APPLIED ANATOMY AND INNERVATION OF THE NOSE

J. Dankmeyer, M. D.,
Leiden, The Netherlands

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EMBRYOLOGICAL INTRODUCTION TO CONGENITAL MALFORMATIONS OF THE NOSE

Zvi Rosen, M.D.

Since the face and nose have a very complex phylogenetic and ontogenetic history, it is understandable why among the developmental abnormalities of the human body, the facial and nasal malformations occur relatively frequently.

Classification of the anomalies is difficult and criteria to be considered are : —

(a) the presumptive time of onset ("teratogenic termination point").

(b) the "cause".

(c) the organs involved.

(d) compatability and incompatability with life.

(e) association with other anomalies, ("causal syngenetic" or "accidental syngenetic").

(f) "degree of severity", which according to Cottle ([1]), may be classified as follows : —

1) arrested growth of substance and of differenciation,
2) accelerated growth of a part, combined with accelerated or normal growth of another part,
3) decelerated growth of a part, combined with accelerated or normal growth of another part,
4) gross fetal deformities originating in the first 6 weeks of intrauterine life,
5) lesser fetal deformities originating in the first trimester of the fetal age.

In view of the above, a knowledge of fundamental embryology is of great importance for better understanding of anomalies, deformities and assymetries of the nose.

In the embryology of the human nose, three phylogenetic stages, which become factors in normal and abnormal development, are recapitulated in order: (a) the piscine stage, (b) the amphibian stage and (c) the mammalian stage. The result of these stages is the development of the nose from a prevalently olfactory organ to a prevalently respiratory organ. ([2], [3]).

The differenciation of the central face begins at the third week of intrauterine life, from five primordia situated around the primitive oral cavity, the "stomodeum": ([4], [5].)

a) Superior to it is the fronto-nasal process,
b) lateral to it the right and the left maxillary processes, and
c) inferior to it the paired mandibular processes.

The origin of the nose begins at the third week of embryonal life — bilaterally above the stomodeum appear thickened epithelial areas, a pair of convex ectodermal masses, called the "olfactory placodes".

During the fourth week, the areas invaginate and become depressed by proliferations of surrounding mesoderm, forming the shallow "olfactory pits" which open ventro-caudally and are separated by the broad fronto-nasal process. This fronto-nasal process (of the five primordia mentioned above) subdivides into secondary processes, differenciating at the end of this stage into: a) a single, median, large, nasal field; b) right and left medial nasal processes (processi globulares) and c) lateral nasal right and left processes, forming respectively the early medial and lateral boundaries of the primitive nasal pits.

Growth and fusion of the maxillary processes with the medial and lateral nasal processes form the inferior boundary of the nasal pits, separating it from the early oral cavity. Blind sacks or "primary nasal fossae" result, communicating with the exterior by "anterior nares" and occluded by epithelial plugs, which disappear by resorption at the end of the fifth week. By dorsal growth of these primary nasal fossae and the rupture through the bucco-nasal membrane, communication with the oral cavity is established, and thereby the "posterior nares" or "primitive choanae" are formed during the seventh week of foetal life.

Laterally, the obliteration of the "naso-optic furrow" takes place by fusion of the lateral processes with the maxillary processes, while medially, the lateral portions of the upper lip are formed by conjunction of the medial nasal processes with the maxillary processes. The central portion of the upper lip and the philtrum are formed by fusion of the medial nasal processes with each other and with the lower part of the fronto-nasal process.

Limitation from below is first outlined at the end of the sixth week by the appearance of the "primary palate". Lateral ridges appear on the medial sides of the maxillary processes, by approximation of which the anterior portion of the palate is formed, fusing anteriorly with the premaxillary process, and posteriorly to this with each other. General broadening of the head shifts the widely separated primitive nasal fossae to the midline of the face, narrowing and modifying the broad mass of the single median fronto-nasal process and ultimately forming the primary nasal septum, which by junction with the palatal ridges divides the nasal cavity into the left and right nasal fossae. On the lateral walls of the nasal fossae the nasal turbinates develop, and from the corresponding nasal meatuses the paranasal sinuses expand and pneumatize into their relative bones. By the end of the eighth week the external nose is fairly well defined.

The nasal capsule and the septum are at the beginning all cartilaginous. Later certain parts persist and are carried over as cartilage into the skeleton of the nose; other parts become ossified as individual bones, and certain parts remain rudimentary. The nasal bones develop in membrane overlying the cartilage, and replace the later-resorbed cartilage, while the ossification

of the septum starts from an important growth and ossification center which exists at the junction of the anterior end of the vomer with the maxilla, premaxilla and the cartilaginous septum.

At the end of the third lunar month of fetal age, the ectoderm gives rise to the epidermis.

To recapitulate: the upper part of the fronto-nasal process gives rise to the dorsum of the nose, while from its lower part, in conjuncton with the medial nasal processes, the following parts originate: — the middle third of the upper lip, the philtrum, the medial crura of the lobular cartilages, the premaxilla, the primary and secondary septum and the perpendicular plate of the ethmoid. From the lateral nasal processes arise the lateral crura of the lobular cartilages, the ethmoid labyrinth and the nasal bones, and the upper lateral cartilages.

Since it is beyond the scope and possibility of this paper to describe all the anomalies of the nose, we shall confine ourselves to their brief enumeration: —

In the nasal integument, the inclusion of dermal tissue along the lines of the embryonic fissures or clefts leads to congenital nasal dermoid cysts and nasal fistulas. ([6], [7], [8].)

n the bony framework we can include the various deviations and assymmetries of the nasal bones, the agenesia, hypoplasia and hyperplasia of the bones; the anomalies of the floor of the nose, and of the pyriform aperture.

The cartilaginous vault and the lobular cartilages show an enormous variety of aberrations from the normal, from the arrest: absence or atrophy of the upper lateral cartilages, of the lateral crura, or of the medial crura of the lower laterals, to the excessive development: broad, hypertrophic or long columella, bifid or cleft tip, and a variety of assymmetries of the tip and nostrils.

In the septum, 3-4-5 septa may be found due to rudimentary persistence of the vomeral organ of Jacobson, and of the paraseptal cartilages, which too are embryonic remnants ([1]). Congenital perforations of the septum were noted, although very rare ([5]). Total absence may occur due to lack of differenciation of mesenchymal connective tissue between the nasal sacks ([1], [5], [9]).

Developmental failures during the differenciation of anterior or posterior nares, and of the primary nasal fossae lead to partial or total atresia, synchiae, and to anomalies such as the one nostril, the triple and the quadruple nares ([6], [10], [11]).

Considering the total nose, various malformations may occur: the double nose — "birhinia"; the unilateral — "arhinia" and the "total arhinia" — all due to faulty organisation on the central embryological structures. The birhinia is not to be confused with the "bifid nose" or "Doggenase" also known as "split" or "cleft" nose, an abnormality due to persistent nasal fissure and lack of development of the nasal septum ([6], [12], [13]).

The lateral fissure of the nose is a very rare malformation. Related to it are the great variety of nasal deformities associated with cleft palate or cleft lip.

A severe degree of disturbed differenciation of the fronto-nasal process and migration to the center of the face is the group of tubular, snoutlike,

nasal appendages called "proboscis" which may occur as a single median proboscis above a regular nose or in the form of a single unilateral proboscis combined with half a nose. ([6], [13], [14], [15]). A very rare condition of a total arhinia with a "bilateral nasal proboscis" has also been described ([16]).

Next there is the cyclopia ([17]) the one-eyed Giant of Greek mythology and Homeric fame, with or without a rudimentary nose, the so-called "median proboscis".

Fortunately, "errors of Nature" are not in the majority and the inheritance of the urge to grow normally is the rule, leading to a normal, harmonious growth and development of the face and nose.

INTRODUCTION EMBRYOLOGIQUE AUX MALFORMATIONS CONGENITALES DU NEZ

Aperçu embryologique.

Le dos du nez nait de la partie supérieure de l'apophyse fronto-nasale. La partie inférieure de cette apophyse, ainsi que les apophyses nasales moyennes, donnent naissance au tiers moyen de la lèvre supérieure, le phyltum, à la partie moyenne du cartilage lobulaire, à l'os prémaxillaire, à la cloison et à la lame perpendiculaire de l'ethmoïde.

Les apophyses latérales sont à l'origine des parties latérales du cartilage lobulaire, du labyrinthe ethmoïdal, des os du nez et des cartilages supérieurs latéraux. La capsule nasale et la cloison sont initialement cartilagineuses et elles s'ossifient ultérieurement.

Malformations.

L'arrêt, l'accélération et le ralentissement du développement de ces structures embryologiques causent des malformations, des asymmétries et des anomalies qui peuvent être résumées comme suit:

Le revêtement cutané : kyste dermoïde et fistule congénitale.

Cadre osseux : agénésie, hypoplasie, hyperplasie, déviations, malformations du plancher et de l'ouverture pyriforme.

Voûte cartilagineuse : celles-ci comprennent les grandes variétés de malformations des cartilages latéraux supérieurs et lobulaires, de la columelle et de la pointe du nez.

La cloison nasale : absence, cloisons multiples et perforation congénitale.

Narines : atrésie totale ou partielle; narine unique, triple, quadruple.

Le nez "en total" : arhynie totale ou unilatérale; nez double, birhynie; nez bifide et fissure latérale du nez; proboscis — unilatéral, bilatéral, médian — solitaire ou avec cyclopie.

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Zvi Rosen, M.D.

Department of Otorhinolaryngology,
Hebrew University,
Hadassah Medical School,
Jerusalem, Israel.