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Developmental nasal anomalies

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Abstract

Despite the complex embryological development of the nose and surrounding structures, significant developmental nasal anomalies are rare. We therefore present our experience in the management of such anomalies (excluding choanal atresia) over the past 10 years.

Fifty cases were surgically treated; two nasopharyngeal cysts, four hairy polyps, two meningoencephalocoeles, seven gliomata, 20 dermoids, two capillary haemangiomata, four fibromas, one fibromyxoma, one mucocoele, one granuloma, one lipoma, two nasal aplasias, two nasal clefts and one nasal web. All cases presented with nasal obstruction and/or as a nasal mass. Computerized tomography and magnetic resonance imaging aided diagnosis determining the extent of intracranial involvement. Various surgical techniques were used ranging from simple excision to the lateral rhinotomy and Goodman's external rhinoplasty approach. Only five patients (10 per cent) suffered complications; two CSF leaks and three recurrences, all treated successfully.

Introduction

Developmental nasal anomalies are are occurring once in every 20,000–40,000 births (Hughes *et al.*, 1980). This is despite the very complex development of the nose and the surrounding structures.

Embryology

The primitive nose develops around the third week of embryonic life. It develops from the frontonasal process, a mesenchymal structure covered with ectoderm, which grows between the floor of the primitive forebrain and the roof of the stomodeum (Fig. 1). During the fifth week primitive nasal pits are produced by invagination of the nasal placodes. The frontonasal process divides into a median and two lateral processes. The maxillary process grows ventrally from the dorsal end of the first arch to fuse with the lateral nasal process forming the nasolacrimal duct. The apices of the maxillary process fuse with the median nasal process converting the pits into the primitive nasal cavity. This is separated from the buccal cavity by the bucconasal membrane which ruptures at the fifth week, to form the primitive choanae which lie posterior to the primary palate. The primary palate is formed from the maxillary processes and the fronto-nasal process.

The definitive palate develops behind the primary palate from the two palatal processes, thus displacing the tongue inferiorly. Mesodermal proliferation occurs between the forebrain and stomodeum as the head increases in size.

A diverticulum (Rathke's pouch) forms at the third week just in front of the bucconasal membrane. It makes contact with the infundibulum developed from the neurohypophyseal bud (Fig. 2) derived from the floor of the forebrain. At the sixth week, the connection between Rathke's pouch and the oral cavity disappears. Rathke's pouch forms the adenohypophysis and the infundibulum, the neurohypophysis of the pituitary gland. The foramen of Lushka represents the site of the embryonic

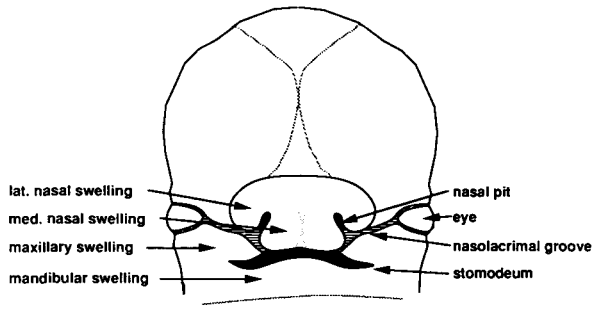


FIG. 1.
Seven week embryo—development of the primitive nose.

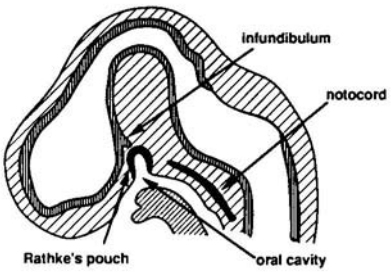


FIG. 2.
Six week embryo—pituitary development.

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Rathke's pouch. The nasal septum develops from a midline ridge from the posterior edge of the frontonasal process and fuses with the palatine processes. Ectodermal epithelium in the roof of the nasal cavities differentiate the olfactory cells which grow into olfactory bulbs. The superior, middle and inferior conchae develop as elevations on the lateral wall of each nasal cavity while diverticulae form the paranasal air sinuses.

After the second month of fetal life the anterior wall of the external nose has a cartilaginous capsule adjacent to the nasal and frontal bones and dura (Fig. 3A). The prenasal space extends from the frontal bone to the tip of the nose between the anterior frontal and nasal bones and the posterior cartilaginous capsule. This space contains firm connective tissue after the dural projection through the foramen caecum has been resorbed and the foramen completely obliterated. Simultaneously the space between the developing nasal and frontal bones (fonticulus frontalis) closes. Thus, developmental nasal anomalies may be derived from ectodermal, mesodermal or neurodermal tissue.

The majority of nasal anomalies present as a nasal mass and/or with nasal obstruction (Hughes *et al.*, 1980; Bradley and Singh, 1982; Myer and Cotton, 1983). Despite the similar presentation, the anomalies vary considerably in nature and position (Gorenstein *et al.*, 1980). Careful examination and imaging is essential to determine the correct surgical procedure and to avoid complications (Bradley and Singh, 1982). Therefore we have reviewed the developmental nasal anomalies which have been treated at The Hospital for Sick Children over the past 10 years.

Discussion

Anomalies

Fifty patients (26 males and 24 females) were treated at The Hospital for Sick Children between 1978 and 1988 (Table I). The age of presentation varied from birth to 10 years of age (median four weeks). All developmental nasal anomalies were included apart from choanal atresia.

Nasopharyngeal cysts (2)

Two girls had nasopharyngeal cysts, one presented at birth with breathing difficulty, the other at one month with feeding and breathing difficulties due to nasal obstruction. Both had large cysts visible in the post nasal space. Both cysts were removed perorally with a cuff of surrounding mucosa.

Midline cysts of developmental origin may be developed from four embryological structures: (i) median nasopharyngeal sinus, thought to have remnants of the notocord adherent to it; (ii) Rathke's pouch (above the buccopharyngeal fascia); (iii) Sessels pouch (rostrad of the buccopharyngeal fascia and caudal to Rathke's pouch); (iv) Thornwaldt's cyst or pharyngeal bursa which is seen in 3 per cent of the population (Myer and Cotton, 1983) and is a potential space between the pharyngeal ectoderm and notocord.

Histologically the cysts are surrounded by ciliated epithelium. The patients normally present with nasal obstruction and poor feeding. Imaging is rarely necessary unless extension into the cranial cavity is suspected. Excision down to periosteum or wide marsupialisation is usually curative (Chaudry *et al.*, 1978).

Hairy polyps 4

Four females presented at birth with airway obstruction due to a polyp arising from the nasopharynx (Fig. 4). Only one patient underwent CT scanning which excluded intracranial communication. The polyps originated from the posterior surface of the soft palate and were removed perorally with a cuff of surrounding tissue.

Hairy polyps are teratoid tumours of the nasopharynx and were first described by Brown-Kelly in 1918. They form part of a broad spectrum of malformations derived from misplaced totipotential cells which were classified by Arnold (1870) into four groups; (i) dermoids or hairy polyps which are derived from epidermal and mesodermal germ layers; (ii) teratomas which are trigerminal in origin and are so well differentiated that organs can be recognized histologically; (iii) epignathi representing the highest order of differentiation to form a parasite fetus normally attached to the sphenoid bone (Tuson, 1971); (iv) teratoids which are similar to teratomas except that they are composed of poorly differentiated tissues.

Hairy polyps histologically consist of ectoderm and mesoderm arranged in a disorganized fashion. The lesion contains skin and its appendages, fat lobules, fibrovascular connective tissue, smooth and striated muscle, minor salivary glands, nerves, lymph nodes, cartilage and bone. Sixty per cent occur in the nasopharynx (Brown-Kelly, 1918), are pedunculated and attached to either the nasopharyngeal surface of the soft palate or the lateral wall. Association with other congenital anomalies may occur (Arnold, 1870; Bradley and Singh, 1982). They classically present at birth, occur predominantly in girls (Brown-Kelly, 1918) and may be asymptomatic or present with complete respiratory obstruction (Foxwell and Kelham, 1958). CT scanning may be useful in differentiating them from neurogenic tumours (Chaudhry *et al.*, 1978; Senac and Segall, 1987). Wide excision of the pedicle is usually curative although there has been one reported case of recurrence (Frech and McAlister, 1969).

Meningoencephalocoeles (2)

Two males are diagnosed as having meningoencephalocoeles. An eight-year old was referred with right sided nasal obstruction, rhinorrhoea and three episodes of meningitis since birth. CT scanning showed a nasopharyngeal meningoencephalocoele communicating with the subarachnoid space via a bony defect in the floor of the sella. A transpalatal removal was performed and despite no evidence of a CSF leak at the time of operation, the patient developed meningitis three months later. The CSF leak was sealed by obliteration of the sphenoid sinus using a muscle graft via an external ethmoidectomy. No further sequelae occurred and the patient had normal pituitary function.

A four-week-old boy presented with unilateral nasal [illegible]ction. Clinical examination and CT scanning sug-

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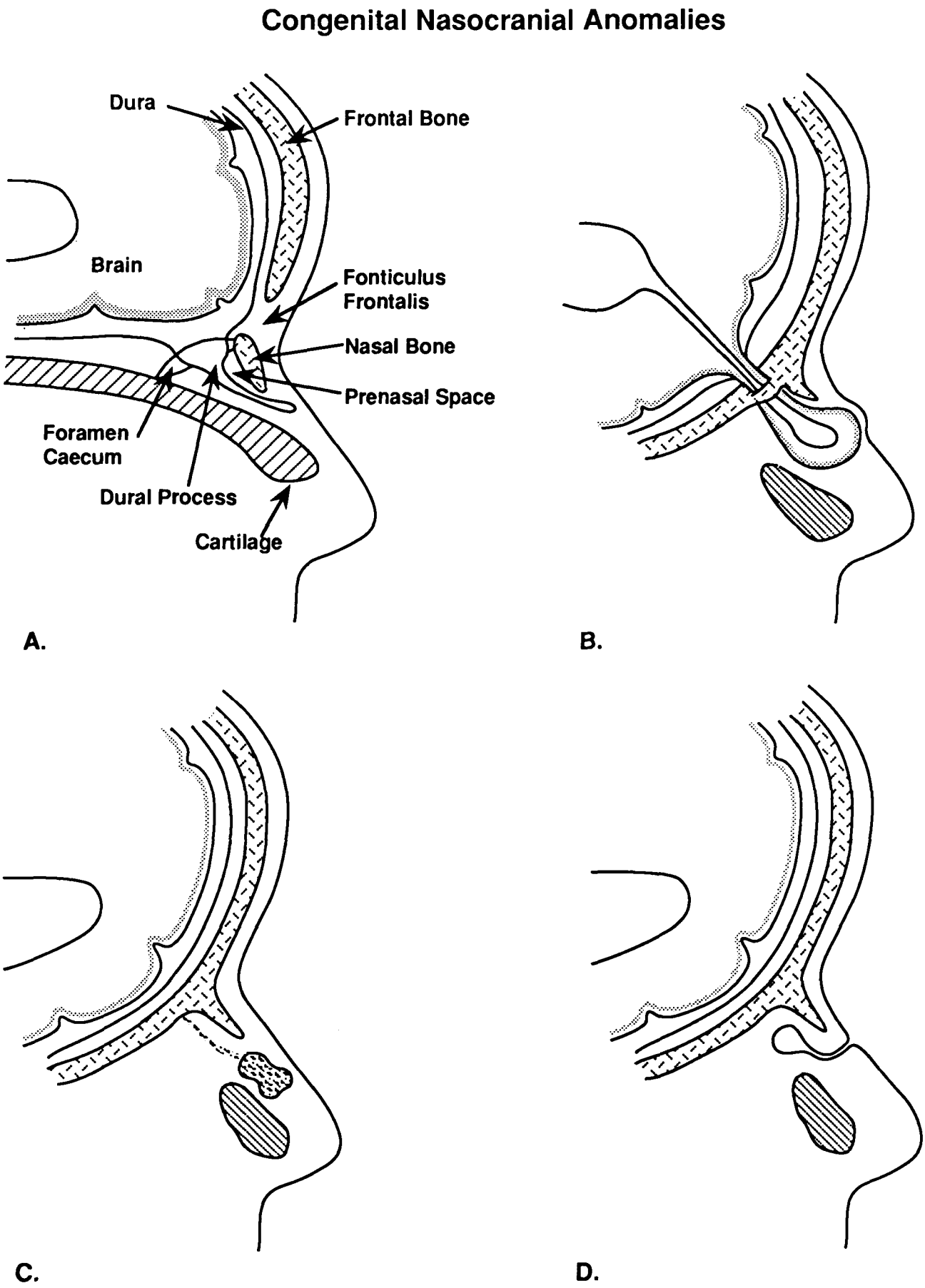


FIG. 3.

Congenital nasocranial anomalies. A—embryonic stage, B—meningoencephalocoele, C—glioma, D—dermoid sinus with cyst.

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TABLE I
DEVELOPMENTAL NASAL ANOMALIES

Dermoids	20
Gliomata	7
Fibromas	4
Hairy polyps	4
Nasopharyngeal cysts	2
Meningoencephalocoeles	2
Capillary haemangiomata	2
Nasal aplasia	2
Nasal cleft	2
Nasal web	1
Fibromyxoma	1
Lipoma	1
Mucocoele	1
Granuloma	1
	50

gested a frontoethmoidal meningoencephalocoele. A frontal craniotomy was normal and further examination of the nasal cavity revealed a bony unilateral choanal atresia which was repaired transnasally.

Meningoencephalocoeles are local herniations of glial tissue through a defect in the skull. The herniation consists of a meningeal sac containing brain tissue and the subarachnoid space which is in free communication with the cranial cavity (Fig. 3B). Three theories of pathogenesis exist (Birnbaum and Owsley, 1968): (i) they occur secondary to adhesions between the brain, dura and skin, arresting bony development; (ii) there is a primary failure in bone formation leaving a defect; (iii) there is increased fluid pressure in one of the ventricles causing herniation of the intracranial contents.

Sixty per cent present as a soft cystic mass overlying the roof of the nose (fronto-ethmoidal type) and 30 per cent as a pedunculated intranasal mass (basal type) (Myer and Cotton, 1983). An association with other anomalies has been noted (Yokota *et al.*, 1986). There may be a history of CSF rhinorrhoea or recurrent meningitis. A probe can be passed lateral but not medial to an intranasal meningoencephalocoele and helps to distinguish it from a polyp. Meningoencephalocoeles may pulsate and expand with compression of the jugular veins (positive Furstenberg's test) although this did not occur in our case and is not totally reliable (Jaffe, 1981).

CT scanning will detect the skull defect and intranasal mass but careful preparation using vasoconstrictors and nasal suction is essential to avoid misleading appearances as demonstrated in our case and others (Hughes *et al.*, 1980).

Biopsy is contraindicated; the surgical approach is dictated by the size of the defect. Anteriorly situated transethmoidal lesions should be excised transcranially (David *et al.*, 1984). Transsphenoidal lesions may be removed transcranially (Yokota *et al.*, 1986), transpalatally (Katz and Lewis, 1971) or by a combined approach (Lewin, 1983). The transcranial approach allows greater visibility, easier dural repair and has less infective complications. However, it may be associated with up to a 50 per cent mortality rate due to damage of vital structures (Yokota *et al.*, 1986). The transpalatal approach preserves the herniated hypothalamic structures and the bony defect may be closed with a muscle graft (Katz and Lewis, 1971). Operative intervention may not be necessary for the less symptomatic transphenoidal lesion in the older patient with no evidence of grave endocrinological or visual disturbance (Shirataki *et al.*, 1976).

Gliomata (7)

Seven patients (four males and three females) presented within the first four weeks of life with a fleshy mass in the nasal cavity (two also had an associated external nasal swelling) (Fig. 5). All patients underwent CT scanning, two of which suggested a defect in the cribriform plate. The median age at operation was two months (range five weeks to ten months). One patient underwent a transpalatal removal as the glioma arose from the nasopharyngeal wall and extended into the parapharyngeal space. One patient with an external nasal swelling had the glioma removed via a longitudinal incision with distraction of the nasal bones. Two excisional biopsies were performed transnasally. In one case this was successful in removing a small glioma from the middle turbinate and in the second the lesion was so

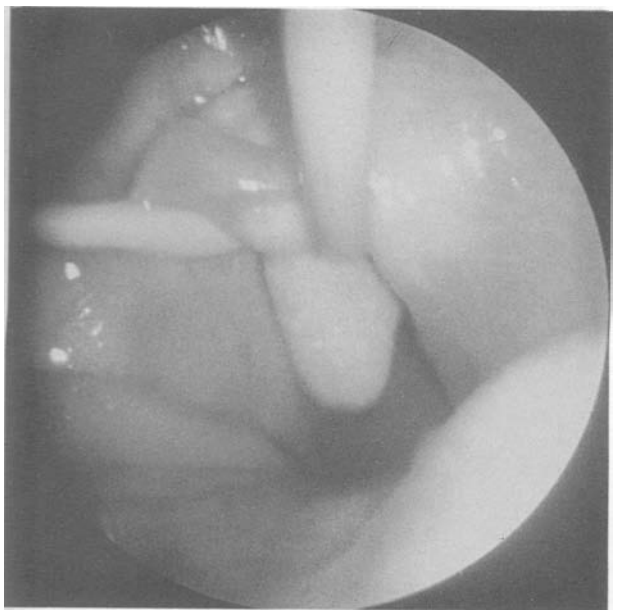


FIG. 4.
Hairy polyp projecting from the post nasal space with soft palate retraction.

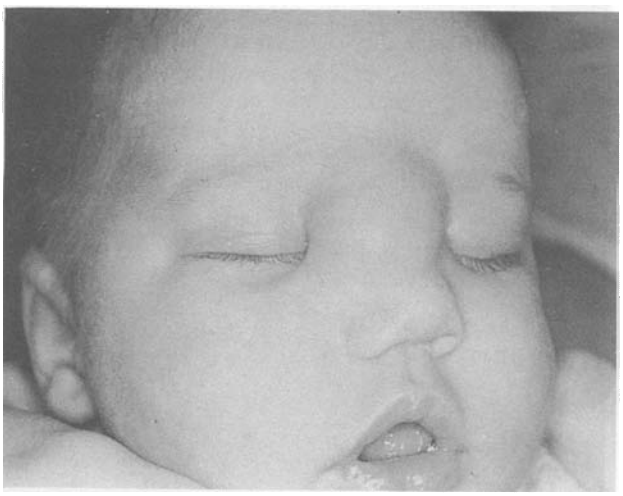


FIG. 5.
Patient with an extranasal glioma.

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extensive (Fig. 6) that it was decided to observe the glioma by serial MRI scanning prior to removal. Three patients had lateral rhinotomies, the gliomas originating from the middle turbinate and in two cases extended over the cribriform plate and onto the septum. Both had an associated defect in the cribriform plate (the dura being intact) which was sealed using a muscle graft. One patient had two further lateral rhinotomies for removal of residual tumour and to repair a CSF leak which subsequently developed.

Nasal gliomas are encephalocoeles which have lost their intracranial connection (Fig. 3C). There are three embryological theories: (i) they are due to faulty closure of the anterior neuropore (Sussenguth, 1909); (ii) are of blastomatous origin due to extracranial separation of embryonal neuroglia (Clark, 1905); (iii) arise from neuroglial cells in the nasal mucosa and grow through the fronto-ethmoidal suture (Anglade, 1920).

Histologically, they are covered with skin or nasal mucosa and are composed of astrocytic neuroglial cells interlaced with fibrous and vascular connective tissue.

They commonly present shortly after birth (Swift and Singh, 1985) and rarely in adult life (Katz and Lewis, 1971). Extranasal presentation occurs in 60 per cent, intranasal as a grey, yellow or purple polyp in 30 per cent and mixed in 10 per cent (Jaffe, 1981). In our series, two were mixed and five intranasal.

The gliomata are firm, incompressible, non-pulsatile and aspiration should be negative although this may be misleading (Birnbaum and Owsley, 1968). CT scanning normally shows a soft tissue mass with no associated intracranial component. However at operation a vestigial fibrous connection to the intracranial space may be found in 20 per cent of cases (Jaffe, 1981).

Extranasal gliomata are best removed by an incision over the glioma. Intranasal gliomata normally arise from the lateral wall of the nose around the middle turbinate. Small lesions may be removed transnasally, while the more extensive lesions involving the cribriform plate are best removed via a lateral rhinotomy. Nasopharyngeal lesions may be removed by a transpalatal route. Complications that may occur include recurrence (10 per cent) and CSF leak (Lowe *et al.*, 1973; Bradley and Singh, 1982). One of our patients experienced both, requiring two further lateral rhinotomies. Therefore careful outpatient follow-up is essential.

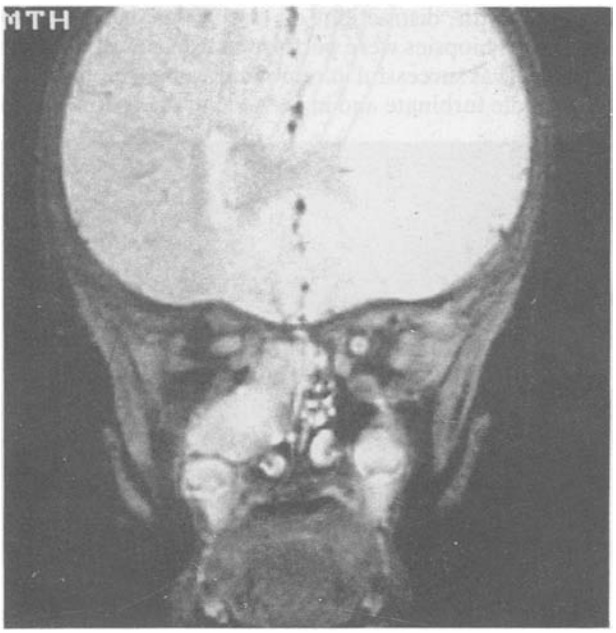


FIG. 6
MRI demonstrating an intranasal glioma.

Dermoids (20)

Twenty patients (12 males and 8 females) presented within 18 months of birth, eleven with a discharging nasal sinus since birth (Fig. 7). Six had an associated cystic swelling and two had protruding hair from the sinus. The remaining nine cases presented as cystic nasal swellings either midline (4) or at the medial canthal region (5). Five cases had further congenital anomalies (talipes, branchial sinus, hypertelorism, hydrocephalus and dysmorphic facies). One case had been operated on twice previously without success. CT scanning showed that 10 had nasal bone defects (Fig. 8) and two had cribriform plate defects.

The median age at operation was two years (range six months and five years). Seventeen patients underwent longitudinal excision with separation of the nasal bones where indicated. Three patients with distal nasal pits underwent a lateral rhinotomy combined with a Goodman's external rhinoplasty (Fig. 9). In two patients, the fibrous tract was traced up to a defect in the cribriform

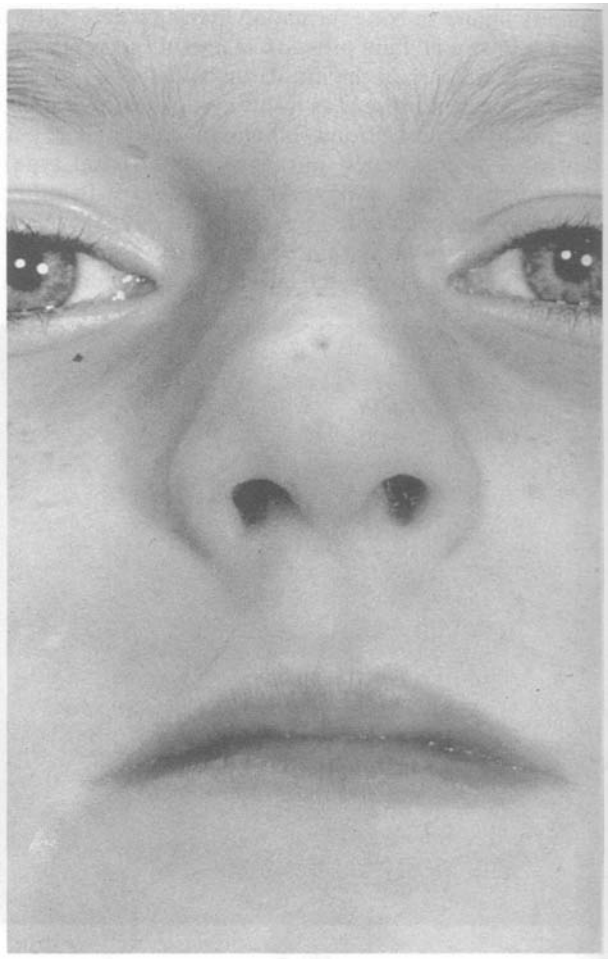


FIG. 7.
Patient with distal nasal pit.

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FIG. 8.

CT scan of nasal dermoid demonstrating erosion of the nasal bones and cystic swelling of the septum.

plate (the dura being intact). Two patients developed recurrent nasal fistulae requiring further excision.

Dermoids are ectodermal cysts which may contain skin, hair follicles, sebaceous glands, and sweat glands. They may have an associated fistula with hair protruding. One to three per cent of all dermoids involve the nose (Hughes *et al.*, 1980). Several embryological theories exist: (i) failure of the anterior neuropore to close; (ii) abnormal separation and obliteration of the prenasal space; (iii) inclusions in facial clefts; (iv) simple inclusion cysts; (v) aberrant development of skin appendages (Hughes *et al.*, 1980; Jaffe, 1981) (Fig. 3B).

The majority present at birth although some remain unrecognized till adult life (Hughes *et al.*, 1980). The cyst itself may become infected, produce intermittent episodes of swelling, inflammation and pain. An expanding cyst may produce pressure necrosis of the surrounding bone (Szalay and Bledsoe, 1972) resulting in an intracranial communication and hence meningitis (Hughes *et al.*, 1980). Characteristic appearances on CT scanning are: (i) fusiform soft tissue swelling within the nasal septum, (ii) widening or disruption of the nasal vault, (iii) a bifid nasal septum, (iv) glabella destruction or a soft tissue mass, (v) periosteal proliferation or pressure necrosis, and (vi) enlarged infrafrontal-intereth-moidal cystic space (Szalay and Bledsoe, 1972). Cribriform plate defects may also be visible as seen in two of our cases.

The timing of surgery is a compromise between the risk of interfering with developing structures and excising prior to the onset of complications; two years of age is ideal. Injection of the sinus tract with methylene blue at the time of surgery may be helpful. The surgical approach is dependent on the position of the cyst and fistula. If both are above the upper lateral cartilages, a longitudinal incision with a medial osteotomy if indicated is ideal. A distal nasal fistula or multiple fistulae may be excised via a lateral rhinotomy combined with a Goodman's external rhinoplasty. This allows excellent exposure for tracing the fistula through soft tissue and performing an osteotomy. The incision heals well with minimal scarring.

In our series, 10 patients required separation of the nasal bones but in only two was the tract traced up to the cribriform plate. A combined bifrontal craniotomy and transnasal excision may be required for extensive lesions however craniotomy carries a five per cent or more risk of epilepsy (Bradley and Singh, 1982).

The two recurrences in our series highlight the importance in complete excision of the tract (Szalay and Bledsoe, 1972; Myer and Cotton, 1983).

Capillary haemangiomata (2)

Two patients presented at six weeks and nine months with a progressive swelling at the glabella region since birth. Only one had an associated cutaneous haemangioma. One patient had both CT scanning and MRI. Only the MRI was consistent with an haemangioma (Fig. 10). The haemangiomata were excised with minimal blood loss through a skin crease incision.

Histologically, haemangiomata are classified as capillary, cavernous, mixed or hypertrophic. However this is of little clinical significance (Batsakis, 1984). They are usually present at birth and rapidly enlarge during the first months of life. Confusion with other lesions such as dermoids may occur in the absence of cutaneous haemangioma (Bradley and Singh, 1982). MRI is the investigation of choice as the STIR (short tau inversion recovery) image highlights the vascular tissue whereas CT scanning may be equivocal. Imaging will also help to exclude a sinus pericranium which communicates with an intracranial sinus via a foramen in the glabella (Birnbaum and Owsley, 1968). Most lesions will regress spontaneously with little or no residual deformity (Thomson and Lanigan, 1979). However if the diagnosis is in doubt, surgical excision may be carried out with minimal blood loss if the feeding vessels are ligated.

Miscellaneous anomalies

Fibromata (4)

Four patients (two males and two females) were found to have nasal fibromata. A two-day-old girl presented with respiratory difficulty, right sided nasal obstruction and proptosis. CT scanning confirmed the presence of an extensive lesion occupying the nasal cavity which was removed via a lateral rhinotomy. Although the dura was exposed the patient made an uneventful recovery.

A two-year-old boy presented with a short history of right sided medial canthal swelling and proptosis. A CT scan confirmed the presence of a localized lesion which was removed via a Howarth's incision.

A three-year-old boy with a cleft palate presented with a progressive swelling localized to the upper lateral/alar cartilage region since birth. The fibroma was removed via an intercartilaginous incision.

A three-year-old girl with Gardener's syndrome presented with a recurrent nasopharyngeal fibroma. The mass, which extended from the nasopharynx into the parapharyngeal wall, was excised perorally.

Fibromyxoma (1)

A 16-month old girl with multiple heart defects presented with a rapidly increasing swelling on the left side of the nose. Extension into the nasal cavity was confirmed by tomography (Fig. 11). The fibromyxoma arose from the nasolacrimal duct and was excised via a lateral rhinotomy.

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FIG. 9.
Exposure of the dermoid tract following a lateral rhinotomy combined with a Goodman's external rhinoplasty approach.

Lipoma (1)

A baby girl presented at birth with a polyp projecting through the nostril (Fig. 12). CT scanning showed the lesion arising from the septum and an associated lipoma of the corpus callosum. A biopsy confirmed the diagnosis of a lipoma which was successfully removed via a lateral rhinotomy at four weeks.

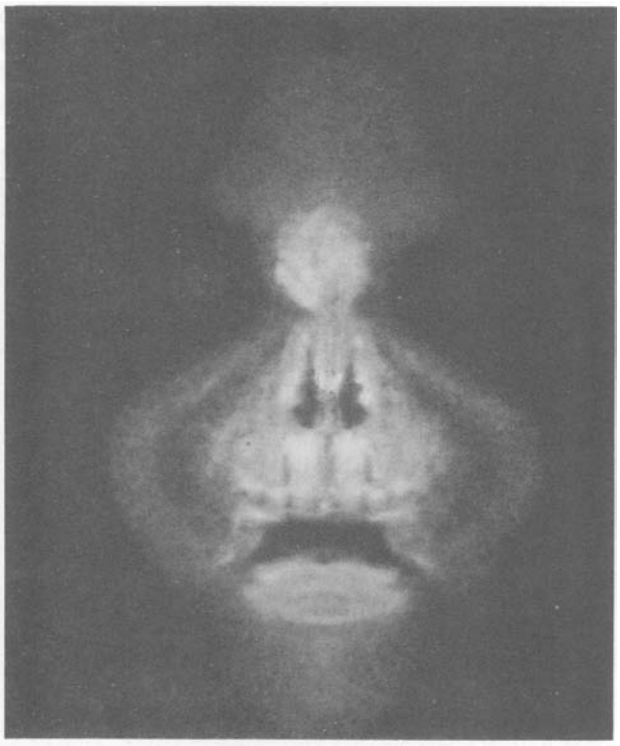


FIG. 10.
MRI of haemangioma. The STIR (short tau inversion recovery) exposure demonstrates the vascular nature of the lesion.

Granuloma (1)

A seven-year-old girl presented with an enlarging external nasal swelling which was removed via a Howarth's incision. The granuloma was found to be arising from the periosteum of the nasal bone.



FIG. 11.
Linear tomography demonstrating a nasal fibromyxoma occupying the expanded nasal cavity.

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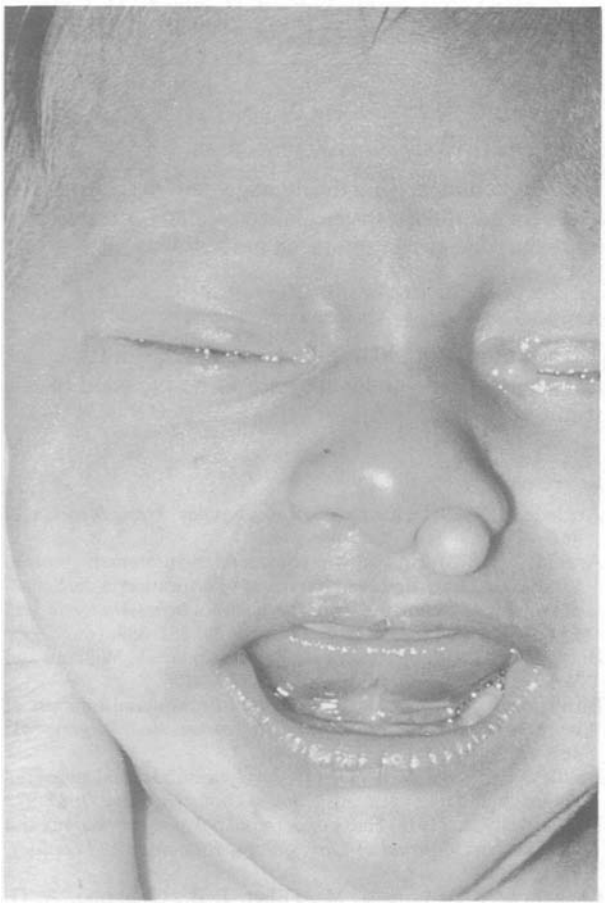


FIG. 12.
Intranasal lipoma projecting from the nasal cavity.

Mucocoele (1)

A previously fit 10-year-old boy presented with proptosis and swelling of the right side of the nose. A CT scan suggested a large ethmoidal mucocoele displacing the orbit. The mucocoele was successfully removed via an external ethmoidectomy.

The variety of other developmental anomalies illustrates the complex development of the nose. The extensive lipoma arising from the septum requiring a lateral rhinotomy has been previously described (Preece *et al.*, 1988). If the anomalies are associated with rapid growth, as in the fibroma, fibromyxoma and granuloma, confusion with malignancy may occur. Heffner has emphasized the importance of careful histological examination of such tissue prior to definitive treatment (Heffner, 1983). Mucocoeles in children are rare and usually occur in the ethmoidal region. Expansion into the orbit via the lamina papyracea occurs rapidly. Ethmoidectomy results in rapid resolution of the orbital tissue displacement. Histologically the mucocoele is lined by cuboidal epithelium and contains opalescent mucus with cholesterol crystals.

External pyramid anomalies (5)

Two patients (one male and one female) presented with total nasal aplasia (Fig. 13). Both were initially managed with an oral airway; however one required a tracheostomy at two weeks. In this case, CT scanning showed obliteration of the nasal cavities. Tissue expanders were inserted under the nasal skin at six weeks, and at two and a half years of age the nasal cavity was reconstructed via a sublabial incision and Portex stents inserted. External nasal reconstruction is planned.

CT scanning in the other child showed a rudimentary central nasal cavity. As the child was a successful mouth breather it was decided to use staged tissue expanders to develop a nasal profile which would be supported by bone grafts. Further implants will allow for growth of the face and create a nasal cavity at 16 years of age.

Two patients (one male and one female) presented at birth with extensive nasal clefts (Fig. 14) one of which was associated with bilateral choanal atresia. This was successfully treated by transnasal drilling and stenting. The cosmetic deformities were corrected by plastic surgery.

A one-day-old boy presented with feeding difficulty and unilateral nasal obstruction due to an anterior nasal web. The web was divided and a nasal stent inserted for three weeks.

Nasal agenesis is rare and is frequently associated with other facial malformations (Gifford *et al.*, 1972). The midface is covered with normal skin with an underlying bony plate. The paranasal sinuses are atretic with an associated high arched palate and absent nasopharynx. Until recently management was aimed at maintaining a nasal airway combined with a nasal prosthesis (Jaffe, 1981). The advent of tissue expanders and 3-D CT scanning has led to multistaged reconstruction resulting in

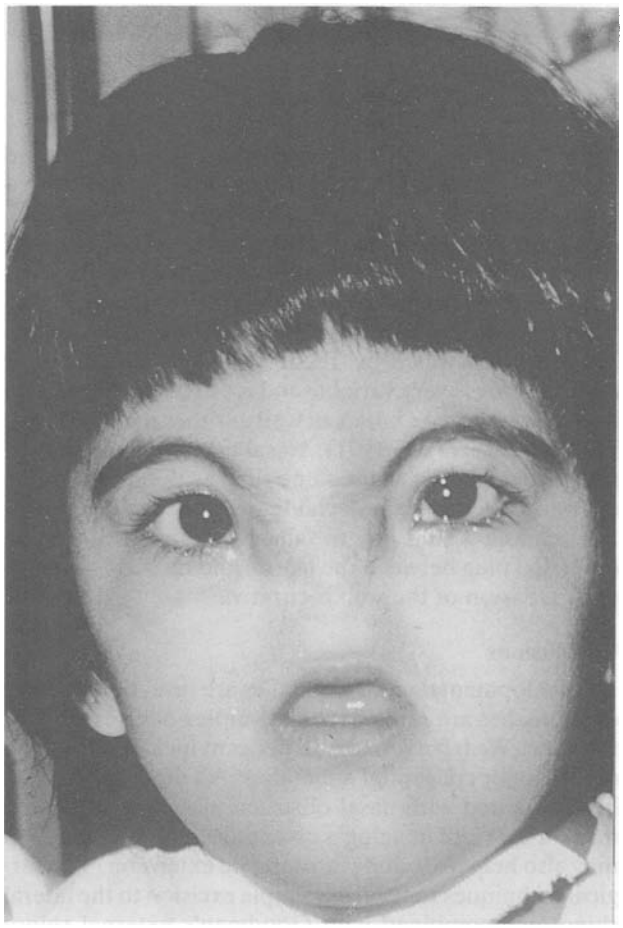


FIG. 13.
Patient with total nasal aplasia.

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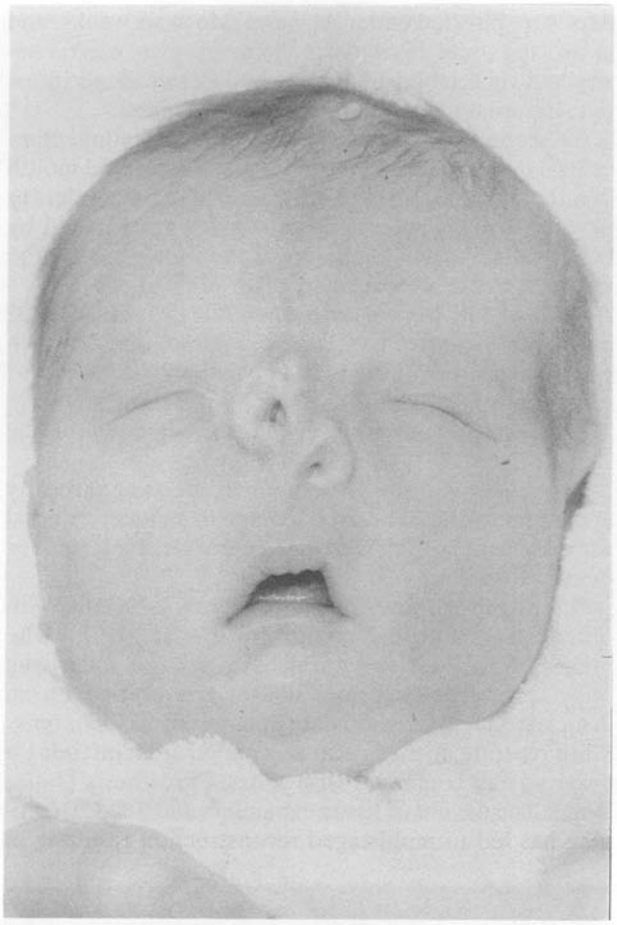


FIG. 14
Patient with nasal clefting.

better cosmesis. Tissue expansion may be started immediately and followed by staged external nasal pyramid reconstruction using bone or silicone implants. The definitive nasal profile is achieved in adolescence. If mouth breathing is established nasal cavity reconstruction may be delayed until palatal growth is completed.

Nasal clefts are but one part of the abnormality of facial clefting classified by Tessier (1976). The degree of nasal clefting is very variable and is often associated with imperforate nasal cavities as well as other major anomalies (Edwards *et al.*, 1971). Nasal cavity reconstruction may be performed at the same time as the repair of the cleft nose by the craniofacial team.

The nasal web is due to failure of canalization of an epithelial plug between the lateral and medial nasal process. Division of the web is curative.

Conclusions

Developmental nasal anomalies are rare, the diversity of anomalies are related to the complex development of the nose. We have found a 28 per cent incidence of associated major congenital anomalies. All the nasal anomalies presented with nasal obstruction and/or as a nasal mass. Therefore imaging is essential to aid diagnosis and may also help to exclude intracranial extension. The surgical techniques range from simple excision to the lateral rhinotomy combined with Goodman's external rhinoplasty approach for the more complex anomalies. We have found a 10 per cent complication rate (Table II) and would recommend that the management of such anomalies be undertaken in specialized units with access to neurosurgical and craniofacial units.

TABLE II
COMPLICATIONS

CSF leak	2 (1 glioma, 2 meningoencephalocoele)
Recurrence	3 (2 dermoids, 1 glioma)

Acknowledgements

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