CASE REPORT

'Trifid Nose'- A rare case of nasal duplication

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Introduction:
Developmental anomalies of the nose encompass a wide and diverse group of conditions. Nasal dysplasia can range from a supernumery nostril to a complete duplication of the nose[1]. These anomalies can be associated with malformations such as facial clefts and can be unilateral or bilateral, most reported cases are unilateral. A number of cases of supernumery nostril have been reported in the literature and although anomalies of the face and nose are not rare in themselves, cases of nasal duplication are extremely rare with only a few reported cases since Lindsay in 1906[2]. Herein we report an extremely rare and unusual case of nasal duplication in the form of a trifid nose.

Key Words: *trifid nose, nasal duplication.*

CASE REPORT-

A two-year old girl presented with a congenital anomaly of the right hemi-face. The child, born at full term without complication was the first-born child to non-consanguineous parents. There was no family history on the maternal or paternal side of congenital malformations. On examination, a fully formed separate duplication of the right nostril was found 2cm to the right of midline. The duplicated nostril had a nasal cavity and fully formed septum. The was not communicating with the ipsilateral nostril (figure 1). A small blind-ending sinus was found at the superior pole of the duplicated nostril, lateral to the nasal bridge at the medial canthus. No other anomaly was found.

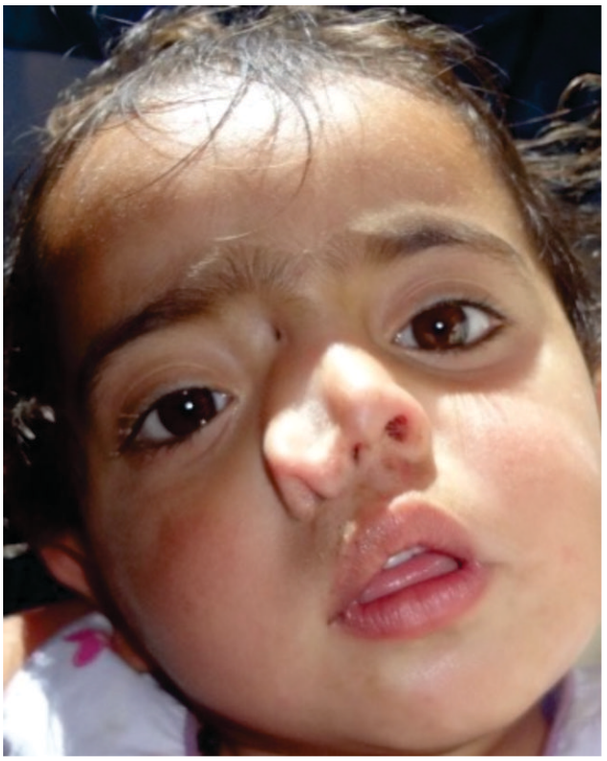

Figure 1- Pre-operative presentation of nasal duplication

The child underwent excision of the duplicated nostril under general anaesthetic (see Figures 2) and the wound was amenable to direct closure. She had an uneventful post-operative course and a successful aesthetic outcome (Figure 3).

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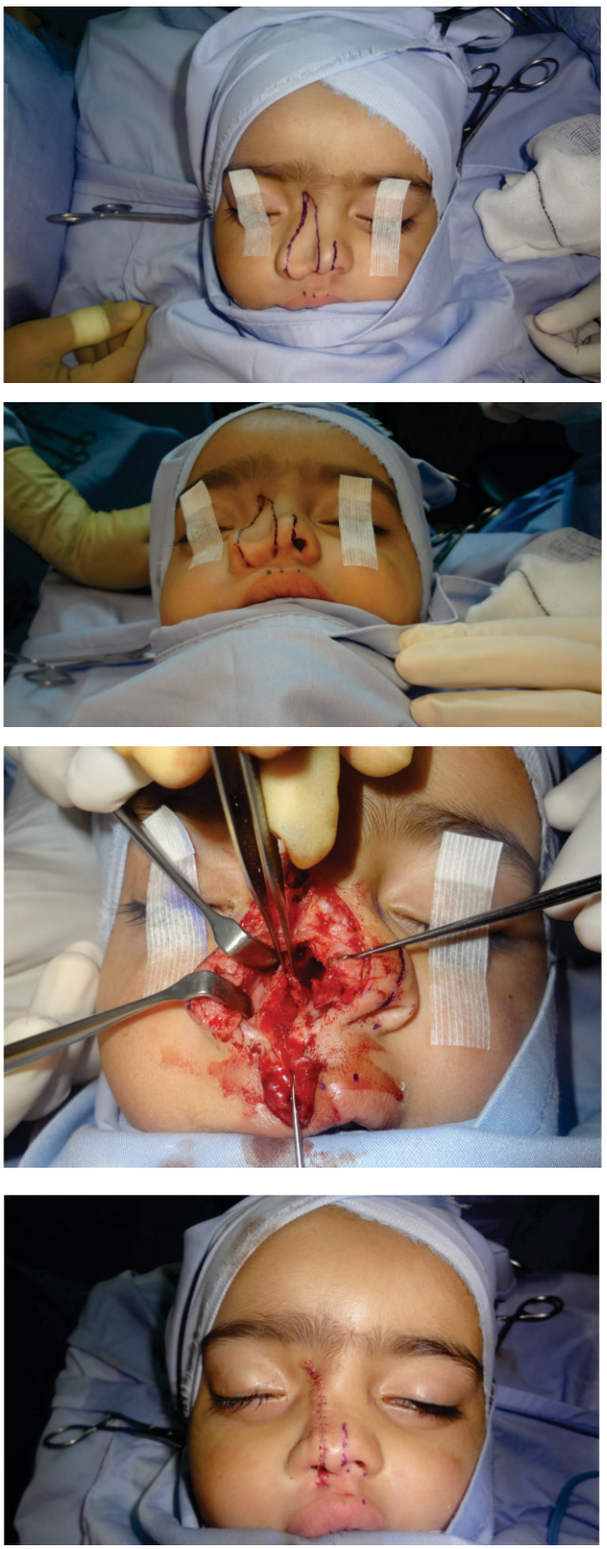

Figure 2
Photographs showing the pre-operative markings, intra-operative excision of duplicated nostril and septal cartilage and the immediate post-operative result.

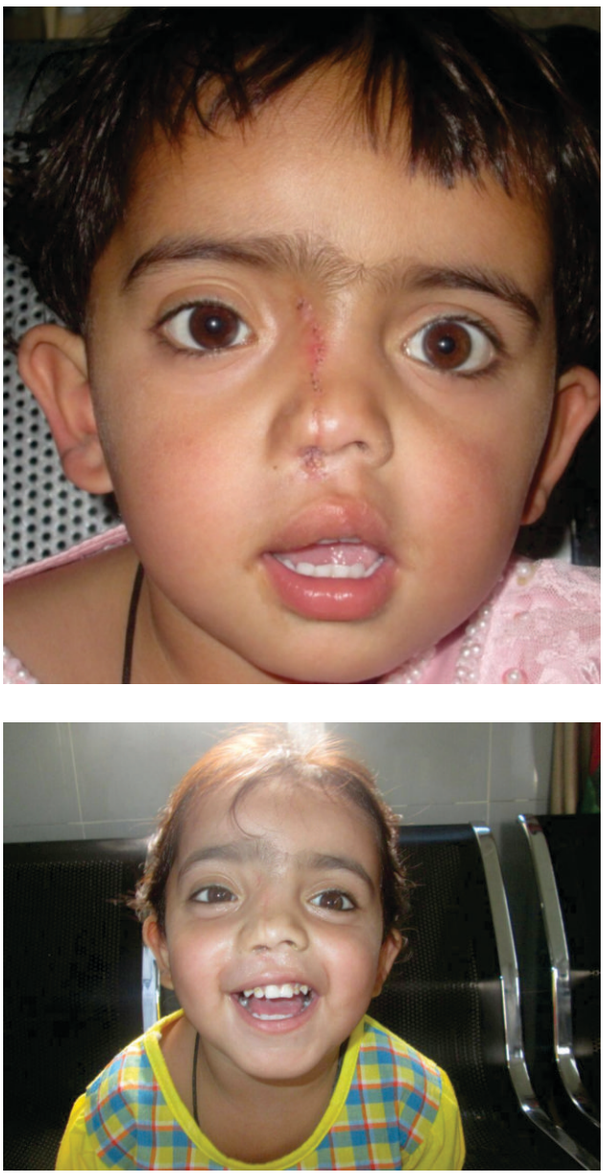

Figure 3: Serial post-operative follow-up pictures showing a pleasing aesthetic result.

DISCUSSION-

The embryological development of the nose and surrounding structures is extremely complex and yet significant developmental nasal anomalies are rare.

The development of the nose starts around the 4th embryological week of development and originates in the bilateral nasal placodes[3].

The nasal placode, which arises from surface ectoderm, develops on the lateral aspects of the frontal prominence. The nasal placodes invaginate around the 5th week of gestation to form the nasal pits that are widely spaced on the anterolateral sides of the developing head of the embryo. As the invagination continues, a tissue ridge surrounding each pit forms the nasal prominences. Prominences on the outer edge of the pits are the lateral nasal prominences; those on the inner aspect are the medial nasal prominences. The depression separating the maxillary swelling from the lateral nasal prominences is known as the nasolacrimal groove, which eventually gives rise to the nasolacrimal apparatus[4]. The middle of the external nose develops from caudal progression of the medial nasal folds, which fuse to form the frontonasal process. Three paired centres of chondrification form the lateral nasal cartilages. Nasal septum bony formation over the cartilaginous capsule occurs during the eighth week.

The maxillary processes fuse with the medial nasal processes, and separates the nasal and oral cavities. The nasal pit invaginates further and breaks through the oral cavity. Development requires enlargement of the nasal cavity, degeneration of existing tissues, and generation of mesenchyme-derived structures. Subsequently, between the 6th and 12th weeks of gestation, the secondary palate is formed as the result of fusion between palatal processes, growing from the oral surface of the maxillary processes. Each merging and fusion site is the potential site of facial or palatal cleft[4].

Losee et al (2004) developed a comprehensive classification scheme dedicated to congenital nasal anomalies which was based on a retrospective review of 261 patients with congenital nasal anomalies and described 4 categories[5].

- 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)

Using the above classification system, our case report would fit in with a Type II hyperplasia and duplication. Duplication of the nose is one of the rarest congenital nasal deformities with only a handful of reported cases in the literature.

The first reported case of a nasal anomaly was over 100 years ago by Lindsay (1906)[2]. It was described to be different from the double nose with two septae and four nostrils and nasal cavities. According to a case report by Erich in 1962[6], during the course of the evolution of the nasal placode, four nasal pits appeared horizontally, each became a nasal sac, and the medial two which were interposed between the two nasal laminae, prevented the laminae from fusing into one nasal septum. This resulted in double nose. Supernumerary nostril is formed when the accessory nasal pit is located so laterally to the nasal lamina that the accessory nostrils are formed above the natural nostril and thus do not disturb the fusion of the nasal laminae. Nakamura in 1987[7] hypothesized that during the proliferation of mesenchymal cells in the lateral nasal process, a concavity or fissure appears in this area accidentally, and thus this lateral nasal process is divided into two segments, resulting in two nostrils and two alae on one side.[7] This hypothesis can

extrapolate the appearance of accessory nostril either above or lateral to the natural nostril or medially, depending on the position of change in the lateral nasal process.

The importance of removing superfluous structures and reconstructing the normal anatomy is stressed to optimize the aesthetic outcome. As demonstrated in our case, timing of surgery is also important when managing nasal anomalies. Excision performed at an early age, in this case pre-school (similar to the approach taken in cleft lip/palate surgery) avoids any serious impact on the nasal cartilages, or adjacent structures, reduces potential anaesthetic complications and also the psychological impact of such an anomaly to the face.

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