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Transsphenoidal Canal (Large Craniopharyngeal Canal) and Its Pathologic Implications

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A vertical conduit in the basisphenoid extending from the floor of the sella to the undersurface of this bone was observed in two children. There was no associated nasopharyngeal mass. A review of the literature on this defect and related subjects suggests a close relation between this lesion and transsphenoidal meningoencephalocele.

Craniopharyngeal canal (CPC) or persistent CPC has long been used to describe a midline, almost vertical conduit in the basisphenoid containing a prolongation of the dura and some vascular structures [1]. The canal extends from the deepest part of the floor of the sella turcica to the pharyngeal surface of the basisphenoid near the alae of the vomer. It is quite small, measuring at most 1.5 mm in diameter, and often is obliterated inferiorly or superiorly. Its incidence in anatomic specimens has been reported to be around 10% during the first few months of life and to be much lower beyond this age, with an estimated incidence of complete canals in older individuals of 0.42% [1]. Because of its small size, a CPC is very seldom seen on skull films even in infancy, and when seen it is no more than a thin line [2]. There are two main theories regarding the origin of this conduit. In the earlier of the two, the finding is related to the development of the anterior lobe of the pituitary gland, representing the remnant of the stalk of Rathke pouch. In the other, first advocated by Arey [1], it represents the remnant of a vascular channel formed during osteogenesis.

CPC has also been used to describe a rarer and much larger bony canal in the same location. This conduit, designated here *large CPC* or *transsphenoidal canal*, measures several millimeters in diameter and is easily identified radiographically both in lateral and submentovertical views of the skull. It occupies most of the floor of the sella turcica and tapers inferiorly. It differs from the first CPC not only in its size but also in the fact that it is prone to be associated with special craniofacial anomalies. It has been suggested that these large craniopharyngeal or transsphenoidal canals are related not to the persistent CPC, but rather to transsphenoidal meningoencephaloceles [3, 4].

We report two cases in which large CPCs were observed on skull films; one of these cases was confirmed by computed tomography (CT) and magnetic resonance imaging (MRI) also. The literature related to this defect and to transsphenoidal meningoencephaloceles is reviewed, and the pathologic implications and possible embryologic origin of the lesion are discussed.

Case Reports

Case 1

An infant girl was born to a mother who throughout her pregnancy had been taking Dilantin and phenobarbital for a seizure disorder. The neonate was diagnosed soon after birth as

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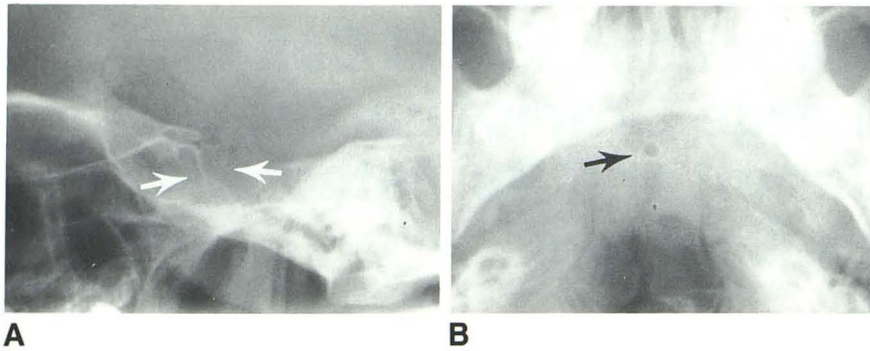


Fig. 1.—Case 1. Vertical defect in central part of sphenoid with origin in floor of sella (arrows) is well seen in lateral (A) and submentovertical (B) projections.

having fetal Dilantin syndrome, with all the typical features of this disorder including hypertelorism, small fingernails and fingertips, and abnormally formed distal phalanges. Skull films at 4 years of age showed a vertical canal through the center of the basisphenoid (fig. 1). It measured 5–6 mm superiorly and 3 mm inferiorly. No mass in the nasopharynx could be demonstrated either clinically or radiographically, and there were no symptoms referable to the pituitary gland.

Case 2

An infant boy was born with multiple congenital anomalies including a midline facial cleft with disruption of the left naris, bilateral cleft lip and a wide palatal bony defect, hypertelorism, slight hypoplasia of the left orbit and forehead, clubfeet, and a deformity of the right hand that was attributed to amniotic constricting bands. The mother had been taking phenobarbital during pregnancy because of a seizure disorder. Skull films and a CT scan of the child's head and face at age 19 months, before craniofacial surgery, showed a vertical canal in the basisphenoid similar to that seen in case 1 (figs. 2A–2D). No mass was demonstrated in the nasopharynx, and there were no symptoms referable to the pituitary gland. At the time of surgery for correction of the craniofacial deformity, several small defects were noted in the cribriform plate with minor herniations of the dura locally. MRI was performed at age 2 years using a Diasonics superconductive unit with an operative field strength of 0.35 T and spin-echo (SE) sequences. MRI showed a normal-appearing anterior third ventricle, suprasellar cistern, and pituitary stalk (figs. 2E and 2F). The pituitary gland appeared to be normally located in the pituitary fossa and there was no visible cerebrospinal fluid (CSF) within the sella turcica. The status of the dura mater at the level of the bony defect along the floor of the sella could not be determined. No mass was demonstrated in the nasopharynx.

Summary of Reported Cases

Cases of large CPC (transsphenoidal canal) and transsphenoidal meningoencephaloceles that have been described in the literature are divided into four categories:

Transsphenoidal Canal Not Associated with a Nasopharyngeal Mass

Our two patients fall into this category. Hypertelorism was present in both. In case 2, a midfacial cleft syndrome with a midline cleft lip and a wide cleft palate was also present, and

a defective ethmoid bone was found at surgery. Silverman [3] described a girl who was followed clinically and radiographically from birth to 5½ years of age. She was born with choanal atresia, hypertelorism, abnormal right ala nasi, and an asymmetry of the palpebral fissures. Larsen and Bassøe [4] reported a 14-year-old girl with a cleft palate, hypertelorism, diabetes insipidus, primary amenorrhea, and a transsphenoidal canal measuring 6–7 mm in diameter. A pneumoencephalogram showed extension of subarachnoid air to the floor of the sella. Kaufman [5] reported two adult patients with a large CPC who had an episode of spontaneous CSF rhinorrhea at the age of 49 and 56 years, respectively. In both, a pneumoencephalogram showed an extension of the subarachnoid air into the pituitary fossa, and in the second patient, also of the infundibular recess of the third ventricle. Apparently, the pituitary gland was located in the pituitary fossa in both instances.

Transsphenoidal Canal Associated with a Nasopharyngeal Mass

Several patients have been described with this bony defect occurring in association with a nasopharyngeal mass. Weber et al. [6] reported a 5-year-old girl who had had a polypike mass in the nasopharynx since birth. The mass extended through a transsphenoidal conduit. Histologically it contained both anterior and posterior pituitary tissue. Tweedie and Keith [7] reported a 6-month-old neonate with a bifid nose, a median cleft lip and palate, and a mass in the nasopharynx. At autopsy this mass was found to be from a funnel-shaped prolongation of the floor of the third ventricle together with the pituitary body, through a "patent CPC" the size of a little finger. Behens and Barr [8] reported a 12-year-old boy with pituitary gigantism since birth who had a transsphenoidal canal and a mass in the nasopharynx that was not further defined. Harwood-Nash and Fitz [9] mentioned a patient with a transsphenoidal canal associated with a mass in the nasopharynx. A pneumoencephalogram showed extension of air within the mass.

Classic Transsphenoidal Meningoencephaloceles

Several patients have been reported with the diagnosis of transsphenoidal meningoencephalocele [10–15] and a defect

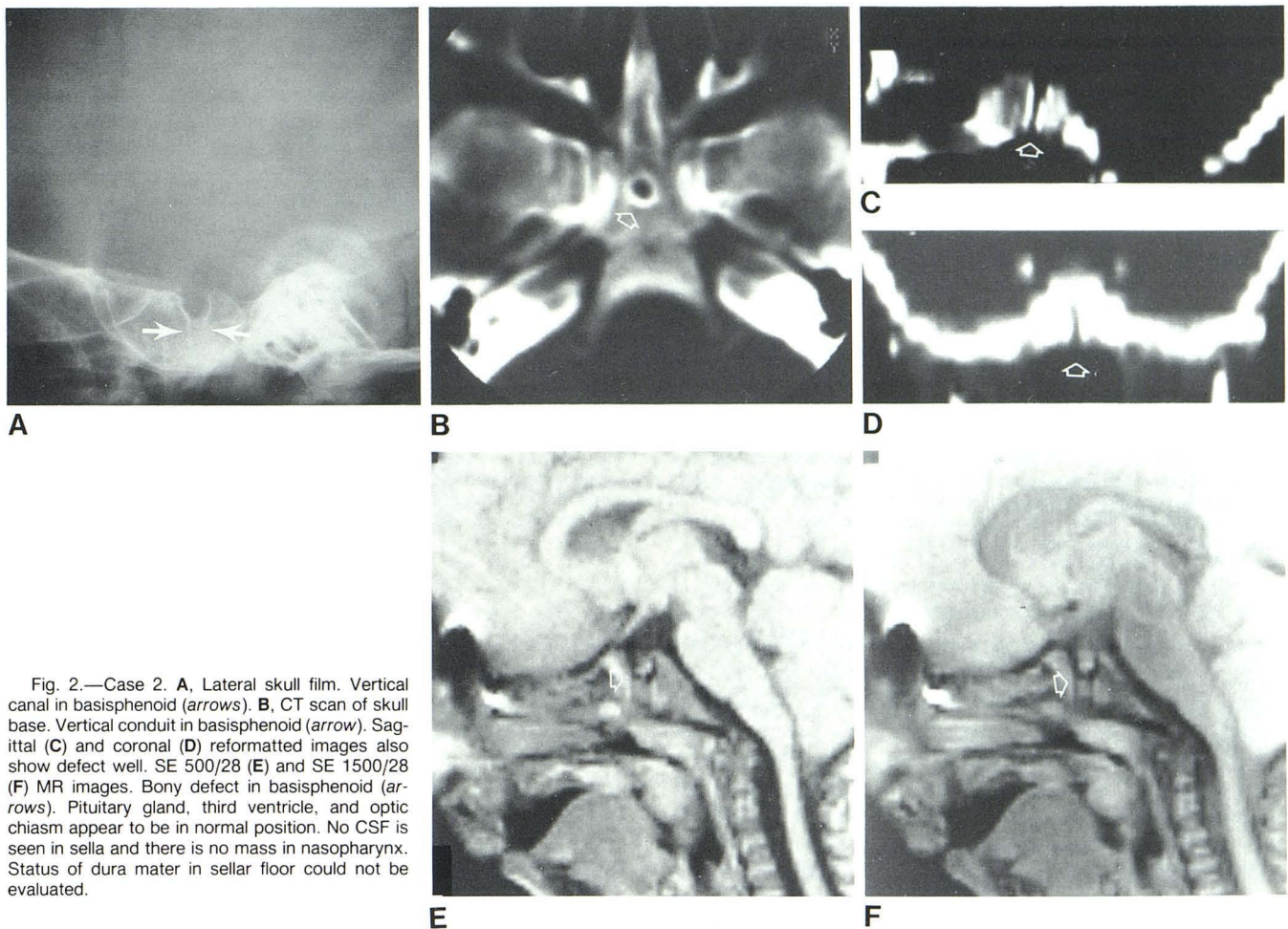


Fig. 2.—Case 2. **A**, Lateral skull film. Vertical canal in basisphenoid (arrows). **B**, CT scan of skull base. Vertical conduit in basisphenoid (arrow). Sagittal (**C**) and coronal (**D**) reformatted images also show defect well. SE 500/28 (**E**) and SE 1500/28 (**F**) MR images. Bony defect in basisphenoid (arrows). Pituitary gland, third ventricle, and optic chiasm appear to be in normal position. No CSF is seen in sella and there is no mass in nasopharynx. Status of dura mater in sellar floor could not be evaluated.

in the basisphenoidal that was quite large, measuring up to 2.5 cm in diameter. All these patients had a mass in the nasopharynx. A pneumoencephalogram showed extension of air into the mass with displacement of the third ventricle through the defect. Hypertelorism was a constant feature. A midfacial cleft and/or a cleft lip and palate, visual defects, and absence of the corpus callosum were reported in some cases. In a case reported by Ellyn et al. [11], the mass was found at time of surgery to represent a herniation of the pituitary gland and the third ventricle. The autopsy findings in the patient reported by Pollock et al. [14] showed that the optic nerves, chiasm, optic tract, hypothalamus, suprasellar cistern, and third ventricle were displaced downward through a large defect in the sella turcica. The supraoptic nuclei were absent.

Sphenoethmoidal Meningoencephalocèles

In reviewing cases of transsphenoidal meningoencephalocèles described in the literature we encountered three patients in whom the sphenoidal defect was quite large and extended forward to involve the ethmoid; two were reported by Pollock et al. [14] and one by Modesti et al. [16]. All three had hypertelorism and a nasopharyngeal mass. In the two cases reported by Pollock et al. [14], a 2 cm and a 3.5 cm midline

defect involved the sphenoid and the posterior part of the ethmoid; in both of these cases, pneumoencephalography showed a downward herniation of a dilated third ventricle. One of these patients also had a midline cleft lip and palate and absence of the corpus callosum. The other patient had a visual defect, a left eye coloboma, and diabetes insipidus. The case reported by Modesti et al. [16] is at the extreme end of the spectrum, showing hypertelorism, cleft lip, a large mass in the nasopharynx, and, at autopsy, a large midline defect involving the crista galli, the entire cribriform plate, body of the sphenoid, sella turcica, and the medial part of both lesser wings of the sphenoid. The nasopharyngeal mass consisted of a sac containing CSF, and it was continuous with the third ventricle. The optic nerves were small and the chiasm, optic tracts, septum pellucidum, and cingulate gyri were absent.

Discussion

It appears from our review that a large CPC, or transsphenoidal canal, may occur in association with a mass in the nasopharynx or without a nasopharyngeal mass. The mass most commonly represents herniation of third ventricle and pituitary gland through the bony defect. Sometimes the mass is formed by a sac filled with CSF, suggesting herniation of

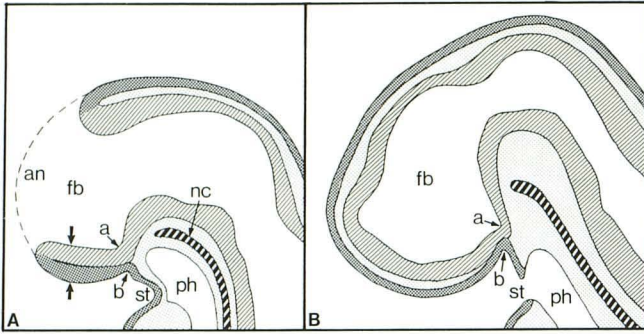


Fig. 3.—Sagittal sections of cephalic end of embryo at 3 (A) and 4 (B) weeks outlining forebrain (fb), anterior neuropore (an), stomodeum (st), pharynx (ph), and notochord (nc). Site of future pituitary gland and basisphenoid is shown at a and b. In A, anterior neuropore is still open, and entire ventral surface of forebrain is adherent to buccal ectoderm (arrows). In B, anterior neuropore is closed and oral membrane is patent; mesenchyma from sides of head has migrated medially separating base of forebrain from buccal ectoderm, except for small area (a,b) immediately cephalad to the remnant of oral membrane, at site of future pituitary gland and basisphenoid, where separation of two epithelia occurs much more slowly. (After [18] and [19].)

the dura (meningocele). The anomaly is considered to be a form of meningoencephalocele, differing from the classical variety (group 3 of this review) only in the size of the bony defect. Hypertelorism, midfacial cleft, cleft lip and palate, abnormal optic tracts, malformed eyes, and absence of the corpus callosum are seen in both conditions. In patients with a large CPC without a nasopharyngeal mass, the anomalies are probably also related to classic transphenoidal meningoencephalocele, and the similarity of the craniofacial anomalies that may be seen in association with these conditions supports this opinion. Our case 2 had a defective ethmoid bone suggesting a relation also with a transethmoidal meningoencephalocele also.

There is little documentation in patients with a transphenoidal canal without a nasopharyngeal mass regarding the status of the dura in the floor of the sella, the status of the diaphragma sellae, and the position of the pituitary gland and third ventricle. An absence of the dural covering in the floor of the sella at the level of the bony defect and an incompetent diaphragma sellae conceivably may predispose to herniation of the pituitary gland, displacement of the third ventricle, or CSF rhinorrhea. The two patients reported by Kaufman [5] apparently had an incompetent diaphragma sellae and a defective dural covering of the bony defect in the sella turcica and bony conduit, resulting in the episode of CSF rhinorrhea in adult life. The diaphragma sellae in these patients probably became incompetent with shrinkage of the pituitary gland due to aging, a possibility suggested by Bergland et al. [17].

In one of the cases reported by Kaufman [5] and in our case 2, the third ventricle and pituitary gland were apparently in normal position. In the second patient described by Kaufman [5] the pituitary gland was apparently in normal position but the third ventricle was displaced downward.

These findings suggest that transphenoidal canals with a nasopharyngeal mass may be associated with abnormalities in and around the sella turcica, although the possibility is not excluded that the defect may also occur as an isolated

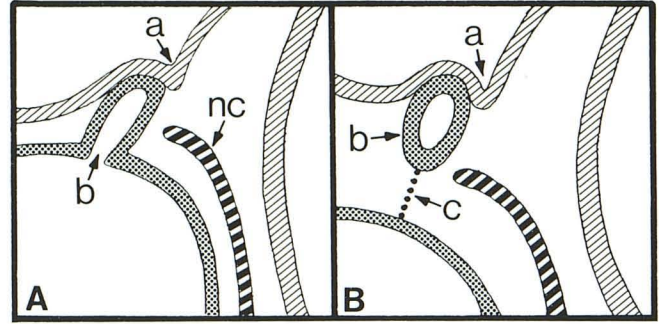


Fig. 4.—A, Sagittal section at level of developing pituitary gland in 6-week-old embryo. At b is a diverticulum of buccal ectoderm (Rathke pouch), representing precursor of anterior lobe of pituitary gland. Comparable diverticulum of forebrain (a) is precursor of posterior lobe or neurohypophysis. nc = notochord. B, Subsequent stage of development (7 week embryo). Local mesenchyma has increased, constricting lower part of Rathke pouch. Constricted inferior part of Rathke pouch at first is thin channel (c) and then an obliterated stalk that soon disappears concomitant with chondrification of local mesenchyma, future basisphenoid.

anatomic variant. CT and MRI of the area may provide valuable diagnostic information. Metrizamide cisternography may be more accurate in evaluating these patients, but probably this invasive procedure should be used only in cases of large CPCs when a nasopharyngeal mass is not excluded satisfactorily on clinical grounds or by noninvasive imaging (e.g., CT and MRI); before adenoidectomy; or in patients who also have diabetes insipidus, gigantism, acromegaly, or other hypothalamus-pituitary abnormalities.

The oldest and most likely theory regarding the embryologic origin of basal meningoencephaloceles, and in all probability also of simple transphenoidal canal, is an abnormality of early fetal life, resulting in an incomplete separation of the buccal ectoderm from the ventral surface of the prosencephalon (forebrain). (See figs. 3 and 4 for the normal development of this area, of the pituitary gland, and of the basisphenoid.) In meningoencephaloceles involving both sphenoid and ethmoid, probably a large area of the neuroectodermal union at the ventral surface of the head is abnormally developed, whereas in isolated transphenoidal meningoencephaloceles, as well as in transphenoidal canals, the area involved is the site of development of the pituitary gland and basisphenoid. The main abnormality in these defects is probably a failure of this peculiar area of the skull base to be invaded by mesenchyma, hence the bony defect. The changes probably occur concomitantly with the formation of the pituitary gland, but not necessarily as a result of a faulty resorption of Rathke stalk.

Although the defect in transphenoidal meningoencephaloceles and large craniofacial or transphenoidal canal seems limited to a small area of the skull base, the association of specific craniofacial and central nervous system (CNS) abnormalities points to a pathologic factor interfering also with the normal development of other parts of the "neuroectodermal plate" along the ventral surface of the forebrain, and affecting structures that in early life are in close contact and undoubtedly interdependent developmentally, such as optic



Fig. 5.—Preoperative CT scan in 3-year-old boy. Transsphenoidal canal (short arrow) and bilateral cleft palate (long arrows).

tract, corpus callosum, cranial base, and maxillary processes. The genetic and teratogenic influences that result in these conditions are unknown. It may be noteworthy that the mothers in our two cases received phenobarbital, and one of them also Dilantin, throughout pregnancy for a seizure disorder.

Our study supports the opinion that a large CPC is related developmentally to transsphenoidal meningoencephaloceles and may indeed be the site of herniation of the pituitary gland and third ventricle. Patients with this defect are prone to have associated craniofacial anomalies and abnormalities of the CNS similar to those seen in classical transsphenoidal encephaloceles. The available data do not demonstrate or disprove the existence of "simple transsphenoidal canal" occurring in the absence of any other abnormality in and around the sella turcica and pituitary gland.

Addendum

After our manuscript was submitted for publication we investigated another patient, a 3-year-old boy, with findings very similar to those seen in case 2. He had a severe midline facial cleft, a grossly defective nose, marked hypertelorism, bilateral cleft lip and palate, and skin tags and scars on the forehead that were attributed to congenital amniotic bands. Congenital constrictions, also attributed to amniotic bands, were present in the distal segment of the left leg and were associated with an absence of most of the left foot. Constricting bands were also present in four fingers of each hand with soft-tissue syndactyly on the right, and there was congenital amputation of four toes on the left foot distal to constricting bands. Lateral skull films showed a transsphenoidal canal

similar to that illustrated in figures 1A and 2A. This bony defect as well as the bilateral cleft palate were also clearly demonstrated on a CT scan of the head and face (fig. 5). At the time of surgical correction for the hypertelorism through a frontal craniotomy, small dural tears were noted in the region of the cribriform plate, possibly congenital, without basal encephalocele or meningocele. The status of the pituitary gland was not ascertained.

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