

## Single Nose/ Mono nostril/ Heminose

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### Introduction

Congenital anomalies of the nose are thought to be relatively rare, affecting approximately 1 in every 20,000 to 40,000 live births.<sup>(1)</sup> Nasal hypoplasia ranges from underdevelopment or partial absence of parts to complete arhinia. Heminasal aplasia, hemi-arhinia or unilateral aplasia of the nose is a rare congenital malformation in which there is absence of half of the external nose together with a variable degree of abnormality in the internal anatomy of the nose as well as the adjacent facial structures. It imposes a major psychological burden to the parents and may have physiological impact on the child.<sup>(1)</sup>

Single nostril is a very rare condition. These mid line facial defects which arise from cleavage anomalies and it is a phenotypic spectrum of Schinzel Giedion syndrome (1978).<sup>(2)</sup> It is rare, severe condition that is present from birth and affects many parts of the body. Features of SGS are mid face retraction, choanal stenosis, prominent forehead. Till 2003, 30 cases reported in literature Pathogenesis of the disease remains unknown. No genetic marker is available.<sup>(3)</sup> This condition include severe intellectual disability; a distinctive facial appearance; excessive hair growth (hypertrichosis); and various birth defects that may affect the skeletal system, genitourinary system; kidneys; and heart. Affected children usually do not survive beyond a few years after birth.<sup>(4)</sup>

We present two extremely rare cases where the children has single nostril and does not conform to any syndrome.

### Case Report

Two children where one child a 10 years old female as in Fig. 1 & 2 and the other 12 years old female as in Fig. 3 & 4 not related to each other, presented to OPD with history of recurrent dacryocystitis of both eyes. They were physically examined and apart for the nasal deformity were found to be other- wise healthy not conforming to the clinical features of SGS mentioned before and they were living well beyond the expected expectancy of SGS. So they could not be assigned the syndrome of SGS. This makes their condition even rarer, with only a related case report present by Tsur et al.<sup>(5)</sup>

Nasal examination revealed distorted internal anatomy with rudimentary turbinates and poorly

pneumatized sinuses. The lacrimal sac opening could not be made out on either side. A dacryocystorhinostomy was done in both the cases with primary stenting with silicon tubes.



Fig. 1



Fig. 2



Fig. 3



**Fig. 4**

### Discussion

Coming to the embryology of the nasal development, it begins in the fourth week of gestation and is mostly complete by the eighth week. Of the five facial primordia, the frontonasal prominence is the primary structure responsible for nasal development. Neural crest cells migrate into the frontonasal prominence and form the olfactory (nasal) placodes which deepen into nasal pits.<sup>(1,6)</sup> These nasal pits are surrounded by mesenchymal cells that proliferate, developing into the horseshoe-shaped medial and lateral nasal processes on each side. The medial processes will ultimately fuse, contributing to the nasal septum and the medial crura of the lower lateral cartilages. The lateral processes develop into the nasal bones, upper lateral cartilages, ala, and lateral crura of the lower lateral cartilages. The nasal dorsum and glabella are derived directly from the frontonasal prominence. The other four facial primordia—the paired maxillary and mandibular processes of the first branchial arch—will ultimately fuse with the medial and lateral processes, completing facial formation by the 14th week of gestation.<sup>(7)</sup>

Initially the maxillary and the lateral nasal prominences are separated by a deep furrow, the naso-lacrimal groove. Ectoderm in the floor of this groove forms a solid epithelial cord that detaches from the overlying ectoderm. After canalisation, the cord forms the naso-lacrimal duct, its upper end widens to form the lacrimal sac. Following detachment of the cord, the maxillary and lateral nasal prominences merge with each other. The naso-lacrimal duct then runs from medial corner of eye to the inferior meatus of the nasal cavity, and the maxillary prominences enlarge to form the cheeks and maxillae. The nose is formed from five facial prominences, the frontal prominence gives rise to the bridge, the merged medial nasal prominences provide the crest and the tip and the lateral nasal prominences form the sides (alae). In the mid and upper face region, the signals emanating from the surface ectoderm and the underlying areas of the neuroepithelium dictate the fate of the mesenchyme. SHH (Sonic Hedge Hog) and FGF8 (Fibroblast growth

factor 8) play a major role in patterning this area, but the specific genetic interactions are not known.<sup>(8)</sup>

Failure of the development of both nasal placodes results in complete nasal aplasia or arhinia while failure of one placode leads to heminasal aplasia or hemiarhinia. Heminasal aplasia, in which unilateral nostril agenesis is present, has been reported in isolation as well in combination with anomalies affecting the ipsilateral face. Radiographic studies have demonstrated an associated absence of the cribriform plate, which is thought to represent a loss of the ipsilateral nasal placode during development.<sup>(9)</sup>

Single nostril is an extremely rare condition, with very few cases found worldwide. When present it is usually associated with other symptoms with a very short life expectancy after birth.<sup>(1)</sup> Our cases are even more rare as they have single nostril but with both living beyond their first decade of life not conforming to features of any syndrome. In this case it is difficult to state that there had been failure of development of a nasal placode. As the structures present were rudimentary, so it may be stated that in these cases there was improper development of a nasal placode rather than the complete failure.

Losee et al (2004) developed a comprehensive classification scheme dedicated to congenital nasal anomalies.<sup>(2)</sup> This was based on a retrospective review of 261 patients with congenital nasal anomalies. Congenital nasal deformities were classified into the following 4 categories:

- Type I - Hypoplasia and atrophy (represents paucity, atrophy, or underdevelopments of skin, subcutaneous tissue, muscle, cartilage, and/or bone)
- Type II - Hyperplasia and duplications (represents anomalies of excess tissue, ranging from duplications of parts to complete multiples)
- Type III - Clefts (The comprehensive and widely used Tessier classification of craniofacial clefts is applied)
- Type IV - Neoplasms and vascular anomalies (Both benign and malignant neoplasms are found in this category)

According to this classification both our cases belonged to Type I category.

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