



Hybrid rehabilitation conducted in a patient afflicted with Goldenhar's syndrome. Case report

Rehabilitación híbrida realizada en paciente con síndrome de Goldenhar. Reporte de un caso

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ABSTRACT

Goldenhar's syndrome, named after Dr M Goldenhar, is normally reserved for cases presenting hemi-facial microsomia. This syndrome normally affects upper and lower jaws, pinna, and adjacent tissues in a hemi-face. The target of this rehabilitation is to restore to its maximum possible extent patient's esthetics and masticatory function. We present intra-oral rehabilitation of a Goldenhar Syndrome patient who attended the maxillofacial prosthesis clinic. The patient exhibited complete edentulism, maxillary and mandibular hypoplasia (this latter compensated with a titanium chain). Prosthetic bi-maxillary rehabilitation was undertaken, bearing in mind circumstances of facial asymmetry, mandibular movement limitations and decreased alveolar ridges. Treatment was mainly focused on improvement of esthetic, phonetic and masticatory functions, which, as a whole are crucial in patient's day-to-day life and quality of life.

Key words: Goldenhar, hybrid rehabilitation.

Palabras clave: Goldenhar, rehabilitación híbrida.

RESUMEN

El síndrome de Goldenhar en honor al Dr. M. Goldenhar suele reservarse para los casos que presentan microsomía hemifacial. Este síndrome suele afectar principalmente maxilar, mandíbula, pabellón auricular, globo ocular y tejidos adyacentes de una hemicara. El objetivo de esta rehabilitación es devolver en medida de lo posible, la función masticatoria y estética al paciente. A continuación se presenta la rehabilitación intraoral de una paciente con síndrome de Goldenhar que acudió a la clínica de PMF presentando, edentulismo total hipoplasia maxilar y mandibular (esta última compensada con una cadena de titanio). Se realizó rehabilitación protésica bimaxilar, teniendo en cuenta la asimetría facial, la limitación de movimiento mandibular y la disminución de los procesos alveolares. El tratamiento fue enfocado principalmente en mejorar la función masticatoria, fonética y estética, que en conjunto son determinantes en la cotidianidad del paciente y su calidad de vida.

INTRODUCTION

Description of this syndrome corresponds to Dr Goldenhar (1952) who established its clinical definition as: association of malformations of the eyeball and ear, particularly of dermoid, epibulbar, pre-auricular appendix and pre-auricular fistula. Gorlin (1963) named it «ocular-auricular-vertebral dysplasia». Other synonyms are «Goldenhar-Gorlin syndrome» «Hemi-facial microsomia» and «First and Second branchial arch syndrome». ¹⁻⁴ Jones (1988) considers it a spectrum and torso as «sequence» since it offers varied degrees of a same initial morphogenesis disorder. ⁵⁻⁷ Mean incidence varies from one in 3,000 to 5,000 live births. ¹

Two types of treatments, surgical and prosthetic, are used to rehabilitate these patients. ⁴ Surgical treatment is preferred: it consists on several surgical interventions to achieve long term results, which

oftentimes are not the expected ones. ⁴⁻⁶ According to the case and involvement degree of patient's anatomical structures, the following treatments can be used: removable partial prostheses, fixed prostheses, full prostheses as well as overdentures.

In general, rehabilitation by means of a prosthesis, is an individualized treatment, which for the patient represents the advantages of being low cost and non invasive. ^{8,9}

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A patient afflicted with «Goldenhar syndrome» was treated at the Maxillofacial Prosthesis Clinic of the UNAM (National Autonomous University of Mexico). Due to bone circumstances presented by the patient, treatment consisted on placement of individualized full prostheses.

Objective

Manufacture of a full upper prosthesis and a hybrid lower prosthesis. The upper denture was hollowed out in order to lighten the device's weight, the lower denture was left with full weight so as to improve stability and restore patient's esthetics and function.



Figure 1. Patient with Goldenhar syndrome.

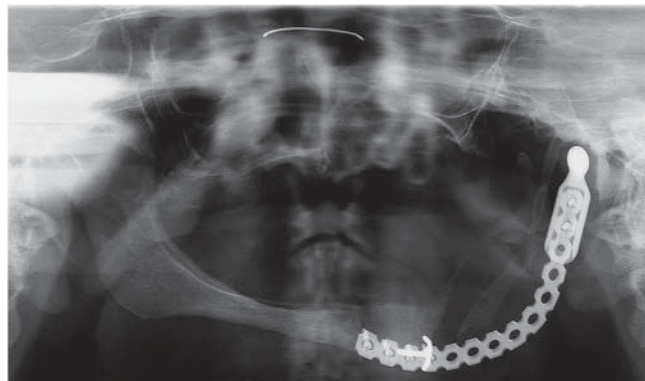


Figure 2. Orthopantomography.



Figure 3. Intra-oral view of lower jaw.



Figure 4. Intraoral view of upper jaw.

MATERIAL AND METHODS

A 64 year old female patient, diagnosed with Goldenhar's syndrome, she had undergone 21 surgeries to improve function and esthetics (*Figure 1*). One of the conducted surgical procedures was a mandibular osseous distraction, which resulted in osteomyelitis. As a result, a hemi-mandibulectomy with titanium chain placement was conducted in order to compensate resulting bone defect. In consequence a practically non-existent residual ridge was obtained (*Figures 2 and 3*). The patient exhibited a «V» shaped palate, with portions of mobile tissue and generalized bone loss causing considerable loss of vertical dimension (*Figure 4*).

Treatment: a hollowed-out full upper denture was manufactured so as to alleviate its weight; a lower denture with «Remanium» bar was manufactured so as to slightly increase the denture's weight without increasing its volume.

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