Personal practice

Congenital choanal atresia*

LEO KIRKEGAARD WINTER

From the University Department of Otorhinolaryngology, Kommunehospital, Aarhus, Denmark

We have treated a total of 15 patients (11 girls, 4 boys) for choanal atresia in the University Department of Otorhinolaryngology during a period of 15 years from 1963-1976. No other cases occurred in their families. One patient is mentally retarded, whereas none of the others have had other congenital defects.

Five patients (4 girls, 1 boy) had *bilateral choanal atresia*. In all 5 cases, the birthweight was low, in 2 patients below 2500 g. Immediately after birth these infants presented severe problems, becoming cyanosed bordering on asphyxiation.

Ten patients (7 girls, 3 boys) had *unilateral choanal atresia*, 7 of them on the right side. The more common symptoms were unilateral persistent purulent nasal secretion, obstruction of one side of the nose, and disturbed sleep, especially with upper respiratory infections.

Methods of treatment and results

The ages at operation of the 5 patients with *bilateral choanal atresia* have varied from 17 hours to 2 months. All operations were performed under general anaesthesia with trans/endonasal perforation of the atretic membrane. A plastic (Portex) tube of an inner diameter of about 3 mm in which an oval hole had been cut in the middle (Fearon and Dickson, 1968; Hogeman and Toremalm, 1968) was then passed through one nostril and led out of the other. The tube was fixed in front of the columella (Fig. 1). The infant then became able to breathe satisfactorily through the nose.

In our 2 last patients, operated on in 1975 and 1976 at the age of 17 hours and 5 days respectively, a rhinopharyngoscope† was introduced through the

mouth into the pharynx, allowing the nasopharynx and the choanal atresia to be inspected (Fig. 2). By this means, a metal bougie can be guided safely under direct vision, and choanal atresia perforated (Winther, 1978).

The nasal tube can easily be cleaned by suction. After 2, or preferably 3, months it is removed. The patients are seen regularly until the age of 1-2 years, at which time the nasal airway has become established. Dilatation with metal bougies under local anaesthesia is done periodically at the follow-up, if necessary, but no failure has so far occurred.

Fig. 1 Infant with bilateral choanal atresia; a tube has been inserted through both sides of the nose.

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†Hopkins rod-lens system; by Storz Endoscopy Company, Tuttlingen, West Germany.

Fig. 2 Congenital bilateral choanal atresia seen through the endoscope, in a newborn infant.

Surgical treatment of the 10 patients, with *unilateral choanal atresia* was also performed under general anaesthesia. Three different methods of approach were used, transpalatal (8 patients, including 1 with recurrence), transseptal (1 patient in whom the atresia recurred), and endonasal (1 patient). At operation a Portex tube was inserted in the affected side of the nose for varying periods of time. Operation has been carried out at ages from $3\frac{1}{2}$ to 20 years. In 2 patients the conditions recurred; they were subjected to reoperation by the same technique without further recurrence. We have been able to trace 8 of these 10 patients, who are all free of recurrence and have satisfactory air passage through both nostrils.

Discussion

The developmental anomaly in choanal atresia is due to failure of breakdown of the buccopharyngeal membrane in the 4th or 5th week of embryonic life. Durward *et al.* (1945) reviewed about 500 published cases. The incidence is reported to be 0.02–0.04% (Parkes and Brennan, 1973). About two-thirds of the cases on record are unilateral. The anomaly is twice as frequent in girls as in boys, and involves the right side twice as often as the left, which is in accordance with our experience.

Some investigators (Flake and Ferguson, 1964; Ransome, 1964) have reported a hereditary tendency, but most cases of choanal atresia seem to be of sporadic occurrence. Concurrent congenital anomalies have been encountered, in some series in as many as 50% (Ewans and Maclachlan, 1971), but no hereditary tendency or associated congenital anomalies were noted in any of our patients.

Bilateral choanal atresia presents an acute, alarming picture with respiratory distress and cyanosis, often leading to asphyxia and suffocation. It is a well-observed fact that newborn infants up to the 3rd to 6th week of life cannot spontaneously breathe through the mouth. The cause of this has not been fully clarified, but the anatomy at this age probably plays an important role (Hough, 1955). Both bilateral choanal atresia and conditions which lead to nasal mucosal oedema can thus bring an infant into a critical state (Simson and Brantley, 1977). Probing both sides of the nose (a Thiemann no. 8 catheter is suitable) establishes the diagnosis, and should lead to immediate steps to provide a free airway, and allow feeding.

Even if careful nursing is established, newborn infants with choanal atresia, especially if *bilateral*, are at constant risk of suffocating, and they are not out of danger until they have 'learnt' mouth breathing after the course of some weeks. Conservative treatment consists of the application of a tongue depressor and simultaneous insertion of a feeding tube, but this requires constant and close observation during the first few weeks of life. Definitive surgical correction, mostly by the transpalatal approach, has by some been then postponed until the age of 1–2 years (Owens, 1965). As regards the type of operation, a transseptal approach is advocated by some, and Carpenter and Neel (1977) have recently reported good results by this approach. Their patients however were all over 8 years, and the authors expressed the view that this method cannot be applied in young children, which seems reasonable in view of the small intranasal dimensions in this age group.

The longer the child lives without surgical correction the more difficult will perforation of the atretic wall become. It is generally reported that 90% of choanal atresia are osseous, and only 10% membranous. In our 5 bilateral cases it was not possible to assess with certainty the thickness of the atretic wall. Seemingly, it was only a few millimetres; it could without difficulty be perforated and also be satisfactorily dilated. At this early age it is perhaps irrelevant to speak of osseous or membranous atresia, since the cranial bones are relatively soft (Weseman, 1973).

Several authors have advised against endonasal perforation of the atretic wall because of the high frequency of recurrence (Hobolth *et al.*, 1967; Owens, 1965) and the difficulty in keeping the tubes clean. In addition, surgical complications have been encountered with intracranial and spinal canal injury.

The technique described above is, as previously pointed out by Hough (1955), Beinfeld (1959), Fearon and Dickson (1968), and Weseman (1973),

a reasonably simple means of correcting a life-threatening condition, which can be performed as soon as the diagnosis has been established.

In contrast to the severe, life-threatening symptoms of the bilateral variety, *unilateral choanal atresia* may escape recognition for several years. It is often detected quite accidentally because of nasal stenosis and the presence of mucus and, sometimes, foul-smelling secretions from the affected nostril. However, if the unaffected side is obstructed for example by secretions or by mucosal oedema, respiratory distress will develop in infants with unilateral choanal atresia and the clinical picture becomes similar to that of bilateral choanal atresia.

The view has sometimes been expressed that unilateral choanal atresia might be left untreated, a view with which we must disagree. The last patient among our 10 unilateral cases, a 6-year-old boy with right-sided atresia (Fig. 3), was successfully operated on in 1976 by the same method as the bilateral cases.

Fig. 3 Congenital unilateral choanal atresia (the right side) seen through the endoscope, in a 6-year-old boy.

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Correspondence to Dr L. K. Winther, University Department of Otorhinolaryngology, Kommunehospitalet, DK-8000 Aarhus C, Denmark.