HYBRID FACIAL OSTEOMAS WITH PROSBOSIS IN PEDIATRICS.

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CONTENTS.

PREFRACE

Osteomas and Prosbocis is an un commons patology in the pediatric patients, it has a full relations with embryology, but one of the biggest enigma is the beginning of this patology, but no many cases, and we reported the first case of this entity.

So we decide to write this book, because this is an unknow patology just the prosbocid, and conjugated with and osteoma is 3 more times more uncommon, in plastic surgery we know all variant of clef lip and palate, but this is related with the patology with study for many years in craniofacial embryology anomalities.

The reconstruction of this cases are not simply, it´s a hard work of many especialities, until Genetic, Pediatric-, Cardialogist, Urologist, Nefrologys, Radiologist, Anestesiologist and Plastic Surgery, having all speciality informes and having a green light to begin the surgery is a hard work.

ACKNOWLEDGMENT.

The prosboscis with osteomas is three times more uncommon than any pathology of clef of lip and palate, in order to present the case to all speciality, and having a piece of knowledgment of everyone of the first Case reported.

We want to thank all the plastic surgery division from the nurses, residents from the first of four year, our Expert in clef lip and palate (M.D. Silverio Tovar), the Genetic Service that help us to conclude a diagnosis, and exclude another signs to not conclude a Syndrome, the Pathology service that help us to exclude cancer, or neoplasm, and the general doctors of the community of the patient to catching us in contact for the evolution of the patient.

ABSTRACT.

Introduction.
Proboscis Lateralis is a rare congenital craniofacial anomaly with a birth prevalence of 1/100,000 to 1/1,000,000.1 It is characterized by a rudimentary nose like tubular appendage arising from either the medial or lateral canthus, nose or chin.

Case Report.
We present and infant that since birth has a prosbosis, during his genetic study it didn't confere about a syndrome, so we have turning to the surgery room, performing a full remove of the prosboscis anomalous and reconstruct the lip, nose and mucosa from both mentioned.

Discusion.
The spectrum of facial malformations was diverse. The most severe facial malformations of cyclopia, synopthalmia, and proboscis with arrhinia have an overall prevalence in patients with Holoprosencephaly ranging from 7.4% to 27%, and they are invariably associated with the alobar subtype.

Conclusion.
The Prosbosis has anomalous origin and one of them was this patient, but having a full knowledge of the disease and complementary imagen and analisys, the surgery became successful, It is not a commun disease, but the technique of the surgery was the key.

Key Words. Malar Osteoma, Surgery, Cleft Repair.

INTRODUCTION.

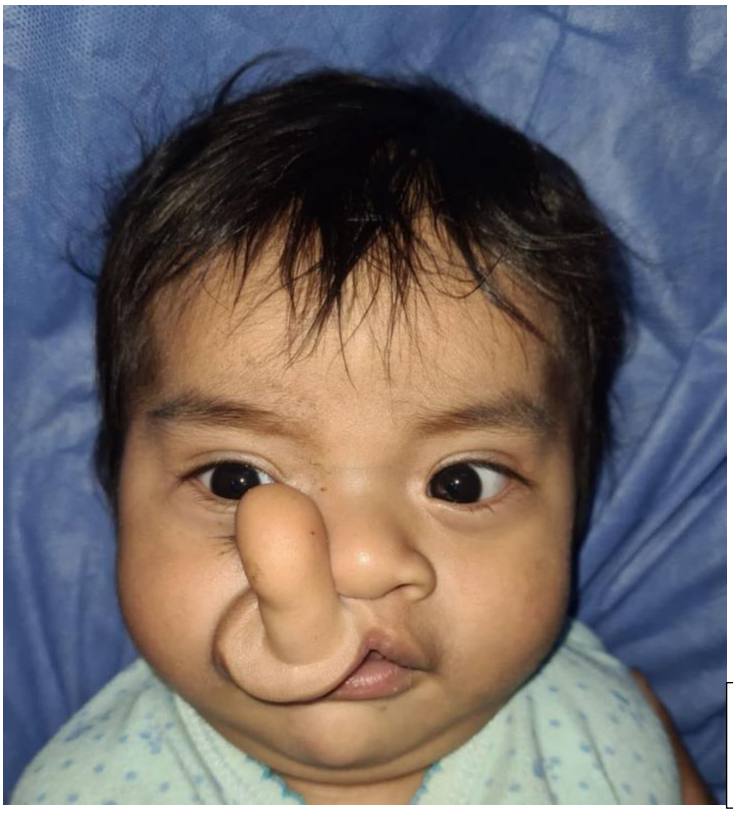
Proboscis Lateralis is a rare congenital craniofacial anomaly with a birth prevalence of 1/100,000 to 1/1,000,000.1 It is characterized by a rudimentary nose like tubular appendage arising from either the medial or lateral canthus, nose or chin. The cylindrical structure can have a small lumen along its length with an opening at the end. Heminasal aplasia, choanal atresia, and paranasal sinus hypoplasia can be present on the ipsilateral side[1].

The real etiology of the anomaly is also unclear. A deficiency in one of the nasal placodes may directly cause the development of heminasal aplasia. Central nervous system and facial bone anomalies may occur with proboscis lateralis, as in most patients with soft-tissue-related issues. There is no consensus about the appropriate time of surgery, but performing the surgery may create an acceptable situation[1].

There are 4 groups of proboscis lateralis (proboscis is a blind ended tube-like structure located in the midface) according to Khoo. Group I affected individuals will have proboscis lateralis with a normal nose. Group II individuals will have proboscis lateralis with an ipsilateral deformed nose. (This is dependant of the patient) Similarly, Group III patients will have proboscis lateralis with an ipsilateral deformed nose, eye, and ocular adnexa and Group IV patients have an ipsilateral deformed nose, eye, ocular adnexa, and cleft lip or palate[1-4].

CASE REPORT & TECHNIQUE.

PATIENT INFORMATION AND CLINICAL FINDINGS



We are reporting a case of proboscis lateralis with Hybrid Osteoma in the General Hospital of Mexico, “Dr. Eduardo Liceaga”, which was seen for the first time on 06/15/2021 by the plastic surgery service.

He was born to a 28-year-old mother, the product of pregnancy 5, is unknown in which week of gestation, born vaginally at home attended by a midwife, apparently without complications. The weight and height at birth of her are unknown. He is the youngest of four siblings, his mother does not report that any of his other children had a congenital malformation.

Imagen 1. Infant boy, with an hybrid prosbosis with osteoma of content.

A 7-month-old male lactating patient, who was referred to this hospital from a rural community in the state from Guerrero in México, began his condition from birth, presenting a bone-dependent tumor, involving skin and cartilage, also presenting hair follicles. [IMAGE 1]

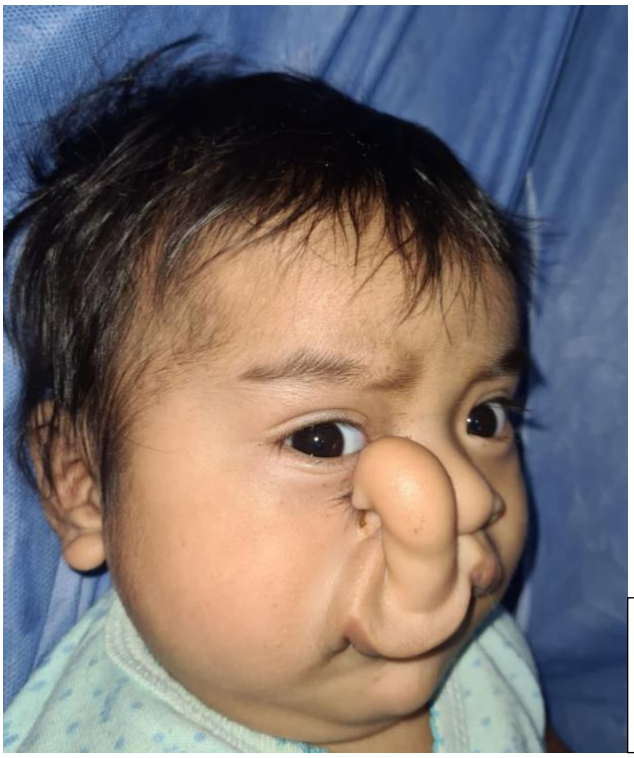


Imagen 2. Pediatric boy, with ¾ picture, admire the prosboscic in all the mid face, no dependand of the suborbital eye.

Cutaneous horn-shaped structure approximately 3 cm long and 3 cm in diameter pedunculated with a circular base on the right upper lip, which emits a cylindrical projection towards the cephalad, approximately 4 cm long and 1 centimeter in diameter, of soft consistency with a cartilaginous bottom, spindle shape, with the presence of hairs, at the tip, which joins the nose in a subunit of the right lateral wall and right wing; short columella deviated to the right, intraoral: the tumor extends towards the gum and ipsilateral palate. [TAC FINDINGS, IMAGE 2,3]

2

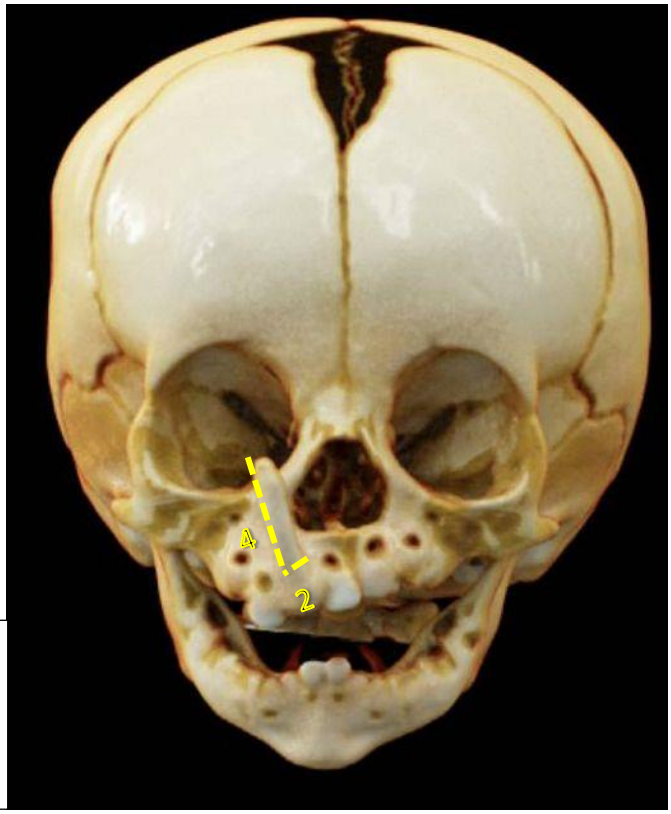
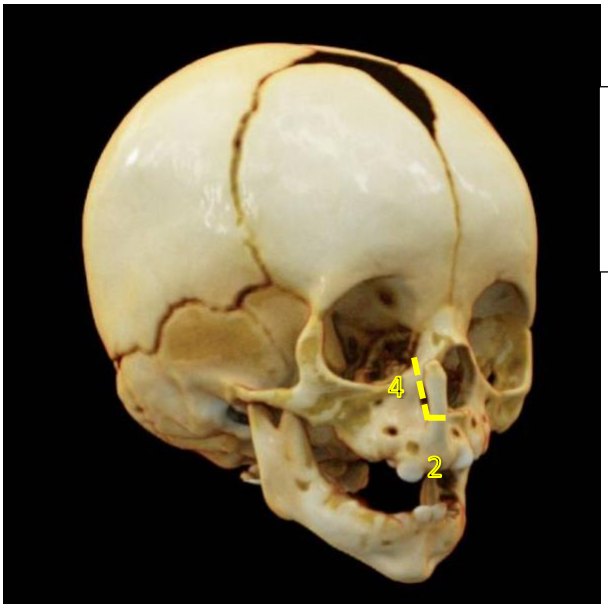


Imagen 3. 3D RECONSTRUCTION OF THE CORONAL IMAGEN OF FACE OF THE PATIENT WITH A OSTEOMA ALL OVER DE MAXILAR, MEASURES FROM IMAGEN OF 4 X 3 X 2 CMS.



[TAC FINDINGS, IMAGE 4]

Imagen 4. 3D RECONSTRUCTION VIEW OF THE ¾ OF THE FACE OF THE PATIENT WITH A OSTEOMA ALL OVER DE MAXILAR, MEASURES FROM IMAGEN OF 4 X 3 X 2 CMS.

There was no history of mixed marriages or family history of birth defects. The prenatal history was negative for exposure to alcohol, drugs or ionizing radiation. Physical examination revealed normal nasal passages. There was no accompanying eye, eyelid, or nose abnormality. Imaging studies showed a horn on the right upper lip.

TECHNIQUE.

During the surgery we begun with a oral intubation, the initially of the surgery was with the dissection of the mucosal flap [Imagen 5] , doing the dissection of the superior maxilla continuing the osteoma a clean cut with the chisel [Imagen 6,7]. Doing the dissection from the medial third of the face of the prosbocid, realize the lateral movement for a total thickness cut of the superior lip, and begin the reconstruction of a hybrid cheiloplasty, and the full remove of the prosbocid of 5 cm of long, 2 cms of width, and 2.5 of thickness [Imagen 8,9,10]. After the full removement and analyze the residual defect of the tumor [Imagen 11].
The reconstruction time began, with the compensation of the excess of tissue of lateral upper right lip, first with the mucosa flap for the vestibular floor and the main compensation, and then the orbicular miorraphy with U stitches, the compensation of the upper lip with the excedent of bermellon subunit and the lesser lateral filtrum with a linear cheiloplasty, and a maccomb dissection of the alar nose for the retraction of the alar nose, and columnella, with monocryl stitches for the regularization, after the compensation we set a nasal shaper that 1 month after the follow-up spontaneously remove [Imagen 12,13].

Imagen 5. The surgery initial mucosal flap, *in the yellow line*, to lift up the dissection of the osteoma.

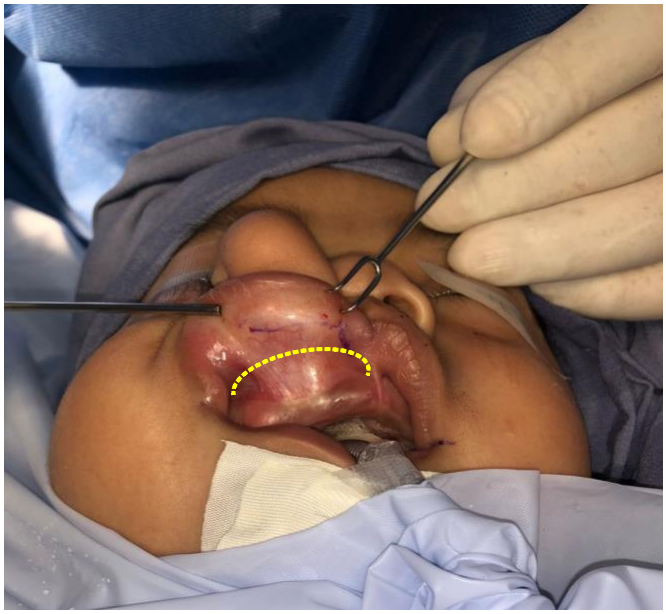


Imagen 6. The dissection of the osteoma from the dissection of the maxilar bone.

Imagen 7. Dissection of the superior lip, all doing an osteotomy.

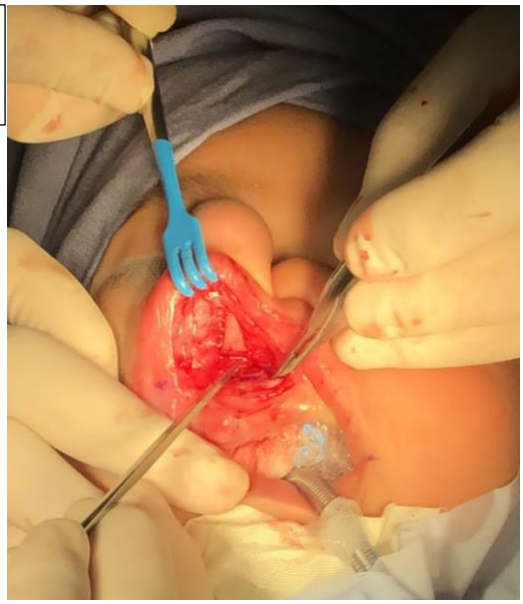


Imagen 8. Free dissection of the prosboscid all over the superior lip and the full cut of the total thickness.

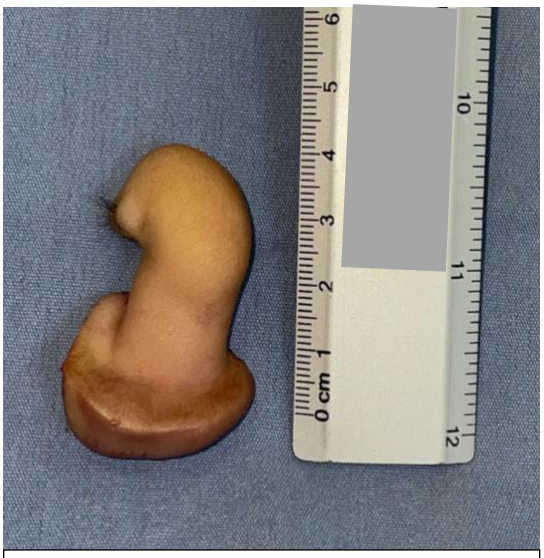


Imagen 9. The prosbocid with hamartoma osteoma component of 5x2x2 cms, from the anterior view.

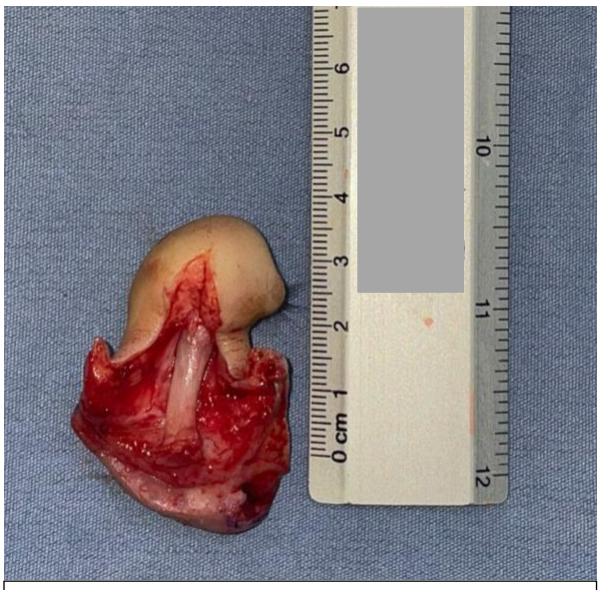


Imagen 10. The prosbocid with hamartoma osteoma component of 5x2x2 cms, from the posterior view.

Imagen 11. The central defect of the tumor in the upper right lip.

Imagen 12. Total reconstruction of the upper right lip (Mucosal flap, miorraphy, and lineal cheiloplasty).

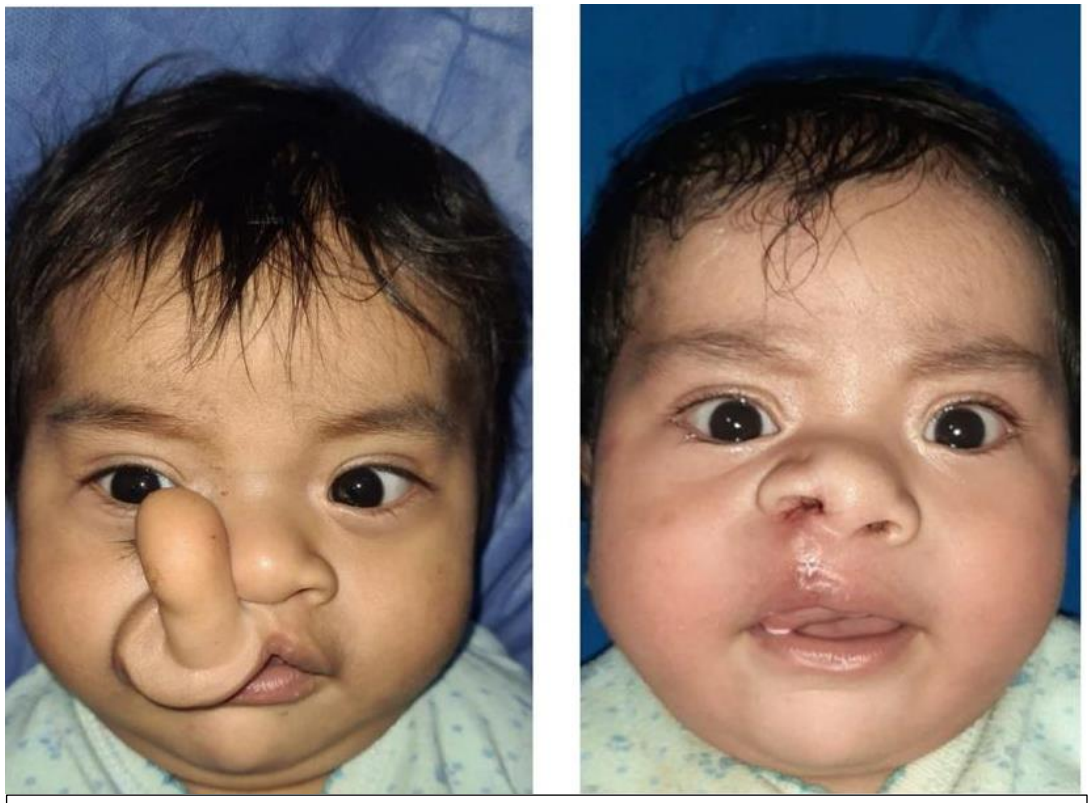


Imagen 13. Comparative of initial versus final result al 1 month of follow up after reconstruction.

During the follow up of the patient, we will reconstruct the nose, in older age, in his follow up consultant the patient is in perfect conditions, the family is happier with his instant result, and grateful with the team of the Plastic Surgery of the General of Hospital, “Dr. Eduardo Liceaga”.

DISCUSION.

The spectrum of facial malformations was diverse. The most severe facial malformations of cyclopia, synopthalmia, and proboscis with arrhinia have an overall prevalence in patients with Holoprosencephaly ranging from 7.4% to 27%, and they are invariably associated with the alobar subtype[1,4-6].

Proboscis Lateralis was first reported by Selenkoffin 1884 where he described the case of a 34 year old Finnish farmer with proboscis lateralis. At the embryological level, proboscis lateralis is thought to be the result of abnormal mesodermal proliferation in the frontonasal and maxillary processes[2,3-6].

Proboscis lateralis, present patterns of anomalies are more severe. Half-nose and proboscis are typically evident deformities of the facial skeleton that can be diagnosed at birth or prenatally. Prenatal diagnosis is important for the psychological preparation of the family. The aims of reconstruction include achieving a pleasing esthetic and a functional result, which is a challenge for plastic surgeons[2].

Often attaches in the upper medial canthal region; however, atypically, it may lie in the lateral canthus, the lateral supraorbital ridge, the midupper eyelid, the root of the nose in the midline, the chin,and just above the nostril [5].

Proboscis lateralis is a rare congenital anomaly among the nasal malformations. It is believed to be a variety of the median cleft.The fusion of lateral and medial nasal processes forms the external nose during embryogenesis. Because the lateral nasal process supplies the material for the proboscis, any anomaly of this structure may cause the deformity. It is commonly unilateral.There are only two cases of bilateral proboscis reported in the literature[5].

According to Boo-Chai, the proboscis used to be extirpated in the early 1900s. Excision of the proboscis may still be adequate for patients in- cluded in group I of Boo-Chai’s classification. According to Denecke and Meyer,Young was the first to publish a description of the correction of proboscis lateralis, in 1949[5].

Treatment options of proboscis lateralis greatly depend on the associated malformations and the age of presentation. The proboscis itself is the ideal donor to reconstruct the heminasal aplasia. Never- theless, Integration of the proboscis tissue in the reconstruction is governed by the proximity of the proboscis to the nose. Reconstruction of the nasal airway in cases ofsevere heminasal hypoplasia and accompanying airway obstruction is dependent on whether or not the unilateral nasal breathing is adequate[1-6].

In the reconstruction of proboscis lateralis tissue, local, forehead, and extended forehead flaps are mainly used.In early interventions, tissue trauma should be kept to a minimum and local tissues should be used as much as possible; the forehead area should be preserved for necessary situations[3-6].

This can require further operations that may affect the growth and mobilization of surgically damaged local and regional tissues. There is no accepted standard for the time of surgery. Some authors argue that surgical intervention should begin in infancy. These rare anomalies can have a devastating influence on the patient and the family, which creates twice the difficulties for the surgeon because of the challenge of the recon-struction and the expectations of the family. The most important point is not only to manage the problem but also to coordinate with the family in terms of treatment and follow-up time. The patient's relatives should be informed that the final outcome after surgery will never be the same as the unaffected side. In such situations, surgery is primarily for functional purposes and esthetic expecta-tions should not be too high[5,6].

CONCLUSIONS.

The prosbosis and osteomas is and hybrid entity that we not know, this is the first case we ever treat in the General Hospital of Mexico, Plastic Surgery Division, so we decide to make a book, like a case report, and inform all the plastic surgery entities about this new entity, and had an antecedent of it. And a perfect technique to offer a surgical treatment, a modified cheiloplasty with a linear closure. Even the patient has a difficult position for a constant follow up at the consult, for the place that the family live.

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ABOUT THE AUTHORS.

To be a plastic surgeon resident is something that is not so easy to become, but when you get there, you realize that doing surgery is not the only thing that you need to know, not everything is perfect, you have to work and deal with abnormalities, even al craniofacial. Clef lip and palate is common altruism surgery we made, and the variants of this disease are not impossible in Plastic Surgery.