NOTES ON A CASE OF CONGENITAL ATRESIA OF THE CHOANA.

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Cases of congenital occlusion of the posterior nares are of interest to rhinologists, not only on account of their rarity, but also because they give a demonstration of the results of complete and permanent nasal obstruction, uncomplicated by any morbid process.

The case which I have had the opportunity of examining is under the care of Dr. Max Scheier of Berlin, and was shown by him to the Laryngological Society of that city; but as it is, in one point, unique, he has kindly permitted me to publish a more complete account of it, and for this I thank him most heartily.

Case.—The patient, an intelligent young woman, at 21, complains that she has been unable to breathe through the right nostril for some years. The history shows that her mother died of phthisis; her father is healthy. She is one of five children, three of whom are alive and in good health. Besides herself, none of the family, so far as is known, exhibited any abnormality. As a child, the patient was healthy; no difficulty was experienced in rearing her. Her father remembers that she had constant discharge from the right nostril; he further definitely states that she did not breathe through her mouth except when she had a cold. She does not snore at night. She herself has noticed her inability to breathe through the right nostril since she was 16. She has also observed that her right side, especially the right breast, is less fully developed than the left.

Condition.—The patient is well developed and nourished; her complexion is rather pale. A slight but definite asymmetry of the body is apparent. The right eye is rather more prominent, and is at a slightly lower level than the left. The right cheek is rather flatter, and

1 From the Poliklinik of Dr. Max Scheier of Berlin.
the right naso-lobial fold is less marked. More noticeable still is the asymmetry of the trunk and lower limbs. The right shoulder is lower than the left, and less clothed with soft parts. The right mamma is smaller, and hangs lower than the left. The upper arm is less developed on the right side, though there is no difference between the forearms and hands. No abnormal curvature of the spine can be detected. The right leg is shorter than the left, and the right foot is rather smaller.

Measurements.

|                | Right. | Left. |
|----------------|--------|-------|
| Upper arm      | 10½ in. | 11 in. |
| Knee, maximum circumference | 13¼, | 14, |
| Lower limb (anterior superior iliac spine to tip external malleolus) | 36, | 37½, |

The patient is right-handed; voluntary movements are carried out against resistance equally well on either side; but she states that the right arm and leg are more easily tired than the left. The sense of smell on the right side is absent; on the left is unimpaired. The senses of taste and hearing are unimpaired, and the tympanic membranes are normal. A complete examination of the nervous system discloses no further abnormality.

Examination of the nose.—During even forced respiration there is no movement of the right ala nasi, nor is there the slightest passage of air through the right nostril.

Anterior rhinoscopy.—On the right side there is a slight deviation of the septum; the anterior end of the inferior turbinated body is large, and blocks the way to further examination. On the left side, the turbinated bodies are of normal dimensions.

Posterior rhinoscopy.—On the right side a pale grey wall is seen blocking the choana and flush with its margin, in it there is an oval depression, rather above the centre, and there are two minute indentations immediately above that. No cicatrices can be seen, and it is apparently covered by a continuation of the pharyngeal mucous membrane. On the left side the turbinated bodies can be seen. On the roof, immediately in front of the posterior nasal opening, a narrow grey strip of tissue is visible, apparently an attempt at the formation of an occluding membrane on this side also. This felt hard when touched with a probe. The distance between the septum and Eustachian orifice is greater on the left side than on the right.

Cocaine was applied to the swollen turbinated body on the right side; it was then possible, after syringing out a large quantity of tenacious mucus, to see the obstructing wall from the front. This, on being per-cussed with a probe, was proved to consist of bone. An attempt was made to estimate its thickness by trans-illumination, but without success. The palate was symmetrical. The measurements, which were carried out exactly after the method of Buser (1), a pupil of Siebenmann, and the author of the most recent paper on the form of the palate, were as follows:—

Height (at level of first molar, in this case the highest point), 24 mm.

Breadth (at same level), 5 cms. Palatal index $= \frac{B}{H} \times 100 = \frac{500}{24} = 48$. 

i.e., we have here a typical low palate, the normal according to E. Fränkel (2) being up to 55.

The measurements of the face were—

Height (naso-frontal suture to cutting edge of central incisor), 7 cms.

Breadth (inter-molar), 13 cms.  

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\frac{\text{Facial index} = \frac{H}{B} \times 100}{13} = \frac{700}{13} = 53.
\]

The face therefore falls into the category of long faces, as the index is over 50. The larynx is normal, the voice is not nasal.

The heart, lungs, and other organs are healthy.

The diagnosis of congenital bony atresia of the choana was here obvious, from the history, from the appearance of the obstruction, and from the absence of any indication of present or past inflammation.

The treatment, which was carried out by Dr. Scheier, consisted in making, under local anaesthesia, a small orifice with a burr in the bony wall. This at first showed a tendency to close, but the margins are now healed. The discomfort from which the patient suffered has thus been entirely relieved. The sense of smell is now slightly present on the right side.

Remarks.—The chief interest in this case lies in the association of asymmetry of the face and body with unilateral nasal atresia. Out of the total number of previously published cases of congenital occlusion of the choana, amounting now to nearly 100, the association of facial asymmetry is noted only in some twelve cases. In Schwendt's (3) collection of twenty-five cases, published in 1889, no instances are mentioned. In Haag's (4) series we find four such—three published by Keimer, Pluder, and Baumgarten, and one by himself. Cohn (5) notes further cases published by Joel, Morf, Onodi, Cresswell Baber, and also one of his own, and in addition to these I find cases published by Wolff (6), Hanszel (7), and Lewy (8). In only one of these cases—that of Haag—was the asymmetry associated with bilateral atresia. In three—Cresswell Baber, Hanszel, and Lewy—the hypoplasia was on the side opposite to the occlusion, and in every case it was limited to the face. The appearances must have been remarkably similar in all these cases; the points usually noted being—slight flattening of the cheek, partial obliteration of the naso-labial fold, and a slight difference in the level of the eyes, the one on the affected side being the lower. Morf (9) also notes in his case, that sweating was readily induced on the affected side. We see, then, that though the description of the facial asymmetry in these cases very closely corresponds to the appearances found in the present one, yet the latter is unique in that the hypoplasia extends to the trunk and limbs.

Ziem (10) in 1879 had indeed observed patients with unilateral nasal obstruction in whom there was impaired development of the face and spinal curvature. Conceiving the possibility of this depending on the nasal obstruction, he carried out a series of experiments on young dogs and rabbits. He rawed and sutured
the margins of the anterior nares in some, and in others he blocked the interior of the nose with a pledget of wool; in each case one side only was so treated. Four to eight weeks later the animals were killed and examined. It was then found that the side of the face corresponding to the obstruction was the smaller, and there was present in addition a definite scoliosis, which Ziem thought was due to the unequally developed head throwing its weight more on one side of the spinal column than on the other. It is certainly conceivable that diminished pressure of air in the nose might prevent proper development of the antrum of Highmore, and so bring about some flattening of the cheek on that side, and also cause the eye to stand at a lower level. We have, however, already seen that clinical evidence negatives this view, for in three cases the hypoplasia was not on the side of the obstruction, and in one case it was associated with bilateral atresia. With regard to the present case, the thought suggested itself that possibly a nervous lesion might explain the hypoplasia of the trunk and limbs, but an examination of that system brought no confirmation of this view. We are therefore driven to the explanation accepted by previous authors, that we have to deal with a developmental abnormality, probably depending on the same causes as the atresia itself.

Cases of congenital occlusion of the posterior nares have been looked on as affording a classical example of the effect of mouth breathing on the form of the palate. This is undoubtedly true in the cases of bilateral atresia, but is not necessarily so, where the obstruction is confined to one side.

There are two main theories regarding the causation of high palate—one that it is due to mouth breathing, adopted by Koerner (11), Waldow (12), Bloch (13), and others; the other that it is merely a characteristic of the long narrow type of face known as the leptoprosopic. This view is advanced by Siebenmann (14) and his pupils. Those cases are naturally not considered where the high palate is found in persons mentally impaired, and in whom it has long been recognised as merely being one of the stigmata of degeneration.

I shall discuss this question only in so far as cases of congenital atresia of the choana throw light on it. Haag (4) found that the palate was said to be low in six cases out of twenty-one in which its condition was noted: he therefore concludes that mouth breathing is not a probable cause of high palate. His six cases, however, include a case of Ronaldson's (15), where the patient died a few hours after birth; the others were all cases of unilateral atresia, and in only two is it stated by the authors themselves—Simon (16), Gougenheim (17)—that they were mouth breathers. Of the cases published since this paper, in only three out of twelve, collected by Cohn (5), is the palate low, one of these—a case of unilateral atresia—cannot be included as it is not stated whether
patient is a mouth breather or not. This gives a total of twenty-eight out of thirty-two, or 87.5 per cent. of high palates. That we are right in excluding those cases where the atresia is unilateral, unless it is definitely stated that there is mouth breathing, is sufficiently shown by the present case, where there is no doubt whatever that the patient usually breathed through her nose. I think these figures allow of the statement that the evidence of cases of congenital atresia of the choana is in favour of the Koerner-Waldow theory.

I should like to draw special attention to the normal hearing and unaltered appearance of the drum membrane noted in my case. This is not an uncommon experience, and is of importance, in view of the tendency in some quarters to find a cause in the nose for every middle ear affection.

Finally, may I be allowed to lay emphasis on the development of a sense of smell on the right side after operation? This seems to be hardly in accordance with nature's law that disease causes atrophy.

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SPONDYLITIS, OR THE "VERTEBRAL TYPE OF ARTHRITIS DEFORMANS."

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From the point of view of nomenclature, there is, I venture to think, one valid argument against our retaining the term spondylitis deformans.

The objection to my mind is this, that the use of the term seems to carry with it the inference that we are dealing with a condition in some way or other distinct from arthritis deformans. As far as my experience, gained at the Mineral Water Hospital, goes, no sharp differentiation is possible between the so-called