

Surgical and prosthetic treatment of congenital absence of the nose: a case report*

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SUMMARY

This paper presents a case of a 5-year old girl with a congenital absence of the nose. Congenital arrhinia is a very rare malformation of the midfacial bones. The difficulties of treating a child with this abnormality are discussed.

Key words: arrhinia, congenital development, nasal surgery, facial prosthetics

INTRODUCTION

The development of the nose and paranasal sinuses during prenatal growth occurs in three main stages. During the first (pre-skeletal) stage – i.e., about the 25th day of pregnancy – the nasal primordium develops from the local ectodermal tissue. At the 33rd day, it is transformed into the nasal sulcus. The primary nasal cavity is formed between the 42nd and 45th day. During the second (chondrocranial) stage growth of cartilaginous structures of the nose starts; this stage ends in the 12th week of pregnancy. During the third stage of osteogenesis, centres of nasal and facial osteogenesis are formed.

The external nose is formed from five mesenchymal processes (or facial processes) that surround the primitive mouth, the bilateral maxillary and mandibular processes and the single frontal process. In between these processes ectodermal buds are formed, that will develop into the olfactory bulbs. During the first period of nasal growth, the nose and oral cavity are separated by the oronasal membrane which gradually disappears, resulting in the formation of the choanae. During the 6th week, the separation of the oral and nasal cavities is completed. Starting in the 6th week of pregnancy, outgrowths are developing on the lateral walls of the nasal cavity, which will be transformed into the nasal turbinates.

Nasal sinus growth is closely related to that of the nasal cavities. Ethmoid and maxillary sinuses are formed during the 2nd–3rd month of pregnancy. At birth, the ethmoid sinuses measure approximately $5 \times 3 \times 4$ mm, whereas the maxillary sinuses measure $7 \times 3 \times 5$ mm each. Other sinuses are absent at this age. So, it is possible that any agent – be it chemical (toxins), physical (radiation) or biological (viruses) – that affects a child's growth during the first six weeks of pregnancy, can cause serious nose deformities or even complete absence of the nose.

Congenital absence of the nose is a severe malformation of the midfacial bones. Global incidence is very low; reports in ENT scientific periodicals are sporadic, and concern single cases. In our ENT clinic, which serves a population of more than 10 million children, we have seen only two children with arrhinia and two others with severe nasal malformations and non-patent noses over a 20-year period. Due to the fact that arrhinia may be caused by several aetiological agents, it is difficult to determine the exact mechanism responsible for its occurrence in a particular patient.

CASE REPORT

On 1st February 1992 a girl F.P. was born before term, in the 33rd week of pregnancy with an almost complete absence of the external nose and atresia of the nares (Fig. 1). She was the mother's fourth child, and was preceded by children aged 11, 15 and 16 years who are all free of congenital defects. The parents are healthy individuals – the mother 33 years of age and the father 38. The only suspect factor is a viral infection (i.e., rhinitis) of the mother during the third month of pregnancy (Gifford et al., 1972). After delivery by natural means, with an APGAR of 7, congenital arrhinia was noticed. Due to the resulting respiratory insufficiency, an oropharyngeal tube was inserted. Feeding was carried out by a gastric tube. At the age of 10 days, the child was transferred to our clinic. A CT scan revealed an absence of cartilaginous and bony elements of the nose and the nasal septum, together with an empty space that was connected with the pharynx. The lachrymal canaliculi were also lacking. In the 8th week of life, the first stage of surgical treatment was performed. The operation was done by bow-lancet cutting, rasping, and drilling. During surgery, we found a nasal cavity without a nasal septum, in agreement with the CT scan. However,

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Figure 1. Photograph demonstrating the absence of the external nose and atresia of the nares at birth.



Figure 2. Result of the third operation after removal of the stent.



Figure 3. Photograph of the silicone prosthesis placed at the age of five years.

contrary to the CT findings, the cavity was a cul-de-sac. Choanae had to be reconstructed by resecting the bone tissue between the nasal cavity and the nasopharynx. Two intubation tubes were used as stents. There were no post-operative complications. The child was evaluated, and discharged with two 4.0 Portex tubes in place (Muhlbauer et al., 1993).

At the age of 2 years and 10 months the child was re-admitted to the clinic. After examination, including CT scans, the second surgical stage was performed. A 2cm-long transverse incision between the eyebrows was made. The bone between the forehead and the nose appeared to be eroded, the nasal bones were flat and asymmetric with displacement to the right. Preserved human cartilage was transplanted under the periosteum of the nasal ridge. The Portex stents were removed. Fibre-endoscopy of the nose and nasopharynx revealed a stenosis in the choanal region.

The third surgical operation was performed at the age of three years. The choanal stricture was addressed through a transpalatine approach, and the nares were widened by cutting the skin around the previous scars and turning them into the nasal cavity. A large stent (8 mm diameter) was left behind.

Five weeks later, the situation was re-evaluated (Figure 2). The choanal openings were sufficient, but the nares were strictured to about 2 mm. They were dilated and the stent was replaced. This was repeated during the subsequent months. Finally, when the child was five years old we have asked the Facial Prosthesis Department at Lodz (Poland) to make a nasal prosthesis, consisting of skin-toned silicone (Figure 3).

The breathing function is satisfactory, but it is still necessary to retain a stent under the prosthesis. According to the needs of facial growth, a further silicone prosthesis will be made every 2-3 years (Weinberg et al., 1993). At the age of 16-18 years, the final phases of surgical treatment will be carried out (Navarro-Vila et al., 1991).

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