

# Nasal Duplication Combined with Cleft Lip and Palate: Surgical Correction and Long-Term Follow-Up

Kanharith Long, MD\*  
Kazuaki Yamaguchi, MD†  
Daniel Lonic, MD‡  
Vanna Long, MD\*  
Vuthy Chhoeurn, MD\*  
Lun-Jou Lo, MD§

**Background:** Diprosopus dirrhinus, or nasal duplication, is a rare entity of partial craniofacial duplication.

**Methods:** The case we present is the first report of diprosopus dirrhinus associated with complete cleft lip and palate. The baby was born in Cambodia at full term by normal vaginal delivery with no significant perinatal and family history. Physical examination revealed significant facial deformity due to the duplicated nose and the left complete cleft lip/palate on the right subset.

**Results:** There were 4 nostrils; both medial apertures including the cleft site were found to be 10–15 mm deep cul-de-sac structures without communication to the nasopharynx. The upper third of the face was notable for hypertelorism with a duplication of the soft-tissue nasion and glabella. Between the 2 nasal dorsums, there was a small cutaneous depression with a lacrimal fistula in the midline. Surgical treatment included the first stage of primary lip and nose repair and the second stage of palatoplasty.

**Conclusions:** The patient was followed up at the age of 10 years showing satisfactory results for both aesthetic and functional aspects. Further management in the future will be required for the hypertelorism and nasal deformity. (*Plast Reconstr Surg Glob Open* 2017;5:e1524; doi: 10.1097/GOX.0000000000001524; Published online 20 October 2017.)

## INTRODUCTION

Diprosopus, or craniofacial duplication, is a rare entity with approximately 100 reported cases since 1900, occurring in approximately 1 in 180,000 to 15 million live births.<sup>1,2</sup> The spectrum of diprosopus varies from complete duplication of facial structures with 2 faces to partial craniofacial duplication on a single head. Diprosopus is considered to be a subtype of the conjoined twins anomaly.<sup>3</sup>

Nasal duplication is an extremely rare subtype of this condition. We report the case of a newborn infant with complete nasal duplication combined with a unilateral cleft lip/palate and the long-term follow-up.

*From the \*Department of Surgery, National Pediatric Hospital, Phnom Penh, Cambodia; †Shonan Beauty Clinic, Ryogoku/Shinsaibashi, Tokyo/Osaka, Japan; ‡MFACE KieferGesichtsZentrum München, Munich, Germany; and §Plastic and Reconstructive Surgery, and Craniofacial Research Center, Chang Gung Memorial Hospital, Chang Gung University, Taoyuan, Taiwan.*

*Received for publication May 3, 2017; accepted August 17, 2017.*

*Copyright © 2017 The Authors. Published by Wolters Kluwer Health, Inc. on behalf of The American Society of Plastic Surgeons. This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.*

DOI: 10.1097/GOX.0000000000001524

## CASE REPORT

This study was performed conforming to Helsinki Declaration. The boy was referred to the National Pediatric Hospital for evaluation of a rare anomaly during a regular cleft surgery mission in Phnom Penh, Cambodia (Fig. 1). The baby was born full term. The mother denied exposure to medication, infection, alcohol, radiation therapy, or consanguinity. There was no family history for congenital anomaly.

Physical examination showed that the oral cavity was a single structure without duplication opposed to the duplicated nose, which presented with synchronous movements. Although there were 4 nostrils, both medial apertures were found to be 10–15 mm deep cul-de-sac structures without communication to the nasopharynx. The left upper lip had 2 distinct Cupid's peaks, whereas there was only 1 Cupid's peak on the right lip. Hypertelorism with duplication of the soft-tissue nasion and glabella was accompanied by a small cutaneous depression with a lacrimal fistula. The oral commissures and the chin were normal. The patient was able to eat with mild regurgitation. Growth and development were noted to be in the lower normal range.

Computed tomography revealed bony deformities including a wide skull base and duplication of the nasal bone. In the cerebral parenchyma, structural anomalies

**Disclosure:** The authors have no financial interest to declare in relation to the content of this article. The Article Processing Charge was paid for by the authors.



**Fig. 1.** The patient at 3 months old, showing nasal duplication with left complete cleft lip and palate on the right subset. A lacrimal fistula was noted in the middle of 2 eyes. Both medial nostrils including the cleft site were cul-de-sac structures without patency to nasopharynx.

were observed including large-sized lateral ventricles without an anterior horn and a fused third ventricle with a suprasellar cistern without signs of hydrocephalus.

At 15 months of age, lip repair and rhinoplasty were performed. Excision of the 2 medial nostrils was designed. Marking lines included half of the philtrum, nasal ala and tip of the middle 2 nostrils, as well as the nasal dorsum on both sides of the noses (Fig. 2). The excision was placed below the nasion and fistula to avoid injury to the structure. The left-side columella was preserved, but the right-side columella was excised with the right lip incision in a curvilinear fashion to facilitate downward rotation. Mucosal excision was limited to preserve tissue for tension-free closure. The dissection revealed a duplication of the lower part of the nasal septum. As for the bony structures, the palatal shelf on the right side extended posteriorly to the choanae and connected to the perpendicular plate of ethmoid bone. The cleft side ala including the right columella and anterior lower nasal septum were removed. The left columella was cut at the base of the anterior nasal spine for mobilization. After these dissections, the reconstruction of the nasal floor with preserved cleft margin mucosa was completed. Muscle repair was achieved, and a small Z-plasty was performed above the Cupid's bow. Nose reconstruction was completed with a Z-plasty in the middle of the dorsum. The postoperative course and wound healing were uneventful.

Palate repair was performed at the age of 24 months, using the 2-flap method in the hard palate and small dou-



**Fig. 2.** Resection of the middle part of the duplicated noses was outlined.

ble-opposing Z-plasty in the soft palate. The buccal fat pad was used on both sides to cover the lateral relaxing incisions.<sup>4</sup> The recovery was smooth.

The patient lived and was regularly followed up in Cambodia (Figs. 3, 4). For the past 10 years, his psychomotor development was within normal limits. Perceptual speech assessment showed adequate resonance. Tearing from the glabellar lacrimal fistula was occasionally observed but not bothering. The patient attended regular elementary school with good peer relationship.

## DISCUSSION

A large area of duplication like complete conjoined twins commonly results in intrauterine death, because of central nervous system, heart or gastrointestinal anomalies.<sup>5</sup> The prognosis for survival is better with partial diprosopus such as the nose, eyes, or mouth.<sup>6</sup> An accepted etiology for the condition is incomplete splitting of a single embryo between the 13th and 25th days after conception.<sup>7</sup> The cleft lip and palate entity occurs at later timing as a result of fusion failure between the 4th and 12th week of gestation, and this happened in the right subset of the patient (Fig. 1). Prenatal diagnosis of diprosopus using ultrasound has been reported.<sup>1</sup>

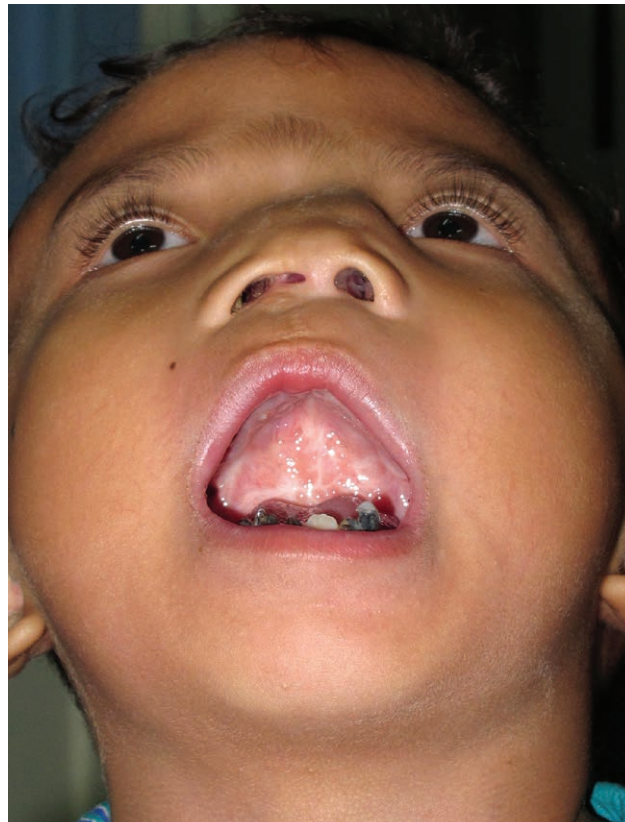
There are various types and locations of duplication such as nose,<sup>8</sup> maxilla or alveolar arch,<sup>1,2</sup> tongue,<sup>6</sup> or mandible. Duplications of these structures are also reported to occur in combination.<sup>9</sup> Some patients were reported to have a cleft lip/palate,<sup>1</sup> and 3 cases of supernumerary nostrils with cleft



**Fig. 3.** Patient was 6 years 6 months old during a follow-up, showing acceptable lip but nasal deformity.

lip and palate were reported in the literature.<sup>10</sup> However, the supernumerary nostrils were not considered a duplication, as they were located above the normal nostrils without an additional nasal septum and appeared to be secondary to nasal pits.<sup>9</sup> Therefore, we consider this patient to be the first reported case of combined complete nasal duplication and unilateral complete cleft lip and palate. From the facial morphology, this boy could also be regarded as 0–14 cleft with nasal duplication. The patient's maxillary teeth and alveolar cleft were similar to those of the regular right cleft lip and palate, instead of the Tessier no. 3 or 4 cleft.

From a psychosocial standpoint, the most important issue for patients with congenital facial anomaly is to obtain acceptable appearance and function to avoid isolation in society. The rationale of decisions about the timing and orders of surgeries was safety of anesthesia and recovery of wounds. As the baby lived in Cambodia and was small, we delayed the first repair until 15 months of age during our next regular mission trip. The second surgery of palate repair was performed at 24 months of age by the local team. Certainly, the timing and orders could change in different scenarios. Decision of the surgical repair was made to excise the 2 central nostrils because the 2 were blind ends without communication to nasopharynx. Another option could be to keep the left nose and completely excise the right one. Using this method, the right side tissue would be moved during the future correction of the hypertelorism. As this is a rare case with individual structural difference, it is difficult to compare the results of different approaches. The outcome was acceptable in this case after the 2



**Fig. 4.** The nostrils were patent in airway. Palatal repair was good with adequate speech function.

surgeries, but the problems of hypertelorism and nasal deformity remained. However, judging from the patient's situation and the overall possibilities of medical care in the country, conservative management of the residual deformities was applied. Regular follow-up is mandatory, and further treatment will be considered after the growth spurt.

*Lun-Jou Lo, MD*

Department of Plastic and Reconstructive Surgery  
Chang Gung Memorial Hospital  
5 Fu-Shin Street, Kwei Shan  
Taoyuan, Taiwan 333  
E-mail: lunjoulo@cgmh.org.tw

### ACKNOWLEDGMENT

*Part of the cleft care for this patient was supported by the Noordhoff Craniofacial Foundation in Taiwan.*

### PATIENT CONSENT

*Parents or guardians provided written consent for the use of the patient's image.*

### REFERENCES

1. Wu JK, Kozakewich HP, Mulliken JB. Partial oral duplication: a defect in ectomesenchymal migration? *Plast Reconstr Surg.* 2003;112:1645–1648.
2. Costa MA, Borzabadi-Farahani A, Lara-Sanchez PA, et al. Partial craniofacial duplication: a review of the literature and case report. *J Craniomaxillofac Surg.* 2014;42:290–296.

3. Machin GA. Conjoined twins: implications for blastogenesis. *Birth Defects Orig Artic Ser.* 1993;29:141–179.
4. Yamaguchi K, Lonic D, Lee CH, et al. Modified Furlow palatoplasty using small double-opposing Z-plasty: surgical technique and outcome. *Plast Reconstr Surg.* 2016;137:1825–1831.
5. Spencer R. Theoretical and analytical embryology of conjoined twins: part I: embryogenesis. *Clin Anat.* 2000;13:36–53.
6. Verdi GD, Hersh JH, Russell LJ. Partial duplication of the face: case report and review. *Plast Reconstr Surg.* 1991;87:759–762.
7. Strauss S, Tamarkin M, Engelberg S, et al. Prenatal sonographic appearance of diprosopus. *J Ultrasound Med.* 1987;6:93–95.
8. Kotrikova B, Hassfeld S, Steiner HH, et al. Operative correction and follow-up of craniofacial duplication. *Plast Reconstr Surg.* 2007;119:985–991.
9. Barr M, Jr. Facial duplication: case, review, and embryogenesis. *Teratology.* 1982;25:153–159.
10. Powar RS, Tubaki VR. Supernumerary nostril with complete unilateral cleft lip: a case report and review. *Cleft Palate Craniofac J.* 2007;44:657–659.