Cephaloceles and Related Malformations

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This chapter addresses the gross pathology and imaging of anencephaly, exencephaly and cephaloceles, and includes the clinical, epidemiologic, and pathologic features that have helped the authors to understand these conditions and to contribute meaningfully to their diagnosis and management. This paper specifically *excludes* the fronto-ethmoidal (sincipital) cephaloceles, the related nasal gliomas and nasal dermal sinuses, and the median cleft face syndrome, because these subjects have already been reviewed, in detail, in the American College of Radiology Self-Evaluation Program No. 28, Neuroradiology Text and Syllabus (1), in texts (2–6), and in articles (7–22).

Definitions

Cranium bifidum occultum designates simple midline or paired paramedian skull defect(s) without prolapse of meninges or brain (23).

Acrania is absence of the calvarium. The skull base may be intact.

Exencephaly is acrania with protrusion of a substantial portion of the central nervous system (CNS) into the amnionic cavity in utero and then into the environment postnatally.

Anencephaly is acrania with absence of most or all of the brain tissue.

Cephalocele signifies a congenital defect in the cranium and dura with extracranial herniation of any intracranial structure (23, 24). Cephaloceles are subdivided into four types (23).

1. *Cranial meningoceles* are cephaloceles in which the structures protruding consist solely of leptomeninges and cerebrospinal fluid (CSF).
2. *Cranial glioceles* are cephaloceles in which the structures protruding consist only of a glial-lined cyst containing CSF (22).
3. *Cranial meningoencephaloceles* are cephaloceles in which the structures protruding consist of leptomeninges, CSF, and brain. Some authors also distinguish cranial meningoencephaloventriculocele in which portions of the ventricles protrude into the hernia sac, perhaps in association with herniating choroid plexus.
4. *Atretic cephaloceles* are formes fruste of cephaloceles that are characterized by a small, noncystic, flat, or nodular lesion situated in the midline of the scalp, either near to vertex (parietal form) or just cephalic to the external occipital protuberance (occipital form) (25).

Cutis aplasia congenita signifies a (multi)focal congenital defect of the scalp, with or without absence of the underlying bone and meninges (26).

Cranium Bifidum

Cranium bifidum is characterized by the persistence of wide fontanelles into childhood (27). The normal frontal and parietal bones form from intramembranous ossification centers that arise laterally and expand toward the midline to enclose the brain (28). The normal fontanelles are transient zones of nonossified tissue that lie between the converging ossification centers. They narrow

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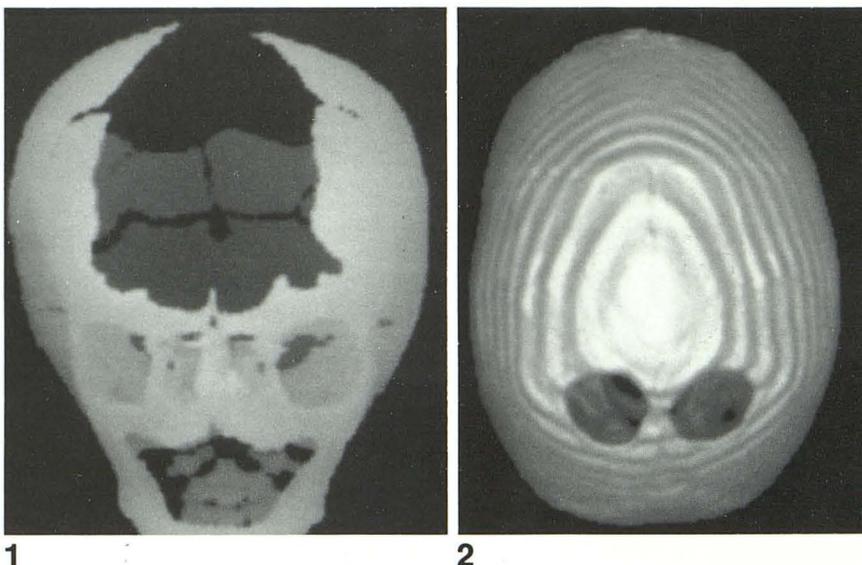
Fig. 1. Cranium bifidum; median cleft face syndrome; Sedano facies D; 3-month-old girl. Anterior view of a three-dimensional bone surface reformation shows deficient ossification of the entire frontal midline and most of the parietal midline.

Fig. 2. Large symmetrical parietal foramina; 12-year-old boy. Vertex view of a three-dimensional bone surface reformation.

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progressively after birth, typically close by 12–18 months (or earlier), and persist into adulthood only rarely (<1%) (27).

In some patients, delay in the ossification of the anterior or posterior parietal bones leads to large anterior, posterior, or confluent frontoparietal defects that persist into early childhood. This is cranium bifidum (Fig. 1) (29, 30). The underlying brain is covered by a fibrous membrane made of dura and pericranium (31). The overlying scalp is usually intact (31). With time, continued ossification of the midline membrane may divide the single defect into two symmetrical frontal or parietal foramina at perhaps 6–8 years of age. These foramina may then close progressively, or persist (Fig. 2).

In approximately 60% of the population, foramina persist in one or both parietal bones as small, approximately 1 mm, ostia to transmit the emissary veins that connect the epicranial veins to the superior sagittal sinus. These are normal. In approximately 1 person in 25,000, the foramina persist as large full-thickness defects in the frontal or parietal calvarium (Fig. 2). Such persistently large foramina appear in families and may show autosomal dominant inheritance (27). They are so common in the Catlin family that they may be known medically by the eponym "Catlin Mark" (32). Since individuals in the same family may exhibit defects at either frontal or parietal locations, the variability in the location of the foramina appears to represent genetic heterogeneity (27).

Clinically, patients with cranium bifidum manifest "soft spots" at the calvarial defect(s). The overlying tissue shows slightly corrugated thick-

ening and may bulge outward when the child cries. Stroking or pressure at the defect may cause unusual pains and severe headaches (32). Vomiting and seizures are rare (31). Mentation is normal. No herniation of brain, no spina bifida, and no other gross cerebral anomaly has been reported with this condition. Midline cranium bifidum may be associated with cleidocranial dysplasia, median cleft face syndrome (Fig. 1), and aminopterin fetopathy (31). Some studies show a slight association with cleft lip and cleft palate.

Exencephaly

Exencephaly is defined as acrania with persistence of a substantial portion of the CNS (Fig. 3) (33–37). It is a rare condition believed by some to be an early form of anencephaly, that, given time, would evolve to frank anencephaly by progressive destruction of the protruding neural tissue. The precise etiology and epidemiology are unknown.

In exencephaly, the flat intramembranous bones of the vault are absent. The skull base and the facial bones are typically preserved. The exencephalic brain is covered by a highly vascular epithelium (33, 34) that may be intimately related to the residual cerebral tissue (35, 36). The dura may fuse with an underlying loose connective tissue that contains tortuous vascular channels and foci of dysplastic neural elements (33). The subarachnoid "space" is obliterated (35, 36).

The cerebral remnant is usually midline and extends from the nasion to the inion. Typically, it is a disorganized mass of nervous tissue, blood vessels, fibrous tissue, and fluid-filled spaces.
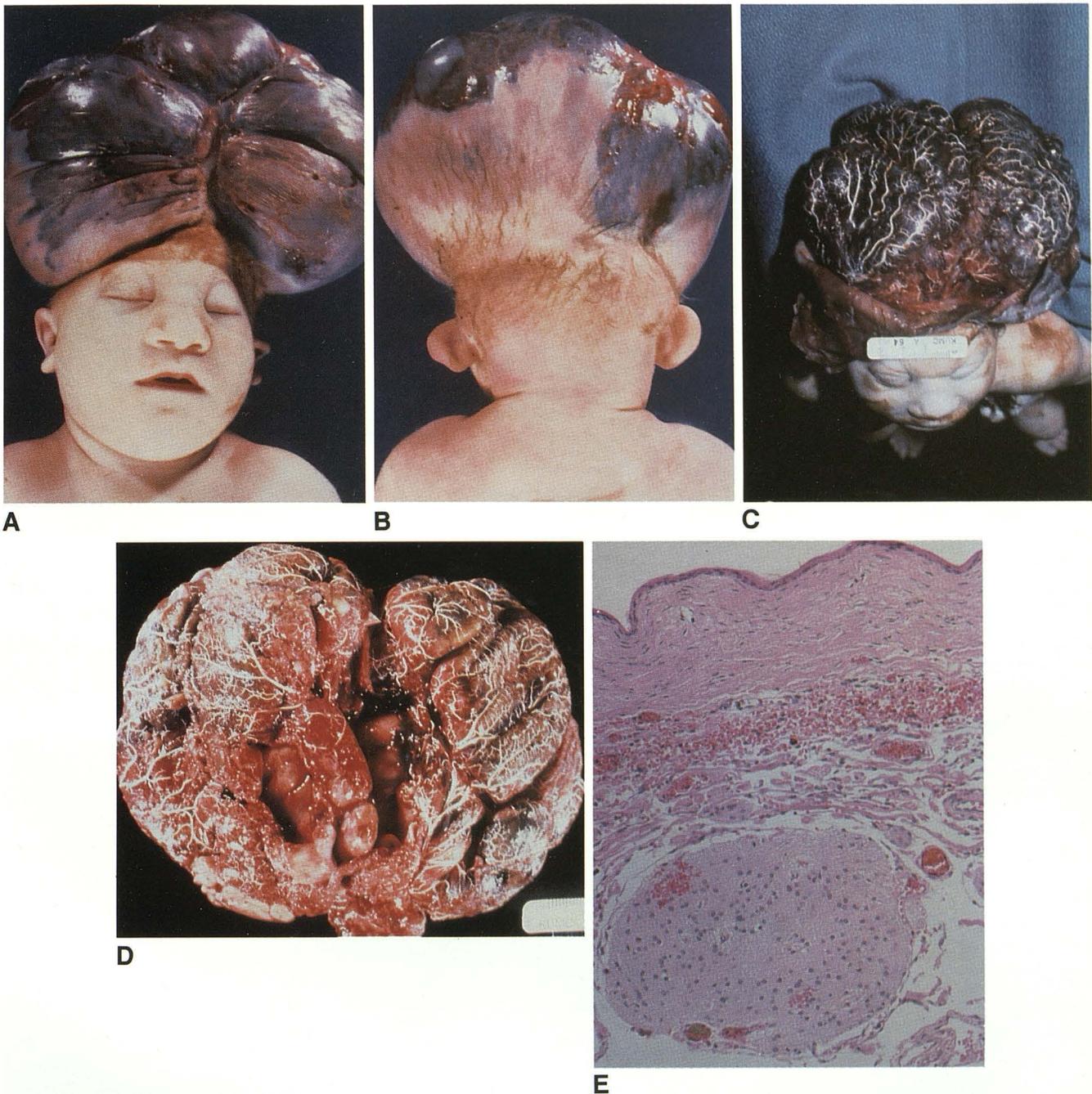
Fig. 3. Exencephaly; Postmortem specimen of a 38-week-gestation, stillborn. (Reprinted with permission from Cox et al (33). Color images and additional material courtesy of Dr John J. Kepes and Dr Michael S. Handler, University of Kansas Medical Center, Kansas City, KA.).

A and *B*, Anterior (*A*) and posterior (*B*) views show a large multilobed exencephaly covered by a highly vascular layer of skin. The face is dysplastic with low-set ears. The flat bones of the cranium were absent.

C, Following intravascular injection of dilute barium, reflection of the skin revealed the outer surface of a 2 to 4-cm thick rind of soft connective tissue that occupied the expected position of the leptomeninges and subarachnoid space—a dermal meningeal membrane.

D, Removal of the meningeal rind revealed the posterior surface of roughly symmetrical cerebral hemispheres with flattened gyri, shallow sulci, and foci of hemorrhage. The ventricular system is open and displays a single chamber lined by numerous multilobular soft-tissue masses. No normal midline structures could be identified. The cerebellar tissue protruded as a single dysplastic bulb to the right of the medulla.

E, Photomicrograph (×100) of the dermal meningeal membrane oriented with skin at the top reveals a loose connective tissue containing numerous tortuous vascular channels and foci of dysplastic neural elements.
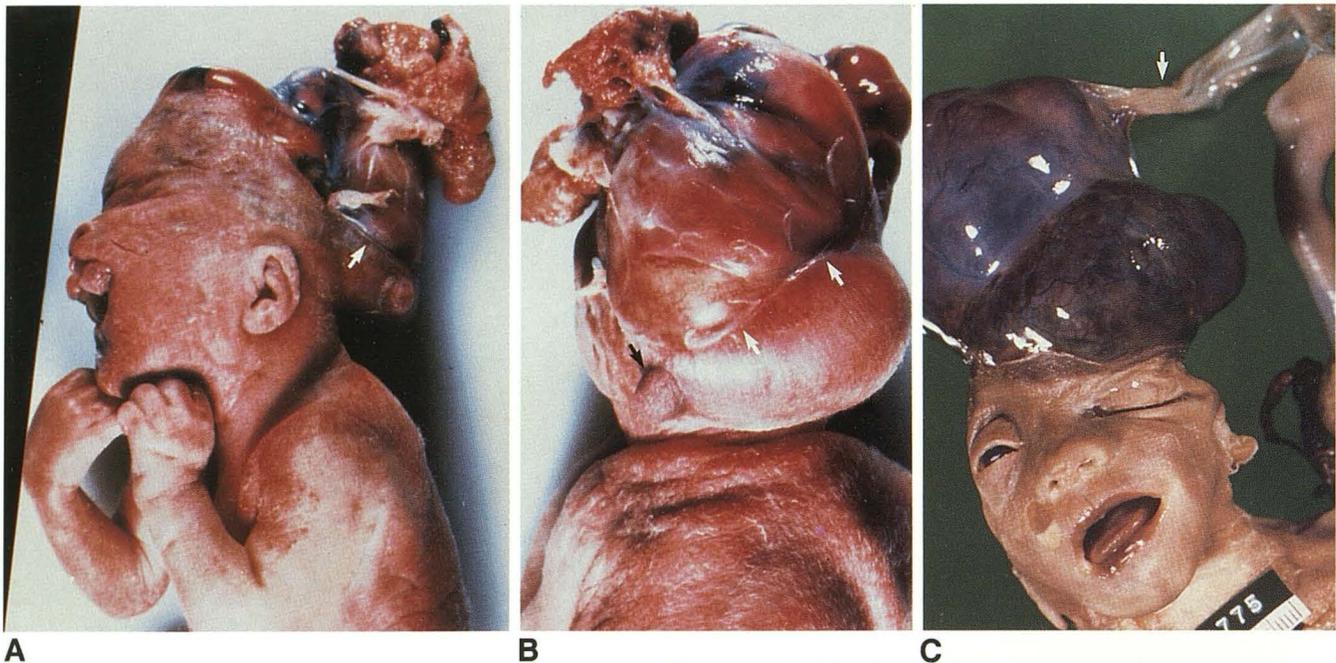
A **B** **C**

Fig. 4. Syndrome of amnionic bands; two specimens. (Cases courtesy of Dr Ellsworth C. Alvord, Jr., Seattle, WA.)

A and *B*, Lateral (*A*) and posterior (*B*) views of a stillborn show acrania, substantial residual dysplastic cerebral and meningeal tissue, and constriction lines (*white arrows*) at the site of crossing bands. A small papilloma lies along the lower margin of the crease (*black arrow*). There is facial-palatal clefting.

C, In this specimen, the exencephaly is continuous superiorly with an amnionic band (*arrow*). The face and orbits are dysplastic.

Fig. 5. Syndrome of amnionic bands with encephalocele. (Reprinted with permission from Naidich et al (5).)

A, Facies. Lateral view of a 12-year-old mentally retarded girl shows a long, thin band-like scar extending across the scalp and face from the temporoparietal region through the cheek and corner of the mouth to the lower lip. The large posterior zone of atrophic skin and absent hair bulged laterally and displaced the ear inferiorly.

B, Noncontrast axial CT reveals a lateral cephalocele underlying the atrophic skin at the asterion.

A **B**
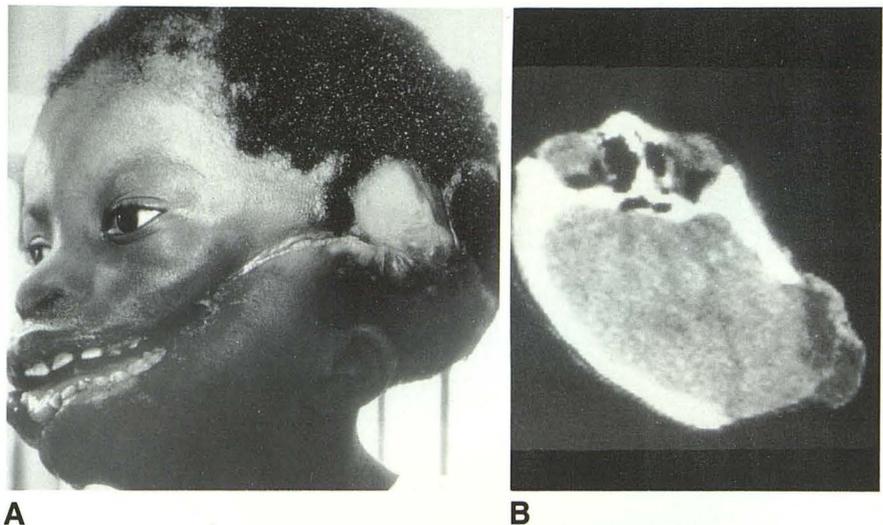
Shallow grooves (pseudosulcations) and "gyri" are seen, but midline structures are rare. Sections through the mass of neural tissue may show a large irregular central chamber lined by numerous multilobular soft-tissue masses without normal midline structures (33) or just a degenerative mass extending deeply (35). Some cases show little or no neuronal differentiation or cortex (35, 36), but others show a "cortex" 2-cm thick with a chaotic arrangement of neurons, neuroblasts, and glial elements (35).

In animal models of exencephaly (35), the basal ganglia, cerebellum, and brain stem are hyper-myelinated and have overdeveloped neuronal cells.

Exencephaly may result from the syndrome of amnionic bands (Fig. 4). Indeed, Hendricks (34) observed amnionic bands in two of four cases of
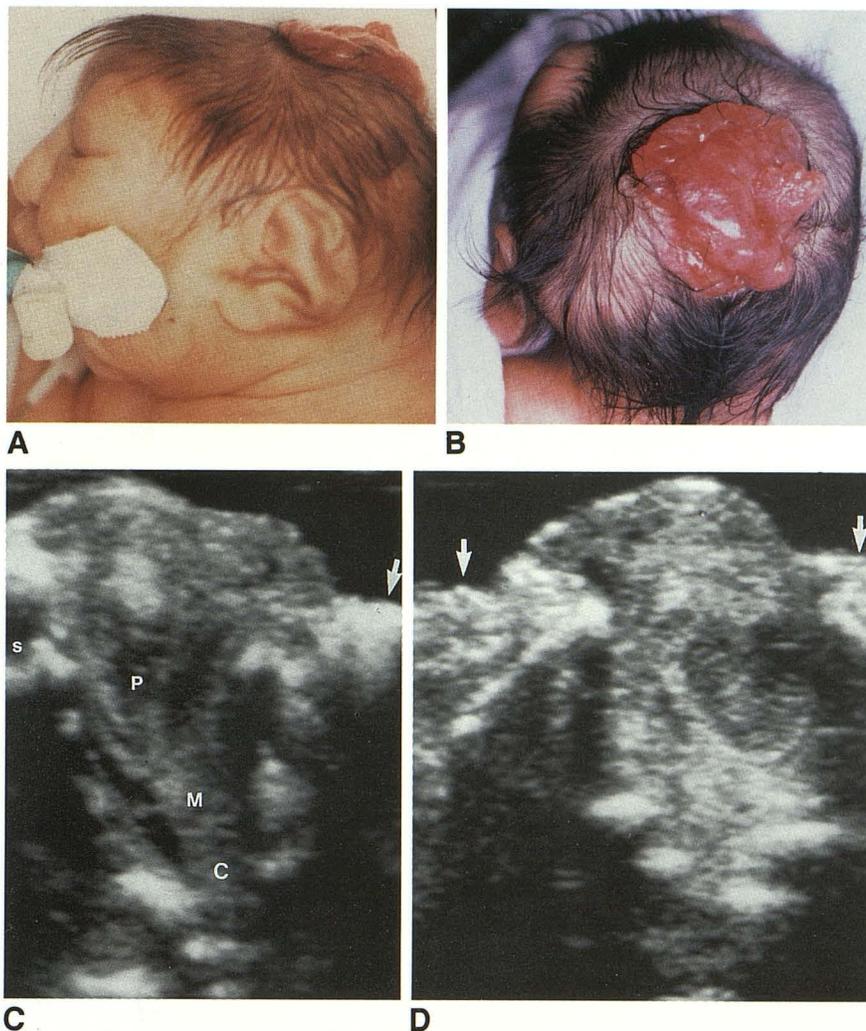
A

B

C

D

exencephaly (one of which had amputated digits as well).

The term *syndrome of amnionic bands* refers to a disruption of normal growth and development resulting from adhesions between the amnion and the affected part, or from constriction of the affected part by a crossing amnionic band. The band may tether a structure, constrict it, "slash" it, or amputate it. Papillomas may arise from the skin near to the attachment of the amnionic band.

Amnionic bands are rare in neonates at term, but are not uncommon in fetuses aborted in the early months of pregnancy (38, 39). The bands may be fine, thread-like strands extending between the fetus and the normal-appearing amnion, or the amnion may be drawn up off the chorion into a broad elongated sheet with its apex attached at the digits, the body, or the head (38). Amnionic bands are believed to be responsible

for many cases of exencephalus, "nonanatomic" encephaloceles, and unusual facial clefts (Fig. 5) (34, 40).

Anencephaly

Anencephaly is observed in 0.5–2.0 per 1000 live births, and accounts for approximately 30% of all major abnormal live births (41). Its incidence is increased in diabetic pregnancies (42). Females are affected predominantly, in a ratio of 3:1 to 4:1. The neonates are stillborn or live only a few days (43). The etiology of anencephaly is unknown. However, anencephaly, cephalocele, and spina bifida occur with increased frequency in the same extended kindreds, suggesting that the pathogenesis may be related, in part, to defects in neural tube closure. Perhaps defective neural tube closure leads to exencephalus which eventuates in anencephaly.

Fig. 7. Anencephaly; complete craniospinal rachischisis. Frontal (*A*) and posterosuperior (*B*) views of this stillborn display absence of the calvarium, convexity of the skull base with poor separation of the anterior and middle cranial fossae, the funnel-shaped posterior fossa leading directly into the wide holospinal spina bifida, continuity of the skin over the supraciliary ridges onto the superior surface and virtual absence of any residual neural tissue. The shallow orbits cause protrusion of the globes. The tongue is relatively large. The upper extremities are characteristically elongated and there are bilateral club feet.

A   **B**

Anencephaly is characterized by absence or underdevelopment of the cranial vault and maldevelopment of the skull base (Figs. 6 and 7). The frontal bones superior to the supraciliary ridge, the parietal bones, and the squamous occipital bone are deficient. The anterior and middle cranial fossae are poorly defined and convex upward. The posterior fossa is usually funnel-shaped.

The lamina cribrosa is imperforate (41). The sella turcica is small. The anterior lobe of the pituitary gland is present in the sella but may be small (41). The posterior lobe may be absent. Shallow orbits are associated with protruding eyes. The globes elaborate optic nerves that end blindly in the orbits.

The skin of the eyelids extends posteriorly over the supraciliary ridges onto the skull base. The skin from the temple and occiput similarly passes onto the skull base to encircle an amorphous, reddish, highly vascular mass of neural and neuroglial tissue designated the area cerebrovasculosa (41). The tentorium is absent. Any residual meninges usually lie flat against the remaining "brain" (42). The cerebellar hemispheres are absent.

The spine is always abnormal in anencephaly (41). Segmentation anomalies are common. The cervical vertebrae may be reduced in number and bifid posteriorly. Wide spina bifida may extend the length of the spinal column as a complete craniospinal rachischisis. The spinal cord may be absent or dysplastic.

Histologically, the area cerebrovasculosa contains predominantly glia with scattered disorganized neuronal elements, ependymal nests, and, perhaps, choroid plexus. Disorganized remnants of cerebellar cortex may also be present. These are enclosed in a vascular connective tissue which represents the leptomeninges and the primitive meningeal vascular plexus (41).

Cephaloceles

Classification

Cephaloceles are classified, in part, by the site of the cranial defect through which herniation occurs (Fig. 8).

Cephaloceles are designated (44):

A. Occipito-cervical, if the skull defect involves the occipital bone, foramen magnum and the posterior arches of C1, C2, etc.

B. Occipital, if the defect lies between foramen magnum and the lambda.

C. Parietal, if the defect lies between the lambda and the bregma.

D. Lateral, if the defect lies along the coronal or lambdoid sutures as far inferiorly as the

anterolateral and posterolateral fontanelles.

E. Interfrontal, if the defect lies between the bregma and the nasal bones.

F. Temporal, if the defect lies along the superior surface of the petrous pyramid.

G. Fronto-ethmoidal (synonym: sincipital), if the defect lies between the nasal bones and the ethmoid bone.

H. Spheno-orbital, if the ostium for the herniation involves the optic foramen, the superior orbital fissure, or a defect in the orbital wall.

I. Spheno-maxillary, if the ostium for the herniation extends through the superior orbital fissure *and* the inferior orbital fissure into the pterygopalatine fossa.

J. Nasopharyngeal, if the defect lies within the ethmoid, sphenoid, or basioccipital bones.

These lesions are subdivided in diverse ways.

For example, occipital cephaloceles are designated *inferior occipital cephaloceles* if the midline defect in the occipital bone lies inferior to the internal and external occipital protuberances, and *superior occipital cephaloceles* if the defect lies above the external occipital protuberance and above the tentorial attachment. Similarly, the nasopharyngeal cephaloceles are divided into transethmoidal, sphenoethmoidal, transsphenoidal, and transbasioccipital forms by the specific site of the ostium. One common classification is summarized by Nager (45) (Table 1).

Overview of Cephaloceles

Epidemiology

Cephaloceles typically present at birth and usually come to medical attention within the first

A

B

C

D

Fig. 8. The spectrum of cephaloceles.

A, Occipital cephalocele; newborn.

B, Posterior parietal encephalocele; 2-week-old boy with large herniation, microcephaly, thinnings of the sac (*white arrow*), erosions of the dome, and increasing size of the sac with time.

*C*, Anterior parietal encephalocele; note the hair at the base and the thinning and erosion at the dome.

D, Interfrontal cephalocele, situated cephalic to the nasofrontal suture. (Case courtesy of Dr Francisco Arredondo and Dr Francisco de Leon, Guatemala City, Guatemala.).

(Fig. 8 continued on next page.)

Fig. 8—*continued. E,* Skin-covered fronto-ethmoidal meningoencephalocele; nasofrontal type. 1-month-old girl with portions of both frontal lobes in the sac.

F, Membrane-covered fronto-ethmoidal meningoencephalocele, nasofrontal type; newborn infant. (Case courtesy of Dr Francisco Arredondo and Dr Francisco de Leon, Guatemala City, Guatemala.)

G, Lateral frontal meningoencephalocele, status post-shunting. Newborn boy with lateral forehead cleft and microphthalmos. The hydrocephalic frontal lobe herniated into the orbit through a defect in the orbital roof.

E

F

G
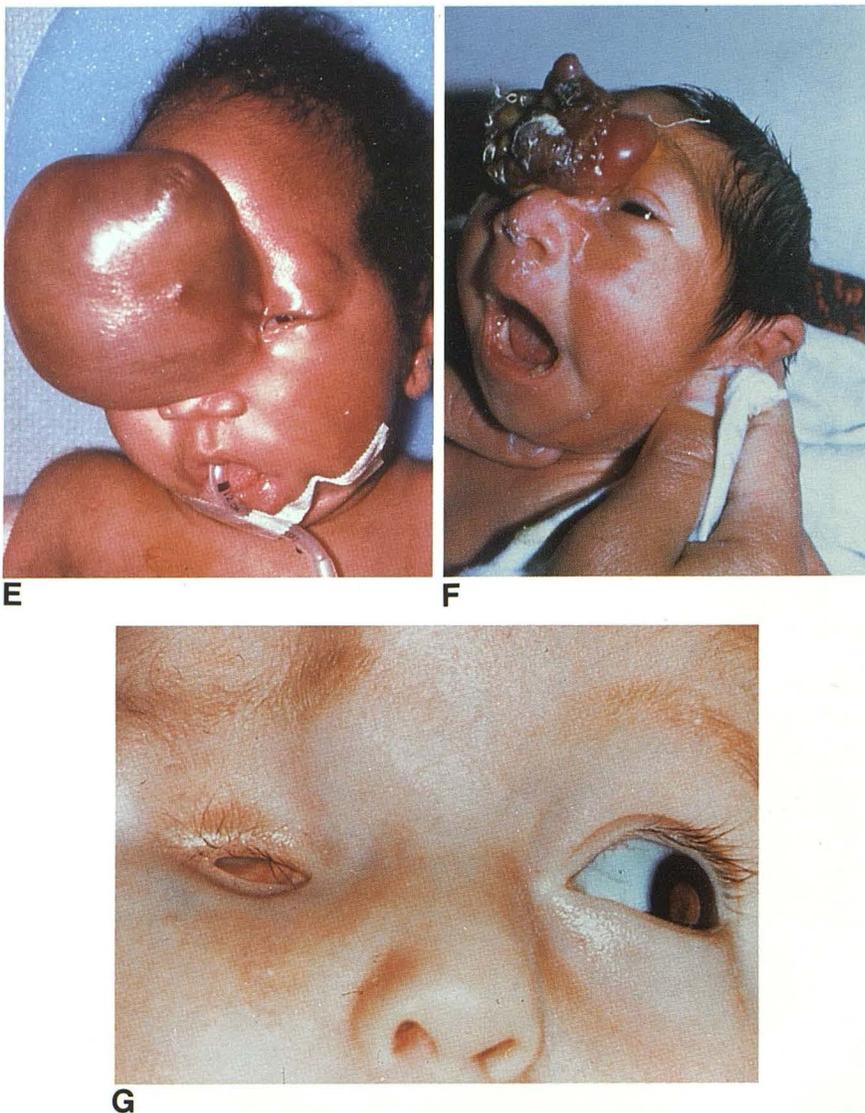
week of life. Occasional lesions—especially basal cephaloceles—escape detection for years and first present in adulthood (46).

The incidence of cephaloceles is approximately 0.8–3.0 per 10,000 births (41, 44, 47). Thus cephaloceles are approximately 10 times less common than myelomeningoceles. Mealey et al (46) found 60 encephaloceles and 559 myelomeningoceles among 76,280 admissions to a children's hospital over a 20 year period. The encephaloceles accounted for approximately 0.07% of all pediatric admissions, and about 10.3% of all craniospinal malformations seen in this period.

The different types of cephaloceles show strong geographic variation, substantial differences in incidence and form with race and sex, and different associations with neural tube defects: among white populations in Europe and North America, occipital cephaloceles constitute 66%–89% of all cephaloceles (44, 48). In Southeast Asia, sincipital cephaloceles are more common (49). Among white Australians, 66.7% of cephaloceles are occipital and only 2.2% sincipital, whereas among aboriginal Australians, 50% of cephaloceles are sincipital (44). Igbos Nigerians have a similar, disproportionately high incidence of sincipital cephaloceles. These variations suggest that at least some cephaloceles are genetic in origin and that the different types of cephaloceles may represent different genetic defects. Table 2 documents the variable incidences of different types of cephaloceles among patients from different geographic locations.

Sex data indicate that occipital cephaloceles occur more commonly in women, whereas parietal cephaloceles and sincipital cephaloceles

TABLE 1: Classification of cephaloceles

Cephaloceles involving the convexity
  A. Cervico-occipital cephalocele
  B. Inferior occipital cephalocele
  C. Superior occipital cephalocele
  D. Parietal (sagittal) cephalocele
  E. Lateral cephalocele
    1. Coronal and pterional
    2. Lambdoidal and asterional
  F. Interfrontal cephalocele

Cephaloceles involving the skull base
  G. Temporal
  H. Fronto-ethmoidal (sincipital) cephalocele
    1. Naso-frontal type
    2. Naso-ethmoidal type
    3. Naso-orbital (anterior orbital) type
  I. Spheno-orbital (posterior orbital) cephalocele
  J. Spheno-maxillary cephalocele
  K. Nasopharyngeal cephalocele
    1. Transethmoidal type
    2. Spheno-ethmoidal type
    3. Transsphenoidal type
    4. Basioccipital type

Note.—Adapted from Nager (45).

show male predominance (22, 44, 46, 50). The male to female ratios are 1/(2.0–2.7), approximately 2:1, and approximately 1.5/1, respectively.

Occipital and parietal cephaloceles appear to be related to neural tube defects. In diverse series, 7% of children with occipital encephalocele show concurrent myelomeningocele (Fig. 9) (46). Three percent of patients with (cervico)occipital cephalocele have diastematomyelia (44) and 3% have a second cephalocele in the parietal region (44). In Simpson's series, two unrelated patients (5%) each had sibs with neural tube defects: one anencephaly; the other an occipital meningoencephalocele (44). Similarly, parietal cephaloceles are associated with the Chiari II malformation in 7%–

33% of cases (25, 51). Sincipital cephaloceles show no association with neural tube defects.

There is no apparent relationship to maternal aging. In 42 large sibships, the encephalocele had no tendency to appear in any specific sibs (50). However, cephaloceles were relatively uncommon in the firstborn and in sibs numbering six or higher.

The relative incidences of cranial meningoceles versus meningoencephaloceles vary widely, partly as a function of the type of cephalocele. In our personal experience, cranial meningoceles are very rare. In literature series, Simpson (44) found that 24% of reported cephaloceles were cranial meningoceles and 76% meningoencephaloceles (Table 3). This high incidence of meningocele appears to be exceptional.

Most cephaloceles occur as isolated anomalies, not associated with syndromes, although they may show some concurrent intracranial or extracranial lesions (41). None of Simpson's 74 cephaloceles had an associated syndrome, but the patients had concurrent cleft palate (3%), microphthalmia (1%), corneal opacity (1%), and tracheo-esophageal fistula (1%) (44). Dermoid cysts were included within the herniation in 5 of 74 cases (7%) (44).

A smaller number of cephaloceles form variable parts of known syndromes. Cohen (52) and Cohen and Lemire (53) have detailed the diverse syndromes associated with cephalocele, listing the syndrome, site of cephalocele, frequency of cephalocele in that syndrome, the features of the syndrome, and the genetic pattern of the syndrome (52). Prominent among these is the Meckel syndrome, an autosomal recessive condition characterized by occipital encephalocele (in 80%), holoprosencephaly, orofacial clefting, microphthalmia, retinal dysplasia, polydactyly,

TABLE 2: Relative incidence of diverse cephaloceles

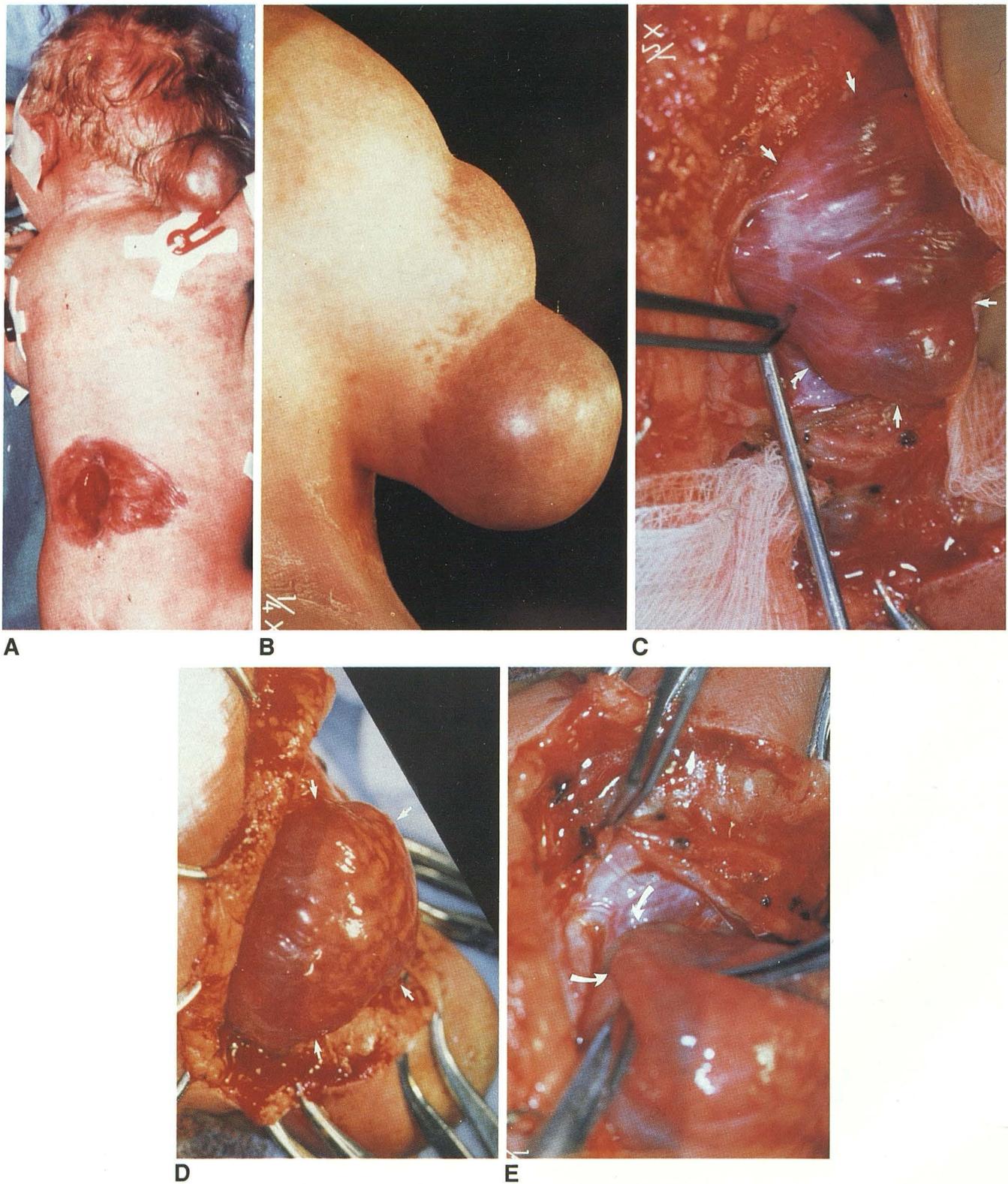|	Series (reference no.)							
	1 (96)	2 (97)	3 (44)	4 (46)	5 (25)	6 (98)	7 (99)	8 (100)
Cephalocele Location	Boston N = 265 (%)	Europe N = 68 (%)	Australia N = 74 (%)	Indiana N = 67 (%)	Japan N = 40 (%)	Western Nigeria N = 36 (%)	Eastern Nigeria N = 5 (%)	Boston N = 84 (%)
Cervico-occipital		11 (16)	2 (3)					
Occipital	196 (74)	34 (50)	34 (46)	55 (82)	14 (35)	25 (69)	2 (40)	63 (75)
Parieto-occipital					4 (10)	3 (8)		
Parietal	34 (13)	6 (9)	13 (18)	3 (4)	15 (38)	1 (3)	1 (20)	9 (11)
Lateral		1 (1)				1 (3)		
Interfrontal	}31 (12)	}16 (24)						}11 (13)
Sincipital			25 (34)	8 (12)	3 (7)	6 (17)	2 (40)	
Nasopharyngeal	4 (2)			1 (1)	4 (10)			1 (1)

Fig. 9. Combined occipital meningoencephalocele and upper lumbar myelomeningocele; 1-day-old girl with an L5-S1 functional level; Uneventful pregnancy and delivery.

A, Posterior view shows the combined lesions. The myelomeningocele was repaired routinely on day 1. Resection of the encephalocele could be deferred because the skin cover was excellent.

TABLE 3: Relative incidences of cranial meningoceles and meningoencephaloceles among 74 mixed cephaloceles

Cephalocele Site	Total Cases	Meningoen	Meningoen-cephalocele
Cervico-occipital	2	2	
Occipital	34	13	21
Parietal	13	3	10
Sincipital	25		25
Total	74 (100%)	18 (24%)	56 (76%)

Note.—Adapted from Simpson et al (44).

polycystic kidneys, cardiac anomalies, and ambiguous genitalia in diverse combinations (53). Table 4 from Cohen (52), summarizes these malformations.

General Pathologic Features

Cephaloceles show variable size, shape, and skin cover. They may be broad-based or pedunculated, tiny nubbins or masses larger than the head (Fig. 10). A few are bilobed (Fig. 10) (50). Their consistency may be cystic to mixed solid-cystic (Fig. 11) (50).

When watched, the lesions may enlarge over time (25%) or disappear following closure of a myelomeningocele and shunting the lateral ventricles (46, 50). Since the intracranial volume is reduced by herniation, children with large cephaloceles tend toward microcephaly. The face is relatively little affected, so cranial-facial disproportion ensues.

Cephaloceles typically have an intact skin cover. Where appropriate to the normal anatomy, hair covers the base and a variable portion of the sac, tending to thin out toward the dome of the sac. The skin usually has no primary defect, but may be thinned and glistening, parchment-like, focally deficient, secondarily ulcerated, or secondarily infected (41, 54). Thinnings and ulcerations (3.5%) tend to occur at the distended dome of the sac (5, 50). Hemorrhage may occur where ulcers erode into vascular structures.

Nervous tissue within the cephalocele connects with the underlying brain. The connection may be a narrow neck of cerebral or glial tissue, or there may be large portions of recognizable brain herniating en block into the sac (41). Other encephaloceles contain disorganized neural tissue, perhaps with fragments of cerebral cortex, islands of glioneuronal tissue containing ependyma, or choroid plexus (41).

Clinical Outcome

In older series of Lorber (55) and Lorber and Shofield (56), approximately 50% of patients died and 76% of survivors were retarded. Significant improvement has occurred since then. At present the clinical success of managing patients with cephaloceles depends heavily on the cephaloceles' site, size and content, and upon any concurrent intracranial malformation (25, 44, 46). For overview of patient prognosis see Tables 5A and 5B.

Specific analysis of reported series (25, 44) indicates the importance of these prognostic criteria. In Simpson's series (44), 17 or 18 patients had meningoceles. All but two of these did well. Twenty patients had recognizable parietal or occipital cortex in the cephalocele. Fifteen of these 20 died or were totally dependent. Another 2 of the 20 had severe intellectual deficit. In 46 cases categorized by size, only two of 26 patients with lesions smaller than 5 cm did badly, whereas 15 of 20 lesions greater than 5 cm did badly. Two of these 15 large cephaloceles were the only two meningoceles that did badly.

Mealey et al (46) operated on 49 occipital cephaloceles. His findings confirm the prognostic importance of the tissue herniating (Table 6). Simpson et al (44) and Yokota et al (22) also provide useful data on clinical outcome (Tables 7 and 8).

Analysis of Cephaloceles by Specific Site

Occipital Cephaloceles

Clinical Findings. Occipital cephaloceles are usually large: >20 cm (16%), 15–20 cm (14%), 10–15 cm (12%), 5–10 cm (30%), and <5 cm (28%)

Fig. 9.—Continued.

B–D, On day 15, the cephalocele was shaved to reveal a double-lobed configuration. Horse-shoe incision around the upper hemicircumference of the lesion was carried deeply to reveal C, a fibrous tissue (white arrows) that resembled dura. This thickened deeply and contained veins at the junction of the superficial with the deeper lobe. Incision through the dura disclosed D, abnormal microgyric cortical tissue (white arrows) filling the deep lobe. The tissue appeared distended by fluid. Incision resulted in gush of CSF with collapse of the dome of cortex.

E, The pedicle of the encephalocele was then traced deeply by careful dissection and cautery to reach the dural-lined bony ostium (curved white arrow). The stalk was transected revealing occipital cortex with an open occipital horn. The ventricle, mantle, and pia were closed appropriately. The dural flap was sealed in watertight fashion and plastic repair was performed on the scalp. At age 25 months, a rib graft was placed to seal the bony ostium.

TABLE 4: Conditions with encephaloceles

Condition	Site(s) of Encephalocele	Reported Frequency of Encephalocele in Condition	Striking Features	Etiology
Syndromes				
Aberrant tissue band syndrome	Multiple, predominantly anterior	Uncommon	Ring constrictions and amputations of digits or limbs, distal syndactyly, irregular or asymmetric encephalocele, microcephaly, microphthalmia, bizarre orofacial clefts, other facial disruptions, tissue bands, various other anomalies	Aberrant tissue bands
Chemke syndrome	Occipital	3/6	Hydrocephaly, agyria, absent cortical laminar structure, cerebellar dysgenesis, retinal dysplasia, corneal opacities, cataracts	Autosomal recessive
Cryptophthalmos syndrome	Occipital	10%	Extension of forehead skin to cover one or both eyes, unusual hairline, ear anomalies, notching of the nasal wings, soft-tissue syndactyly of hands and/or feet, genital anomalies	Autosomal recessive
Dyssegmental dwarfism	Occipital	2/10	Lethal dwarfism, short broad tubular bones with metaphyseal widening, accelerated carpal bone maturation, bowing of legs as well as thighs and forearms, short broad pelvis with widely flared iliac wings, vertebral anomalies, small thorax, cleft palate, micrognathia	Autosomal recessive
Frontonasal dysplasia	Frontal	Constant	Ocular hypertelorism, widow's peak, anterior cranium bifidum occultum, widely set nostrils with lack of elevation of the nasal tip, notching of nostrils, other abnormalities	Most cases sporadic; some familial; probably etiologically heterogeneous
Knobloch syndrome	Occipital	4/5	High myopia, vitreoretinal degeneration, retinal detachment, meningocele, normal intelligence	Autosomal recessive (presumed)
Meckel syndrome	Occipital	80%	Polydactyly, polycystic kidneys, holoprosencephaly, microphthalmia, retinal dysplasia, cardiac anomalies, orofacial clefting, ambiguous external genitalia, other abnormalities	Autosomal recessive
Pseudo-Meckel syndrome	Occipital	?	Arhinencephaly, absent corpus callosum, Arnold-Chiari defect, no evidence of retinal dysplasia, cleft palate, congenital heart defects, accessory spleen, clubfoot, hallucal hammertoes	t(3p +)

TABLE 4—*Continued.*

Condition	Site(s) of Encephalocele	Reported Frequency of Encephalocele in Condition	Striking Features	Etiology
von Voss syndrome	Occipital	2/2	Aplasia of corpus callosum, hypoplastic olives and pyramids of the medulla oblongata, phocomelia, urogenital anomalies, thrombocytopenia	?
Warfarin syndrome	Occipital	Uncommon	Nasal hypoplasia, bone stippling, limb shortening, low birth weight, optic atrophy, mental retardation, seizures, hydrocephaly	Warfarin during pregnancy
Associations				
Encephalocele/absent corpus callosum	Parietal Transsphenoidal Other	4/13 3/10	Absent corpus callosum; optic nerve abnormalities may occur with transsphenoidal encephalocele; encephalocele/absent corpus callosum association can occur with various syndromes including holoprosencephaly	
Encephalocele/clefting	Transsphenoidal Other	4/8	Cleft lip, cleft palate, both cleft lip and cleft palate; less commonly oblique facial cleft; common and bizarre orofacial clefts can occur with various syndromes	
Encephalocele/craniostenosis	Occipital Frontal	2/20 Rare	Craniostenosis; association can occur alone or with other anomalies and syndromes	
Encephalocele/Dandy-Walker; Arnold-Chiari	Occipital Parietal	Uncommon Uncommon	Dandy-Walker defect or Arnold-Chiari defect or both	
Encephalocele/ectrodactyly	Occipital	2/2	Ectrodactyly	?
Encephalocele/hemifacial microsomia	Occipital	Uncommon	Unilateral dysplastic ear, ear tags and/or pits, unilateral hypoplasia of mandibular ramus, various other anomalies especially ocular, skeletal, and cardiac defects	Most cases sporadic; some familial; probably etiologically heterogeneous
Encephalocele/hypothalamic pituitary dysfunction	Transsphenoidal	?	Different patterns of hypothalamic/pituitary dysfunction; sometimes optic nerve abnormalities, ocular hypertelorism, cleft lip/palate	Presumed embryonic secondary
Encephalocele/iniencephaly; Klippet-Feil	Occipital	Constant with iniencephaly apertus; rare with Klippel-Feil anomaly	Iniencephaly apertus, Klippel-Feil anomaly, various other anomalies	?
Encephalocele/meningomyelocele	Occipital Frontal	Rare	Meningomyelocele	?

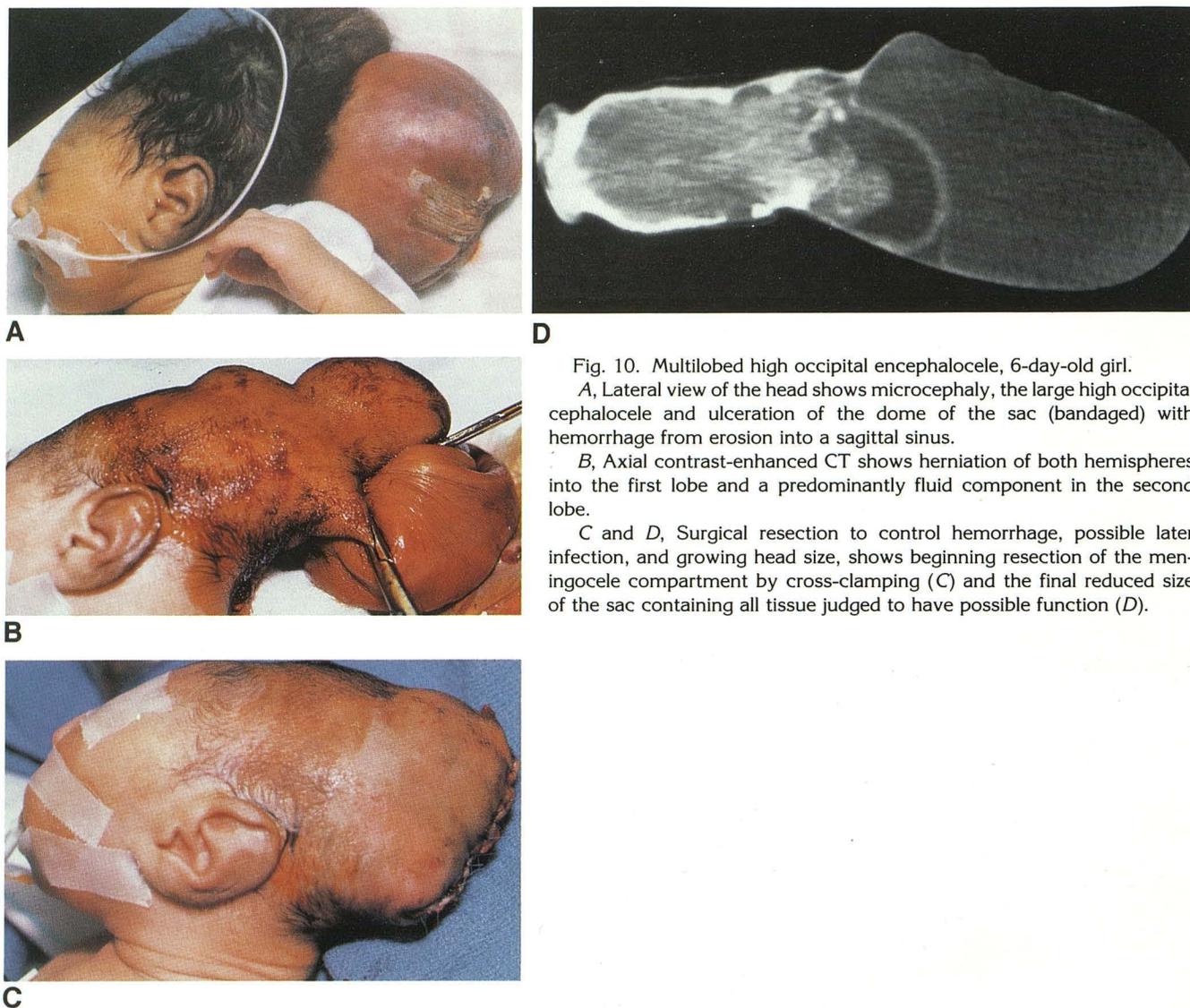Note.—Adapted from Cohen (52).

A

D

B

C

Fig. 10. Multilobed high occipital encephalocele, 6-day-old girl.

A, Lateral view of the head shows microcephaly, the large high occipital cephalocele and ulceration of the dome of the sac (bandaged) with hemorrhage from erosion into a sagittal sinus.

B, Axial contrast-enhanced CT shows herniation of both hemispheres into the first lobe and a predominantly fluid component in the second lobe.

C and D, Surgical resection to control hemorrhage, possible later infection, and growing head size, shows beginning resection of the meningocele compartment by cross-clamping (C) and the final reduced size of the sac containing all tissue judged to have possible function (D).

(Fig. 9) (50). The incidence of microcephaly varies from 9% to 24% (44, 50). At the time of presentation, 80% of patients had no apparent neurologic deficit. Eleven percent had spastic paraparesis, 2% were blind, and 7% had developmental delay (50). Hydrocephalus may result from a Chiari III malformation or aqueductal stenosis (54). The incidence of hydrocephalus varies from approximately 20% to 65% (44, 57), and is higher with occipital meningoencephaloceles (65%), than occipital meningoceles (19%) (55, 57).

Contents of the Sac. The contents of the occipital meningoencephalocele sac vary widely. No unifying concept presently enables us to predict sac content on the basis of any other findings. The following statistics and cases have been selected to convey the diversity of pathology one may expect to encounter in this condition. Thus:

1. Simpson (44) found that 37% of occipital meningoencephaloceles contained cerebral tissue but no cerebellar tissue; 21% had cerebral and cerebellar tissue (Fig. 12); 5% had cerebellar tissue only (Fig. 13), and 37% had nodules of glia or other dysplastic neural tissue.

2. In five cases, Leong and Shaw (58) found the sac contained 1) only choroid plexus connected to the diencephalic roof; 2) a herniated portion of the diencephalic roof, a pedicle of mesencephalon, and the right temporal lobe; 3) both occipital lobes and the entire cerebellum; 4) the entire telencephalon posterior to the central sulcus with a poorly developed brain stem and

TABLE 7: Clinical outcome of 68 cephaloceles

Site	Type	Normal or Slight Disability	Significant Mental Deficit	Died or Totally Dependent	Uncertain	
Cervico-occipital	Meningocele	1	1			2
Occipital	Meningocele	11			1	12
	Meningoencephalocele	10	1	9	1	21
Parietal	Meningocele	3				3
	Meningoencephalocele	1	1	6	2	10
Sincipital	Meningoencephalocele	12	5	1	2	20
Total		38 (56%)	8 (12%)	16 (24%)	6 (9%)	68

Note.—Adapted from Simpson et al (44).

TABLE 8: Clinical outcome of 33 cephaloceles

Cephalocele Type	Cephalocele Location	Total Cases	Outcome		
			Normal	Educable	Died or Retarded
Encephalocele	Occipital	8	2 (25%)	0	6 (75%)
	Parietal	4	0	0	4 (100%)
Meningocele	Occipital	5	4 (80%)	0	1 (20%)
	Parietal	6	2 (33%)	2 (33%)	2 (33%)
Atretic	Occipital	5	5 (100%)	0	0
	Parietal	5	0	0	5 (100%)
Total		33	13 (39%)	2 (6%)	13 (55%)

Note.—Adapted from Yokota et al (25).

metrically herniating structures draw the attached brain toward the ostium, so the residual intracranial content is rotated, elongated posteriorly, and shows progressive constriction toward the hernia orifice. The tissue appears "banded" at the orifice and then re-expands within the sac (Fig. 12C). The rotation and constriction force the remaining intracranial structures to assume new locations and relationships with respect to the bony anterior, middle and posterior fossae, and with respect to the dural partitions. Frequently, there is "skew" deviation of the hemispheres such that, for example, one occipital lobe herniates externally into a midline posterior ostium drawing its own hemisphere posteriorly and medially, while the contralateral hemisphere moves anteriorly and across the midline to occupy both anterior fossae. The middle fossae become occupied predominantly by frontal lobes, while the temporal lobes come to lie posterior to the petrous ridges superior to the low set tentorium. The temporal lobes may even penetrate below the residual tentorium to lie in the usual position of the pons (47).

The convolutional pattern of the brain is grossly normal, after allowance is made for secondary displacements and distortions (47). Pach-ygyria and agyria are usually absent. Samples of isocortex are normal, except that the herniating tissue may show ischemic changes due to strangulation of the blood vessels at the hernia neck. There may be extensive new and old infarcts and occasional hemorrhage (41). The cerebral tissue at the hernia neck may be converted into a glial scar.

The hippocampal allocortex may be abnormal, with variable degrees of faulty convolution, inversion or absence of the dentate fascia, disorganization of the pyramidal cell layer, and, in 80%, rudimentary to absent amygdalae (47). There may be microgyria of the medial temporal cortex.

Deep Structures. The optic nerves are often stretched, post-fixed, kinked, atrophic, or barely identifiable (47). The mammillary bodies are abnormal in 60%, and may be absent entirely, absent unilaterally, or distorted (47).

The corpus callosum is usually present, but can be partly or completely absent. The anterior commissure, septum pellucidum, and fornices are absent in 80% of cases, and partially absent in many others (47).
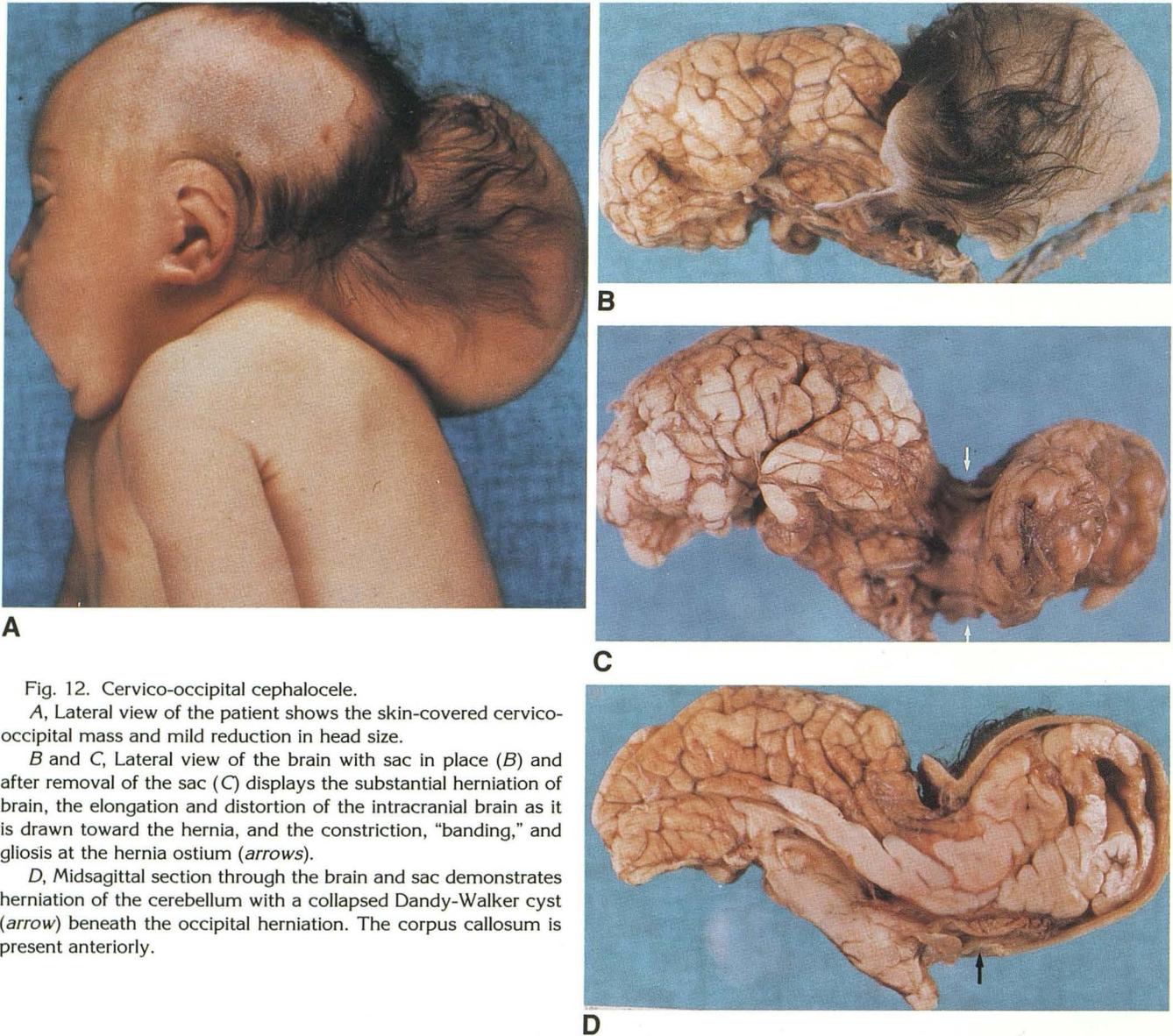
A

B

C

D

Fig. 12. Cervico-occipital cephalocele.

A, Lateral view of the patient shows the skin-covered cervico-occipital mass and mild reduction in head size.

B and C, Lateral view of the brain with sac in place (B) and after removal of the sac (C) displays the substantial herniation of brain, the elongation and distortion of the intracranial brain as it is drawn toward the hernia, and the constriction, "banding," and gliosis at the hernia ostium (arrows).

D, Midsagittal section through the brain and sac demonstrates herniation of the cerebellum with a collapsed Dandy-Walker cyst (arrow) beneath the occipital herniation. The corpus callosum is present anteriorly.

The basal ganglia are usually normal, except for the distortion and asymmetry resulting from unequal herniation of the two hemispheres (47). The thalami are usually normal except for occasional fusion across the midline (20%) and occasional herniation of the thalamus partway into the sac (20%). The substantia innominata is well preserved. In 40%, it is abnormally prominent (47).

Ventricles. Hydrocephalus may affect the entire ventricular system or be limited to the extracranial portion of the ventricles (62, 63). The ventricles typically show distortion and compression. The walls tend to fuse wherever they come into apposition and may be densely gliotic (47). Abnormal communications occur between the lateral and third ventricles, or between the lateral ventricle and the interhemispheric fissure (47).

Masses of aberrant neural tissue may extend out from the brain into the third ventricular cavity (47). Most often this aberrant tissue is in the form of strands or bundles that lie free in the ventricular cavities. Occasional bands or islands of similar tissue may be seen in the neighborhood of the brain stem and cord (47). Islands of malformed neuroectodermal tissue may also be found in the leptomeninges, associated with abundant sinusoidal capillaries, consistent with fetal leptomeningeal vascularization (41).
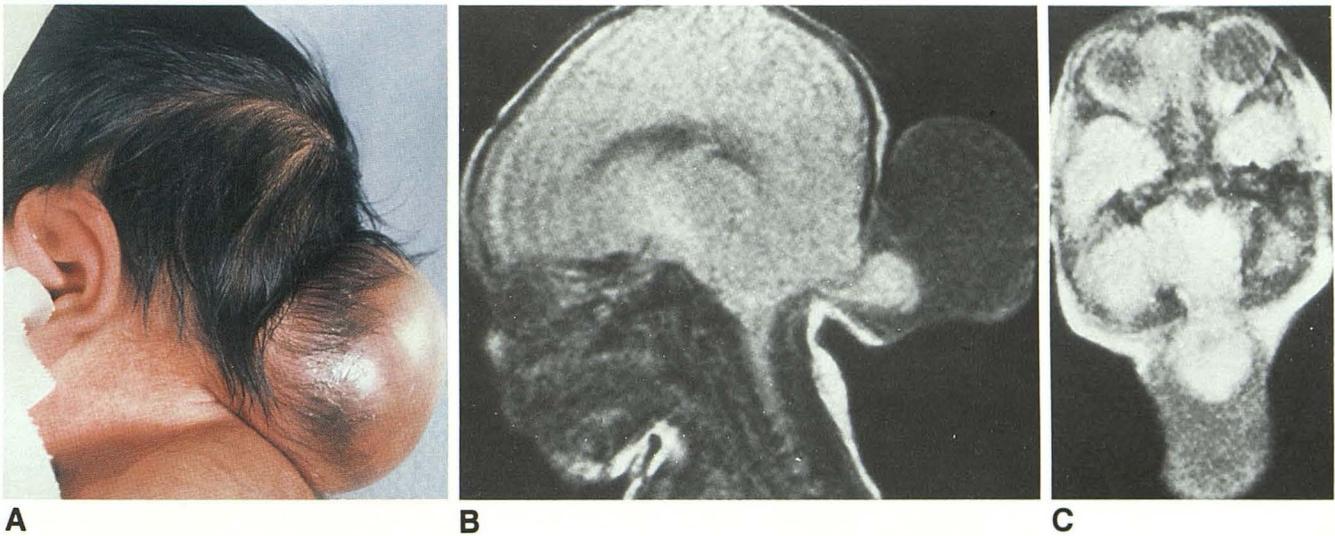
A   B   C

Fig. 13. Occipital meningoencephalocele; 3-day-old girl with a defect in the squamous occipital bone.

A, Lateral view of the patient reveals an 8-cm cephalocele with hair at the base and thinner skin at the dome, resembling a tonsure. The skin of the back recurves onto the inferior surface of the sac with a characteristic hairpin turn.

B and C, T1 MR; sagittal (B) and axial (C) sections show the high signal subcutaneous fat recurving from the back onto the inferior surface of the cephalocele and then becoming deficient around the dome. The cephalocele is predominantly fluid-filled, but contains tissue that is directly continuous with the vermis through the bony ostium.

D, At surgery, the CSF within the sac was slightly xanthochromic. The cerebellar vermis (white arrows) and the medial portions of the hemispheres were evident in the depth of the sac and appeared "healthy." No tissue was resected. Rather the sac was reduced in size to accommodate the viable tissue. A ventriculo-peritoneal shunt was placed at age 2 weeks.
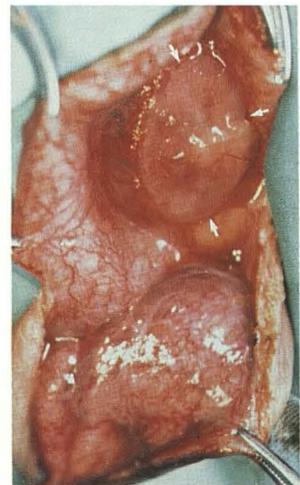
D

Hindbrain. In detailed studies of five occipital encephaloceles, Karch and Urich found that only one of the 10 cerebellar hemispheres was even moderately preserved (47). No cerebellar tissue could be identified in two of five cases (40%), and only herniated cerebellar rudiments were seen in three (60%). The residual cerebellar tissue showed densely gliotic folia devoid of neurons. The vermis was absent in all five cases (100%). The two hemispheres were separated by a midline cleft bridged by a thin glial membrane. Dandy-Walker malformation has been seen in 6% of low occipital meningoceles (44) and 17% of occipital cephaloceles (59, 64).

The brain stem may lie entirely intracranial, herniate partially, or herniate nearly completely with the hindbrain. It typically shows an S-shaped distortion: 1) The midbrain courses posteriorly and inferiorly, and usually shows partial fusion of the tectum. The aqueduct may be replaced by a midline cleft that divides the stem into asymmetrical halves. 2) The poorly differentiated pons and medulla recurve anteriorly. As a consequence, the floor of the fourth ventricle that should lie dorsal superior to the brain stem recurves anteriorly with the stem and thereby comes to lie posterior-inferior to the recurved stem. It then forms the inferior surface of this part of the stem. Frequently, cleft-like fissures extend from this surface into the stem. 3) The caudal brain stem then again curves posteriorly toward foramen magnum to complete the S.

In these cases, the residual rudimentary cerebellar tissue tends to lie ventral and lateral to the stem. An abnormal plexus of vessels, especially venules, surrounds the stem.

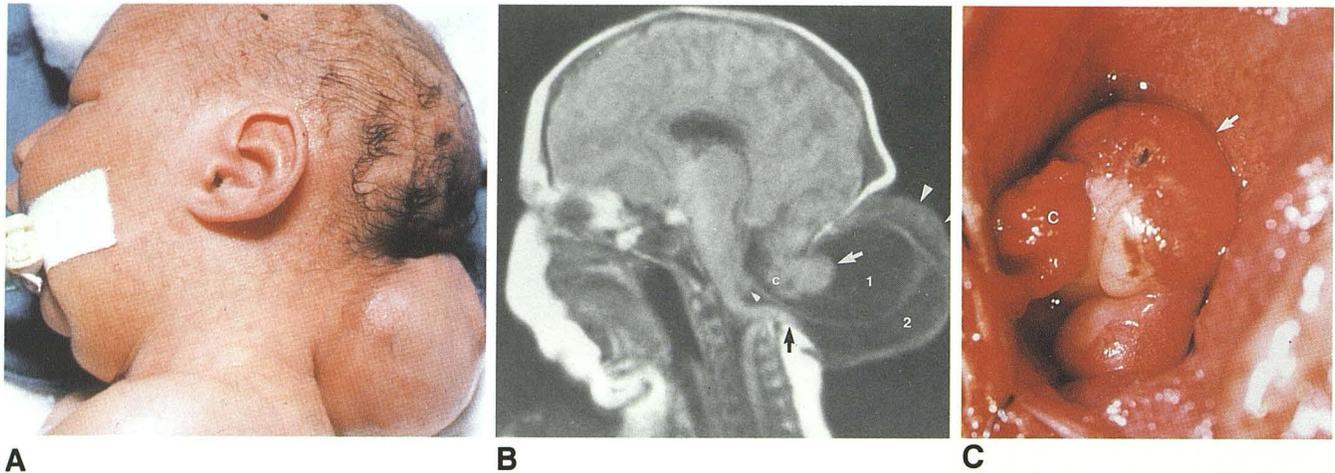**A**   **B**   **C**

Fig. 14. Cervico-occipital meningoencephalocele; 4-day-old girl with a 2 × 4 cm skin-covered cephalocele. The child became apneic and respirator-dependent following birth.

*A*, Lateral view shows the low cervico-occipital position of the cephalocele.

B, Sagittal T1 MR demonstrates the cervico-occipital bone defect with posterior spina bifida, herniation of the occipital lobe (*white arrow*) into a fluid compartment (*1*) through a defect in the tentorium, a nubbin of choroid plexus (*C*) that could be traced to the lateral ventricle on the serial sections, dysplasia of the cerebellum (*arrowheads*) in the midline and at the superior surface of the sac related to a second fluid compartment (*2*), and sharp S-shaped kinking (*black arrow*) of the cervicomedullary junction. It was believed that compartment *1* was an occipital horn ventriculocele containing choroid plexus and occipital lobe. Compartment *2* was the cyst of a Dandy-Walker malformation with the cerebellar hemispheres (*large arrowheads*) displaced laterally and superiorly, the vermian remnant (*small arrowhead*) displaced anteroinferior to the occipital ventriculocele, and the floor of the fourth ventricle inverted and facing inferiorly at the underside of the S curve.

C, Entry into fluid compartment *1* revealed the occipital lobe herniation (*arrow*) and choroid plexus (*C*) of the occipital ventriculocele. The gyral sulcal pattern could be traced through the defect into the intracranial occipital lobe.

Fig. 15. Parietal meningoencephalocele; 2-month-old boy.

A, T1 sagittal MR shows herniation of recognizable portions of the temporal and parietal lobes into the sac.

B, Coronal proton density MR shows bilateral hydrocephalus with extension of the ventricle into the sac.

A   **B**
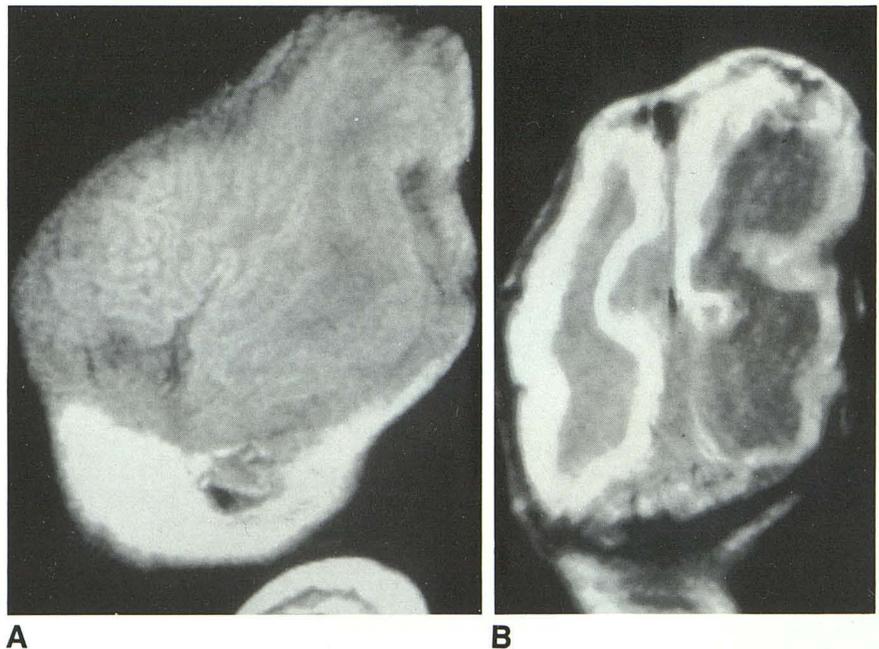
The fiber tracts of the stem are similarly distorted and hard to identify. The pyramidal systems could not be identified in any of the five cases of Karch and Urich (47). The medial lem-nisci were relatively well preserved. The inferior olives were absent in 60%, present on one side only in 20% and deformed bilaterally in 20%. The cerebellar peduncles could not be identified

A

B

C

Fig. 16. Bilobed parietal meningoencephalocele; 2-day-old, full-term boy with un-complicated pregnancy and delivery and a ostium secundum defect of the heart, foramen ovale type.

A and B, Patient photographs reveal a large 8 × 9 × 10 cm bilobed anterior parietal cephalocele with microcephaly and low-set ears.

C and D, Gross brain specimen in left lateral view (C) and coronal section (D) just anterior to the stalk reveal the narrow pedunculated origin of the cephalocele, herniation of recognizable portions of both cerebral hemispheres into the sac, rotation of the herniating segments so they are seen to lie anteroposterior within the sac on true lateral views (C), additional nubbins of tissue in the cephalocele, agenesis of the corpus callosum, azygous anterior cerebral artery and single draining vein, and hemorrhage within the ventricles and subarachnoid spaces. The residual intracranial contents were composed predominantly of temporal lobes.
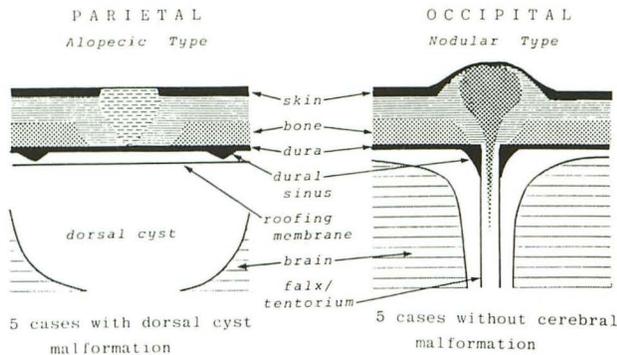
D

Fig. 17. Atretic meningocele; diagrammatic representations of parietal and occipital types. (Reprinted with permission from Yokota et al (25).)
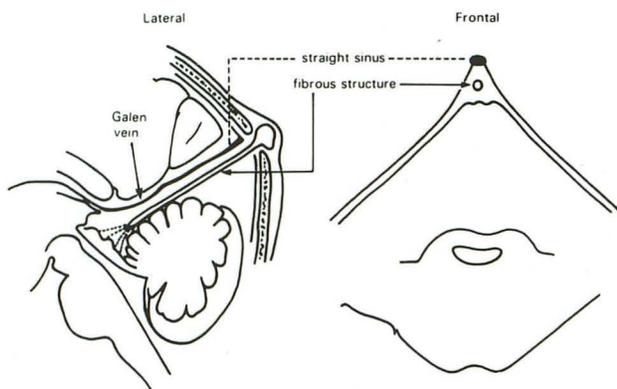

Fig. 18. Atretic meningocele; meningocele manque. Diagrammatic representation of the course of the fibrous structure between the vertex and the tectum and the relationship of the fibrous cord to the embryonic position of the tentorium and the dural venous sinuses. (Reprinted with permission from Inoue et al (29).)

in any of the cases (47). Instead, there were raw gliotic threads and strands of tissue at the expected positions of the peduncles.

Chapman et al (48) summarized diverse data to conclude that, when an occipital cephalocele is associated with a hindbrain malformation,

- herniation occurs below the torcular regardless of the extent to which supratentorial structures are involved;
- the fluid-filled sac communicates with the fourth ventricle as a ventriculocele;
- a *midline* ventriculocele is usually situated caudal to the developing cerebellum;
- variable degrees of cerebellar and tectal dysplasia are the rule;
- extreme cases may show no identifiable cerebellum; and
- if the evagination arises from the aqueduct, posterior aqueductal recess, or intervening
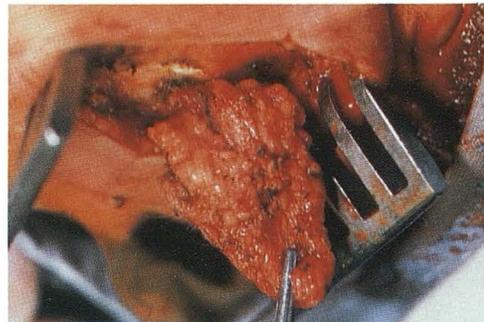
Fig. 19. Atretic meningocele with posterior fontanelle dermoid; newborn male.

A, Vertex view shows a membranous angiomatous cutaneous defect at the posterior fontanelle, with anomalous whorling of the surrounding hair. Angiography demonstrated a bifid superior sagittal sinus beneath the lesion and large vein passing from the lesion into the sinus.
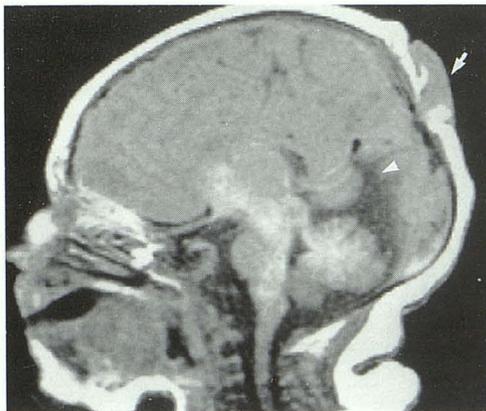
B, At surgery, the skin was incised elliptically to encompass the cutaneous lesion. An underlying hemangioma was cauterized and the lesion was dissected to disclose a stalk (*arrow*) representing the penetrating vein. This was coagulated and transected. Elliptical flaps of pericranium were carried over the sinus and sutured. The wound was then closed in layers.

C, The resected specimen contained angioma and dermoid.

isthmus, the degree of deformation and reduction in size of mesencephalic and rhombencephalic structures may be extreme.

A

B

Fig. 20. Atretic meningocele; two patients.

A, Six-week-old boy. Patient photograph shows the typical flattened appearance of an atretic meningocele and the elongated abnormally whorled hair that surrounds it. At surgery, the skin was incised elliptically to encompass the lesion. Sharp dissection uncovered a stalk which extended through a skull defect to course inferiorly in the intracranial midline. The stalk contained no dermal elements.

B, Sagittal T1 MR shows a flattened lesion at the vertex (arrow), dehiscence of high-signal fat and low-signal bone beneath the lesion and an abnormally high falcotentorial junction (arrowhead). The vertex lesion appears aligned with the quadrigeminal plate along the course expected for an embryonic tentorium.

Spinal Cord. The shape of the spinal cord is nearly always distorted by torsion and side-to-side compression, especially toward the cervico-medullary junction. Hydromyelia is present in many cases, five of five in the series of Karch and Urich (47). The enlarged central canal may extend into the dorsal median fissure to form a cavity with a triangular cross section. A rich vascular plexus often surrounds the brain stem and upper cervical cord (49). Islands of aberrant neural tissue are commonly observed in the region of the brain stem and cord (49).

Subsets of Occipital Cephalocele. Two rare subsets of occipital cephalocele require special mention.

1. The posterior fossa ventriculocele is an occipital cephalocele that consists nearly entirely of a cyst communicating with the fourth ventricle (65, 66). The cephalocele may be high or low occipital. The cyst may arise from the midline rhombic roof or from the lateral recess of the fourth ventricle. Callosal dysgenesis and hydrocephalus may accompany the ventriculocele.

2. Inverse cerebellum with occipital cephalocele (synonym: tectocerebellar dysraphia with occipital encephalocele) is a malformation in which a complex occipital encephalocele contains hypoplastic cerebellar hemispheres connected to the brain stem by a dorsal extension of the midbrain tectum (67–69). The vermis is absent. The cerebellar hemispheres extend laterally and ventrally to (partially) cover the olives and the pons. The fourth ventricle and aqueduct are open to the superior surface of the brain stem. The tectum of midbrain is broadened, forked, and lacks the corpora quadrigemina. The aqueduct and tectum extend posteriorly through the foramen magnum and the ostium of the encephalocele. Each of the hypoplastic cerebellar hemispheres appears to arise separately from one fork of the cleft stem and each contains a large ventricular structure. The corpus callosum may be deficient. Gyral abnormalities are common, particularly focal polymicrogyria.

Surgical Correction. Surgery is performed to protect the child from ulceration of the sac with subsequent infection or hemorrhage, to prevent the sac from expanding with time, to effect cosmesis and—perhaps—to provide as homeopathic an environment as possible in that hope that such an environment may facilitate more nearly normal brain function.
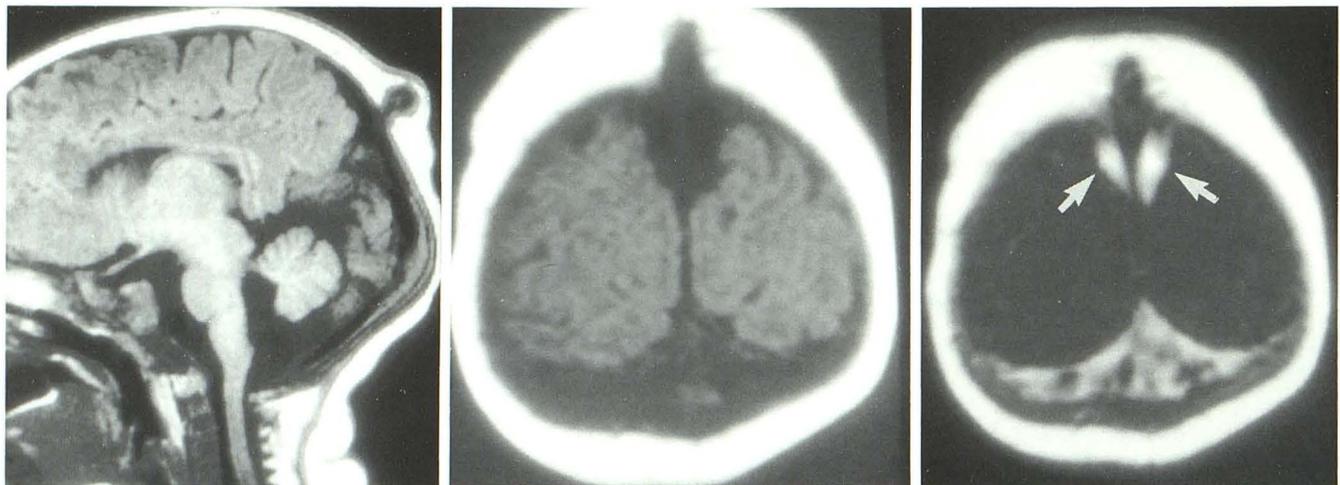
Fig. 21. Atretic meningocele.

A, Sagittal T1 MR displays the small, skin-covered meningocele within the scalp at the vertex, the dehiscence in the bone, and the alignment of the brain surfaces along the trajectory from the meningocele to the quadrigeminal plate. No falcotentorial junction is seen *in the midline*. The posterior interhemispheric fissure is wide. Cisterna magna is large.

B and *C*, Coronal T1 MR (*B*) and gadolinium-enhanced T1 MR (*C*) show the wide interhemispheric fissure and the bifid halves (*arrows*) of the superior sagittal sinus.

D, MR angiography demonstrates the fenestration in the superior sagittal sinus where it is traversed by the cord.

Fig. 22. Atretic parietal meningocele; 1-day-old girl.

A, Sagittal T1 MR shows the small, bulging, cephalocele (*arrow*) with dehiscence of high-signal subcutaneous fat and low-signal bone immediately beneath the lesion. The trajectory of the vessels and fibrous structures appears to pass from the vertex toward the quadrigeminal plate along the line that corresponds to the embryonic position of the tentorium. The falcotentorial junction is abnormal with large superior vermian cisterns.

B, Coronal spoiled gradient recalled image. The vertex lesion has a deep extension (*small white arrow*) that passes intracranially in the midline between halves of a bifid superior sagittal sinus (*black arrows*).

C, Coronal image further anteriorly shows the discontinuity in the dura and in the venous sinuses where the stalk (*black arrow*) traverses the falx.

A

B

Fig. 23. Cutis aplasia; two cases.

A, Newborn girl with macrocrania and severe hydrocephalus requiring shunt diversion. There were dextrocardia, transverse liver, and absent spleen. The lesion healed spontaneously.

B, Newborn girl with macrocrania and ventricular dilatation.

With true meningoceles, surgery is begun with a transverse elliptical incision extending partway up the neck of the sac to preserve tissue for closure (54). The extent of the cranial defect is exposed and defined. The dura is closed in watertight fashion and the overlying skin is closed separately. There is no need to repair the bony defect.

With true meningoencephalocele, the limiting factors are the presence of (potentially) functional neural elements, the unusually small size of the

posterior fossa and cranium that severely limit the available intracranial space, and the blood vessels in and around the neck of the herniation (48). Most surgeons resect the obviously abnormal tissue. To provide room to return herniated tissue intracranially, they then attempt to enlarge the posterior fossa or to decompress the skull by ventriculostomy. This is usually difficult. An attempt is made to enlarge the bony ostium to prevent vascular strangulation and to close the *soft* tissue over any residual herniated functional brain once the excess soft tissue has been excised. Further surgery at a later age could potentially effect more complete return of brain to calvarium.

Outcome of *occipital* cephaloceles is variable (Tables 7 and 8). In Simpson's series of 34 occipital cephaloceles, six of 34 were treated "expectantly"; five died. The survivor was alive at age 25, but profoundly retarded and totally dependent (44). Twenty-seven of 34 patients had simple excision of the cephalocele with dural closure. Two required later cranioplasty to cover pulsatile defects. One of 34 had conservative closure with duraplasty. Five of 34 required shunts for hydrocephalus (44). Of two cervico-occipital cephaloceles, one had resection of bilocular meningoceles, and one was not treated. The meningocele decreased in size with time.

Parietal Cephaloceles

Clinical Findings. Parietal cephaloceles present in the midline between the lambda and the bregma. Like occipital cephaloceles, they show diverse size, shape, position, and content. Typically, they show hair at the base, may be tender (possibly because of dural stretching), and rarely are multiple (22, 51).

In Yokota's (25) series of 15 parietal cephaloceles: four of 15 (27%) were encephaloceles; four of 15 (27%) were meningoceles; two of 15 (13%) were gliaceles; and five of 15 (33%) were atretic meningoceles.

In this small series, there was male predominance for the encephaloceles (four of four) and meningoceles (three of four), equal sex ratio for the gliaceles (one of two), and possible female predominance for the atretic meningoceles (three of five). Microcephaly is present in approximately 20% of patients with parietal cephalocele (44).
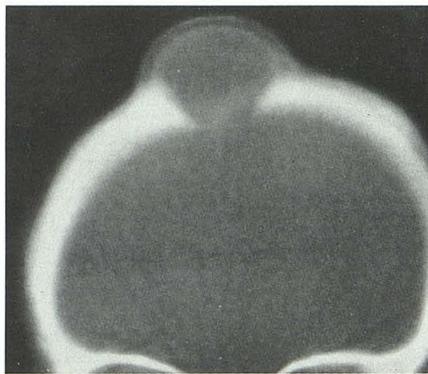
In Yokota's series, 33% died, 40% showed marked retardation, 13% were educable, and 15% had normal development (25). The two
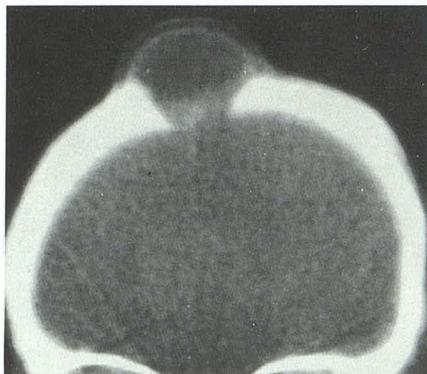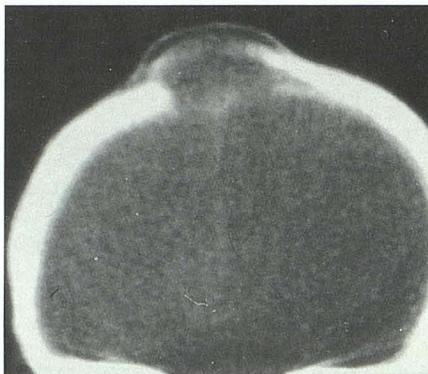
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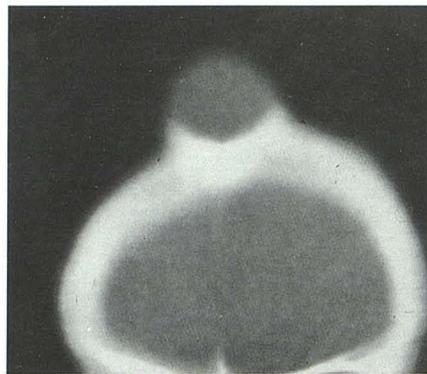
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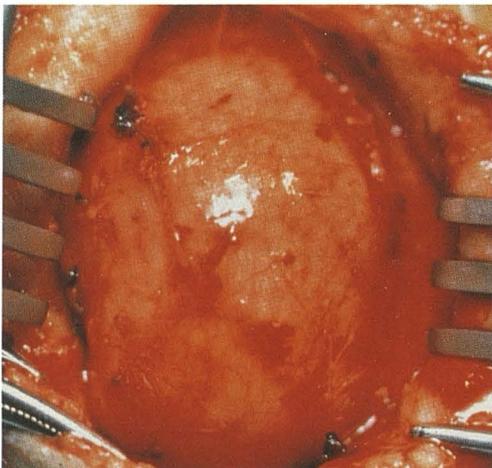
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normal patients both had meningoceles with no associated cerebral malformations.

Osseous Structures and Dura. In parietal cephaloceles, the bony defect typically lies in the midline. It is more commonly situated near to the posterior fontanelle or to the anterior fontanelle than along the midportion of the sagittal suture (45). Beneath the bony ostium, the two leaves of dura that form the falx may fail to fuse together focally, leaving a long tract or fenestration through the falx and a focal dural dehiscence leading to the ostium. The posterior part of the interhemispheric fissure is typically widened as a consequence (59). The separated dural leaves may each insert into the ipsilateral tentorial leaf, forming a tentorial cleft or fenestration as well.

The dural venous sinuses deflect around one or both sides of the ostium and tract creating a fenestrated superior sagittal sinus and/or torcular. The straight sinus may be absent, short, or fenestrated.

The superficial lesion may have open communication with the intracranial space, or not. Yokota found such open communication in three of four parietal *en*cephaloceles (25). Of the four meningoceles, open communication was found in one, partial communication in two, and no communication in one. Of the two glioceles, one had open communication with the intracranial space. None of the five atretic meningoceles communicated freely with the intracranial space.

Hemispheres. Recognizable parietal cortex extended into the cephalocele sac in 60% of Simpson's parietal meningoencephaloceles (Fig. 15) (44). The sac contained only small glial nodules in 30% of cases (44). In Yokota's four meningoencephaloceles, the herniation was too large to determine the precise anatomic structures involved (25).

Ventricular Dilatation. Ventricular dilatation was seen in utero in 47% of cases, postnatally in 20% of cases, and following surgery in an additional 7% of cases (Table 5B) (25).

Associated Cerebral Malformations. Parietal cephaloceles are commonly associated with midline brain anomalies (Fig. 16). In small series, these have been reported as agenesis of the corpus callosum and/or Dandy-Walker cyst in 38% (70), agenesis of the corpus callosum and/or lobar holoprosencephaly in 40% (44), Chiari II malformation in 33% (51), and Dandy-Walker malformation in 67% (51). Yokota found midline dysgenesis varying from callosal agenesis to dorsal cyst in 60%, vermian agenesis in 13%, Chiari II malformation in 7%, and normal cerebral structure in only two cases with pure pedunculated meningoceles (13%) (25).

Specifically analyzed by lesion type in 15 parietal cephaloceles, Yokota's data reveal (25):

- among four meningoencephaloceles, one Chiari II malformation with thoracolumbar myelomeningocele, and one median cleft face syndrome with syndactyly;

- among four meningoceles, one diencephalic cyst; one midline porencephaly, and two normal brains;

- among two glioceles, one diencephalic cyst with vermian agenesis, and one midline porencephaly;

- among five atretic meningoceles, one diencephalic cyst, three midline porencephalies, and one callosal agenesis with vermian agenesis.

Surgical Correction. In Simpson's series of 13 parietal cephaloceles, one was treated expectantly and died (8%), four were treated with conservative closure and duraplasty. Such duraplasty is more difficult when the superior sagittal sinus lies close to the dural defect or forms a circumferential venous channel. In one case, division of a seemingly unimportant sinus to enlarge cranial capacity resulted in seizures from cortical venous infarct (44).

Fig. 24. Anterior fontanelle dermoid; 8-month-old girl with a slowly growing tense mass at the anterior fontanelle. The overlying skin appeared normal.

A, Frontal view of the face. The anterior fontanelle mass (*white arrow*) is characteristically obscured by normal hair.

B–F, Coronal contrast-enhanced CT demonstrates a tense round lucent nonenhancing well-encapsulated mass that lies deep to the cutis, remains separate from the cutis, rests within a beveled bony fossa, abuts directly on the roof of the enhancing superior sagittal sinus, and depresses the sinus and dura with slight widening of the epidural space in *E*.

F–H, Surgical exposure and resection.

F, Midline skin incision and bilateral subcutaneous dissection discloses the dome of the capsule of the dermoid.

G, Careful undermining and reflection exposes the vascular pedicle (*arrows*) from the base of the dermoid to the dural roof of the superior sagittal sinus. This has been coagulated and transected, permitting the dermoid to roll out of the fossa.

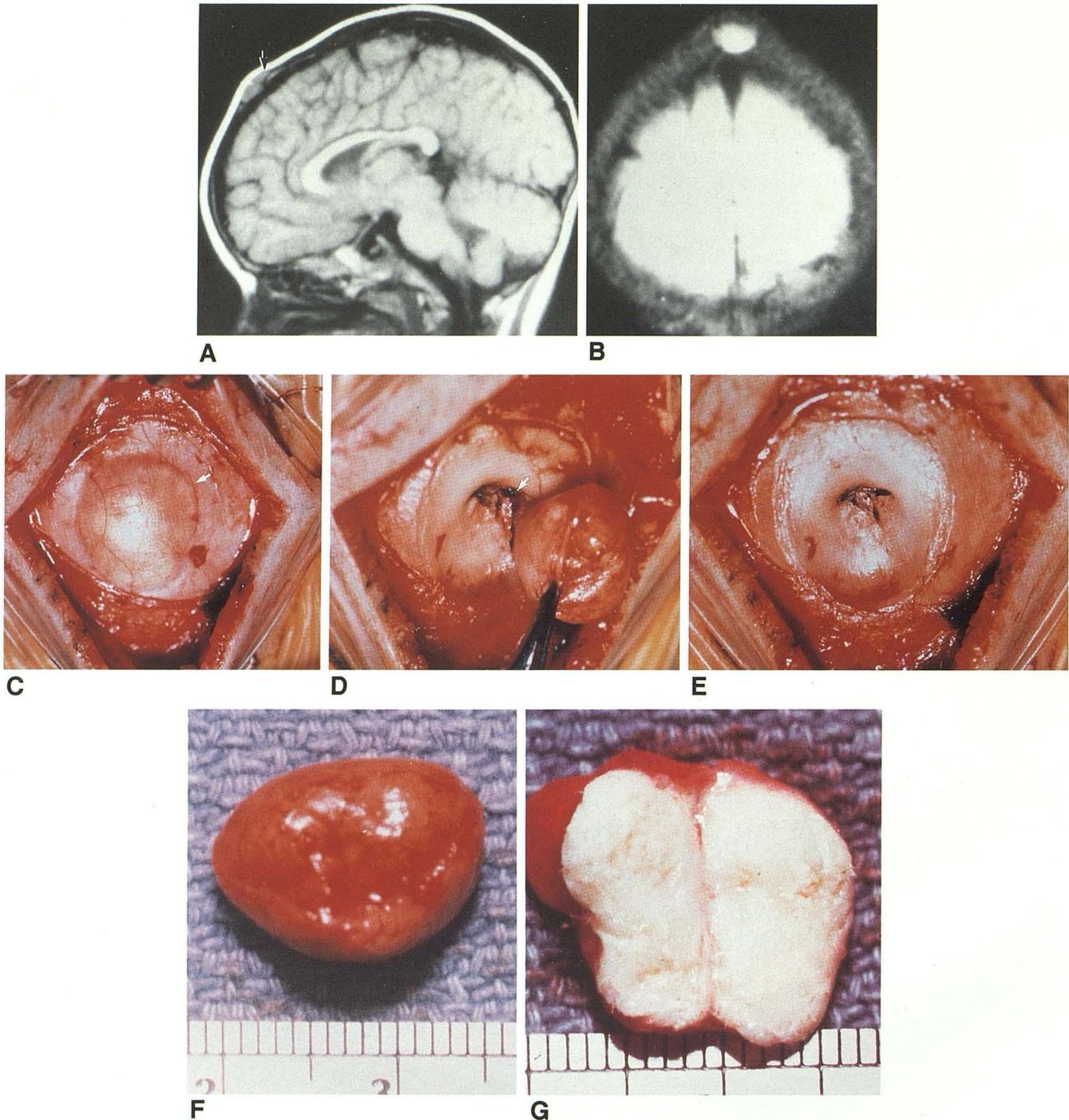*H*, Incision discloses the hair and cheesy content of the dermoid.

Fig. 25. Anterior fontanelle dermoid; 2-year-old girl with a single 2 × 2 cm mass that is growing firmer. The overlying skin is normal.

A, Sagittal T1 MR displays a well-defined mass (*arrow*) situated within a depression in the bone at the anterior fontanelle. The overlying skin and subcutaneous fat are normal. The lesion is isointense with brain. It does not extend intracranially. Midline brain structures are normal.

B, Coronal T2 MR shows very high signal from the well-defined rounded subcutaneous mass. The flow void in the subjacent superior sagittal sinus is normal.

C–G, Surgical exposure and resection.

C, The skin was incised transversely. The subcutaneous tissue was retracted and the periosteum denuded to expose the dome of a 2 × 2 cm smooth well-encapsulated yellow-brown midline cyst (*arrow*).

D, The mass was shelled out with a periosteal elevator to expose the small vessel that penetrated the inner table toward the superior sagittal sinus.

Atretic Meningoceles

Atretic meningoceles are regarded as formes fruste of meningoencephaloceles. They commonly occur in the parietal and occipital regions, but manifest differently at the two sites (25). The anatomic arrangements of the atretic parietal and occipital meningoceles are illustrated in Figures 17 and 18.

Atretic *parietal* meningoceles usually present externally in the midline, near to the vertex, as hairless, well-marginated round-oval lesions 5 mm–15 mm in size (25). They may be covered by atrophic skin or by a glistening, parchment-like membrane. Typically, they appear bullate or cystic just after birth, but transform over days to a flat alopecic lesion (25). Histologically, the skin lesion is nonspecific fibrous tissue devoid of cutaneous adnexae and neural tissue.

Beneath the skin lesion, the calvarium shows a sharply marginated ostium. The dura characteristically seals the osseous defect, separating the alopecic lesion from the intracranial cavity (100% of cases) (25). In Yokota's series of five atretic parietal meningoceles, the underlying brain always manifested midline anomalies, such as midline porencephaly (60%), a diencephalic cyst (20%), or callosal agenesis (20%) (25).

Atretic *occipital* cephaloceles usually present externally in the midline just cephalic to the external occipital protuberance as nodular, non-cystic lesions that are 10–15 mm in diameter and covered by smooth, scarred epithelium or sparsely haired skin (25). From the lesion, a fibrous core 2–3 mm in diameter extends intracranially through a small cranial defect at the occipital midline just above the inion (external occipital protuberance) (25). The fibrous core terminates at the dura, or at the falcotentorial tissue after piercing through the dura. Typically, the superior sagittal sinus bifurcates to the transverse sinuses proximal to the usual site, permitting the core to pass under (caudal to) the distal end of the superior sagittal sinus, *or* the superior sagittal sinus is fenestrated. Microscopically, the central fibrous cord is composed of meningofibrous tissue devoid of any neural elements and devoid of any elements suggestive of a dermal sinus tract.

In our experience, the atretic lesions observed in our patients have originated at the posterior fontanelle and exhibited features that resemble those of the occipital form more closely than those of the parietal form (Figs. 19–22).

Lesions clinically similar to the atretic occipital cephalocele have been found entirely within the scalp with no connection to the skull. These have been reported as ectopic glial tissue in the scalp (71). Sinus pericranii is typically an off-midline lesion. Some of its cutaneous manifestations, however, resemble those of the atretic meningoceles (1).

Cutis Aplasia Congenita

Cutis aplasia congenita is an unusual condition that presents with focal aplasia of the scalp, with or without deficiency of the underlying bone and meninges (26). Approximately 300 cases have been reported (72). The significance of this lesion lies in the differential diagnosis of atretic meningocele.

Cutis aplasia congenita may be sporadic (80%) or inherited as an autosomal dominant or autosomal recessive trait (20%) (26, 72). It has been observed at identical sites in identical twins (73). It may be an isolated lesion, or occur as part of a multiple congenital anomaly. Trisomy 13/15 was observed in 33% of patients with cutis aplasia congenita in Kosnik's series (73), but not in others' series (72). The sex incidence varies widely among different series (72, 73).

Typically, the lesion presents in the midline as a one or more hairless well-marginated defect(s) varying in size from a few mm to 8 cm (occasionally larger) (Fig. 23) (72, 73). Some lesions are temporal, parietal, or retroauricular, rather than midline. Approximately 10% of cases show defects in the cutis at other sites. The defect is usually oval or round, less commonly dumbbell, horse-shoe, or Y-shaped.

The defect may be confined to the scalp (partial thickness) (67%) or extend more deeply to involve periosteum, skull, and dura (33%). The skin at the edge of the lesion has a flattened epithelium. Skin appendages are absent. The surface may be smoothly membranous, ulcerated, or


Fig. 25—*Continued.*

E, After resection of the dermoid, the bony fossa is seen to be a partial thickness depression in the bone, not a full thickness defect as it was in the prior case.
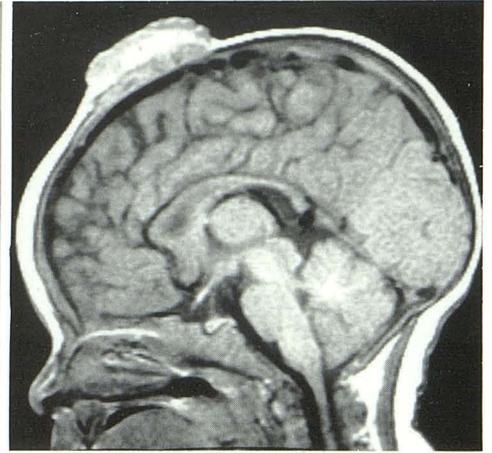
F and *G,* View of the outer capsule (*F*) and midline incision (*G*) show the smooth thin-walled cyst filled with homogeneous material. Histology disclosed a keratinizing squamous epithelium with surrounding adnexal structures. The cyst content was keratinaceous debris.

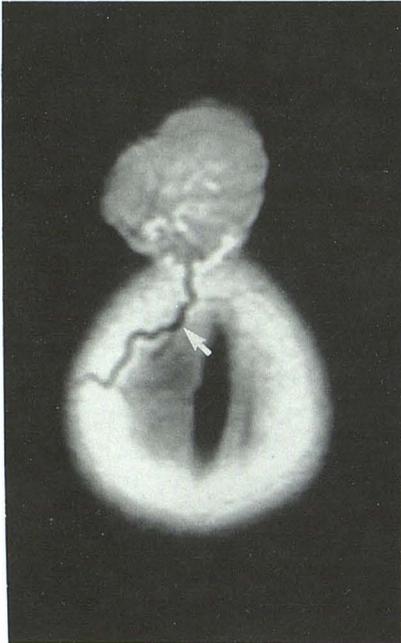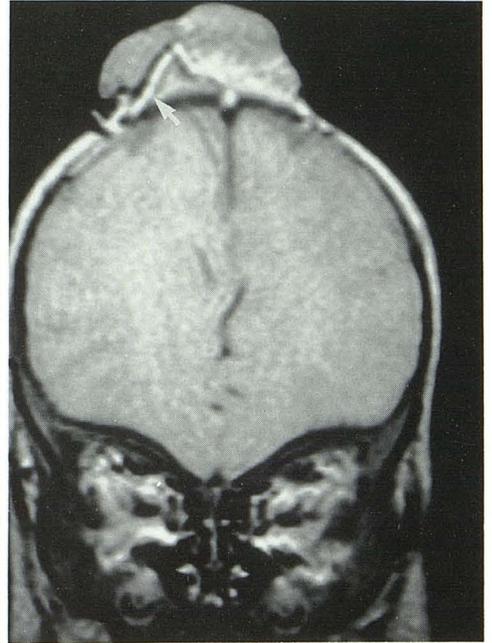(Portions of these figures have been reprinted with permission from Naidich (83).)
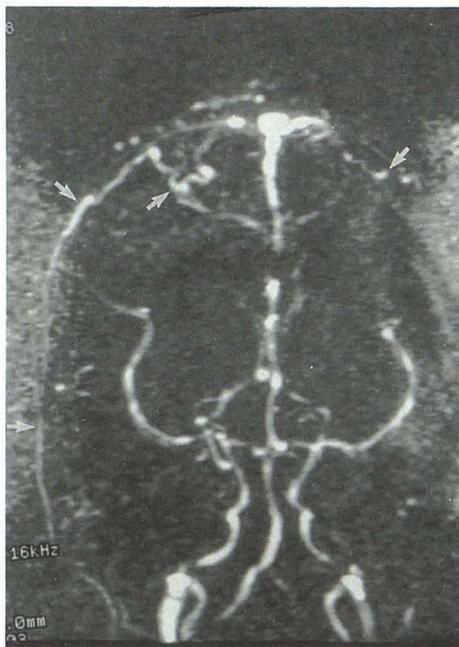
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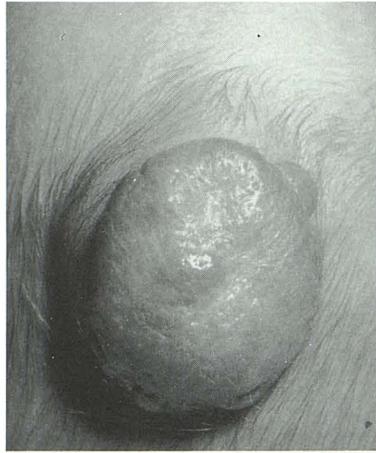
B

C

D

E

F

G

H

already scarred at birth. In full thickness lesions, the superior sagittal sinus and cortex may be visible through the arachnoid.

The lesion may heal spontaneously or require extensive reconstructive surgery. It may erode into the superior sagittal sinus leading to hemorrhage (26). It may become infected leading to osteomyelitis and meningocephalitis.

Cutis aplasia has been associated with CNS malformations such as meningocele, lipomyelomeningocele, holoprosencephaly, hydranencephaly, and colobomas (26, 72–74). It has also been associated with a host of chromosomal and nonchromosomal non-CNS malformations detailed in Reference 26.

Lesions of the Anterior Fontanelle

Lesions at the anterior fontanelle include meningoencephaloceles, dermoids, scalp angiomas, and protruding pacchionian granulations.

Dermoid cysts constitute approximately 23% of all scalp lesions and are distributed widely over the scalp (75). Dermoid cysts of the anterior fontanelle account for 4.8% (75) to 27% (76) of all head and neck dermoids and are regarded as the most frequent type of dermoid seen in neurosurgical practice (76). They usually present at birth or within the first few months of life. Females predominate 2:1 (76–79). Nearly all are sporadic. No family history has been reported (77). Early literature suggested increased incidence of these dermoids in black Africans (77, 80, 81), but increasing experience indicates that they occur in all ethnic groups (76, 79).

Any part of the anterior fontanelle may be affected (76), but the anterior angle of the fontanelle is the most common site (76, 82). Identical cysts may also occur just anterior to, posterior to, or at a distance from the anterior fontanelle, near to the vertex or occiput (78).

Clinically, anterior fontanelle dermoids present as solitary, round, nontender, slightly mobile, or sessile masses situated at the anterior fontanelle (Fig. 24) (77). The overlying skin is normal with no dimple or sinus (76, 80). Some of the lesions are fluid-filled and fluctuant; these transilluminate. Others are solid and opaque. The lesions may become tenser with crying (77, 79). A few (11%) are pulsatile with bruits.

Cyst size varies from 1 cm to $10 \times 15 \times 6$ cm. The lesions may enlarge very slowly over time, but there is no good correlation between lesion size and patient age (79). Affected children are otherwise normal, with normal growth and development, and suffer no concurrent malformations of brain or body.

Surgical exploration documents that the mass lies in the subgaleal or subperiosteal space. It is well encapsulated and separate from the overlying skin. Typically, it rests within a well-corticated bone depression and has a small pedicle that attaches to the bony base or to the roof of the superior sagittal sinus. This must be coagulated and severed prior to delivery of the cyst. The inferior pole of the mass may compress the subjacent tissue slightly. It can be stubbornly adherent to the superior sagittal sinus, leading to sinus damage at surgery. However, the cyst has never been reported to extend into the sinus or to extend intracranially. The mass is usually excised in toto and does not recur. Malignant degeneration has never been observed, but remains a potential. Infection of the cyst is rare and, if present, is usually iatrogenic after diagnostic cyst puncture (76).

Histologically, the lesion is a thin-walled dermoid cyst lined by keratinizing squamous epithelium containing skin appendages such as hair follicles, sebaceous glands, and sweat glands. There are variable amounts of collagen in the capsule (78). Giant-cell foreign body reaction typical of cholesterol-induced inflammation may be

Fig. 26. Anterior fontanelle hemangioendothelioma; 4-month-old boy.

A and *B*, Lateral (*A*) and vertex (*B*) views show a well-defined, highly vascular lesion at the anterior fontanelle. There is some elevation of the adjacent skin but no other abnormality.

C–F, MR images: *C*, Midsagittal gadolinium-enhanced, T1 MR; *D* and *E*, axial gadolinium-enhanced T1 MR; and *F*, coronal noncontrast proton density MR all demonstrate an ovoid vascular-enhancing mass closely applied to the outer table of the calvarium. A very prominent superficial temporal artery branch (*arrows*) supplies the lesion. The dural venous sinus is normal. The subcutaneous fat is slightly elevated.

G, Coronal compressed 3-D phase-contrast MR angiogram demonstrates the enlarged superficial temporal branch (*white arrows*), other less prominent supply (*single white arrow*), and the normal superior sagittal sinus.

H, At surgery, Doppler was used to define all arterial (*A*) and venous (*V*) pedicles. Blood supply was easily controlled. The scalp was incised circumferentially and undermined, easily separating the lesion from the underlying pericranium. Histologic diagnosis was hemangioendothelioma. Scalp angiomas and arteriovenous malformations can be identified by imaging studies and distinguished from the angiomatous nevi associated with cephaloceles. (Fig. 26H courtesy of Dr S. Gonzalez-Arias, Miami, FL.)

present (81). The cyst contents vary from clear liquid to thick white cheesy material and may evolve with age. Adeloye and Odeku (77) reported that the small cysts contained uniformly clear colorless fluid with low pH, low protein, low sodium, low chloride, and high potassium. Larger cysts are yellow to yellow-brown with high pH, high sodium and chloride, and low potassium, protein, and glucose. The color may result from chromidrosis—the secretion of pigment—containing sweat by apocrine sweat glands (78).

Imaging studies vary with patient age and precise lesion location. In infants with open fontanelles and small cysts, studies may show only the soft-tissue mass. In older children with closed fontanelles and larger masses, the studies usually demonstrate a scalloped bone defect beneath the mass. The defect may be shallow, moderately deep, or full-thickness. The depth of the defect does *not* correlate with the size of the cyst and may not increase over time (78). The shape of the depression has been postulated to be congenital (78). Calcification is not reported, but can be expected, rarely, as a consequence of inflammation or formation of calcium soaps.

Computed tomography (CT) scans through the mass perpendicular to the skull demonstrate the intact skin; the separate round well-encapsulated lesion, associated bone defect, and the exact relationship of the mass to the superior sagittal sinus.

Magnetic resonance (MR) may show a lesion isointense with CSF on the T1 image mimicking a meningocele (31) or a lesion that is isointense to brain on T1 and high signal on T2 (Fig. 25) (83).

Anterior fontanelle angiomas and arteriovenous malformations may mimic cephaloceles and other anterior fossa lesions with intracranial connections (Fig. 26).

Temporal Cephalocele. The majority of temporal cephaloceles result from infection or surgery (75%). Some are posttraumatic (15%) (84). *Congenital* temporal cephaloceles are uncommon protrusions of meninges and/or brain through one or multiple defects in the tegmen tympani and tegmen antri (Fig. 27). They constitute approximately 13%–21% of all temporal cephaloceles (84, 85).

Congenital temporal cephaloceles appear to result from congenital dehiscences of the tegmen tympani and tegmen antri. At birth, the temporal bone consists of a combination of membranous and cartilaginous bone in various stages of ossification. Pneumatization in the attic begins at about the 34th week when mucosa begins to invade the marrow-containing bone. It progresses rapidly post-partum. Formation of the air cells involves a sequential mucosal advancement, marrow resorption, and bone remodelling that continues to adulthood. Secondary to remodeling, small bony dehiscences frequently occur in the tegmen tympani and antri. Thus, Ahren and Thulen (86) found that 21% of 94 consecutive postmortem specimens had congenital defects of the tegmen. Six percent had multiple (greater than five) significant defects (84). An additional 16% had very thin cortical bone separating the mastoid air system from the middle fossa dura (87). This work was confirmed by Lang (88). If later (subclinical) infection weakens the exposed dura overlying these bony defects, then temporal lobe herniation may ensue. Pacchionian granulations may also play a role in formation of temporal cephaloceles.

Clinical symptoms of temporal cephaloceles include recurrent/persistent CSF otorrhea and rhinorrhea, recurrent meningitis, intermittent serous otitis media, and progressive conductive hearing loss (89). Occasionally, the mass is seen behind the tympanic membrane or in the mastoid cavity. Facial paralysis and temporal lobe seizures are rare. Sex incidence may be equal, or the cephaloceles may be slightly more frequent in females (male to female ratio = 2:7 in one series (90)).

The most frequent sites of true herniation are the anterior epitympanum and the mastoid antrum (90). There may be multiple pits or incomplete depressions in the roof, in addition to the one or two true dehiscences that do extend through the bone. In Kemink's series (90) of surgically confirmed cases, the seven patients exhibited: patient 1, a single hole in the tegmen antri; patient 2, a large hole in the tegmen antri plus multiple incomplete pits in the tegmen tympani; patient 3, two distinct holes in the tegmen tympani; patient 4, two distinct holes in the tegmen antri, plus one in the floor of the middle fossa and one in the nasal roof; patient 5, three holes in the tegmen tympani; and patients 6 and 7, multiple holes.

The multiplicity of defects and their wide dispersion significantly influence the extent of presurgical workup and surgical repair.

Unlike the other cephaloceles described in this report, the congenital temporal cephaloceles typically exhibit a defect in the dura at the site of the herniation (90). Thus, the herniating brain
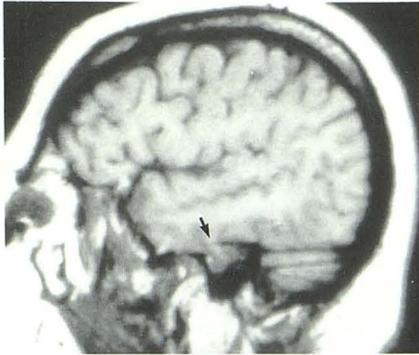
Fig. 27. Post-surgical temporal cephalocele; 30-year-old woman with 6-month history of headache following right mastoidectomy. Sagittal T1 MR shows herniation of the temporal lobe (*arrow*) and fluid into the petrous pyramid via a defect in the tegman tympani. (Case courtesy of Mahmood F. Mafee, MD, Chicago.)

does not have a dural sheath. Typically, the arachnoid sheath is partly deficient and confined to the neck of the ostium. The brain may displace the epithelial lining of the mastoid and middle ear cleft as it herniates and, thereby, appear to be encapsulated.

The herniating brain is composed of mature neural tissue with microglial proliferation surrounded by a "capsule" of fibrovascular reaction (91). The lesion may fill much of the middle ear cleft and even extend down the eustachian tube

(90). The herniating mass may also erode adjacent bone such as the geniculate fossa and horizontal facial nerve canal, the ossicles, the cochlear wall at the promontory, the lateral wall of the epitympanum, and the bony semicircular canals (91).

Surgical Correction. Temporal encephaloceles are treated surgically as follows (90):

The patient is placed on the table, affected ear upward. Retroauricular incision is made and the ear is reflected forward. Following wide mastoidectomy, the extent and number of encephaloceles, dural holes, etc, is determined. Adhesions are separated, but the cephaloceles are not yet elevated.

A middle temporal skin incision is made and fascia harvested. The temporal fossa is entered and the dura is elevated from the floor of the middle fossa. The encephaloceles are identified intracranially and reduced or excised. Part of the temporal squama is harvested and refashioned into a bone graft to bridge any bony defects in the floor of the temporal fossa. Two layers of dried temporal fascia are placed, under and over the bone graft. Large dural defects are sutured and the patient is closed.

Lateral Cephaloceles. The lateral cephalocele develops along the coronal or lambdoid suture, as
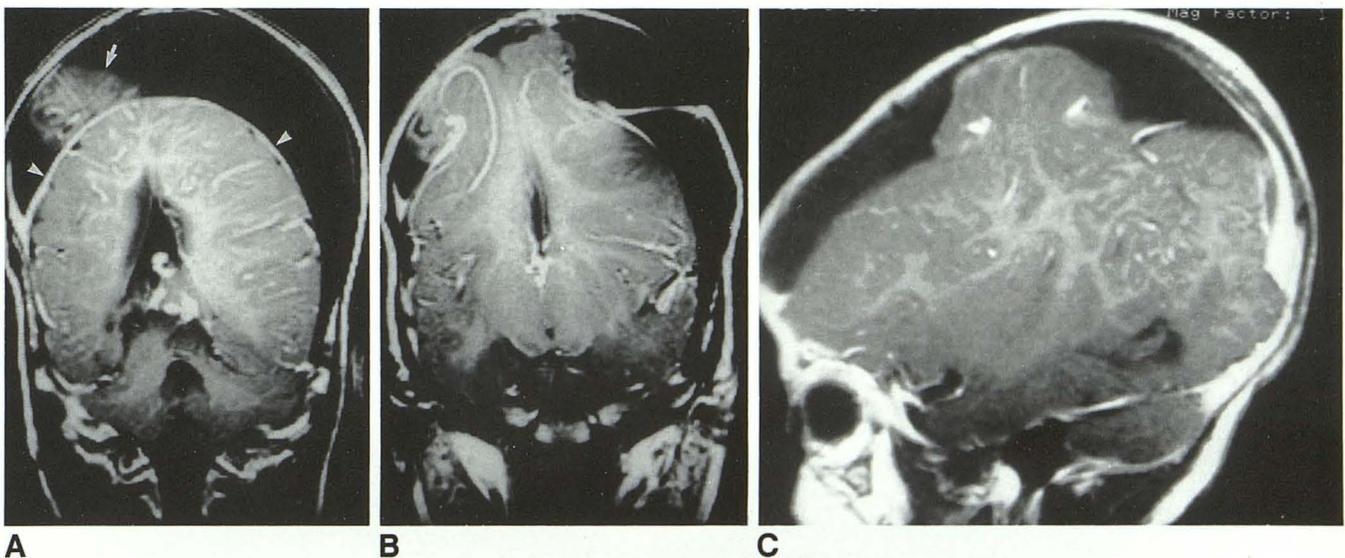

A   B   C

Fig. 28. Internal encephalocele, 1-year-old girl. Shunt decompression of massive hydrocephalus led to craniocerebral disproportion with bilateral large subdural spaces.

A, Coronal contrast-enhanced T1 MR at the posterior edge of the lesion shows the brain (*arrow*) external to the enhancing subdural membrane (*arrowheads*).

B, Coronal and *C,* sagittal contrast-enhanced T1 MR at the midpoint of the lesion demonstrates direct extension of gray matter, white matter, vessels, and interhemispheric fissure into the subdural space, external to the enhancing membrane, to form a mushroom-shaped intracranial "internal" encephalocele. (Case courtesy Sharon E. Byrd, MD, Chicago.)

far inferiorly as the anterolateral and posterolateral fontanelles (Fig. 5B); it is very rare. Arseni and Horvath (92) found four temporal lesions among 419 meningoencephaloceles in Hungary (0.95%). Martinez-Lage et al (93) reported one presumed meningocele at the asterion (junction of parietal, temporal, and occipital bones). Nagulich et al (94) reported eight cephaloceles at the pterion (junction of the frontal, sphenoid, parietal, and temporal bones). Seven of the eight had one ostium. One of the eight had two adjacent ostia. Left and right sides were affected equally. Seven of the eight patients were female.

Seven of these pterional lesions were meningoceles; the one meningoencephalocele was seen in a 10-month-old girl, had 2 cystic cavities, and contained atrophied cerebral tissue, possibly ectopic. The eight masses varied from 0.5–4.0 cm. One of the eight cephaloceles was pedunculated. Six of the eight had normal skin cover that was loose or tightly stretched. Two were enclosed in a finely vascularized membrane.

Tranmer (95) reported a newborn boy with a skin-covered 8 × 9 × 10 cm cystic mass that was either a teratoma or a meningoencephalocele originating at a bone defect in the pterion and floor of the middle fossa and extending into the subcutaneous tissue. A pedunculated portion of the temporal lobe extended through the bone defect into the base of the mass and was supplied by an anomalous branch from the cavernous segment of the internal carotid artery, distinct from the middle cerebral artery. The majority of the mass was cystic and composed of meningeal tissue, choroid plexus, and ependyma. In some areas, respiratory epithelium was found adjacent to normal brain tissue. The dome of the mass was supplied by hypertrophied external carotid branches. The lesion displaced facial structures anteroinferiorly, displaced globe forward stretching the optic nerve, displaced a rudimentary ear inferiorly, and extended posteroinferiorly into the precervical and lateral cervical region to C5.

"Internal" Cephaloceles. Very rarely, trauma or surgery may alter the normal intracranial anatomy to create new compartments with new membrane partitions that provide novel opportunities for brain herniation. In patients with large subdural spaces, defects in the inner subdural membrane may permit herniation of the brain into the subdural space as an "internal" intracranial encephalocele (Fig. 28).

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