

IMAGES

Nasal Reconstruction of a Frontonasal Dysplasia Deformity Using Aesthetic Rhinoplasty Techniques

Sin Young Song¹, Jong Woo Choi¹, Han Wook Lew², Kyung S Koh¹

Departments of ¹Plastic and Reconstructive Surgery and

²Pediatrics and Genetics, Asan Medical Center, Ulsan University College of Medicine, Seoul, Korea

Correspondence: Jong Woo Choi

Department of Plastic and Reconstructive Surgery, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 05505, Korea

Tel: +82-2-3010-3600, Fax: +82-2-476-7471, E-mail: pschoi@amc.seoul.kr

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Frontonasal dysplasia (FND) is an unusual congenital condition that presents with a broad range of phenotypes [1]. For this reason, only a small number of cases and the management thereof have been reported in the literature. No consensus has been established regarding the appropriate technique for correcting associated nasal deformities. The clinical picture is highly variable, but the major characteristics include hypertelorism (an increased distance between the eyes), a broad nasal root, a large and bifid nasal tip, and a widow's peak [1,2]. Occasional abnormalities include accessory nasal tags, cleft lip, ocular abnormalities (coloboma, cataract, and microphthalmia), conductive hearing loss, basal encephalocele (an opening of the skull with protrusion of the brain), and/or agenesis of the corpus callosum [1,2]. The majority of affected individuals have normal intelligence. The etiology of the condition remains unknown, but seems to be linked to defective embryologic development of the nasal capsule [1,2]. The disorder is thought to be sporadic in most cases [2]. The correction of severe nasal deformities is a valid consideration for FND patients for whom improvement is possible; however, the optimal timing and method for this procedure are

still controversial.

A boy with non-consanguineous parents and no family history of FND was born by normal spontaneous vaginal delivery. His birth weight was 2.370 kg and the antenatal period was uneventful. On physical examination, the boy was found to have a wide intercanthal distance and a nose deformity characterised by small deformed nostrils and a bifid nose (Fig. 1). Genetic counseling was provided, and the prognosis and appropriate treatment strategies were explained to the parents. Genetic analysis revealed that the patient had a normal *ALX3* gene sequence, indicating a sporadic occurrence of FND (Table 1). If the patient desires the exact gene sequence, it is possible to analyze the *ALX1* or *ALX4* genes, but the parents of the patient did not want to do so in this case. The parents wanted the child to undergo surgical correction as soon as possible to allow for optimal recovery and adaptation. A course of early nasal reconstruction was chosen, and nasal reconstruction based on the principles of aesthetic rhinoplasty was performed at the age of two years and nine months.

The patient presented with mild hypertelorism, a broad nasal root, a large and bifid nasal tip, and a widow's peak. Accessory nasal tags, cleft lip, ocular abnormalities, conductive hearing loss, and basal encephalocele were not observed.

The major obstacles to restoring the nasal anatomy in this patient included a very short columella,



Fig. 1. Images of a child with features of frontonasal dysplasia, including true ocular hypertelorism, broadening of the nasal root, lack of nasal tip formation, and nose deformity with small nostrils and irregular dorsal skin.

Table 1. Mutation details and clinical features of subject with *ALX3* mutation

Sample ID	Karyotype	Country of origin	Method ^{a)}	Exon	Exon-intron boundary	Point mutation	Additional clinical features
K308406	46XY	Korean	Normal	Normal sequence	Normal sequence	None on slicing junction	Small deformed nostril Large and bifid nasal tip Hypertelorism Broad nasal root Widow's peak

^{a)}(1) Phytohaemagglutinin-stimulated culture technique, GTL-banding; Peripheral blood gene analysis; Count 20 cells in metaphase and analysis more than 5 cells in metaphase with 550 band-level resolution; (2) No information of *ALX1* or *ALX4* gene; (3) Known polymorphism of homozygote (IVS1(+33) C>T; c.*68_69ins28 was found in polymerase chain reaction-sequencing.



Fig. 2.
Preoperative markings. Columellar reconstruction was performed using V-Y advancement of the upper part of the upper lip.



Fig. 3.
Conchal cartilage was harvested and underdeveloped areas of lower lateral cartilage on both sides were reconstructed. Additional conchal cartilage was used for derotation of the nasal tip. The upper lateral cartilage was approximated for dorsal heightening. A 1–4-cm AlloDerm was trimmed and folded for dorsal augmentation.

increased alar width, separated lower and upper lateral cartilages, bifid dorsum with severe skin indentation, and a deficient nasal framework. Two strategies were devised to overcome these obstacles. First, the deficient nasal framework was reconstructed by grafting additional cartilages and using appropriate suture techniques. Second, the deficient soft tissue envelope was repositioned on both sides with de-epithelialized bifid nasal soft tissue.

First, various aesthetic rhinoplasty techniques were used, including tip extension suture, derotation graft, septal extension graft using conchal cartilages, and interdomal and transdomal sutures.

A V-Y patterned transcolumella infracartilagenous incision was performed and the scroll area was subsequently dissected in order to allow lower lateral cartilage mobility, as is done in aesthetic rhinoplasty in East Asian adults (Fig. 2). Dissections were

performed along the lower and upper lateral cartilage, which were highly deficient in terms of growth and dimension. In order to reconstruct the lower lateral cartilages, conchal cartilage was harvested from the right ear and attached to the septum, using polydioxanone II (PDS II, Ethicon, Somerville, NJ, USA) 4-0 sutures, in order to function as the septal extension graft. Additionally, we used an AlloDerm (LifeCell, Branchburg, NJ, USA) for dorsal augmentation (1 × 4 cm in size). The upper lateral cartilage areas were approximated in order to narrow the horizontal nasal width, and a derotation graft of conchal cartilage was fixed with PDS II 4-0 sutures, in order to minimize the cephalic rotation of the nasal tip (Fig. 3).

Second, the widespread dorsal redundant soft tissue was de-epithelialized and moved to the central part of the nose in order to augment the nasal

dorsum. Finally, the bifid nasal skin was closed.

After six months, the patient underwent scar revision and minor contouring on the nasal dorsum. Fusiform excision was used to remove the previous scar on the nasal dorsum, and undermining and advancement of the skin flap was performed.

Multistage craniofacial surgery is justified for FND, and is generally performed at six to eight years of age, resulting in satisfactory cosmetic and functional results [3]. However, a second procedure is often needed, as nose development is completed at the age of 14 years or later [3]. Pham et al. [4] reported three novel cases of frontorhiny that were treated with reconstructive open rhinoplasty, with special attention paid to columellar contouring, eradicating bifidity, columellar lengthening with V-Y closure, and enhancing tip projection. The nasal dorsum and underdeveloped nasal tip present unique clinical challenges, and the authors employed a staged definitive rhinoplasty that yielded positive results at a two-year follow-up. Although definitive nasal correction is often performed at older ages, severe nasal deformities have the potential to significantly impact a child's psychological development. This impact leads many parents to request nasal correction as soon as possible. Such a request was the impetus for the early surgery described in this report.

In order to correct the nasal deformity of this patient, various aesthetic rhinoplasty techniques were used, such as a V-Y transcolumella incision, derotation graft, tip extension suture, septal extension using conchal cartilage, and interdomal suture techniques [5]. Our strategy proved successful. Although various surgical methods have been developed for nasal correction in patients with FND, the technique presented herein was feasible and satisfactory. The operation resulted in the early improvement of the patient's appearance, and no complications occurred (Fig. 4). Although the patient may require future secondary rhinoplasty to enhance the projection of the tip and refine the nose shape, the early correction of the deformed nasal skeleton and soft tissue seems to be beneficial for social adaptation and the development of an improved self-image in growing children. Two years after the surgery, the growth of the nose appeared normal.



Fig. 4. Images of the child eight months after the operation.

by recessive mutations in the ALX3 homeobox gene.

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