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Results of surgical treatment of choanal atresia

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SUMMARY

During a period of 19 years we have treated 65 infants with choanal atresia. Thirty of them showed a bilateral atresia, thirty-five a unilateral (29 left-sided, 6 right-sided). Fifty patients were girls, fifteen boys. In 27 cases the abnormality was combined with other anomalies. Most infants have been operated upon in the first six months of their life by a transpalatinal approach in general anesthesia.

From the results the following conclusions can be made: early treatment is the method of choice; restoration of nasal permeability, although minimal, allows normal nutrition and diminishes a number of respiratory infections, due to choking. The transpalatinal approach permits a direct control of the surgical area. It does not inhibit the growth of the hard palate and the function of the soft palate. Failures are caused by difficulties to keep the new passage open.

Congenital choanal atresia is de developmental anomaly resulting from the action of noxious factors on the embryo in the period from the 3rd to the 6th weeks of pregnancy. In view of its location the anomaly is rarely recognized. This location is also the cause of difficulties in clinical examination of choanae. Even during autopsy this region is usually disregarded and thus there is no reliable information about the prevalence of this change. Choanal atresia belongs to malformations endangering the life of the newborn. The anomaly requires surgical treatment since the babies not operated upon in due time die after labour from dyspnoea or several weeks later from cachexia. Nutritional disturbances and aspiration pneumonia frequently recurrent cause death of the infants with this anomaly. However, cases surviving even to adult life have been reported.

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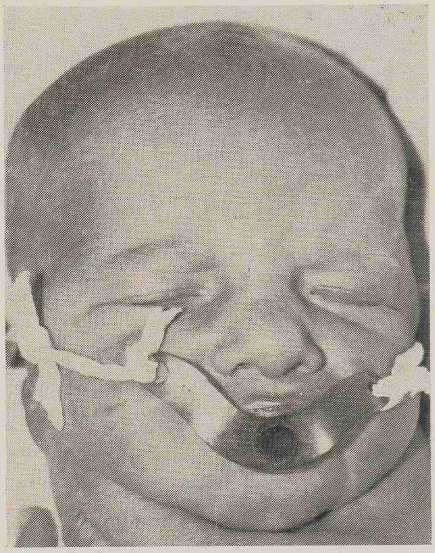

Figure 1. Tracheostomy tube used for securing mouth breathing before operation of choanal atresia.

The first aid before the operation includes facilitation of respiration by stimulation the baby to crying, depression of the mandible and introduction of an oropharyngeal tube into the mouth. This tube may be substituted by a common rubber catheter, a properly modified dummy, or tracheostomy tube (Figure 1). Intubation or tracheotomy are performed in exceptional cases. In the Warsaw centre we have never been forced to perform tracheotomy or intubation in infants with this anomaly. Frequently it is necessary to feed the babies with a tube. But all these methods cannot protect the babies against disturbances of respiration and nutrition. The child fails to thrive and shows evidence of hypoxia. Surgical treatment is indispensable. In a period of 19 years we treated 65 children with unilateral or bilateral choanal atresia. In 30 cases atresia was bilateral and in 35 cases unilateral, that is left-sided in 29 and right-sided in 6 cases. The group comprised 50 girls and 15 boys. The youngest child was treated at the age of 10 days. In 38 cases choanal atresia was the only anomaly, in 27 cases it coexisted with other malformations of the face or remote organs. Most children were operated upon within the first 6 months of life under general anaesthesia, with an approach through the hard palate. Since 19 years we have been using an incision resembling that introduced by Abulker. The mucoperiosteal flap is separated up to the posterior border of the horizontal lamina of the palate, and from the pharyngeal surface of the plate obturating the choanae. Now a window is made in the horizontal lamina of the palate and the mucosa is separated from the plate obturating the choanae. The bony block of the osseous obturating plate is separated now with a chisel

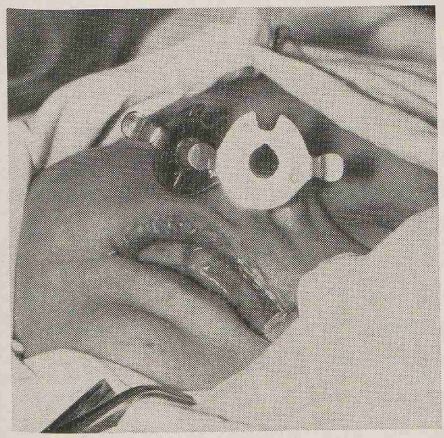

Figure 2. Two tracheostomy tubes introduced through the nasal cavities into the pharynx immediately after removal of choanal obstruction.

and it is taken out together with the posterior part of the nasal septum. In this way a relatively wide nasopharyngeal channel is created. The obturating plate resembles a biconcave lens from 1 to 12 mm thick in its central part which is thinnest. A tracheotomy cannula is inserted into this newly created channel through the nasal cavity down to the throat by the method of Stoner (Figure 2). This cannulation permits the baby to start immediately breathing through the nose. Single nylon sutures are laid on the margins of the surgical wound on the palate. Since metallic tubes cause easily development of granulation tissue on the posterior wall of the pharynx they are replaced after several days with soft plastic tubes.

The results of the operation immediately after it are good: the nutritional state improves, the baby gains weight, ventilation becomes normal. No operative deaths or serious postoperative complications were observed. One child aged 8 months died 8 months after the operation from multiple congenital anomalies (congenital heart disease, absence of one ureter etc.), 2 other children died due to Pneumocystis carinii infection: one 2 months after the operation, the other 3 months after it. Two infants with bilateral atresia not treated surgically died because of that anomaly. We have no information about one child discharged from our hospital. One child with unilateral atresia was not operated upon because of lack of parental consent. In one case otitis media developed after the operation.

For evaluation of therapeutic results we examined 30 children aged from 6 to 10 years who had been operated upon in infancy. This group comprised 14 children with bilateral and 16 with unilateral anomaly. At the same time 30 children matched for age were examined for comparison – they had never had choanal atresia. The examination showed that all children with the history of choanal atresia had narrowing of the choanae, in 12 cases

reoperation was necessary. In the newborn the channel is 2 mm long and its diameter is 3-5 mm. In cases in which the choanae fail to develop the development of the nose has stopped in the 7th week of foetal life. This is the cause why it is so difficult to create such small a channel and to maintain it after the operation. Late control examinations carried out in 35 children after more than 6 months from the operation showed impairment of nasal patency in about 50% of cases due to development of secondary atresia. This required dilation of the created channel or perforation of the secondary membrane. This fact should, however, not restrict the indications to operation. The operation should be done immediately after recognition of the anomaly. Transpalatal reoperation was done also in many cases, and perforation of the secondary membrane through the nose even more frequently.

The radix nasi in the children with past choanal atresia examined at the age of 6-10 years was relatively broad, but the configuration of the nose was generally not different from that in healthy children. Abnormal shape of the nose was observed only in children with coexistent other anomalies of the face. The nasal septum was usually straight in children with isolated atresia but in children with coexistent other more advanced abnormalities the septum was deviated or broken impairing additionally nasal patency. In children with unilateral anomaly the nasal cavity was significantly wider on the side of atresia in comparison with the side of normal choanal even many years after the operation restoring choanal patency.

It is thought generally that the transpalatal operation of choanal atresia can be performed only in children aged over 6 months. We carry out this operation earlier, as soon as the child has been admitted and the diagnosis has been established. We operated upon children aged 1, 2 and 3 months. In all, 30 infants aged less than 6 months were treated surgically in this way. From the viewpoint of surgical technique it is easiest to carry out this operation when the palate is yet very short. Control examinations of children operated upon at the age below 6 months failed to demonstrate any greater deformities of the hard palate in relation to children operated upon at a higher age. The palate of children with bilateral atresia was high and narrow. In children with unilateral anomaly, mainly in those with isolated malformation, there was no palatal deformity, but the hard palate was fairly frequently asymmetrical. Apart from one case of cleft palate we never noticed any abnormalities in the structure or mobility of the soft palate in the children treated surgically for choanal atresia.

The palatine and pharyngeal tonsils were small in all children with atresia in comparison with healthy children of the same age. It is possible that this underdevelopment of tonsils is connected with less intense stimulation in these children breathing rather through the mouth than nose. The incidence

of infections was similar in children with atresia as in those without it, or it was perhaps even smaller. The serum immunoglobulin A, M and G level was within the normal range accepted for that age group.

The development of nasal sinuses corresponded to the age of the children, with the exception of those with high-grade maxillofacial anomalies.

Ten children had hearing disturbances, 5 of them had conduction hypoacusia at the level of about 30 dB due to otitis media. In 5 children less pronounced hearing inpairment was found connected with middle or external ear anomalies associated with faciomandibular or faciocranial dysostosis. Karyotype investigations showed normal chromosomes.

In this group of 65 children with atresia only two sisters were found and in these cases familial occurrence of the anomaly could be possible.

Follow-up examinations demonstrated that many children with initially doubtful prognosis as to survival and mental development developed later normally. In only 4 out of 30 children examined at the age of 6-10 years mental retardation was diagnosed. One of them had Down's syndrome and three had multiple anomalies impairing mental development. The remaining 16 children had mental and somatic development compatible with their age.

The reported investigations suggest the following conclusions. Surgical treatment should be carried out possibly early. Restoration of even minimal nasal patency ensures normal nutrition of the child, normal development, and prevention of respiratory infections and aspiration pneumonia. The transpalatal operation gives a good insight into the region of the anomaly and ensures a safe course of the operation. The transpalatal approach causes no disturbances in the growth of the hard palate or in the mobility of the soft palate. All 65 children treated surgically from transpalatal approach had uneventful postoperative course. Therapeutic failures included difficulties in maintenance of choanal patency after its restoration.

RÉSUMÉ

Au cours de 19 ans, nous avons traité 65 enfants atteints d'imperforation choanale. Parmi 65 enfants, 30 presentaient l'obturation bilatérale et 35 l'obturation unilatérale dont 29 gauches et 6 droites. Il y avait 50 filles et 15 garçons. Dans 38 cas l'anomalie était isolée et dans 27 cas elle coexistait avec d'autres vices de la région facio-craniale, ou avec çeux des organes distants. La plupart des enfants ont été operés pendant les six premiers mois de la vie, sous anesthesie générale en accèdant par le palais dur. Les résultats de notre étude permettent de tirer des conclusions suivantes: Le traitement opératoire à la periode la plus précoce de la vie est un traitement de choix. Le retablissement de la perméabilité nasale, même à un degré minime, permet à l'enfant la nutrition normale êtant une condition de déve-

loppement normal et previent l'inflammation des voies respiratoires inférieures et du tissus pulmonaires, dûe à l'avalement de travers. L'operation par le palais permettant le contrôle direct du champ opératoire présente la securité. Elle ne freine pas la croissance du palais dur et la fonction du palais mou. Des echec ont été causé par les difficultés du maintien de la perméabilité du canal formé des choanes.

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