. Schubert J, Schmidt R, Syska E. B group vitamins and cleft lip and cleft palate. *Int J Oral Maxillofac Surg* 2002;31:410-3.
22. van Rooij IA, Swinkels DW, Blom HJ, Merkus HM, Steegers-Theunissen RP. Vitamin and homocysteine status of mothers and infants and the risk of nonsyndromic orofacial clefts. *Am J Obstet Gynecol* 2003;189:1155-60.
23. Strong SM. Adolescent dentistry: multidisciplinary treatment for the cleft lip-palate patient. *Pract Proced Aesthet Dent* 2002;14:333-8.
24. Sykes LM. Prosthodontic treatment of the edentulous adult cleft palate patient. *SADJ* 2003;58:64,68-72.
25. Strong SM. Adolescent dentistry: multidisciplinary treatment for the cleft lip-palate patient. *Pract Proced Aesthet Dent* 2002;14:333-8
26. Savion Y, Sharon-Buller A, Kalisker Y, Kalisker N, Sela M. The use of Dental D as an alternative for chrome-cobalt removable partial denture: a case report. *Refuat Hapeh Vehashinayim* 2001;18:30-1.
27. Pham AV, Abarca M, De Mey A, Malevez C. Rehabilitation of a patient with cleft lip and palate with an extremely edentulous atrophied posterior maxilla using zygomatic implants: case report. *Cleft Palate Craniofac J* 2004;41:571-4.
28. Iida T, Mukohyama H, Inoue T, Oki M, Suzuki R, Ohyama T et al. Modal analysis of the maxillary dentition in cleft lip and palate patients before and after bone grafting. *J Med Dent Sci* 2001; 48:87-94.
29. Kawakami S, Yokozeki M, Horiuchi S, Moriyama K. Oral rehabilitation of an orthodontic patient with cleft lip and palate and hypodontia using secondary bone grafting, osseo-integrated implants, and prosthetic treatment. *Cleft Palate Craniofac J* 2004;41:279-84.
30. Isono H, Kaidi K, Hamada Y, Kokubo Y, Ishihara M, Hirashita A, et al. The reconstruction of bilateral clefts using endosseous implants alter bone grafting. *Am J Orthod Dentofacial Orthop* 2002;121:403-10.