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Branchial cleft and pouch anomalies

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Abstract

We present a retrospective study of 106 patients with branchial cleft and pouch anomalies who presented to the Hospital for Sick Children between 1948 and 1990. The relevant embryology of the branchial apparatus is summarized and a theoretical description of individual anomalies given.

Second branchial cleft sinuses were the most common anomalies, and the majority were managed simply, with adequate excision and a low recurrence rate. Five cases of first branchial cleft anomalies are presented, emphasizing the delay in diagnosis, the need for complete excision to prevent recurrence, and for a parotidectomy incision to protect the facial nerve from damage. The two third branchial pouch anomalies presented with a cystic neck swelling, one with recurrent infection and discharge, and the other with stridor. In both, the diagnosis was made at operation. The single fourth branchial pouch cyst was an unexpected finding in a patient with stridor.

Embryology

Six paired branchial arches appear between the fourth and sixth week of intra-uterine development. Each branchial arch consists of a core of mesenchyme covered externally by ectoderm and internally by endoderm. The mesenchyme forms bone, cartilage and muscle, and is supplied by an aortic arch artery and a nerve. Adjacent arches are separated by ectodermal clefts externally and endodermal pouches internally.

In fish, the clefts and pouches approach each other to form a closing membrane, which ruptures to form gill clefts. In man, the membrane is not ruptured and the cleft or pouch is gradually obliterated by invasion of surrounding mesenchyme. The fifth arch disappears and the sixth arch is rudimentary.

Figure 1 summarises the branchial derivatives.

The first branchial arch divides into maxillary and mandibular processes, and between the mandibular process and the second arch lies the first branchial cleft. The pinna develops around the dorsal end of the first branchial cleft from six tubercles. The three tubercles of the mandibular process form the tragus and the crus and superior rim of the helix, and the three tubercles of the second arch form the remainder of the pinna. The ventral part of the first cleft is obliterated, leaving the dorsal end to deepen to form the external auditory canal and surface of the tympanic membrane (Fig. 2). The first pouch forms the Eustachian tube and middle ear cavity.

The second arch enlarges to form an operculum (containing platysma) and covers the succeeding arches and clefts, which sink to form a depression, the cervical sinus. This sinus is soon obliterated from within outwards.

The palatine tonsil develops from the second pouch,

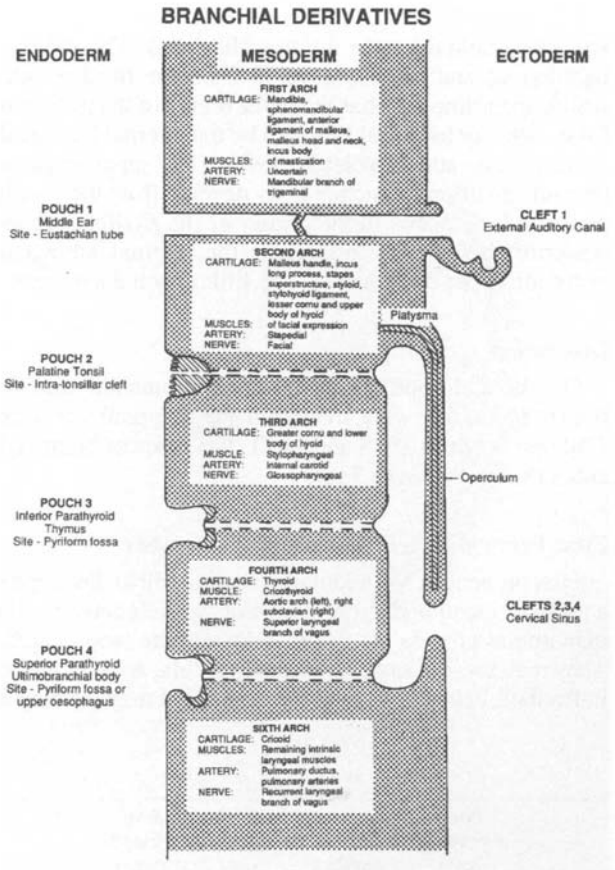

Fig. 1
Summary of branchial derivatives.

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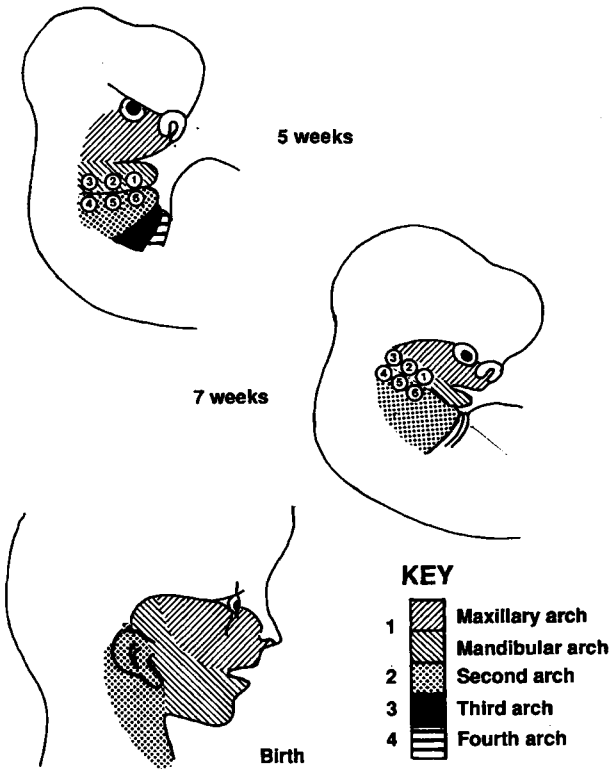

FIG. 2
Development of the external ear.

which is marked by the intratonsillar cleft. The inferior parathyroid and thymus develop from the third pouch, which opens into the pharynx in the region of the pyriform fossa, anterior to the fold formed by the internal laryngeal nerve (Burge and Middleton, 1983). The superior parathyroid and ultimobranchial body develop from the fourth pouch, which opens in the region of the pyriform fossa posterior to the fold formed by the internal laryngeal nerve, or upper oesophagus. The fifth pouch disappears.

Discussion

One hundred and six patients with branchial cleft or pouch anomalies were treated at the Hospital for Sick Children between 1948 and 1990. The types of branchial anomaly are shown in Table I.

First branchial cleft anomalies (five cases)

First branchial cleft anomalies are thought to develop as a result of incomplete obliteration of the cleft between the mandibular process of the first arch and the second arch. They may present as a cyst, sinus or fistula. A cyst may be in front of, below or behind the pinna, and may develop a discharging sinus after infection. A sinus will have an opening in the upper neck or in the floor of the external auditory canal, and a fistula will have an opening in both of these sites.

TABLE I
TYPE OF BRANCHIAL ANOMALY

	Pouch sinus or cyst	Complete fistula	Cleft sinus or cyst	Cleft cartilage remnant	Total
First	–	–	5	–	5
Second	–	2	90	6	98
Third	2	–	–	–	2
Fourth	1	–	–	–	1
				Total	106

The first branchial cleft anomalies have been classified as Type I or Type II by Work (1972).

Type I: Considered to be a duplication of the cartilaginous external auditory canal. A cystic mass in the postauricular area extends medially and anteriorly along the external auditory canal (Fig. 3). It usually passes lateral to the facial nerve and ends at the bony meatus. The lining is of skin plus adnexae, indicating ectodermal origin. No external opening is present except after infection and discharge.

Type II: Considered to be a duplication of the cartilaginous external auditory canal and pinna. A sinus passes from an external opening high in the neck along the anterior border of the sternocleidomastoid muscle, superficial or deep to the facial nerve and closely related to the parotid gland, to end either blindly near the floor of the cartilaginous external auditory canal, or to open into the canal in the same area as a complete (collaural) fistula (Fig. 3). The lining is of skin and cartilage, indicating ectodermal and mesodermal origin.

Case reports

Case 1 (Type I)

A 5-year-old boy with a one-year history of a cystic swelling in the floor of the external auditory canal and deep to the lobule of the pinna on the right side. The swelling was explored and removed via a postaural incision, which showed extension of the cyst down the external auditory canal to the bony meatus. Histology—skin lined with hair appendages.

Case 2 (Type I)

A 4-year-old girl with a discharging sinus in the right preauricular region for six months, and drainage of a low postaural abscess on four occasions. On examination there was anterior meatal wall fullness, and a sinus between the tragus and the lobule as well as behind the lobule (Fig. 4). Both sinuses were excised in continuity with a 2 cm diameter cyst which lay anterior and parallel

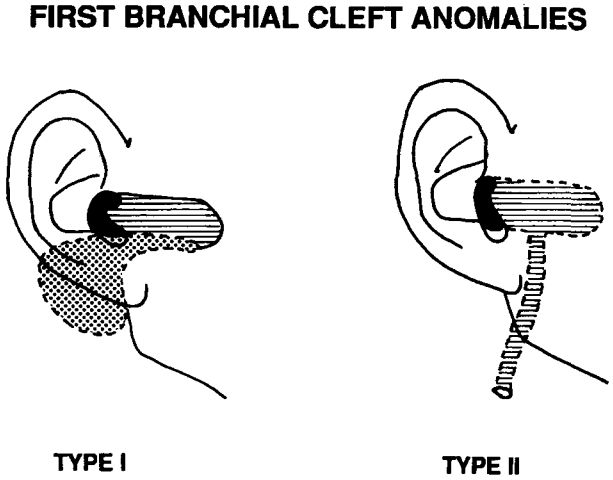

FIG. 3
Classification of first branchial cleft anomalies.

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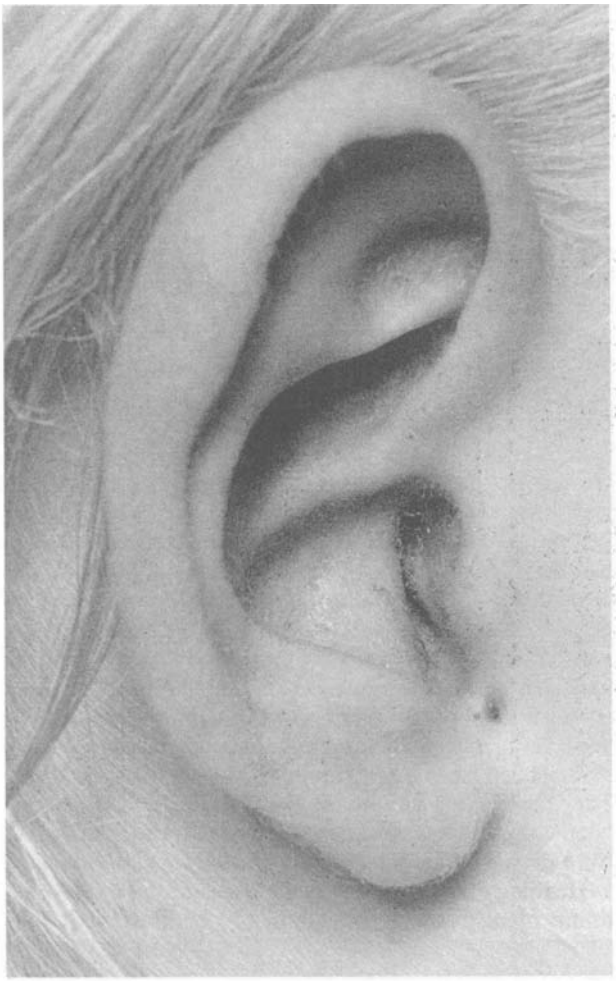

FIG. 4
Case 2: Anterior meatal fullness and cleft sinus—Work type I.

to the external auditory canal and extended down to the bony meatus. This operation was complicated by a transient facial weakness due to retraction. Histology-skin lining with adnexal structures.

Case 3 (Type I)

A 4-year-old boy with a discharging sinus between the right tragus and lobule since the age of five months. Four previous operations to excise per-auricular sinuses in the same region. A sinus tract was found parallel and anterior to the external auditory canal down to the bony meatus. Histology-fibrocartilage and fibrofatty tissue.

Case 4 (Type II)

A 3-year-old boy with a one year history of an infected swelling on the right side of the neck just posterior to the angle of the mandible. Incision and drainage of a neck abscess had been performed on two occasions, followed by excision of a tract leading from the external opening in the neck, superficial to the facial nerve, to an opening in the floor of the external auditory canal. The excision was repeated on two subsequent occasions because of recurrent discharge from the neck. No histology available.

Case 5 (Type II)

A 2-year-old boy presented at the age of six months with discharge from the left ear with a swelling in the left parotid region (Fig. 5). The ear was examined under anaesthetic six times with findings of a normal tympanic membrane, with granulations and polyps in the floor of the external auditory canal. The area was explored via a post-aural incision, confirming a defect in the inferior part of the tympanic plate, and the lesion was finally excised via a parotidectomy incision. A cystic swelling in the region of the angle of the mandible led to a tract passing deep to the facial nerve to the floor of the external auditory canal. The operation was complicated by a complete facial palsy which recovered over the next three months. Histology-a cystic lesion lined with skin and adnexae, with cartilage in the wall.

These five cases illustrate many of the points made by Work in 1972 concerning the diagnosis and management of first branchial cleft anomalies:

1. Diagnosis may be difficult (Randall and Royster, 1963)—all five cases presented before the age of four with periauricular swelling or sinuses, a mass in the external auditory canal, or discharge from the ear with a normal tympanic membrane. The average time taken for the correct diagnosis to be made was four years. Imaging by conventional radiology, CT or MRI scanning was of limited value, especially where previous surgery had been performed.
2. Many patients undergo incision and drainage for infection before definitive surgery is carried out (Belenky

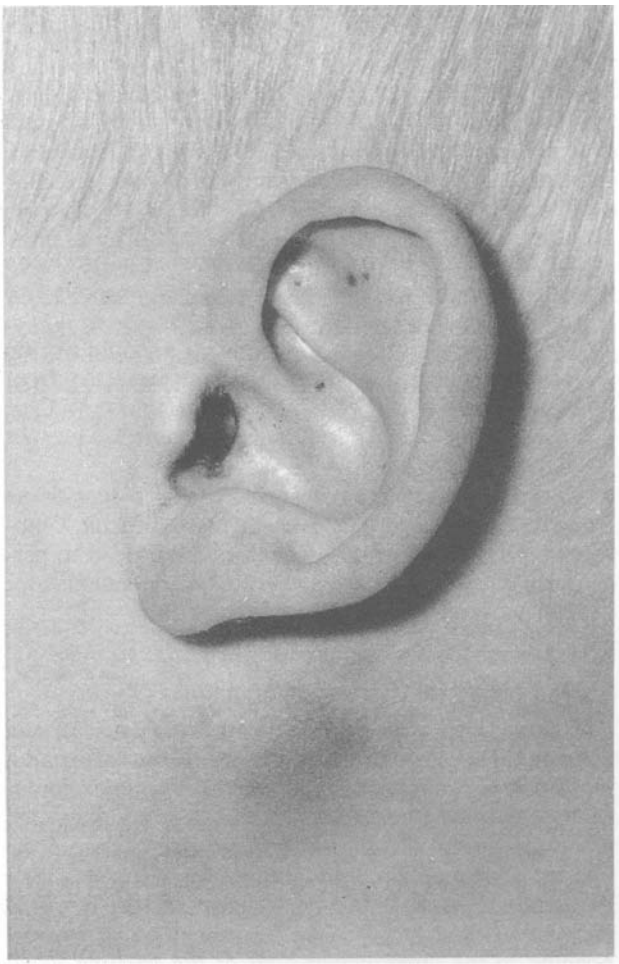

FIG. 5
Case 5: Ear discharge and parotid region swelling—Work type II.

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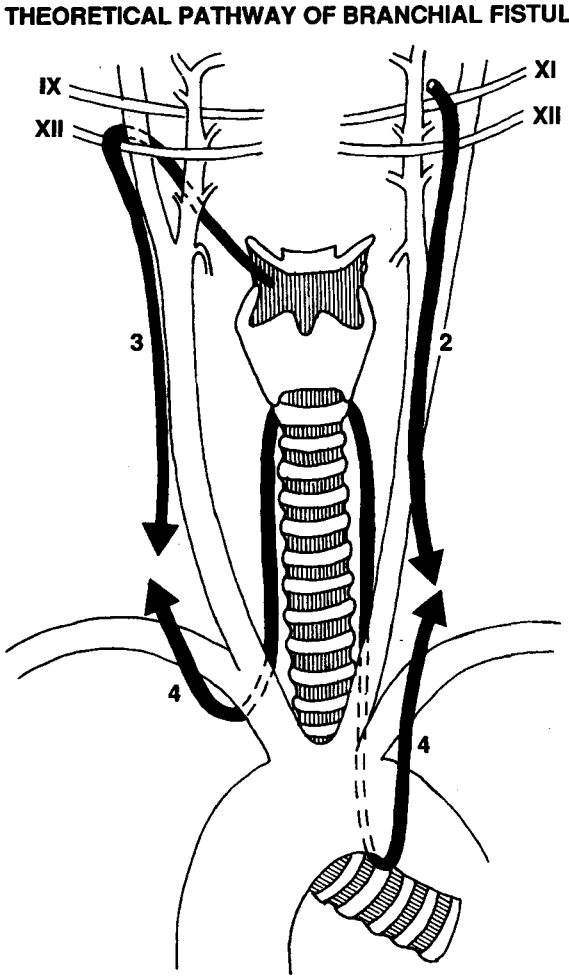

FIG. 6
Theoretical pathway of branchial fistulae.

and Medina, 1980)—on average, incision and drainage had been carried out 1.4 times before definitive surgery.

3. The operation site must be adequately exposed and the facial nerve must be identified and protected from injury (Olsen *et al.*, 1980, Miller *et al.*, 1984; McCrae *et al.*, 1983)—facial nerve damage occurred in two out of five patients.
4. Recurrent infection or operations lead to scar tissue and subsequent surgery is made more difficult. Complete surgical removal of the lesion will lead to permanent cure—an average of 2.4 operations for excision were needed for permanent cure.

Second branchial cleft/pouch anomalies (98 cases)

A persistent fistula of the second branchial cleft and pouch would be expected to pass from an external opening in the mid or lower neck in the line of the anterior border of the sternocleidomastoid muscle, deep to platysma, along the carotid sheath, then passing deep between the internal and external carotid arteries after crossing over the hypoglossal and glossopharyngeal nerves. It would then pass below the stylohyoid ligament to an internal opening in the intratonsillar cleft of the palatine tonsil (Fig. 6).

There were 98 patients in this group (53 female, 45 male). Eighty-four patients had a unilateral cleft sinus (Fig. 7), six had bilateral cleft sinuses (Fig. 8), two patients had a complete branchial cleft and pouch fistula and six had cartilaginous remnants in the neck without a sinus opening.

Figure 9 shows the age at presentation; 78 per cent presented by the age of five and in the vast majority there was a history of intermittent discharge and infection of a neck sinus from birth. In seven per cent there had been incision and drainage of an associated neck abscess.

Figure 10 depicts the site of sinus openings in the neck; 60 per cent were on the right and 40 per cent on the left, 63 per cent were in the lower third of the neck in the line of the anterior border of the sternocleidomastoid muscle.

A family history of branchial sinuses was found in six patients, and an association with preauricular sinuses in the same patient in five (three of these being in patients with bilateral branchial cleft sinuses).

The operation notes were examined to determine the length of the tract. In 49 the tract was 'short' and in the other 49 extended as far as the carotid bifurcation. In 37 the tract was followed as far as the wall of the pharynx, and in two the tract entered the pharynx in the region of the tonsil. In one of these cases a pre-operative sinogram had shown free flow from the neck sinus to the pharynx, and in the other a catheter was passed along the whole length of the tract and had entered the pharynx in the tonsillar fossa. The variable length of the tract influenced the number of 'step-ladder' incisions for complete excision: 68 cases were dealt with by one, and 30 needed two incisions.

Histology was available in 73 cases only; 42 showed a lining of respiratory epithelium (20 of these with sub-

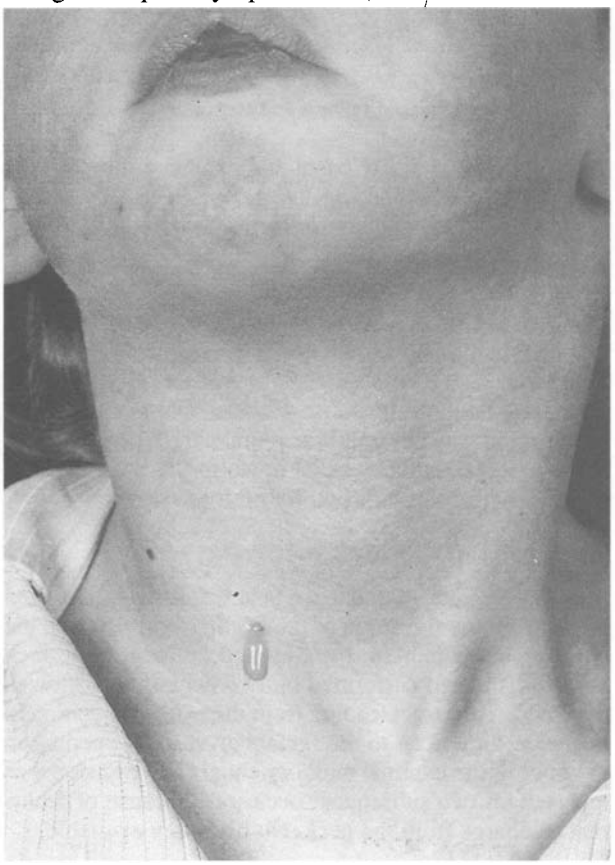

FIG. 7
Unilateral discharging second cleft sinus.

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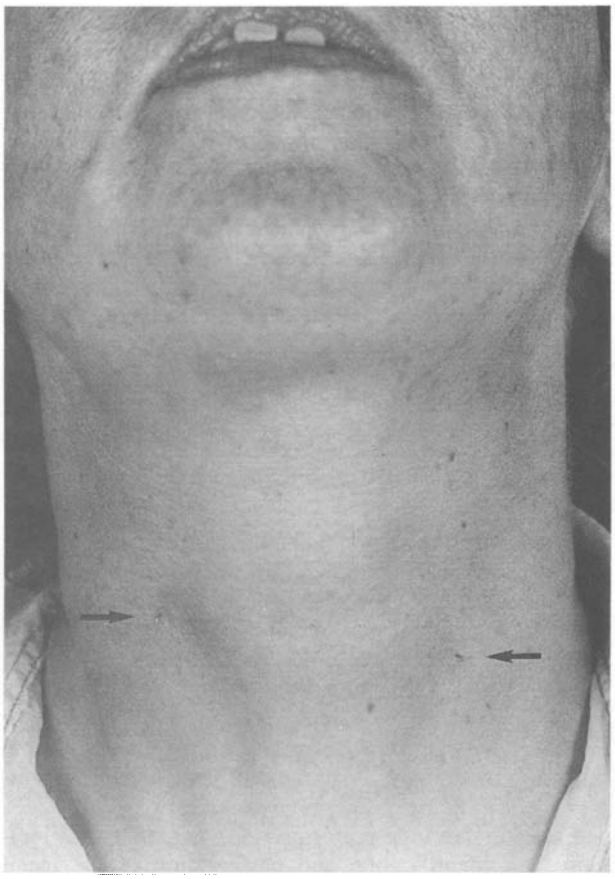

FIG. 8
Bilateral second cleft sinuses.

mucosal lymphoid tissue), 22 showed squamous epithelium, and nine were mixed respiratory and squamous epithelium.

The recurrence rate following surgery was 3 per cent.

Third branchial cleft/pouch anomalies (2 cases)

A persistent fistula of the third branchial cleft and pouch would pass from an external opening in the lower neck in the line of the anterior border of the sternocleidomastoid muscle, deep to platysma, along the carotid sheath then pass deep posterior to the internal carotid artery, between the glossopharyngeal nerve above and hypoglossal nerve below, through the thyrohyoid membrane to enter the pharynx in the region of the pyriform fossa (Fig. 6).

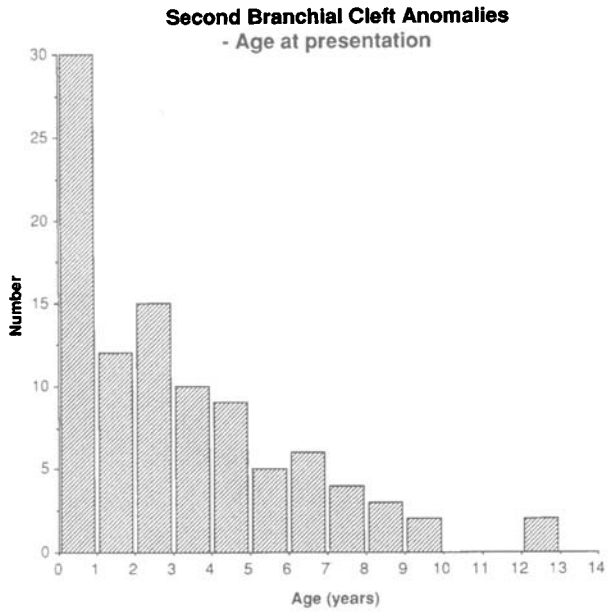

FIG. 9
Second cleft anomalies—age at presentation.

Case reports

Case 6

A 10-year-old girl presented with a four year history of recurrent discharge and swelling on the left side of the neck. Incision and drainage of a neck abscess had been performed at the age of six, and excision of a left sided neck mass had been performed on three separate occasions. On examination there was a neck scar and a cystic swelling between the larynx and the middle third of the sternocleidomastoid muscle on the left, but no sinus opening. Barium swallow showed a sinus from the left pyriform fossa (Fig. 11). Excision of the cystic mass deep to platysma was performed, and a sinus tract 7 cm in length was found to extend up the carotid sheath to the bifurcation. Four months after this operation she developed a discharging sinus on the left side of the neck and she underwent a left neck exploration with *en bloc* dissection of reactive lymph nodes. No recurrence after two years.

Case 7

A 2-week-old boy presented with a history of stridor from birth and a right-sided neck cyst since five days of age. On examination there was a 10 cm diameter right sided neck cyst, and soft inspiratory stridor. A CT scan (Fig. 12) showed a loculated cyst on the right, adjacent to the pharynx and larynx. At operation the cyst was found to contain pus, and was closely attached to the pharynx in the region of the pyriform fossa; laterally it was densely adherent to the carotid sheath from which it was separated with difficulty. Culture of the pus grew *E. coli*. The operation was complicated by a right vocal cord palsy which recovered over the next two months and a right Horner's Syndrome which did not recover.

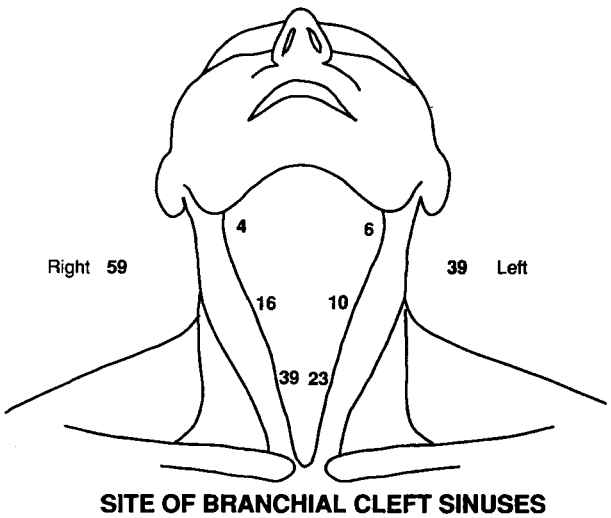

FIG. 10
Site of second cleft sinus openings.

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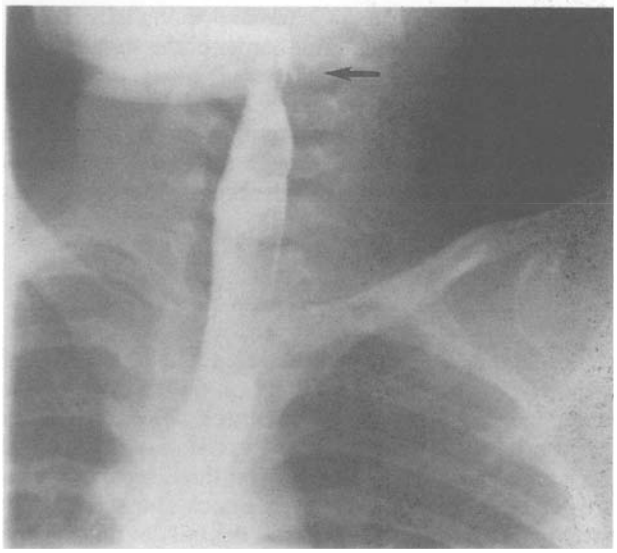

FIG. 11
Case 6: Barium swallow showing sinus in left pyriform fossa.

Case 6 was considered to be a third branchial pouch sinus and case 7 a third branchial pouch cyst. No external opening was present in either case. In the first case previous surgery made pre-operative assessment difficult. In the second case the large cystic mass precluded a preoperative diagnosis. The diagnosis was made at the time of surgery.

Fourth branchial cleft/pouch anomalies (1 case)

A persistent fistula of the fourth branchial cleft and pouch is theoretically possible but has never been demonstrated. From an internal opening at the apex of the pyriform fossa the fistula would pass between the thyroid and cricoid cartilages, and descend between the trachea and recurrent laryngeal nerve. On the left the fistula would loop around the aortic arch and on the right around the right subclavian artery (Fig. 6). Thereafter, the tract should rise in the neck posterior to the common carotid artery before passing over the hypoglossal nerve and descending to an external opening in the lower neck in the line of the anterior border of the sternocleidomastoid muscle (Liston, 1981).

Case 8

A 19-month-old boy presented with a history of stridor since birth with ten admissions to hospital with exacerbations of the stridor. Microlaryngoscopy revealed a large cyst filling the left pyriform fossa, which was deroofed. Open exploration of the pharynx and larynx revealed a cystic mass deep to the left lobe of the thyroid gland, extending via the cricothyroid membrane into the region of the pyriform fossa cyst. This cystic mass was excised.

Histology showed a mixed stratified squamous and respiratory epithelial lining to the cyst, with muscle, cartilage and thyroid tissue in the wall. The operation was complicated by some left cricoarytenoid joint fixation.

This case is probably a fourth branchial pouch cyst. There was no external opening and the diagnosis was made at operation.

Fourth branchial pouch anomalies are rare, with only 31 cases reported in the last 20 years (Takimoto *et al.*, 1990). Over 90 per cent occur on the left side (Godin *et al.*, 1990) and present in childhood. Some cases are associated with recurrent acute suppurative thyroiditis (Tovi *et al.*, 1985; Narcy *et al.*, 1988) with a sinus leading from the pyriform fossa to the left lobe of the thyroid gland. A barium swallow may demonstrate a sinus in the pyriform fossa, and endoscopy may reveal a sinus opening or cystic swelling in the same region. Excision may be facilitated by the insertion at endoscopy of an embolectomy catheter in the sinus opening (Feldman *et al.*, 1990; Godin *et al.*, 1990), and the sinus should be excised in its entirety, with purse-string closure of the pyriform fossa opening and partial removal of the left lobe of the thyroid if necessary.

Conclusions

The diagnosis of a first branchial cleft anomaly is to be considered in any patient with a history of recurrent peri-auricular swelling, a sinus or sinuses in the peri-auricular region or high in the neck in the line of the anterior border of the sternocleidomastoid muscle, a mass in the external auditory canal, a dimple or depression in the floor of the canal, granulations or polyps in the floor of the canal, or chronic discharge from the ear in the presence of a normal tympanic membrane. Imaging is of little benefit, especially when there has been previous surgery.

Excision should be performed via a parotidectomy incision with identification and preservation of the facial nerve and complete excision of the tract. The possibility of a facial nerve palsy must be explained to the patient. A

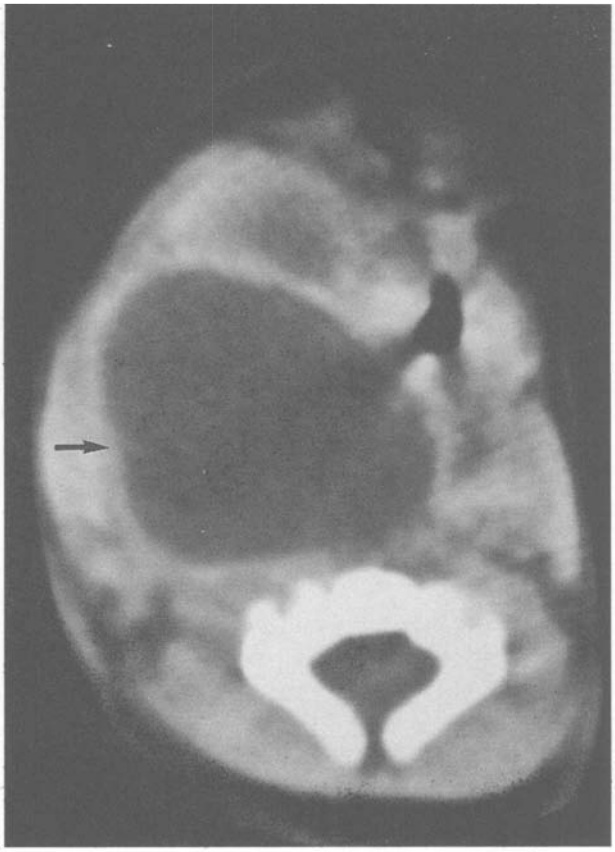

FIG. 12
Case 7: Cyst adjacent to larynx.

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sinus in the floor of the external auditory canal should be laid open and packed to reduce the risk of stenosis.

Second branchial cleft and pouch anomalies are by far the commonest. Most present by the age of five with a history of recurrent discharge or infection of a sinus in the lower part of the neck. There is a slight female predominance, and they are more common on the right side. In most cases there is a unilateral cleft sinus. A complete fistula to the tonsillar region is rare. A family history of cleft sinuses was found in six per cent. Associated preauricular sinuses were found in five cases. In three, bilateral preauricular sinuses and bilateral branchial cleft sinuses coexisted. In the other two cases the preauricular sinus was found on the side of the branchial cleft sinus.

Pre-operative assessment of the patient should include a search for bilateral lesions and any other congenital defects. Imaging is again of little benefit. Surgery may be delayed in an infant, waiting in an uncomplicated case until the age of two or three. Any infective episode must be allowed to subside after adequate antibiotic treatment before surgery takes place. The sinus tract may be delineated by the use of an embolectomy catheter, although the tract may not be canalized throughout its length, and in practice identification is not difficult as these tracts are invested in a layer of striated muscle which makes them quite substantial and fairly easy to follow. The external opening is excised with an elliptical incision and dissection of the sinus carried superiorly as far as possible, with a second 'step-ladder' incision if required. In half of the cases the sinus extended to the level of the carotid bifurcation, and in over a third to the wall of the pharynx. The recurrence rate following surgery was three per cent.

Third and fourth branchial pouch anomalies are rare, and the diagnosis should be considered in any child with recurrent neck abscesses (especially with associated stridor), and any child with recurrent acute suppurative thyroiditis (usually on the left side). Initial endoscopy or barium swallow may reveal a pyriform fossa cyst or sinus opening. Treatment should involve full excision of the sinus or cyst to prevent recurrence and may require a partial thyroidectomy.

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