Endoscopic fenestration of choanal atresia

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Abstract

We report two cases of unilateral choanal atresia and one of choanal stenosis treated successfully by endoscopic fenestration, with no recurrence at a mean follow-up of 18 months. Rigid endoscopy and axial CT scanning confirm the clinical diagnosis and this technique avoids the need for stenting or prolonged post-operative stay.

Key words: Choanal atresia; Endoscopy; Surgery

Introduction

Historically the first description of choanal atresia is attributed by Otto (1830) to Roederer in 1755, with the first successful surgery performed by Emmert in 1851 and reported by him in 1854 (Evans and MacLachlan, 1971). He perforated a bilateral bony congenital choanal atresia transnasally with a curved trocar on a seven-year-old boy after preliminary practice on the hard palate of the corpse of a child! (Pirsig, 1986).

Congenital choanal atresia (CCA) describes narrowing of the anterior or posterior nasal apertures but the term is generally used with respect to posterior occlusion, which may be membranous (10 per cent) or bony (90 per cent). The incidence of presentation of (posterior) CCA to Great Ormond Street Hospital over a 20-year review is 1 in 5000; 45 per cent were bilateral and of the unilateral cases 71 per cent were right-sided. The sex incidence was equal although a female predominance of 2:1 is often stated (Maran and Lund, 1990). The aetiology is considered to be a persistence of the embryological bucconasal membrane which separates the nasal cavity from the stomatodaeum until it breaks down at the seventh week, allowing communication through the primitive posterior nares (Evans and MacLachlan, 1971). Cinnamond (1987) also described a further possible aetiology due to persistence of epithelial cells in the nasal cavities which proliferate between the sixth and eighth weeks.

Congenital choanal atresia may occur in association with other anomalies and Pagon *et al.* (1981) proposed the mnemonic CHARGE to describe the association in varying combinations of coloboma, heart disease, atresia of choanae, retardation of growth and/or CNS anomalies, genital hypoplasia and ear abnormalities or deafness. Kaplan (1985) found that 59 per cent of 41 patients with CCA had at least one other anomaly whilst 72 per cent of 50 cases reported by Morgan and Bailey (1990) had coexisting abnormalities.

Bilateral CCA typically presents as a neonatal emergency because newborn babies are obligate nasal breathers. Indeed the reflexes to facilitate breathing through the open mouth in response to nasal obstruction only develop weeks to months after birth although an infant will mouth breathe if the mouth is opened either during crying or if an artificial oral airway is inserted (Cinnamond, 1987). Ronaldson (1881) first described the classical respiratory cycle of CCA of increasing distress until the child opens its mouth to cry whereupon the asphyxia wanes.

Traditionally the failure to pass a soft transnasal catheter in a suspected case would be considered diagnostic of CCA although the turbinate or adenoids may impede passage and Cinnamond (1987) advocates auscultation over the nostrils to assess airflow. Other simple clinical tests include absence of misting on a metal spatula or of movement of a wisp of cotton wool in front of the nostrils (Morgan and Bailey, 1990). Lateral radiography after instillation of radiopaque dye in the supine position can establish the diagnosis but CT scanning indicates whether the atresia is membranous or bony and demonstrates the thickness in addition to excluding other differential diagnoses such as encephaloceles or dermoids (Maran and Lund, 1990).

A securely taped neonatal oral airway is a satisfactory emergency management, combined with nasogastric feeding, particularly if surgery is scheduled in the immediate future (Cinnamond, 1987). McGovern (1953) reported use of a rubber oral nipple to facilitate both breathing and feeding. If such measures are unsuccessful or intended surgery is delayed then endotracheal intubation or even tracheostomy is required, although the latter is rarely necessary except in the presence of associated craniofacial anomalies (Pirsig, 1986). Elective repair of bilateral CCA should generally be performed in infancy or childhood, typically as soon as general anaesthesia can be tolerated, either as a preliminary or definitive treatment (Pirsig, 1986).

Bilateral CCA may however present late – Rizzo *et al.* (1989) and Candan *et al.* (1991) reported diagnosis at the

age of 17 and 16 years respectively, whilst Evans and MacLachlan (1971) referenced Hart (1926) who described presentation at the age of 21 years.

In contrast unilateral CCA may present in late childhood or early adulthood with unilateral nasal obstruction or rhinnorhoea; Booth and Drake-Lee (1991) reported diagnosis at the age of 36 years. The history of nasal blockage and rhinorrhoea is often lifelong and indeed two of our cases had undergone turbinate surgery elsewhere. Untreated cases may develop speech difficulties and even problems during kissing! (Evans and MacLachlan, 1971). Gross-Isseroff et al. (1989) reported permanent olfactory deficits in three patients who underwent 'late' repair of bilateral CCA but normal olfactory acuity in one after 'late' unilateral repair and postulated that early stimulation may be required for the normal development of olfaction.

Whilst clinical tests are helpful, axial CT scanning after preliminary prepartion with vasoconstrictor drops and suction is the investigation of choice (Morgan and Bailey,1990; Stansbie, 1992) and indicates whether the atresia is bony or membranous. Flexible or rigid fibreoptic nasendoscopy using topical anaesthesia indicates the diagnosis and permits 'palpation' of the obstruction (McIntosh, 1986). Dehaen and Clement (1985) and Morgan and Bailey (1990) respectively report use of 110° and 120° Hopkin's rod telescopes to visualize the post-nasal space whilst the atretic plate is perforated.

Treatment categories for CCA include transnasal, transpalatal, transeptal and transantral approaches, with the first two by far the commonest (Cinnamond, 1987). Reported techniques include use of the CO_2 laser (Illum, 1986), external rhinoplasty (Koltai et al., 1992), Le Fort 1 osteotomy (Thaller and Kawamoto, 1988; Resouly et al., 1990) and a sub-labial trans-septal approach (Krespi et al., 1987). Four cases of endoscopic repair of CCA were reported by Stankiewitz (1990) and a further case by El-Guindy et al. (1992).

Case histories

Case 1

A 16-year-old girl was referred with a lifelong history of right-sided nasal blockage and rhinorrhoea, varying from clear to mucopurulent. Her past medical history included closure of an atrial septal defect in early childhood and a right antral washout performed elsewhere at the age of nine years due to her persistent rhinorrhoea. Coronal paranasal CT scans showed a hypoplastic right maxillary sinus and middle turbinate but rigid endoscopy indicated right choanal atresia. She underwent endoscopic transnasal fenestration and nasal patency remained good after 21 months follow-up.

Case 2

A seven-year-old boy was referred with a lifelong history of right-sided nasal blockage and watery rhinorrhoea. Clinical examination was unremarkable although spatula examination indicated no nasal airway on the right side. Axial CT scanning showed right choanal atresia, as seen in Figure 1. Endoscopic transnasal correction was performed without post-operative stenting or nasal dressing and 18 months later his right nasal airway remained patent.

Case 3

A 29-year-old lady presented with a lifelong history of right nasal obstruction and rhinorrhoea. At the age of 15 years she had undergone trimming of both inferior turbinates. Clinical examination confirmed an absence of the nasal airway on the right side and rigid endoscopy indicated a right bony-hard choanal stenosis with only a pinpoint opening. She underwent endoscopic repair of her right choanal stenosis and 15 months later nasal patency remained good with no rhinorrhoea.

Surgical technique

Preliminary installation of 5 ml of Moffat's solution (comprising 2 ml 4 per cent cocaine solution, 2.5 ml 8.4 per cent sodium bicarbonate solution and 0.5 ml 1/1000 adrenaline) is performed in the anaesthetic room. Use of 25 per cent cocaine paste is avoided due to the tendency to produce smearing of the lens of the. Hopkin's rod telescope.

The patient is positioned prone with the table elevated to approximately 30°. Nasal endoscopy with the 0° telescopes is performed to confirm the diagnosis, check patency and the post-nasal space on the asymptomatic side and assess whether the atresia is membranous or bony.

The mucoperiosteum over the atretic plate and the posterior (vomerine) septum is infiltrated with 2 ml of 2 per cent lignocaine with 1/80 000 adrenaline using a modified spinal needle. A curved sickle knife is used to incise the mucosa over the septum posteriorly; this is elevated backwards from the incision to the atretic plate using Howarth's and Hill's elevators. The bony septum is perforated posteriorly and the contralateral mucosa similarly elevated away from the vomer.

Next, the intervening vomer is removed using a combination of Tilley-Henckel and Blakesley forceps back to the atresia. Mucosa is dissected anteriorly from the plate as far as possible to preserve sufficient to create flaps to line the fenestration. A cruciate incision over the plate may be necessary, although extensive preservation of this mucosa is technically difficult. The very postero–inferior 'submucous resection' is widened laterally to excise the atresia with a combination of Blakesley and Stammberger forceps but full fenestration into the post-nasal space also requires incision of the mucosa behind the bony plate. Finally the preserved mucosa can be fashioned to cover the bony edges.

None of the three cases required post-operative nasal packing or stenting. All cases were discharged home 24 hours after surgery and experienced no complications.

Discussion

Morgan and Bailey (1990) reported a long-term success rate of 84 per cent for transnasal repair of CCA although the initial success rate for bony atresias was 13 per cent compared to 58 per cent for membranous atresias. The latter group did however achieve a 100 per cent long-term success rate. The mean number of dilations required for both groups was two and only revision cases were stented. The early complication rate was 10 per cent, principally palatal fistulae and there was a 10 per cent late complication rate related to stenting producing alar or columellar ulceration.

FIG. 1
Axial CT scan showing right choanal atresia.

Singh (1990) advocated transnasal repair due to the shorter stenting time, possibility for early surgery even at 24 hours after birth, early discharge and the ability to breast feed immediately after surgery. However he suggested currettage for bony CCA in preference to drilling and cautioned against excessive mucosal removal or damage and stenting with a soft Portex tube for six weeks under broad spectrum antibiotic cover. He subsequently advocated use of a Gruber aural speculum for exposure and operating from the tonsillectomy position (Singh, 1991). The importance of partial removal of the posterior vomer to prevent restenosis was highlighted by Maniglia and Goodwin (1981).

From an extensive literature review Pirsig (1986) found similar restenosis rates for transnasal and transpalatal surgery. Although transpalatal surgery offers better visualization of the atretic area and more accurate placement of mucosal flaps the surgery is longer, entails greater blood loss and longer convalescence along with risks of creation of a palatal fistula and violation of the developing hard palate which has an essential growth spurt within the first five years. He advocated avoidance of this route under the age of six years, ideally until after puberty, to prevent maldevelopment of midfacial structures. The trans-septal approach may be useful for correction of unilateral CCA in older children via either a hemitransfixion or sub-labial approach and permits repair of any concomitant septal deformity, resection of the posterior part of the vomer and placement of mucosal flaps over the raw surfaces of the choanae with the intention of preventing restenosis (Pirsig, 1986). Coniglio *et al.* (1988) advocated use of the

transpalatal approach for CCA with CHARGE criteria to permit more radical fenestration.

Various stents have been used in the management of CCA including an endotracheal (Portex) tube (Maniglia and Goodwin, 1981). Foley catheter (Bartal, 1988), a modified tracheostomy tube (Sculateri and Stool, 1988) and short intranasal stents secured sub-labially (Grundfast *et al.*, 1990). Different types of stenting material have also been used including metal, rubber, polyethylene and silicone but use of soft stenting material is most likely to produce long-term patency. If stents are employed they should probably be maintained for at least six weeks for primary and 12 weeks for secondary repairs. The value of lining opened choaenae with mucoperiosteal flaps to prevent restenosis is uncertain and may not always be technically possible whichever approach is used (Pirsig, 1986).

Disadvantages of stenting include discomfort, unsightliness and complications such as septal or columellar damage, formation of intranasal synechiae and infection of the paranasal sinuses or nasal cavity (Bartal, 1988; Grundfast *et al.*, 1990; Morgan and Bailey, 1990).

The Le Fort 1 osteotomy may provide good exposure for adult CCA but is unsuitable for children because of the risks of subsequent dental and maxillary maldevelopment (Thaller and Kawamoto, 1988; Resouly *et al.*, 1990).The external rhinoplasty approach has been reported in five cases of unilateral CCA, although two patients developed restenosis within two months which required transnasal correction (Koltai *et al.*, 1992).

Illum (1986) described the use of the CO_2 laser in CCA,

commending the ease of surgery, minimal patient discomfort, short hospitalization, ease of revision (if required) and avoidance of the need for a stent. Muntz (1987) similarly found the CO_2 laser effective in most cases of CCA except in the presence of septal deviation, inferior turbinate hypertrophy, craniofacial abnormality and a high arched palate. However prolonged stenting (beyond two weeks) appeared to reduce the success rate, although 'statistical support was lacking'.

The safety of transnasal correction is improved by good vision and was initially enhanced by use of an operating microscope and later by fibreoptic endoscopes transorally and transnasally. 'Blind' transnasal surgery has been associated with such complications as CSF leak, brain injury, Gradenigo's syndrome, meningitis, subluxation of cervical vertebrae and death (Pirsig, 1986).

Stankiewitz (1990) reported four cases of endoscopic repair of CCA which underwent silicone rubber stenting initially for two months. Both bilateral CCA's required endoscopic revision; at the time of publication one case still awaited revision surgery, despite repeated home dilations, but the other case remained patent six months after endoscopic revision and two weeks of stenting. One case of unilateral CCA initially treated unsuccessfully by transnasal puncture at one year underwent successful endoscopic repair with stenting for two months. Patency remained good after two years. Another case had right CCA and left choanal stenosis: the right side was repaired endoscopically whilst the left side was dilated and both were stented for two months. At eight months the right side had narrowed but nasal function was unimpeded. The ages at which these patients underwent definitive endoscopic CCA repair were five days, 10 months, seven years and six days respectively.

Stankiewitz (1990) commended the possibility for precise surgery using the endoscope, i.e. good visualization, minimal blood loss, facility for direct placement of flaps and reduced stenting time. He employed a 2.5 mm endoscope for newborn infants, otherwise a 4 mm endoscope. An axial CT scan was advocated to confirm the diagnosis and assess the atresia; thick bony atresias with a large lateral component may be better approached transpalatally rather than transnasally. Similar contraindications may apply to endoscopic surgery as those discussed by Muntz (1987)—particularly severe septal deviations, craniofacial abnormalities and hypertrophic turbinates. Endoscopic transnasal surgery does however permit adequate removal of the vomerine septum under direct vision to reduce restenosis, as recommended by Maniglia and Goodwin (1981). Indeed Stankiewitz (1990) attributed two of his failures to inadequate resection.

El-Guindy et al. (1992) also described a case of endoscopic repair of CCA which resulted in long-term patency after two months of Portex stenting. They commended the excellent vision, precision permitted for surgery and flap preservation, early recovery and short hospitalization afforded by the endoscopic technique.

The diagnosis of unilateral CCA should be considered in cases of unilateral blockage and rhinorrhoea, particularly if there are any associated anomalies such as cardiac abnormalities, which are 'risk factors' – two of our cases had previously undergone nasal surgery elsewhere for symptoms from their CCA!

Rigid endoscopy and axial CT scans are important for establishing the diagnosis; coronal scans in our first case were unhelpful and rigid endoscopy was diagnostic in this and our third case. The benefits of endoscopic fenestration of CCA – particularly for unilateral obstruction, as in our three cases, include precise surgery, a shorter hospital stay and avoidance of packing or stenting.

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