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Nose underdevelopment – etiology, diagnosis and treatment

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Data Interpretation
E – Manuscript Preparation
F – Literature Search
G – Funds Collection

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ABSTRACT:

Introduction. Clinical picture of nose underdevelopment is very polymorphic. It can exist from partial disturbances of nose development, through unilateral hypoplasia of the nose to congenital absence of the nose (arhinia).

Aim. The aim of this study is to present own experience of treatment of patients with different types of nose underdevelopment.

Material and method. Plastic Surgery Out-patient Clinic has under care 13 patients with isolated nose underdevelopment. In this group there were 5 men and 8 women. The patients' age, at the time of first consultation, was from 6 months to 19 years.

Results. The most frequent type of defect in analyzed group was congenital saddle nose (6 persons). Three patients were diagnosed with absence of the quadrilateral cartilage of the nasal septum, two with isolated underdevelopment of the alar cartilage and one with bilateral underdevelopment of the nasal bones. The most advanced disturbance was observed in 6 months old girl, who had unilateral hypoplasia of the nose. Nine patients were surgically treated.

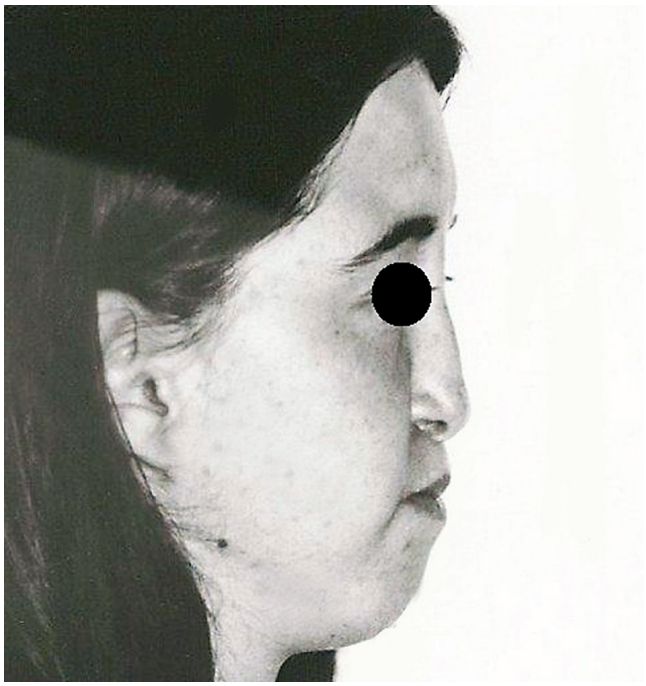
Conclusions. Nose underdevelopment is rare congenital anomaly and is characterized by variable clinical picture. Medical history and precise physical examination are fundamental in proper diagnosis. Disturbances in nose development can lead to serious functional consequences and require individual plan of treatment.

KEYWORDS: nose underdevelopment, clinical picture, surgical treatment

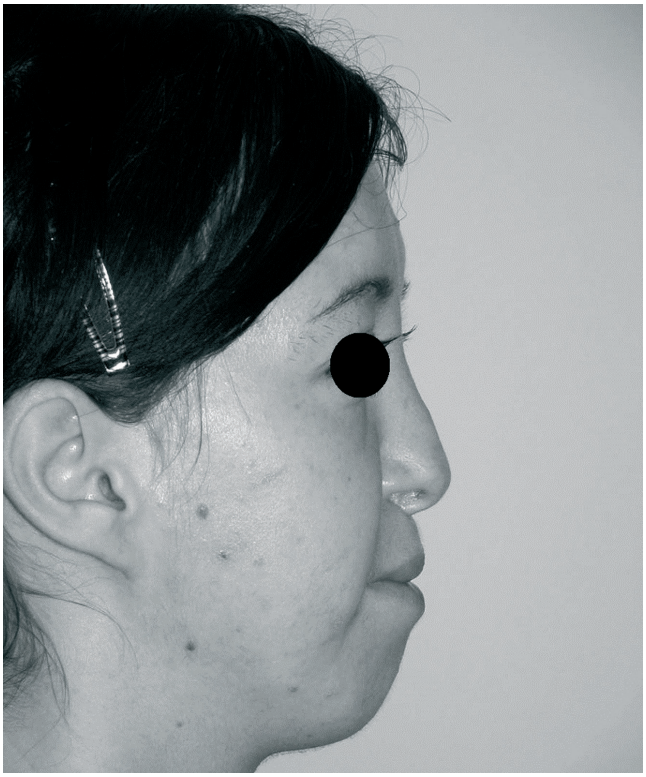
INTRODUCTION

Embryological development of the nose starts in the third week after fertilization and can be estimated as complete in about eighth week, when fundamental parts of the fetal face are developed [1,2,3]. On day 24, the face consists of 5 processes, placed around a primitive mouth (stomodeum): a superior frontal process, paired bilateral maxillary processes and paired bilateral mandible processes [2]. Paired processes develop from the first branchial arch. In the third gestational week, the ectoderm of the frontal prominence starts to proliferate and forms the nasal placodes [4]. In the fifth week the central region of the nasal placodes invaginates and forms the primitive nasal pits, from which the nostrils are developed. The primitive nasal pits further penetrate into the primitive oral cavity, the region surrounded by the frontal, maxillary and mandibular prominences, and form the olfactory pits, from which the nasal cavities are developed [4]. Simultaneously, mesenchyme around the nasal placodes proliferates and builds paired medial and lateral nasal processes [3]. In the sixth and seventh week medial nasal processes fuse which results in formation of the dorsum and tip of the nose and the invaginated part of the medial nasal prominence form the nasal septum [3]. The proper fusion between medial and lateral nasal prominences is responsible for forming the alae of the nose, and correct connection between frontal and medial nasal processes determines development of the nasal bones [1,2].

Congenital anomalies of the nose include a broad spectrum of defects, being a result of abnormalities in the above mentioned developmental process. These conditions range from partial deformities of the nose, such as isolated absence of the nasal bones, absence of the columella, absence of the septal cartilage or alae cartilage, through hemi aplasia of the nose to complete absence of the nose (arhinia) [1,4,5,6,7,8]. Various degrees of nasal malformation present different clinical pictures. In its mild forms the defect causes mainly aesthetic problems. Nose located in the central region of the face has a significant influence on general appearance



Ryc. 1. An 18-year-old patient without the quadrilateral nasal cartilage.



Ryc. 2. The same patient at the age of 20 after surgical treatment.

and self-esteem and even small disturbances can be easily seen and evaluated [9]. In most advanced cases of arhinia, severe airway obstruction and inability to feed occur [2]. All cases required airway management in the neonatal period, most commonly surgical tracheostomy, which allows the infant to breath and eat [2,7].

The surgical plan of treatment performed in patients with nasal underdevelopment is established individually and the methods of nose reconstruction vary in relation to the type of involvement [4]. If the defect is limited to small regions, minor operations using small flaps will improve the patient's appearance [10]. If the external nose is completely absent, large flaps usually with cartilage or bone grafts are necessary for reconstruction [4].

The aim of this study is to present own experience and treatment of patients with different types of nose underdevelopment.

MATERIAL AND METHOD

Plastic Out-Patient Clinic and Plastic, Reconstructive and Aesthetic Surgery Department have under their care 13 patients with isolated nose underdevelopment. Each patient underwent clinical examination, and photographic documentation was performed. On the basis of ambulatory cards, hospital histories and photos the data base of patients with nose defects was created. The collected data included: age, sex, type of anomaly and methods of introduced treatment.

In the analyzed group there were 5 men and 8 women. The patients' age, at the time of first consultation, was from 6 months to 19 years (on average 11 years and 8 months).

On the basis of patients' documentation and created data base the retrospective analysis of performed treatment was conducted.

Due to the descriptive character of the study, the statistical tests were not accomplished.

RESULTS

The collected data are presented in Table 1. Two patients were diagnosed with isolated underdevelopment of the greater alar cartilage; in a female it was unilateral, in a male – bilateral /patient 1 and 2/. The girl was stated to have underdevelopment of the left alar cartilage, which included medial crus, lateral crus and dome. Her left ala was lowered and asymmetry of the nostrils was seen. In a boy, underdevelopment of the greater

Tab. I. Patients with isolated nose underdevelopment treated at our Clinic

No	Sex	Age	Type of nose underdevelopment	Surgical procedures	Additional remarks
1	K	6 years	Left-sided underdevelopment of the greater alar cartilage	Qualification to surgery in later years	Lowered left ala and nostril asymmetry
2	M	13 years	Bilateral underdevelopment of the greater alar cartilage	Cartilage grafts from both auricles to elevate the alae of the nose at the age of 15	Flat alae of the nose
3	K	9 years	Absence of the quadrilateral nasal cartilage	Septum reconstruction with costal cartilage graft at the age of 10, hip bone graft at the age of 18	Flat nose
4	K	18 years	Absence of the quadrilateral nasal cartilage	Hip bone graft in the shape of letter L at the age of 19	Flat nose
5	K	8 lat	Absence of the quadrilateral nasal cartilage, underdevelopment of both medial crus and dome of the greater alar cartilage	Qualification to surgery after 18 years of age /parents' decision/	Flat nose with sunken tip
6	M	14 lat	Congenital saddle nose	Qualification to surgery after 18 years of age	Presence of pseudo-hump, widening of the dorsum and drooping of the tip
7	K	19 years	Congenital saddle nose	Hip bone graft, osteotomy and tip nose plasty at the age of 19	Presence of pseudo-hump, widening of the dorsum and drooping of the tip
8	M	18 years	Congenital saddle nose	Hip bone graft, osteotomy and tip nose plasty at the age of 19	Presence of pseudo-hump, widening of the dorsum and drooping of the tip
9	K	16 lat	Congenital saddle nose	Hip bone graft, and tip nose plasty at the age of 16	Congenital syphilis
10	K	8 years	Congenital saddle nose	Hip bone graft, osteotomy and tip nose plasty at the age of 18	Presence of pseudo-hump, widening of the dorsum and drooping of the tip
11	M	19 years	Congenital saddle nose	Hip bone graft, osteotomy and tip nose plasty at the age of 19	Presence of pseudo-hump, widening of the dorsum and drooping of the tip
12	M	10 months	Bilateral underdevelopment of nasal bones	Qualification to surgery after 18 years of age	Flat nose, increased frontonasal angle
13	K	6 months	Left-sided hemi-aplasia of the nose	Reconstruction of left nostril with nasolabial flap at the age of 3	Further surgeries depending on indications

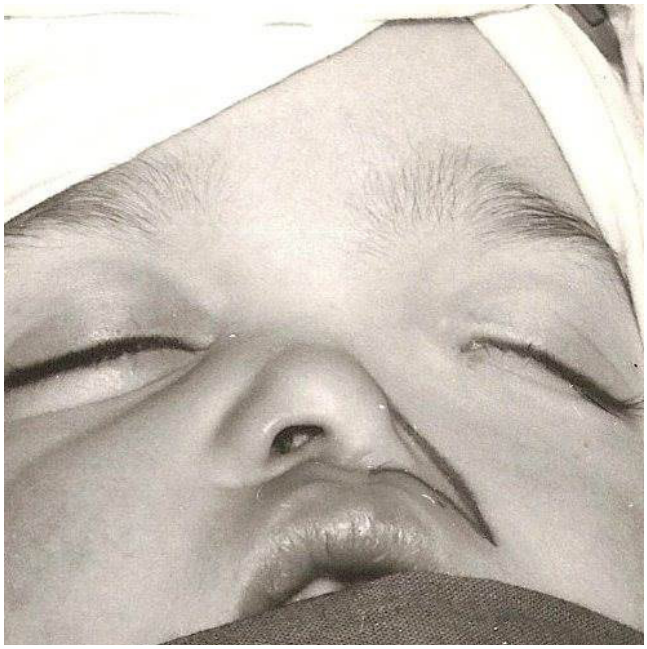
alar cartilages was concentrated mainly in both lateral crura, causing a flat shape of the alae. The male patient was operated on at the age of 15; the alae of the nose were elevated by cartilage grafts taken from both auricles.

Three patients had a flattened nose due to the absence of the quadrilateral nasal cartilage (Fig. 1a). Additionally, in one patient, underdevelopment of both medial crus and dome of the greater alar cartilage was observed, and her tip of the nose was sunken /patient 5/. This patient will be qualified to surgical procedures after 18 years of age. Another patient in this subgroup had the septum reconstructed by using a costal cartilage graft. However, after 18 years of age /when the process of face growth was completed/ the lift effect of the nose was insufficient and a hip bone graft was introduced /patient 3/. The last patient from this subgroup was operated on in the age of 19; a hip bone graft was performed in the shape of letter "L". The longer part of the graft was put to cover the dorsum of the nose, while the shorter one to support the columella of the nose; this provided a proper nose exposition /patient 4/ (Fig. 1b).

The most frequent type of defect in the analyzed group was congenital saddle nose (6 persons), and in one patient it was due to syphilis. The clinical picture in these patients was manifested with the presence of pseudohump, widening of the dorsum and drooping of the tip. Almost all patients in this subgroup underwent surgical procedure with the use of a hip bone graft, osteotomy and tip nose plasty. Such operations were performed after 18 years of age. The only exception was the patient with congenital form of syphilis; she did not have a broad nose bridge, so osteotomy was not necessary and surgery was done earlier at the age of 16 /patient 9/.

In one patient bilateral underdevelopment of the nasal bones was observed /patient 12/. He had a small, flat nose and in-

Ryc. 3. One-and-a-half-year-old female with unilateral nose hypoplasia.



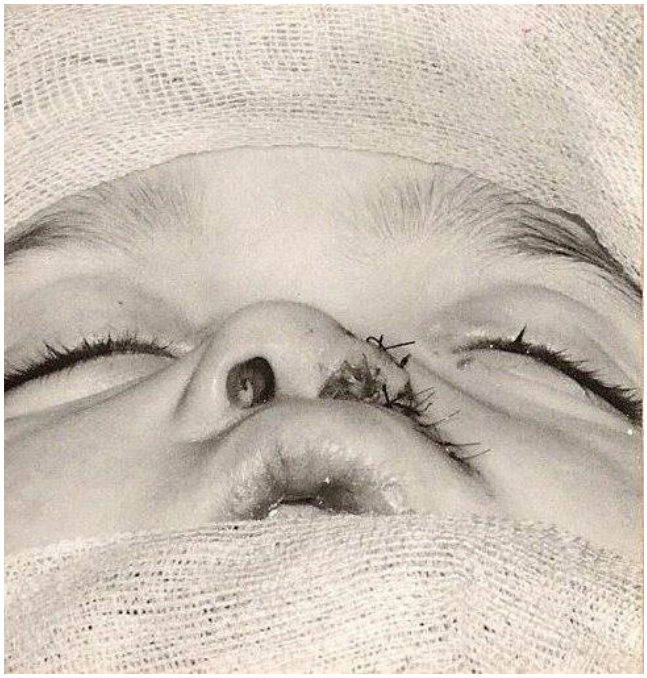
Ryc. 4. The first stage of surgical treatment - reconstruction of the left nostril with the use of a nasolabial flap (at the age of 3).

creased frontonasal angle. However the development of the maxilla was correct. This patient will be qualified to surgical procedures after 18 years of age.

The most advanced abnormality was observed in 6-month-old girl /patient 13/. She was diagnosed with left-sided hemi aplasia of the nose. Normal structures on the right side enabled her proper breathing (Fig. 2a). Operative treatment was introduced by performing reconstruction of the left nostril with the use of a nasolabial flap (Fig. 2b and c). Further surgeries are to be done depending on indications.

DISCUSSION

Congenital anomalies of the nose are rare, with an incidence of one in every 20 000 to 40 000 live-born infants [11]. In the available literature there are mainly casuistic descriptions of patients with different forms of nose underdevelopment (hypoplasia or lack of particular parts of the nose, unilateral nose hypoplasia or arhinia) [1,2,4,5,8,10]. The pathogenesis of nose underdevelopment is poorly understood. It has been postulated that the lack of development of the nose results from failure of the medial and lateral nasal process to grow, abnormal migration of neural crest cells or overdevelopment and early fusion in the medial nasal processes [2,7]. Udagawa et al. and da Silva Freitas et al. claimed that the absence of one nasal placode seems to lead to right- or left-sided hemi-nasal aplasia



Ryc. 5. Early effects of surgery.

[8,12]. Most cases are sporadic. However, genetic hypothesis and familial cases have been described [1]. Considering that our patients did not have other congenital deformities of the body, it is unlikely that genetics was responsible for patients' malformations.

Diver and Hill paid attention to the fact that congenital deformities of the nose may not be present until adulthood [6]. During face development and child maturation some asymmetry and hypoplasia of particular parts of the nose become visible. That is why some patients can present to outpatient clinics even in late adulthood [13]. We made similar observations in our group. Six patients presented for a medical examination at the age of over 14 years, and the mean age was over 11 years. In more advanced malformations that age was lower (example of a 6-month-old girl with unilateral nose hypoplasia). Variable disturbances, which can appear during nose development, are responsible for different symptoms in the clinical picture. Tunçbilek described a patient without the quadrilateral nasal cartilage. She had a flattened nose, increased frontonasal angle and decreased nasolabial angle, but a normal maxilla and nasal bones. Those findings were not compatible with the diagnosis of Binder's syndrome, which is a congenital malformation characterized by a retruded midface, as a result of maxillary hypoplasia [5]. Similar findings were recognized in 3 of our patients who were diagnosed with the lack of the septal cartilage (Fig. 1a). The picture of a flattened nose was also found in one patient without nasal bones. In all of these patients, the described features were isolated, so the diagnosis of Binder's syndrome was not compatible. Andrade et al. presented a group of patients with a saddle nose. Authors reminded that saddle nose is usually caused by a trauma or excessive resection of the septal cartilage. Nevertheless, there are other causes, such as syphilis, leishmaniosis, leprosy, and congenital factors [14]. In this context, it is very common to see a pseudo-hump due to depression of the cartilaginous dorsum, frequently accompanied by a widening of the bony bridge and drooping of the tip [14]. The same clinical picture was observed in 6 of our patients with a congenital saddle nose (Tab. 1).

In the surgical treatment of nasal underdevelopment the methods for nose reconstruction vary from case to case according to the degree of the defect [1,4,7]. Satisfactory correction of a saddle nose can often be obtained by pseudo-hump reduction, narrowing of the nasal bridge by osteotomies of the lateral walls, cartilage or bone grafting of the dorsum and/or columella, and tip remodeling procedures [14]. In patients with hemi-aplasia of the nose different procedures can be applied. Authors prefer to use nasolabial flap, forehead flap or free-tissue transfer with autogenous cartilage or bone graft [1,4,6,8,12]. Similar methods were introduced in our group of patients (Tab. 1).

The goals of nasal reconstruction include restoration of the functional and aesthetic aspects, which still remains a challenge for plastic surgeons [12]. Special attention must be paid to small nuances in particular procedures which should be individually modified depending on the type of deformation and patient's psychomotor development [15].

CONCLUSIONS

Nose underdevelopment is a rare congenital anomaly and is characterized by variable clinical pictures.

Medical history and precise physical examination are fundamental in proper diagnosis.

Disturbances in nose development can lead to serious functional consequences and require an individual plan of treatment

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