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CASE REPORT

Osteogenic mandibular distraction in Nager's Syndrome. Case report

Distracción osteogénica mandibular en síndrome de Nager. Reporte de un caso

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ABSTRACT

Osteogenic distraction is used nowadays for tissuelengtheningdue to the bone formation that occurs during the progressive segment separation after corticotomy, being very useful in dentofacial anomalies, especially in severe hypoplasia. We present the case report of a patient with Nager's syndrome, acrofacial dysostosis of the preaxial type, severe mandibular hypoplasia and oligodonthia who was treated by means of distraction osteogenesiswith ramus osteotomies to lengthen the mandible. He was treated with a Spring Bite-type orthopaedic appliance after the osteotomies. We conclude that treatment with mandibular distraction should be comprehensive and supported with the use of miofuncional advices.

RESUMEN

La distracción osteogénica es actualmente utilizada para el elongamiento tisular, gracias a la neoformación ósea que ocurre durante la separación progresiva de los segmentos después de la corticotomia de los mismos. Se ha utilizado con excelentes resultados en anomalías dentofaciales especialmente en hipoplasias mandibulares severas. Reportamos un paciente con síndrome de Nager, disostosis acrofacial del tipo preaxial con hipoplasia mandibular severa y agenesias dentales, quien fue tratado con distracción mandibular a través de corticotomías en ramas mandibulares y posterior manejo ortopédico funcional con aparatología tipo Spring Bite. Con la presentación de este caso podemos sugerir que el manejo de la distracción mandibular debiera ser apoyado con el uso de aparatología miofuncional.

Key words: Nager's syndrome, postaxial acrofacial dysostosis, mandibular distraction, mandibular hypoplasia and oligodonthia.
Palabras clave: Síndrome de Nager, acrofacial disostosis preaxial, distraccion mandibular, hipoplasia mandibular, oligodoncia.

Nager's syndrome was described for the first time by Nager and De Reynier in 1948; there have been 100 cases reported in the literature up to now.[1-3]

It belongs to the vast group of otofacialmandibular disostosis such as the Treacher Collins Syndrome, Nager's Syndrome, Pierre Robin Anomaly, Hemifacial Microsomia, among others. These are malformations associated with a hypoplasia or agenesis of the earlobe and mandibular hypoplasia among other facial deformities which can be found isolated or in association with other malformations.[3,4]

Nager's syndrome is a preaxial acrofacial disostosis that consists in facial malformations associated with radial effects (absence of the radial or tibial axis-first metacarpian and first toe).[1-5] It has a recessive autosomal genetic pattern and an alteration of the 9q32 chromosome, 1q12q21 deletion with an average neonatal birth rate of 20%, growth delay of 10% and usually normal intelligence.[2-4]

With craniofacial characteristics in 25% of the cases, it presents cygomatic and maxillaryhypoplasia, severe mandibular micrognathia, outwards and downwards palpebral fissures, absence of the lower lid eyelashes, lower lid coloboma, broad nasal bridge, depressed tip of the nose, limited mandibular movements secondary to alterations in the mandibular ramus and the temporomandibular joint, macrostomia, cleft lip and palate, soft palate agenesis, short soft palate, high and narrow palate, dysplasic earlobes, atresia of theear meatus, conductive deafness, enamel hypoplasia and oligodonthia. In the muscular skeletal system, it is characterized by radial anomalies, 75% of them being radial hypoplasia or aplasia, sinostosis of the carpal bones, absence of the 5th metacarpian, agenesis of toes and anomalies of pelvic limbs. Cardiovascular anomalies such as Fallot tetralogy

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and/or ventricular septum defect may be present.[1-5] It can also be associated with vesicoureteral reflux or renal agenesis.[3]

Acral deformities associated with a facial disostosis allow it to be differentiated from the Treacher Collins syndrome, the Nager syndrome and other dysplasiasof the 1st and 2nd facial arch *(Table I)*.[3,4]

OSTEOGENIC DISTRACTION

Osteogenic distraction is a method for bone lengthening that allows the correction of deformities and bone deficiencies with the subsequent correction of the soft tissues[6] by means of a distracting appliance.

It was first used by Dr. Codevilla in 1905, when he performed femur osteotomies. This technique remained forgotten for several decades until Dr. Ilizarov in 1950 made it popular in the field of trauma and orthopedics.[8,9] Its clinical and systematic application in craniofacial deformities began with McCarthy in 1992[9-11]mainly by using it in children with hemifacial microsomias for mandibular distraction.[11-14] Its indications have broadened for the correction of facial asymmetries of diverse etiologies such as severe maxillaryand mandibular retrognathias.[9,10]

Osteogenic distraction is a biological procedure of new bone formation by applying constant traction forces during a period of time. These forces are applied with a distraction device on a bone area that has been previously weakened by corticotomy.[6-8] Thedistraction device is an expansion screw that has been universally graduated in such a way that every 360 degree turn will provide a 0.5 mm[18,19] movement; all this process is under biological principles such as: vascular preservation, adequatelatency period, distraction rhythm and consolidation period. During this last phase the objective is to keep the bone segment immobile to achieve a correct organization and condensation of the elements that will offer the characteristics of resistance to the newly formed tissue.[6,7,18]

Physiologically, the process of distraction begins when the loading stimulus is detected by the osteoblast thus triggering a fast and continuous signaling cascade; in this process the bone growth and differentiation is established, the osteoblastic proliferation is followed by cell differentiation and finally by the mineralization of the extracellular matrix; also, specific factors have been identified associated with the beta 1 transforming growth factor (TGFB-1), the insulinic growth factor(IGF-1) and the E2 prostaglandin (PGE2). The created bone gap is initially filled by fibrillar connective tissue with collagen fibers oriented parallel to the force vector of the distraction forces.[6-8]

Once the tissue neoformation objective has been accomplished, a tissue regeneration phase of this newly formed tissue follows.[18,19]

The success of the distraction will depend on numerous factors such as: small incisions, preservation of the periostium and vascularity, latency period without distraction of 5 to 7 days, an expansion rhythm of 1mm once a day, a stabilization period or consolidating phase of 8 to 12 weeks and finally, a remodeling period.[18]

On this last period one can have more certainty on the formation of new bone tissue as well as in the histodistraction; in this stage the distraction appliance may be removed. Equally important is to verify this process by periodical imagetechniques to monitor the correct function and evolution of the distraction.[19-22]

Difficulties have been found such as the distractor selection, the determination of the direction of the vector, the site for the osteotomy and patient cooperation.[23,24]

The orthodontist should be present during the complete process of study and treatment of these patients, playing an important role in the diagnosis, treatment planning and postsurgical management.

The wide variety of mandibular anomalies associated with a syndromic deformity makes it difficult to predict the treatmentresult, in spite of the surgical, orthodontic and physiotherapeutic management. Alsorelapse appears to be inevitable and overcorrection may not compensate central growthalterationsand poor muscular function.Nevertheless, osteogenic distraction has proved to be the most useful method for solving breathing and swallowing problems in patients with severe mandibular hypoplasia asides of improving facial esthetics.[9,10]

In mandibular retrognathias there is a severe hypoplasia of the mandibular ramus, body and chin so by creating new bone in the posterior part of the mandible (body and ramus) a more anterior positioning of the mandible is obtained,however a moreeffective chin is not always obtained.[13, 14]Once growth has ended the convenience of performing only a mentoplasty or mandibular osteotomies must be reconsidered to achieve the esthetic results.

It is so that osteogenic distraction has quickly become the treatment of choice in craniofacial syndromes with severe mandibular deformities because it is possible to perform during childhood opposite to conventional treatments which may only be performed upon completion of growth; this statement has been controversial due to the fact that multiple studies also report successful results by performing mandibular osteotomies in children.[8,19,25]