PART

I

AXIAL SKELETON

CHAPTER A

SKULL

A-1. CRANIAL VAULT DEVELOPMENT

The skull vault arises from the embryonic blastemal (membranous) desmocranium surrounding the developing brain. Neural crest cells are induced by overlying ectodermal tissue to move from the edges of each side of the neural plate folds to form mesenchymal condensations adjacent to specific areas of the developing brain, beginning with the forebrain. Each mesenchymal condensation is associated with a specific part of the brain. By the thirteenth day, these mesenchymal condensations transform into membranous curved plates on each side of the growing brain to form the primordial cranial vault: frontal, parietals, and interparietal squamosa of the occipital bone (Fig. A-1.0). By the seventh week, these bones begin to ossify directly as bony spicules that radiate outward from developing osseous centers within the membranous tissue. The frontal bone ossifies from a pair of osseous centers, one on each side separated by the fetal metopic suture that normally fuses as the two halves grow together after birth by age 2. Each parietal generally ossifies from two osseous centers, one above the other. The squamosal or interparietal portion of the occipital ossifies from a complex of ossification centers with the number varying according to genetic instructions. As the interparietal occipital ossifies, it fuses at the mendosa line with the expanding lower portion of the occipital of the chondrocranium as it ossifies from the cartilage.

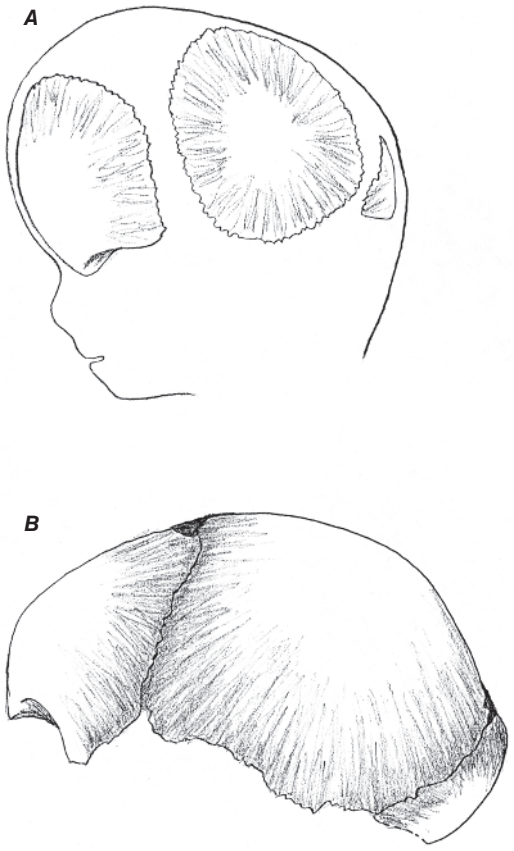

FIGURE A-1.0. Calvaria development: frontal, parietals, occipital interparietal or squamosa—(*A*) embryonic membranous bones; (*B*) newborn bones.

Atlas of Developmental Field Anomalies of the Human Skeleton: A Paleopathology Perspective, First Edition. Ethne Barnes.

The primitive cranial bones are separated by seams and spaces of connective tissue—sutures and fontanelles. These separations allow the cranial bones to be molded during descent through the birth canal. Postnatal cranial bone growth closes the small fetal sagittal fontanelle before birth. The posterolateral (mastoid) fontanelles close by the end of the first year. The posterior and anterolateral (sphenoid) fontanelles close by 3 months and the anterior fontanelle by 18 months.

CRANIAL VAULT ANOMALIES

A-1.1. Extra Ossicles

These commonly form within sutures, especially the lambdoidal suture. Less often they develop within the fontanelles (Fig. A-1.1). Multiple and variable extra ossicles (especially along the lambdoidal suture) also occur with cleidocranial dysostosis, a disturbance in the

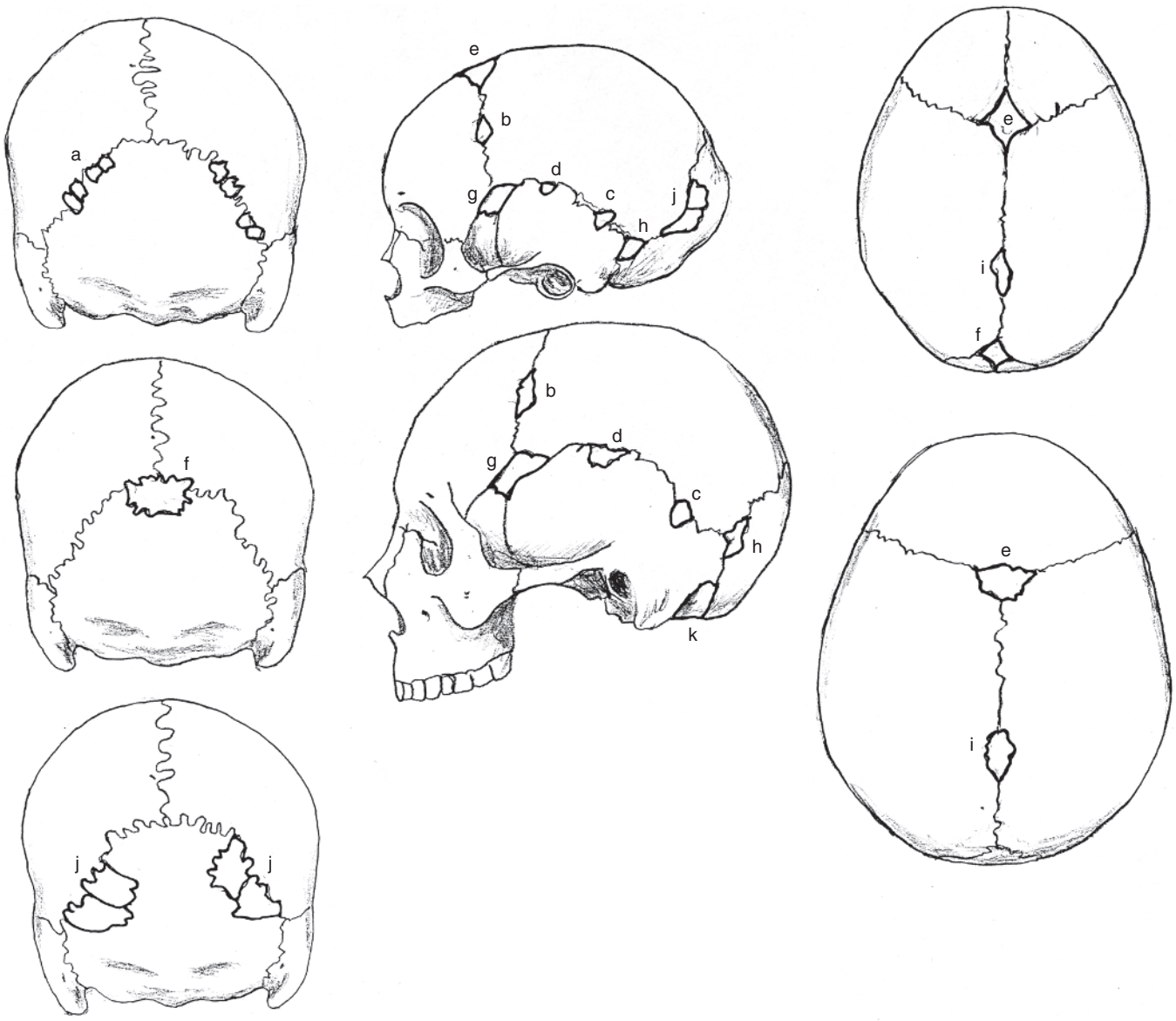

FIGURE A-1.1. Extra ossicles: (newborn and adult) (a) lambdoidal ossicles; (b) coronal ossicles; (c) parietal notch ossicle; (d) temporal squamosa ossicle; (e) bregma (anterior fontanelle) ossicle; (f) lambda (posterior fontanelle) ossicle; (g) epipteric (anteriolateral fontanelle) ossicle; (h) asterion (posterolateral fontanelle) ossicle; (i) obelion (fetal sagittal fontanelle) ossicle; (j) occipital interparietal ossicle; (k) retromastoid ossicle within the chondocranium between the mastoid and occipital.

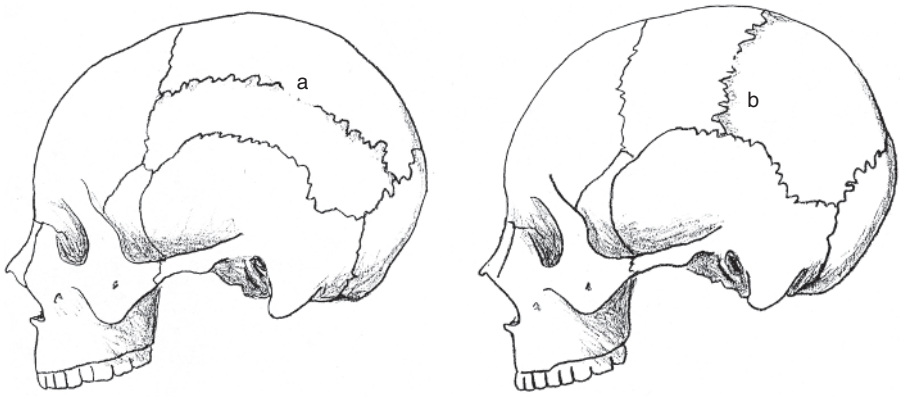

FIGURE A-1.2.1. Extra parietal sutures: (a) horizontal; (b) vertical (Anderson 1995; Shapiro 1972).

membranous bone tissue development that also affects the membranous development of the clavicle.

A-1.2. Extra Sutures

These usually result from the failure of the membranous parts of the same primordial cranial bone to coalesce completely or partially prior to ossification, isolating separate ossification centers within the bone (Fig. A-1.2.1). The metopic suture dividing the infant frontal usually grows together but often remains in place throughout life. The fetal mendosa line between the membranous squamosa and cartilaginous occipital base that normally disappears before birth sometimes remains in place as a complete or incomplete suture. The persistent complete suture gives the appearance of an extra large interparietal occipital bone, commonly referred to as the inca bone (Fig. A-1.2.2).

A-1.3. Sutural Agenesis

This is the failure of sutures to develop (completely or partially) between opposing membranous cranial bones. The lack of bony separation can lead to various forms of cranial deformation, especially with more than one type of suture affected (Figs. A-1.3.1–A-1.3.3).

A-1.4. Parietal Thinning

This is the failure of diploe space to develop within the superior–posterior region of the parietal. This creates a somewhat ovoid depression on the surface of the cranium (Fig. A-1.4.1). It has been noticed in children as well as adults. Most often it appears bilateral, but can be seen unilateral, and the affected area is quite thin, becoming more pronounced with age (Fig. A-1.4.2).

A-1.5. Enlarged Parietal Foramina

This is the failure of membranous bone development within the region of the parietal foramina outlets for Santorini's emissary veins, often associated with some form of lambdoidal and or sagittal suture agenesis (Hoffman 1976). The defects are covered with fibrous membrane instead of bone, ranging in shape from large ovoid to elongated slit bony openings (Figs. A-1.5.1–A-1.5.3).

A-1.6. Inclusion Cysts

These form during embryogenesis with failure of overlying primordial ectodermal cells to retreat from developing cranial bones, usually occurring midline anywhere between the frontonasal region and foramen magnum of the skull base. The anterior fontanelle and the area near the occipital protuberance are the most common sites, sometimes forming in the roof of the eye orbit or greater wing of the sphenoid (Fig. A-1.6.1). Size varies from a few millimeters up to around 10 cm. Tiny cysts often go undetected. Depending on the timing of the event of epidermal entrapment, developing cysts can derive from undifferentiated epidermal cells alone or with differentiated deeper dermal cells. Epidermoid cysts generally contain keratohylin within a capsule of stratified squamosal epithelium and continue to grow slowly by desquamation. Dermoid cysts may contain oil, sebum, cholesterol, and hair follicles, and increase slowly in size by glandular secretions along with epithelial desquamation. Both types can occur internally or externally, leaving cystic depressions within the affected bone. Epidermal cysts frequently occur within the dura mater covering of the brain, while dermoid cysts usually develop within the cranial diploe or between the periosteum and scalp. The cystic bony imprint

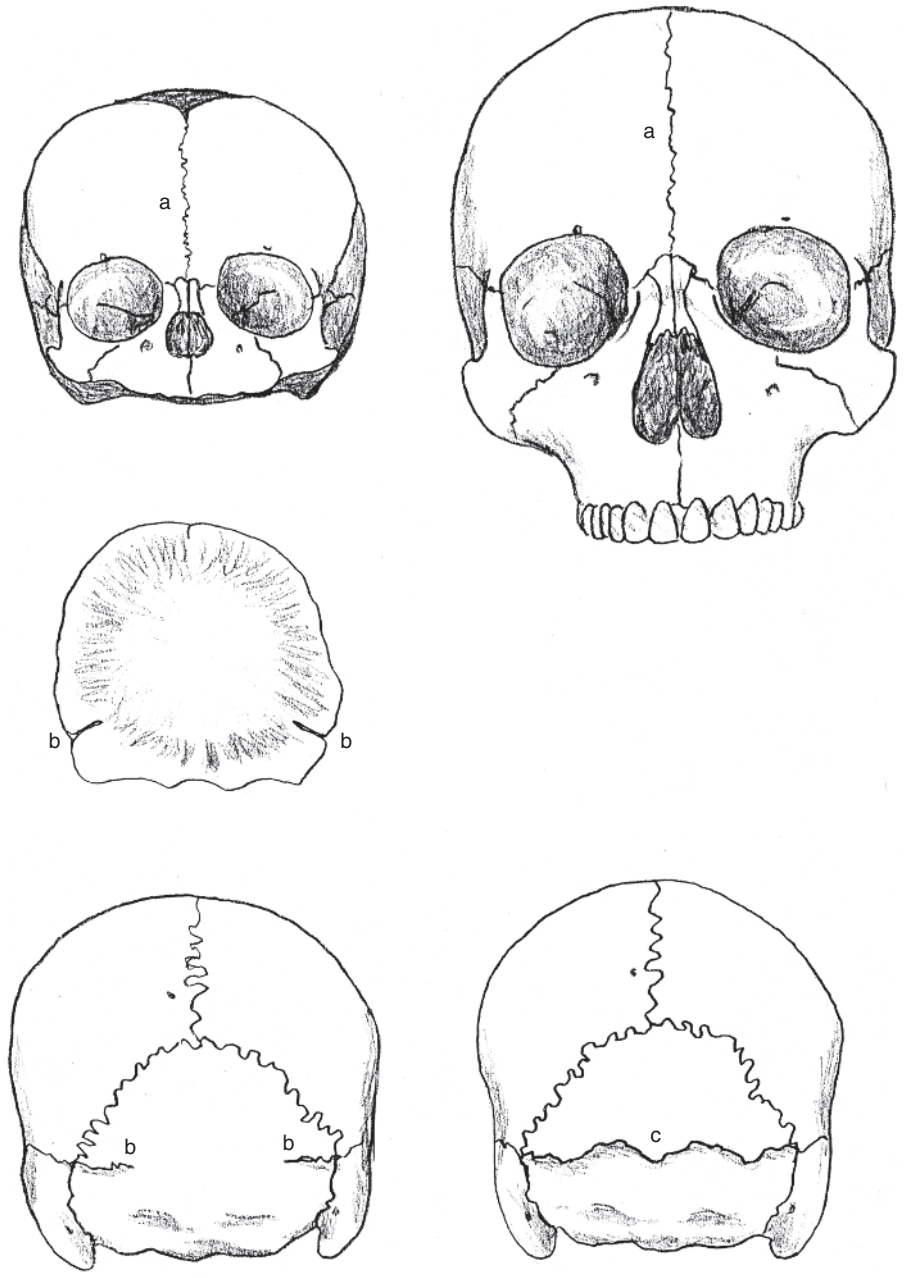

FIGURE A-1.2.2. Extra sutures: (newborn and adult) (a) metopism—retention of infantile suture; (b) remnant fetal mendosa suture; (c) complete retention of fetal mendosa suture (also known as inca bone).

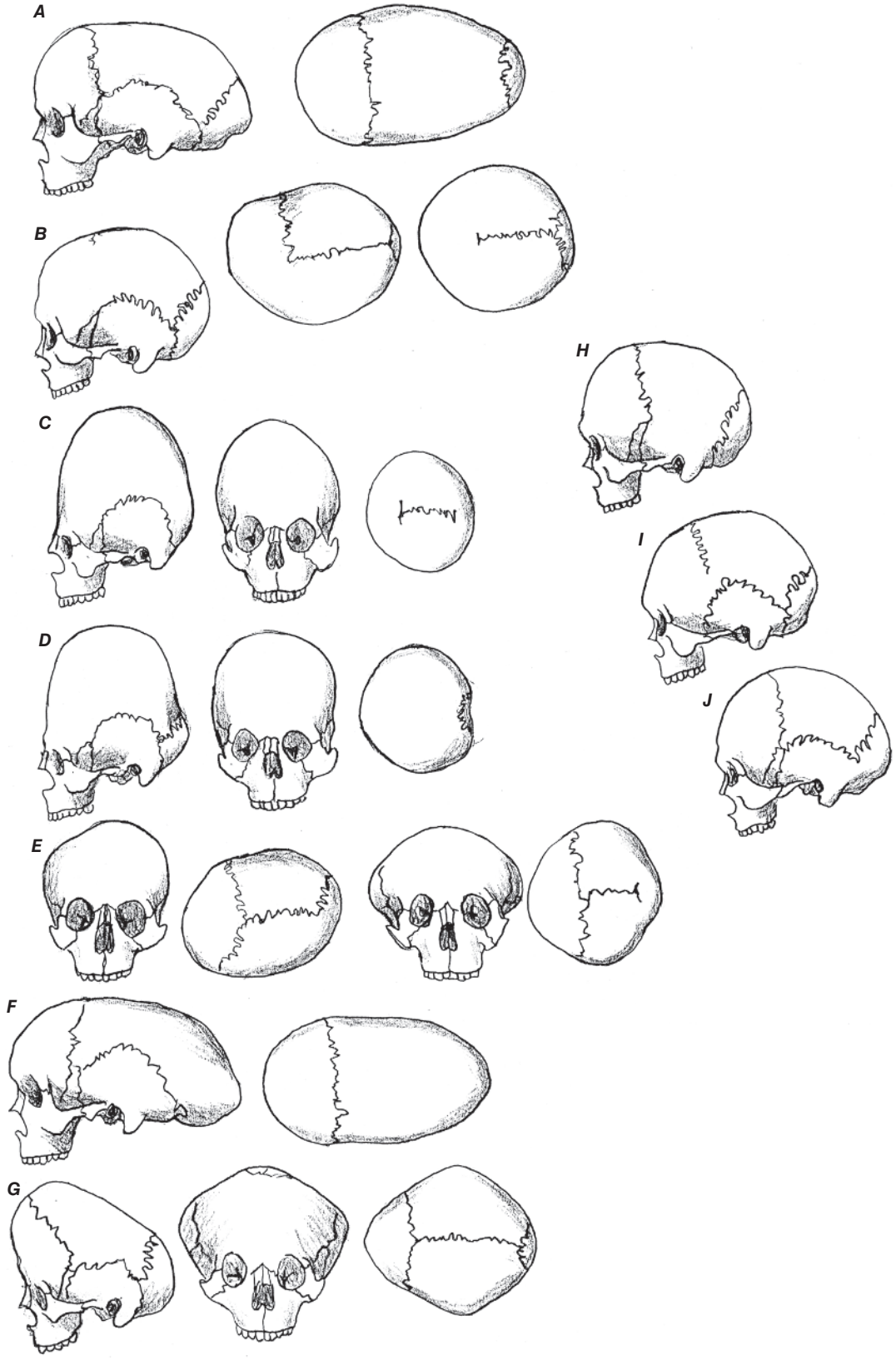

FIGURE A-1.3.1. Sutural agenesis: (*A*) sagittal suture agenesis—scaphocephalic deformity; (*B*) coronal suture agenesis—unilateral or partial, may have plagiocephalic deformity, bilateral—brachycephalic form; (*C*) coronal and lambdoidal suture agenesis—oxycephalic (tower skull) deformity; (*D*) coronal and sagittal suture agenesis—oxycephalic (tower skull) deformity with bulging occipital; (*E*) lambdoidal suture agenesis—unilateral or partial, may have plagiocephalic deformity, bilateral—brachycephalic form; (*F*) lambdoidal and sagittal suture agenesis—dolichocephalic form; (*G*) metopic suture agenesis—trigonocephalic deformity; (*H*) temporal squamosa suture agenesis—no deformity; (*I*) sphenofrontal suture agenesis—no deformity; (*J*) temporoccipital suture agenesis—no deformity.

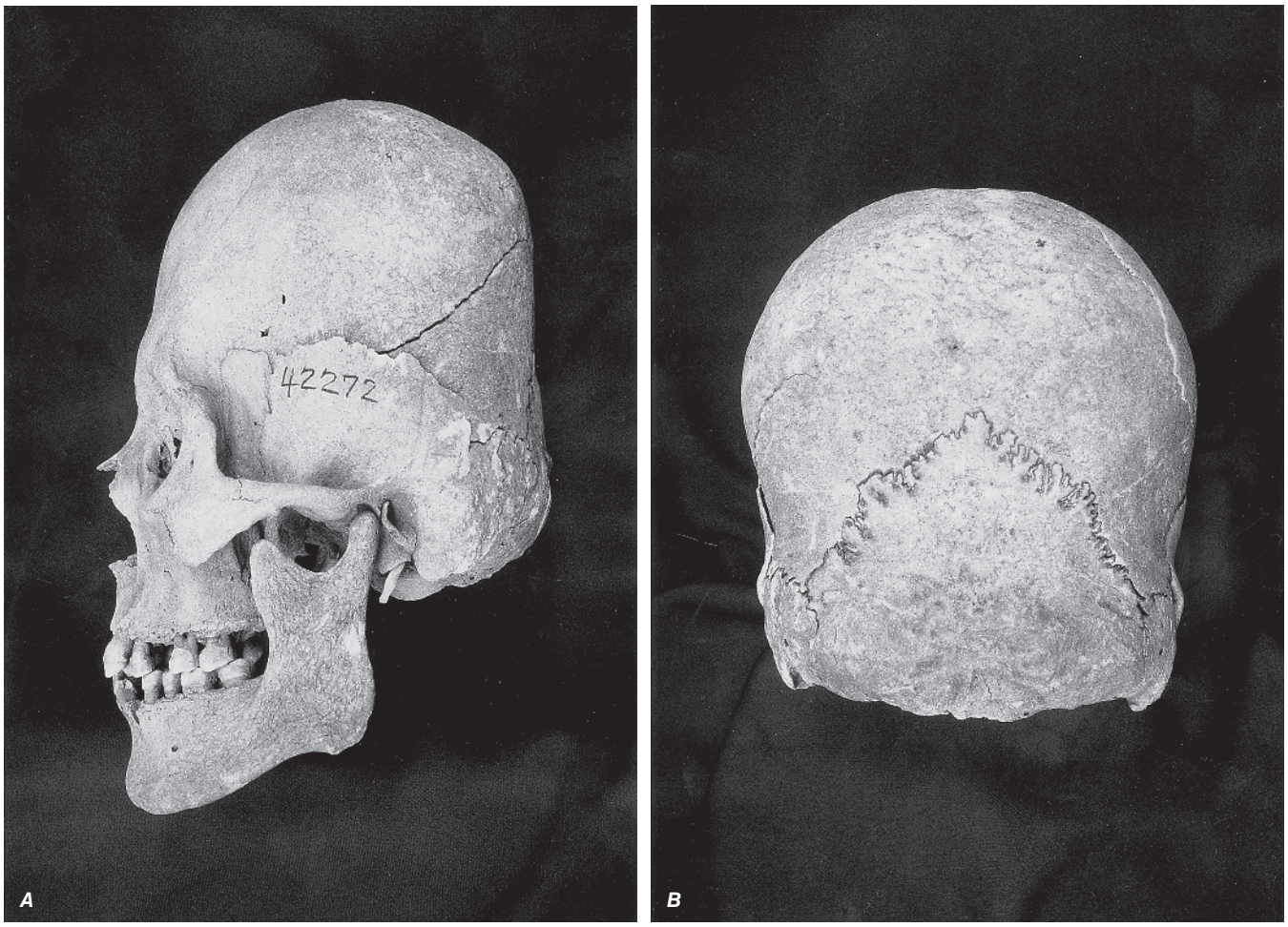

FIGURE A-1.3.2. Sutural agenesis—oxycephaly: coronal and sagittal sutures absent, adult female, Little Colorado River, AZ; (*A*) lateral and (*B*) occipital views (Field Museum).

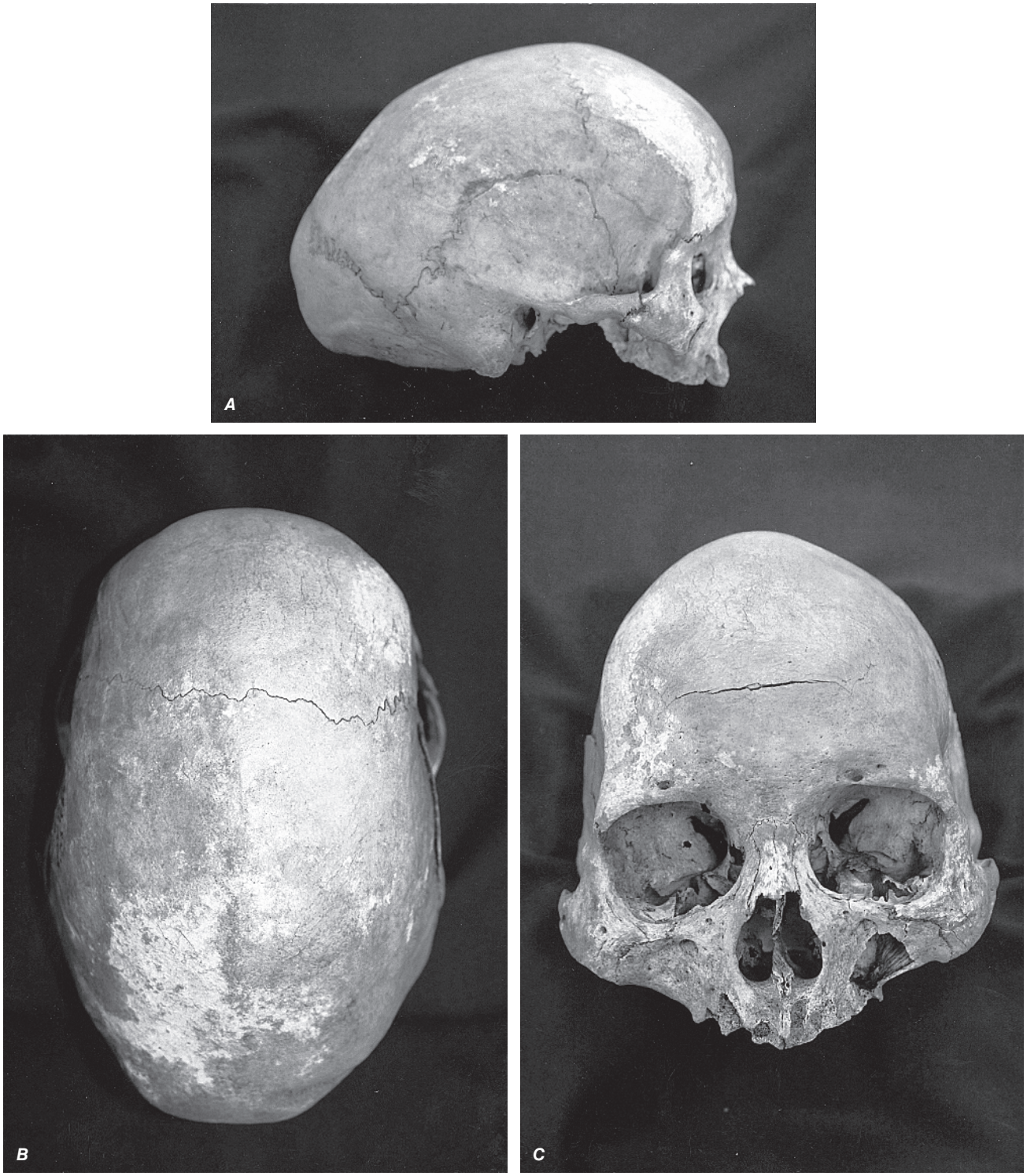

FIGURE A-1.3.3. Sutural agenesis—scaphocephaly: sagittal suture absent, adult male (NMNH 293841), Cerros, Peru; (*A*) lateral, (*B*) top, and (*C*) facial views.

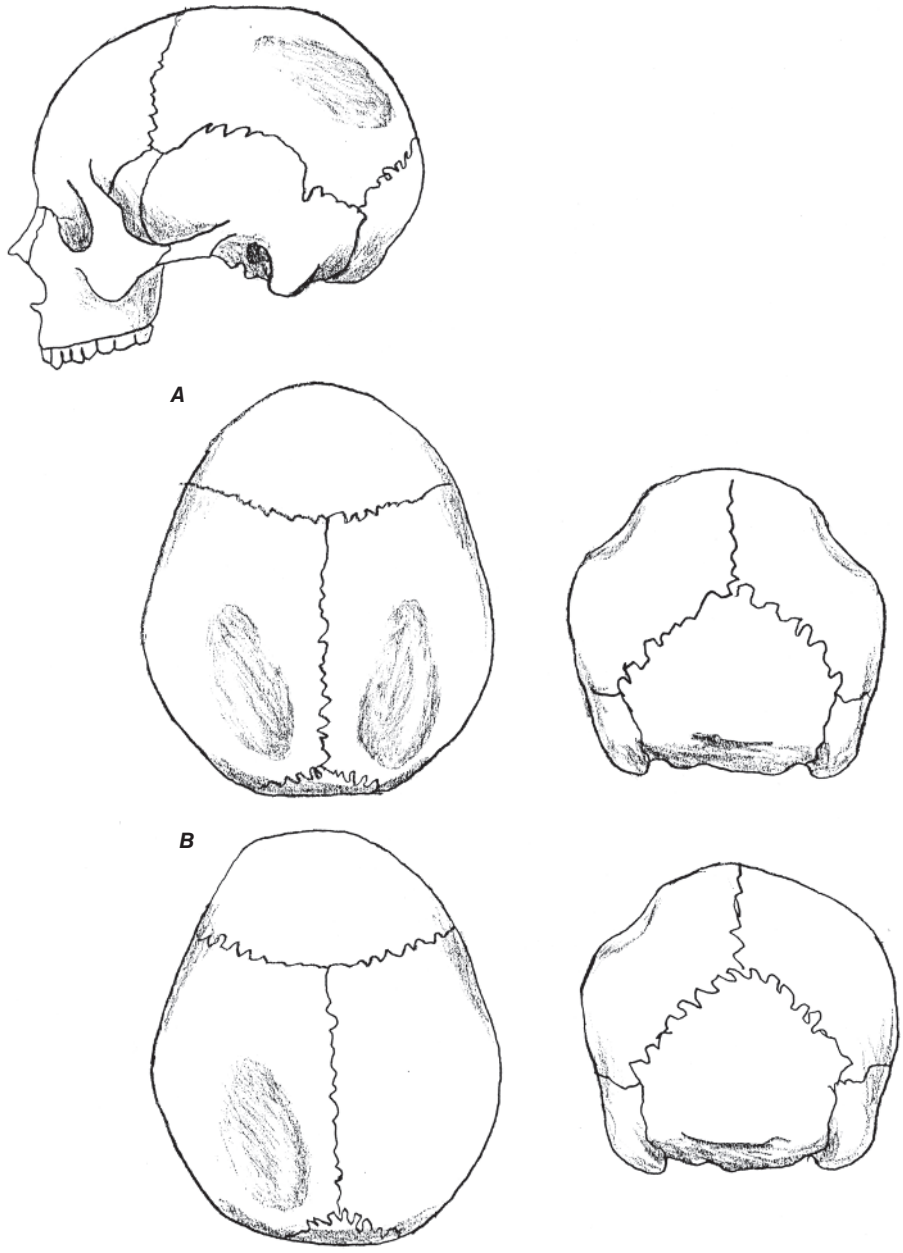

FIGURE A-1.4.1. Parietal thinning: (*A*) bilateral; (*B*) unilateral.

FIGURE A-1.4.2. Parietal thinning close-up: right side, adult male (NMNH 294027), N. Sta. Lucia, Peru.

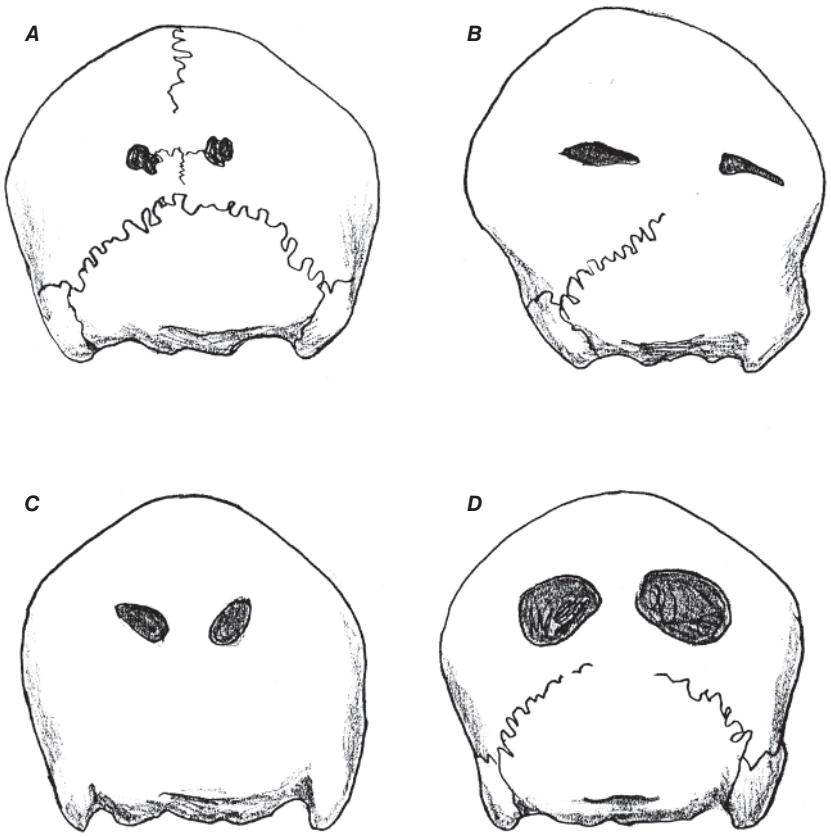

FIGURE A-1.5.1. Enlarged parietal foramina: (*A*) cruciform shape with localized sagittal suture agenesis; (*B*) slit shape with coronal, sagittal, and unilateral right lambdoidal suture agenesis; (*C*) lozenge shape with sagittal and lambdoidal suture agenesis; (*D*) large ovoid shape with partial sagittal and lambdoidal suture agenesis (Hoffman 1976).

FIGURE A-1.5.2. Enlarged parietal foramina: adult male (NMNH 276981) with agenesis sagittal and lambdoidal sutures, Ponce Mound, Santa Clara county, CA.

FIGURE A-1.5.3. Enlarged parietal foramina slits: 4-year-old child (NMNH 276982) with agenesis sagittal suture, right side of the lambdoidal suture, and midportion (bregma area) of the coronal suture, near Palo Alto, CA.

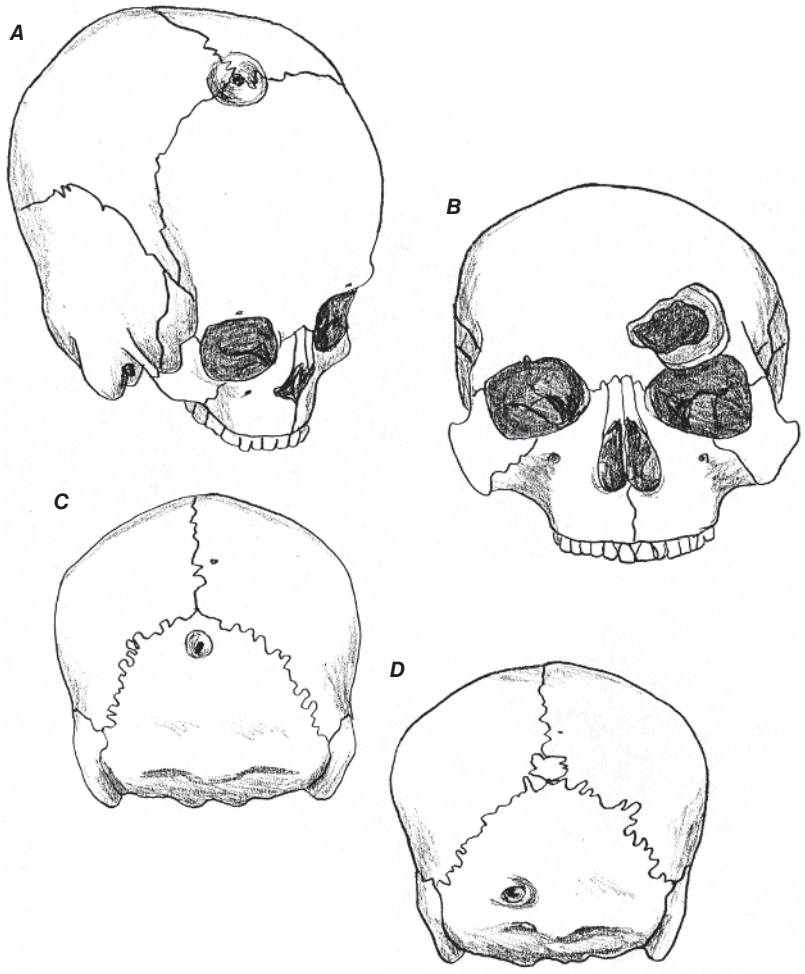

FIGURE A-1.6.1. Cranial inclusion (dermoid) cysts: (*A*) at the bregma; (*B*) involving the supramedial margin left eye orbit; (*C*) below the lambda; (*D*) above the nuchal ridge, left of center.

leaves a rounded depression with a sharp border, often with a thin bony floor (Fig. A-1.6.2). Sometimes an external dermoid cyst communicates with an internal ectodermal cyst by way of a dermoid sinus through a rounded opening in the floor of the external cyst, penetrating through all layers of the bone (Fig. A-1.6.3), or ending blindly within the bony diploe (Rubin et al. 1989; Scheie and Albert 1977).

A-1.7. Cranial Neural Tube Defects

These occur with neurulation disturbances in the cranial end of the developing neural folds and tube. If the neural folds fail to fuse, the developing brain and upper spinal cord develop abnormally with exposure to the amniotic fluid—craniorachischisis, and the stimulus for the corresponding bone development is impaired, including cervical and thoracic neural arch development that leaves them widely spaced in a flayed appearance. Fetal death usually occurs with this disorder in the first trimester (Dudor 2010).

Developmental failure of the anterior neuropore to form causes failure of the brain tissue to form above the brain stem—anencephaly, resulting in death before or shortly after birth. Development of the membranous cranial vault stimulated by the developing brain is severely impaired with severe reduction and deformity of the bones within the desmocranium. The developing chondocranial base is also affected with deformities, especially midline structures (Dudor 2010).

Developing brain tissue can also be disrupted with delay in closure of the anterior neuropore, a postneurulation defect. This allows brain tissue and or the meningeal brain covering to protrude through an opening of overlying developing bone tissue. The abnormally placed tissues are encased in the covering epidermal tissue, appearing as a skin-covered cyst. Whether brain tissue or just the meningeal brain

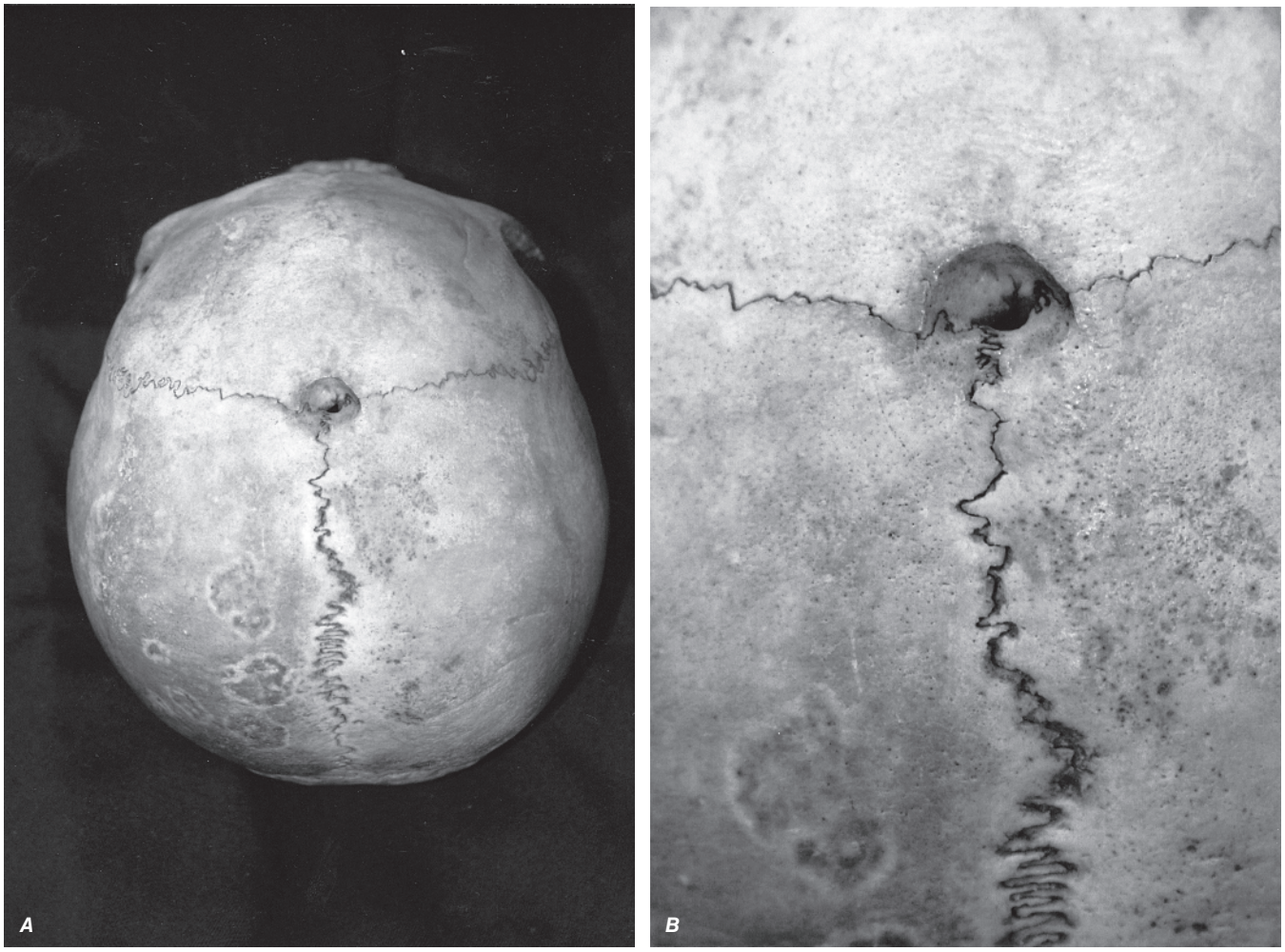

FIGURE A-1.6.2. Cranial inclusion (dermoid) cyst at the bregma: (*A*) adult female (NMNH 264629), Chicama, Peru; (*B*) close-up (Barnes 1994:53; I mistakenly identified this a meningocele).

covering is involved in the defect depends on the timing of the delay. When only the meninges protrude through the adjacent developing bone, it is known as a meningocele, and when the brain tissue is involved, it is an encephalocele. They usually develop along the sagittal plane from the nasion root to the base of the occipital, sometimes occurring in the roof of the orbital angle or root of the sella turcica (Fig. A-1.7.1). Individuals with a meningocele can survive into adulthood, while those born with an encephalocele usually do not survive infancy. Encephaloceles generally develop at the base of the skull and can be quite large with the affected bone appearing bifurcated (Lemire 1988). The anterior fontanelle or bregma region is a common site for a meningocele, leaving a depressed cystic impression of varying size on affected bone with an irregular opening through the bony floor (Fig. A-1.7.2). The saucer-shaped or rounded bony depression has well-defined raised borders surrounded by an outer flange of bony buildup responding to pulsations from the meningeal tissue (Webb and Thorne 1985).

A-1.8. Hydrocephaly

This is an abnormally enlarged cranium caused by increased accumulation of cerebrospinal fluid within the ventricles of the brain resulting from interference with its normal circulation and absorption throughout the spinal and cerebral chambers. This can be due to developmental disturbances, particularly spinal neural tube defects (spina bifida) or defects in the subarachnoid spaces or aqueduct of Sylvius. Disturbances can also arise before or after birth from injury, infection, or brain tumors. Therefore, developmental cause and effect may be difficult to establish. Developmentally related hydrocephalics can survive beyond birth. Normal neonate cranial circumference ca. 35 cm will increase almost 1 cm per month until reaching ca. 46 cm at 1 year while

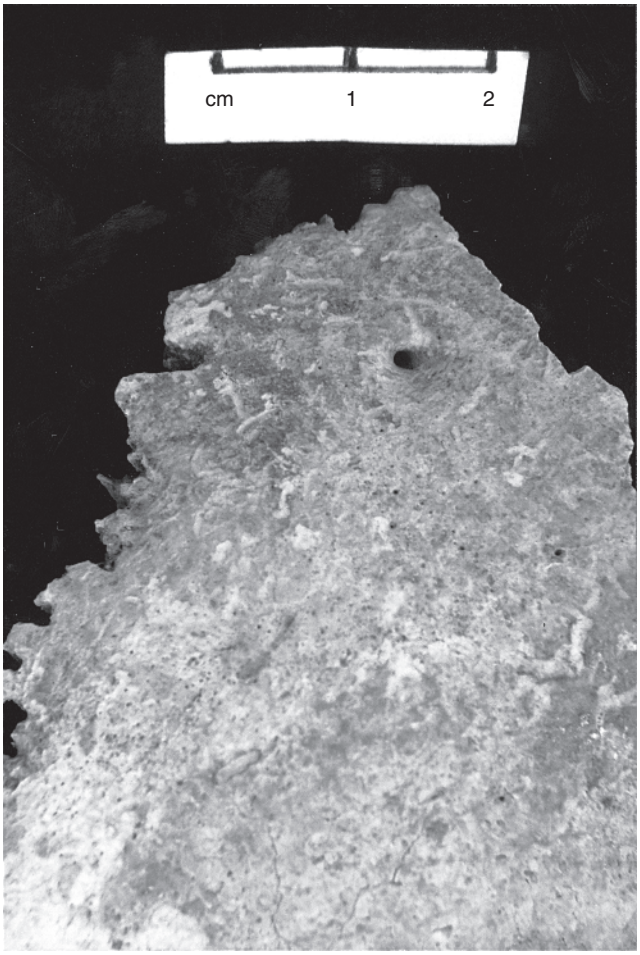

FIGURE A-1.6.3. Cranial inclusion (dermoid) cyst near the lambda: occipital of an adult male, Frankish Corinth, Greece.

the hydrocephalic skull will be much larger (Warkany 1971). The cranium is unusually large and globular in shape with enlarged anterior fontanelle that fails to close. The face appears disproportionately small with roof of eye orbits appearing raised upward (Fig. A-1.8). Signs of developmental defects affecting the aqueduct of Sylvius with inadequate circulation of fluid can be delayed until early adolescence.

A-1.9. Microcephaly

An abnormally small cranium resulting from a defective developing small brain is known as microencephaly, with the forebrain and occipital lobes primarily affected. The cranial sutures usually remain in place as with normally developing calvaria. Head circumference in affected older children and adults falls below 46 cm (about the size of a 1-year-old) with brain weight less than 900 g (Goodman and Gorlin 1983; Warkany 1971). The cranium has a conoidal ("pinhead") shape from narrow, receding frontal and shortened occipital that is

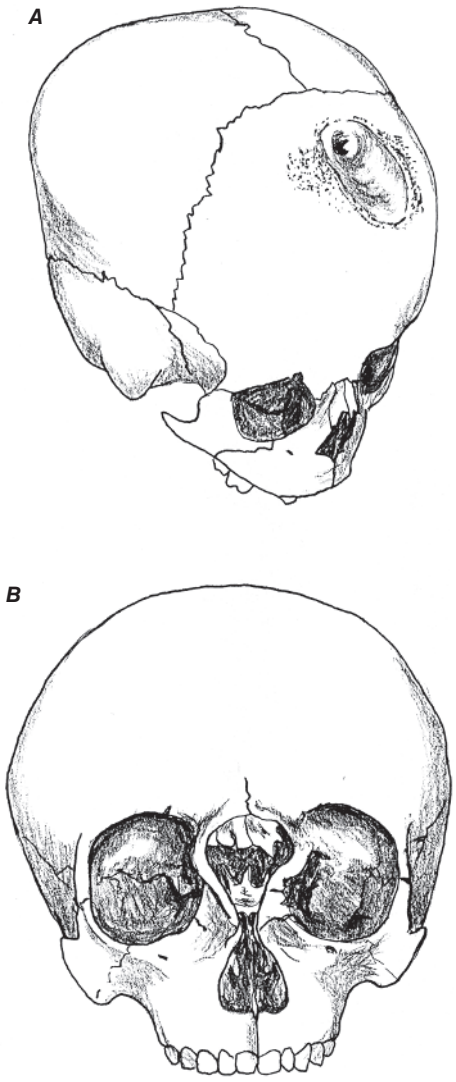

FIGURE A-1.7.1. Cranial neural tube defects: (*A*) frontal meningocele; (*B*) nasal region encephalocele (drawn from Ortner 2003:455).

often flattened (Fig. A-1.9.1). The face appears large compared with the abnormally small-sized skull (Fig. A-1.9.2). The affected individual suffers severe mental retardation (idiot) and can live into adulthood, able to perform simple duties or tricks. Microcephaly can be familial.

A-2. FACE DEVELOPMENT

The membranous bones of the upper midface evolve from the bulging viscerocranium below and forward of the evolving blastemal desmocranium, as neural crest cells migrate into the region from the mid- and hindbrain areas. The center swells to form the frontonasal prominence responsible for the upper midregion of the

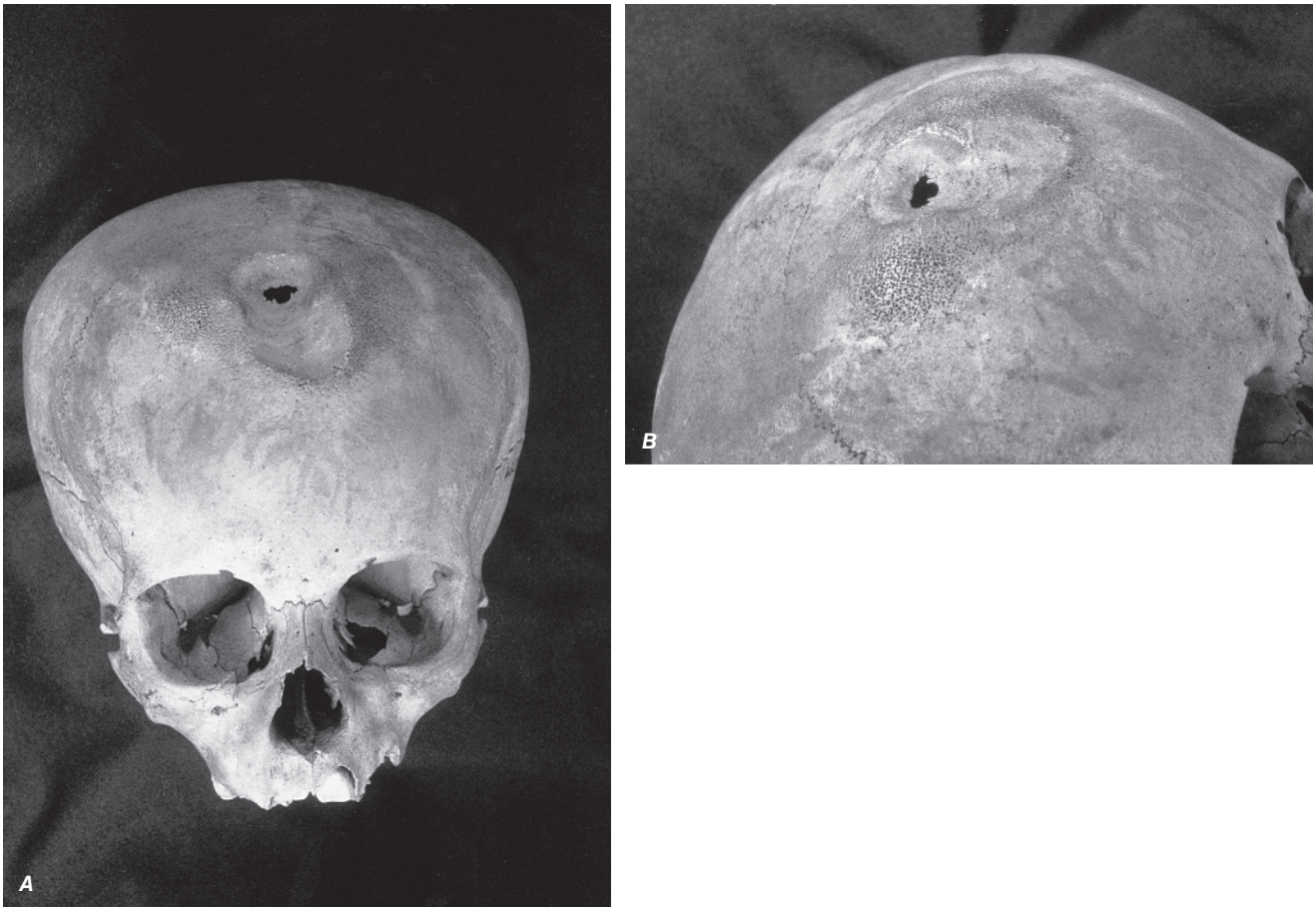

FIGURE A-1.7.2. Meningocele neural tube defect: (*A*) near the bregma on frontal, 7- to 8-year-old child, Ancon, Peru (Field Museum); (*B*) close-up.

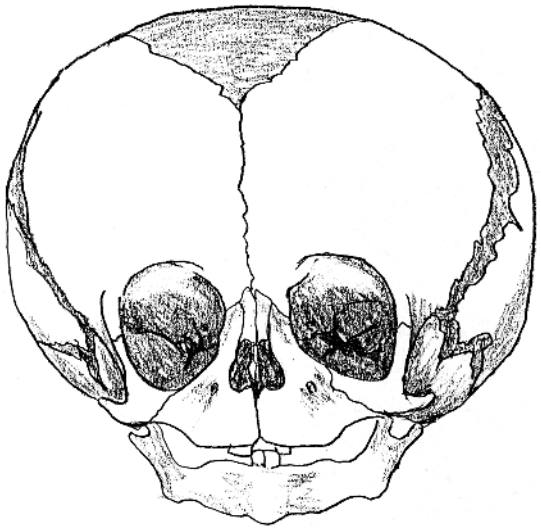

FIGURE A-1.8. Hydrocephaly: infant (drawn from Aufderheide and Rodriguez-Martin 1998:57).

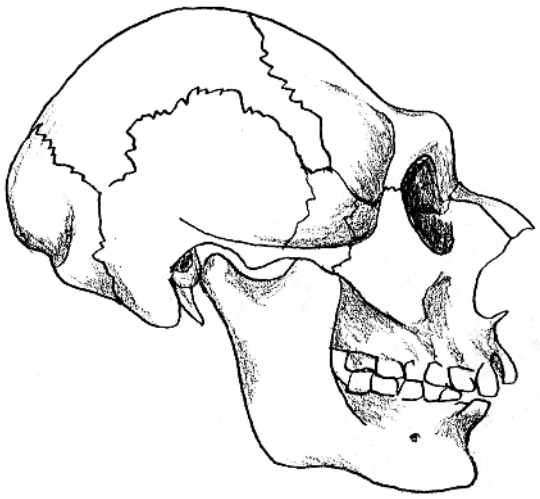

FIGURE A-1.9.1. Microcephaly: adult (drawn from Brothwell 1981:169).

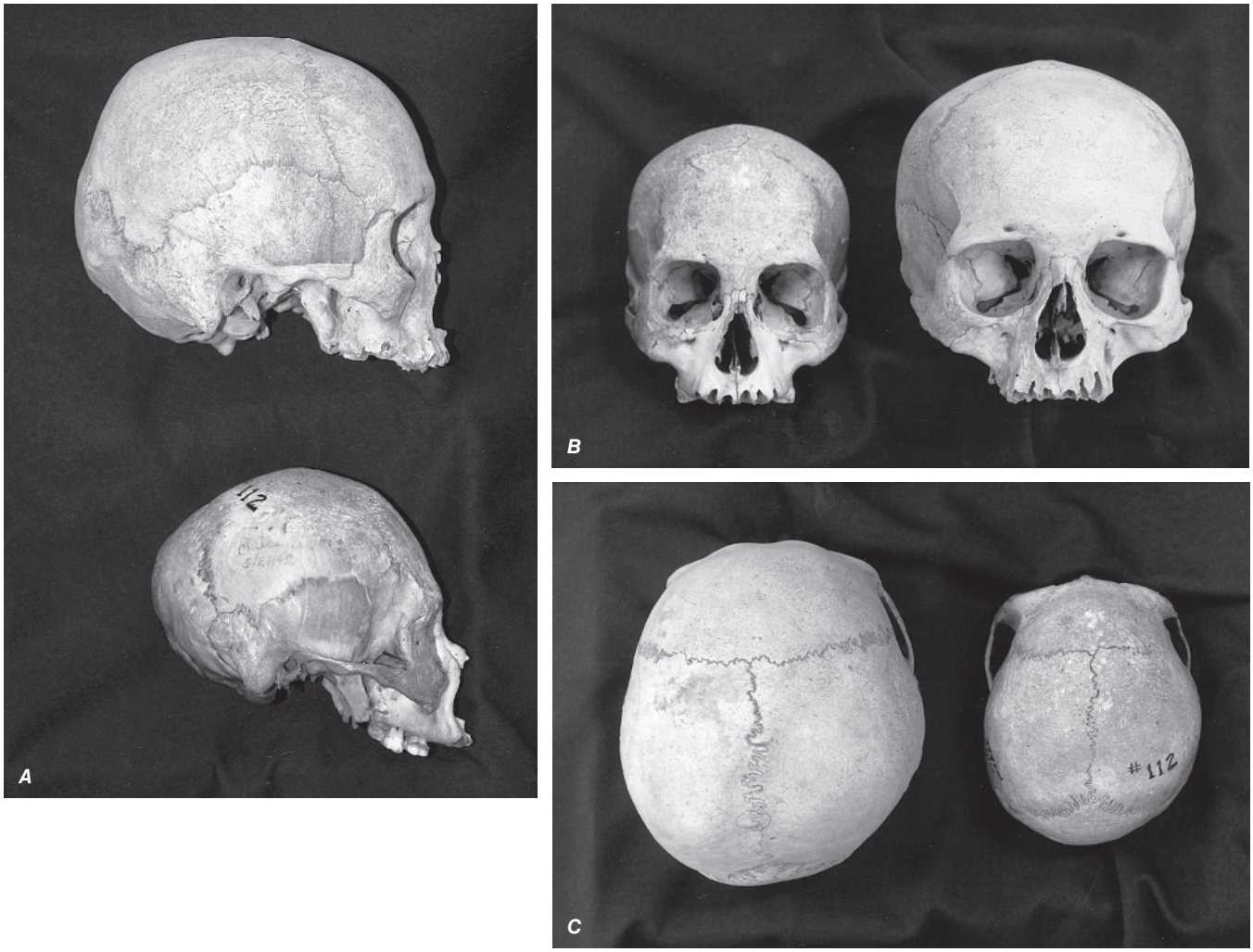

FIGURE A-1.9.2. Microcephaly: adolescent female (NMNH 379510), Chicama, Peru, with normal adult female skull. (*A*) lateral, (*B*) faces, and (*C*) skull tops.

face, while the first pair of pharyngeal or branchial arches appearing along each side (Fig. II) forms the lateral and lower portions of the face—the maxilla, mandible, zygomatics as well as the temporal squamosa, parts of the sphenoid, palatine bones, and malleus and incus ear bones. By the end of the fourth week, the face begins to take shape around the primitive mouth (stomodeum) as the five merging developmental fields—the frontonasal prominence and paired maxillary and mandibular prominences from the first pharyngeal arch—come together. Paired nasal placodes soon appear on the frontonasal prominence to form the nasal pits with raised rims dividing the prominence into paired lateral and medial nasal parts. The medial nasal parts immediately join with the formation of the primordial nasal bones and bony septum formed by the ethmoid and vomar bones within the primitive nasal cavity. The inferior portion expands bilaterally below the nasal cavity to form the intermaxillary process, where the paired halves of the bony premaxilla take shape and fuse together. The premaxilla unites with the two halves of the primordial maxilla to form the upper jaw and palate. Ectodermal grooves develop between the lateral nasal processes and maxillary prominences as they move toward each other, forming nasolacrimal grooves that become the nasolacrimal ducts bounded by the lacrimal bones as the membranous bones meet and fuse together (Fig. A-2.0.1). Between the eighth and ninth weeks the medial walls of the maxillae extend downward to form palatine shelves as the developing tongue moves downward, joining with the small primary palatal extension from the premaxilla. As the two halves expand and come together, they form the primordial membranous bony palate (Fig. A-2.0.2) while continuing to position into place and join with the horizontal plates of the palatine bones. The posterior portion of

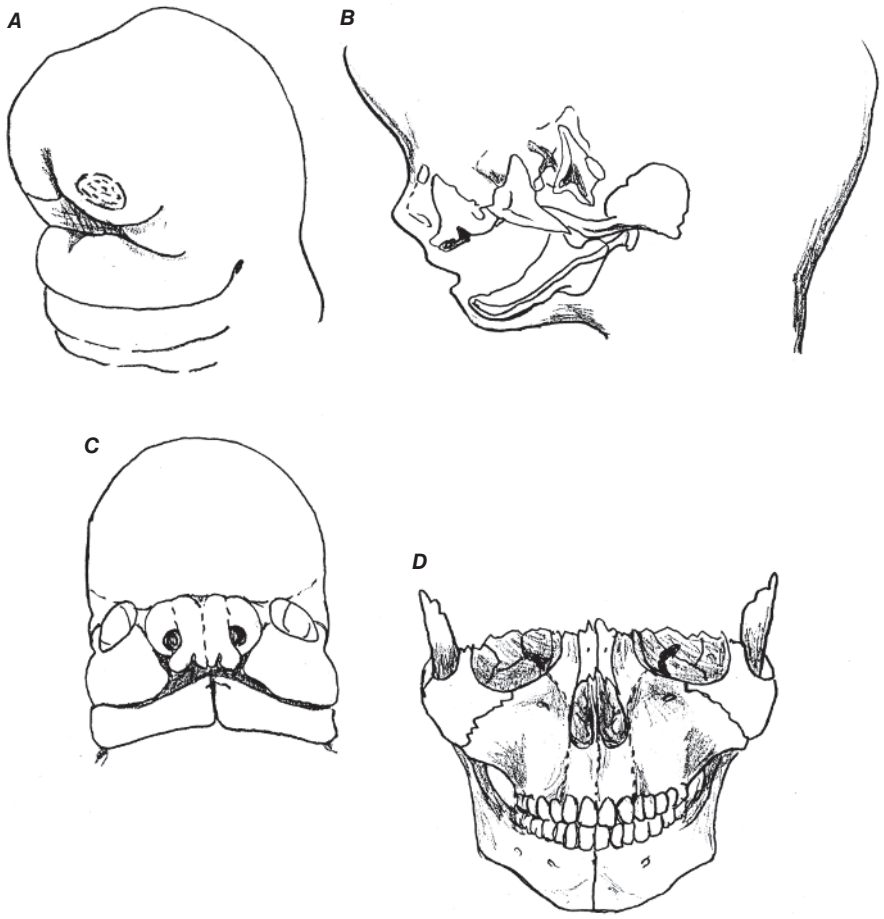

FIGURE A-2.0.1. Facial development: (*A*) frontonasal prominence with the left nasal placode, paired maxillary and mandibular prominences from the first pharyngeal arch surround the primitive mouth (30 days); (*B*) corresponding developing membranous facial bones (9 weeks); (*C*) frontonasal prominence subdivides into medial and lateral nasal parts as maxillae move toward the center and mandibular parts come together (7 weeks); (*D*) corresponding developmental fields (dotted lines) within a mature bony face.

the palatine bones does not ossify but develops into the soft palate and uvula.

Development of the two halves of the mandible depends on the placement of cartilaginous bars known as Meckel's cartilage, extending throughout each mandibular prominence, attracting neural crest cells to form the membranous mandibular bony tissue around them (Fig. A-2.0.1B). Six ossification centers appear in each mandibular half by the eighth week. One ossification center appears in the lower border, in front of the alveolar process, one in the distal end of Meckel's cartilage in the region of the symphysis, one for the coronoid, one in the cartilage for the condyle and top of ramus, one at the mandibular angle, and one for the inner alveolar plate. They all coalesce together by the twelfth week except for the cartilage-bound condyle that develops into two parts by adolescence, separated by a fibrous strip to help mold it into shape. As the fibrous strip recedes, the condyle fuses to the mandible.

FACIAL ANOMALIES

A-2.1. Facial Clefts

These are very rare, arising as the frontonasal process median and lateral prominences, and maxillary prominences grow toward each other to unite. Ectodermal tissue overlies all of the developing mesenchymal facial parts and forms grooves between them. As the mesenchymal prominences move toward each other, they penetrate the ectodermal tissue in the grooves and unite. Failure of penetration at the critical threshold time of union leaves a fissure between the opposing prominences. Hypoplasia with developmental delay affecting one or both of the adjacent facial parts is often the cause with the resulting cleft formation reflecting the degree of developmental delay (Fig. A-2.1.1). The cleft is usually unilateral, complete or incomplete, and fissures can occur anywhere in the facial region where two parts

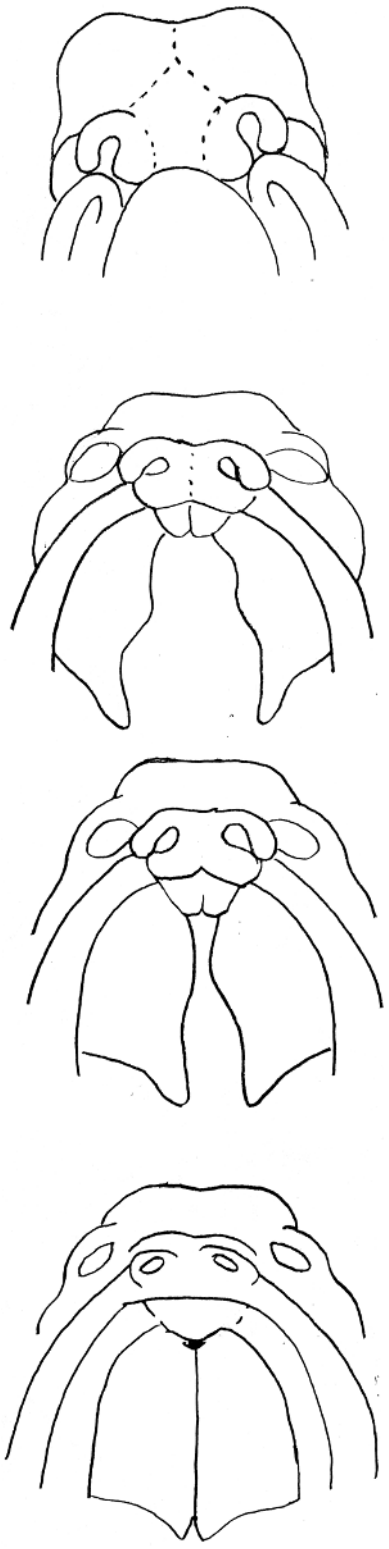

FIGURE A-2.0.2. Embryonic development of the palate: top to bottom—the inferior portion of the two subparts of the frontonasal process unite and extend downward to form the primary palate between the developing maxillary palatine shelves that grow toward each other, uniting with the premaxillary primary palate.

are programmed to meet. With failure of the maxillary and lateral nasal prominences to unite, a nasomaxillary cleft forms between the maxillary and lateral nasal prominence, from the oral cavity to the eye orbit. The naso-ocular groove between the lateral and median nasal prominence becomes the nasolacrimal duct, but when the two parts fail to reach each other at the critical threshold time, the two become divided by a naso-ocular cleft that may extend to the oral cavity (Fig. A-2.1.2). Bilateral clefts can be symmetrical or asymmetrical, and severe forms are generally associated with developmental disturbances of the brain and do not survive fetal life. Mild forms of facial cleft formation often reach adulthood with the cleft usually covered by fibrous tissue (Burdi et al. 1988; Mladick et al. 1974).

Midface hypoplasia of one or both parts of the median nasal prominence creates a median or midline facial cleft as the two parts fail to come together and fuse on time (Figs. A-2.1.1C and A-2.1.2C), often leaving a wide gap between the eye orbits (hypertelorism). The premaxilla may have central notching of the nasal border or a cleft extending between the central incisors. The nares are wide and the nasal root is broad with wide nasal bones attached at odd angles, frontal sinuses are atypical or absent, and the vomer and perpendicular plates of the ethmoid are usually hypoplastic.

Severe median clefts from aplasia or severe hypoplasia are usually associated with brain defects and are very rare and fatal during the perinatal period, with the eye orbits forced close together (hypotelorism) as the bones of the nares and premaxilla fail to develop (DeMyer 1967).

A-2.2. Nasal Bone Hypoplasia/Aplasia

Small or absent nasal bones can occur independent of other facial developmental disturbances resulting in unilateral or bilateral expressions (Fig. A-2.2).

A-2.3.1. Cleft Lip

It is actually a cleft premaxilla, including its primary palate. This usually involves unilateral or bilateral hypoplasia or aplasia of the premaxilla, leaving a separation between adjacent bony structures. Mild forms of clefting appear as notches between affected bony parts. Since the incisors are contained within the premaxilla, wherever the cleft penetrates the alveolar margin, the development of the associated incisor or incisors is disrupted, most often with agenesis. Incomplete expressions are usually expressed as unilateral hypoplasia with some union of the two premaxillary halves. The alveolar margin may be thin and directed upward with central incisors disturbed or absent. The nares on the hypoplastic side may appear lower than the other side. Expressions of cleft lip are varied as clefts can form between the maxilla and premaxilla or between the two

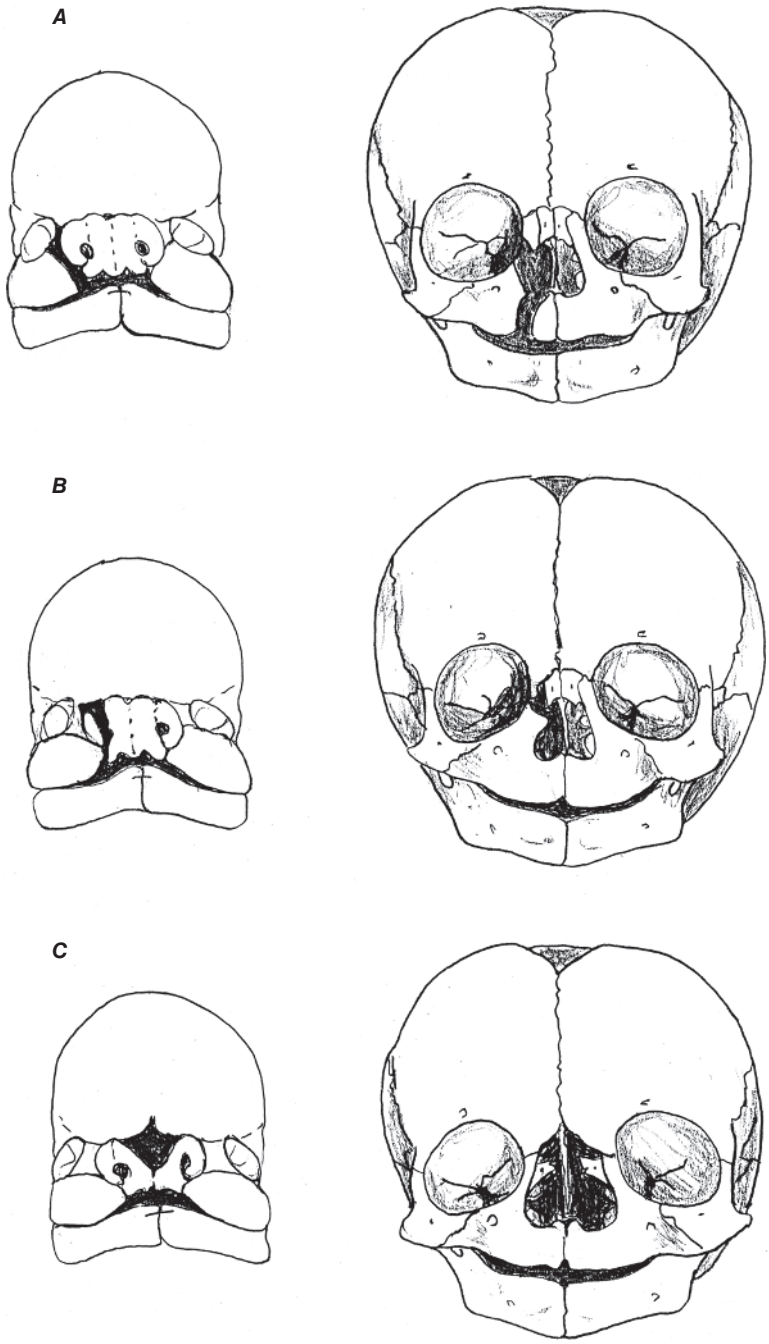

FIGURE A-2.1.1. Facial cleft development: from embryo to newborn—(*A*) nasomaxillary cleft; (*B*) naso-ocular cleft; (*C*) median cleft.

halves of the premaxilla. Unilateral cleft lip forming between the maxilla and premaxilla, most often on the left side, is more common than bilateral expressions, and this type has a greater chance of survival. Severe bilateral cleft with remnant premaxilla usually presents as a rounded ball-like extension between the clefts with all incisors absent or grossly distorted (Fraser 1963). Midline cleft lip from failure of the two subparts of the premaxilla to unite can vary from a slight cleft to a wide cleft with agenesis of one or both halves. Mild expressions of midline clefts appear as notches or indentations between the central incisors (Fig. A-2.3.1).

A-2.3.2. Cleft Lip with Cleft Palate

The majority of cleft lip (cleft premaxilla) expressions are accompanied by cleft maxillary palate. Premaxilla hypoplasia or aplasia, complete or incomplete, often disrupts the development of the adjacent maxillary palate (Figs. A-2.3.2–A-2.3.6).

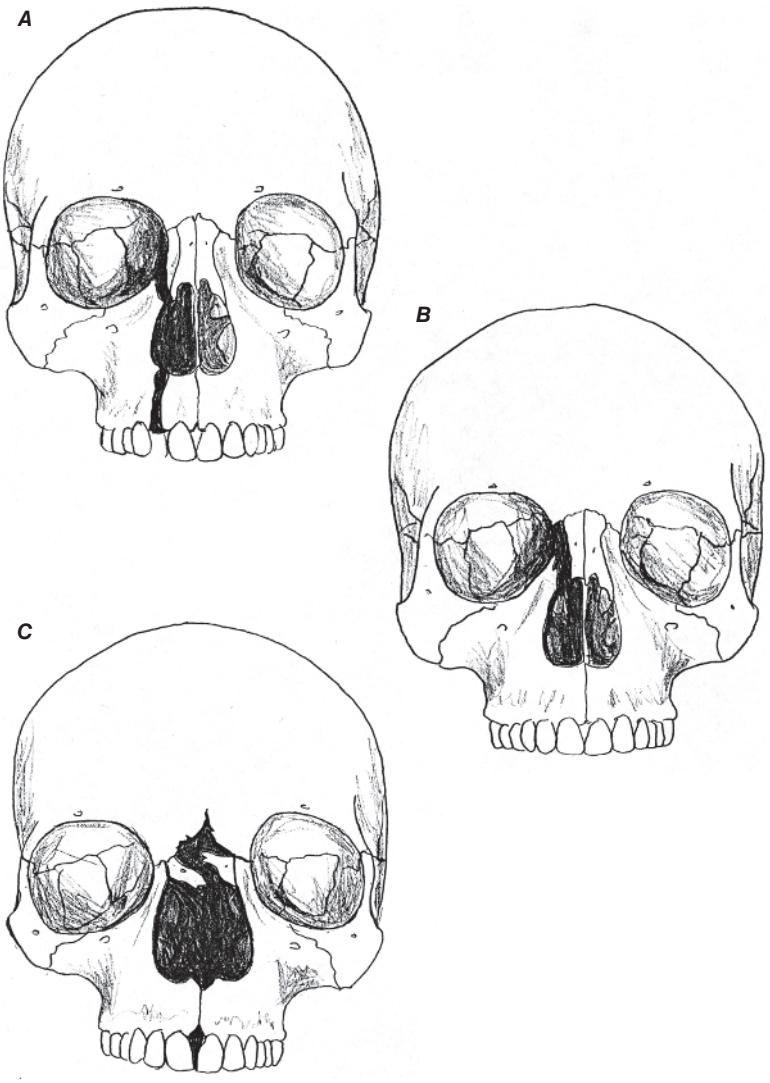

FIGURE A-2.1.2. Facial clefts: (adult) (*A*) nasomaxillary cleft; (*B*) naso-ocular cleft; (*C*) median cleft with wide nares and hypertelorism.

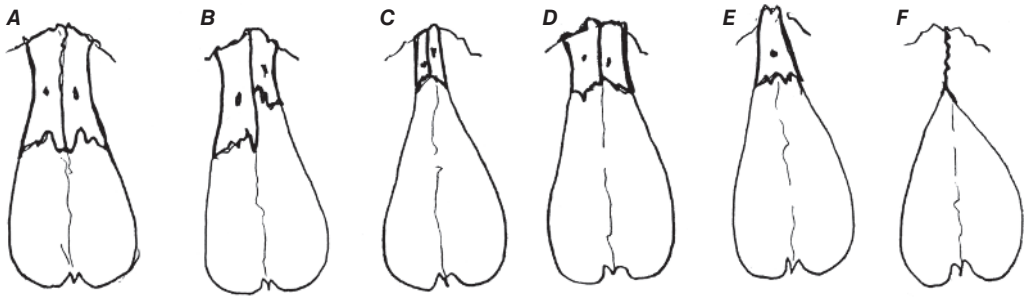

FIGURE A-2.2. Nasal bone hypoplasia/aplasia: (*A*) normal; (*B*) unilateral left hypoplasia; (*C*) bilateral severe hypoplasia; (*D*) bilateral mild hypoplasia; (*E*) single nasal bone from unilateral aplasia; (*F*) bilateral aplasia.

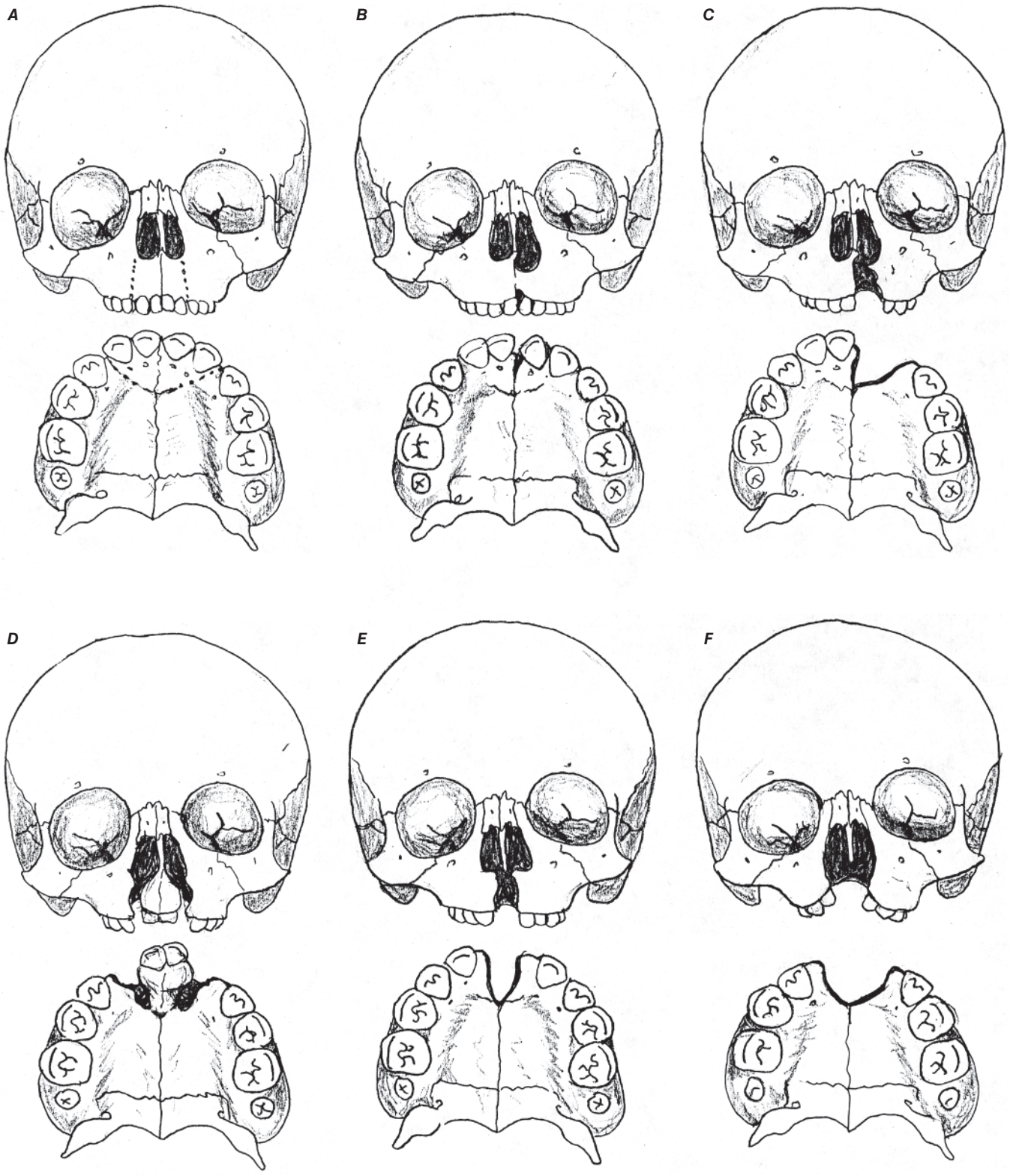

FIGURE A-2.3.1. Cleft lip (cleft premaxilla): (young child) (*A*) normal with dotted lines outlining the premaxilla; (*B*) incomplete unilateral left cleft; (*C*) complete left unilateral cleft; (*D*) bilateral cleft; (*E*) midline cleft; (*F*) agenesis of the premaxilla—wide cleft.

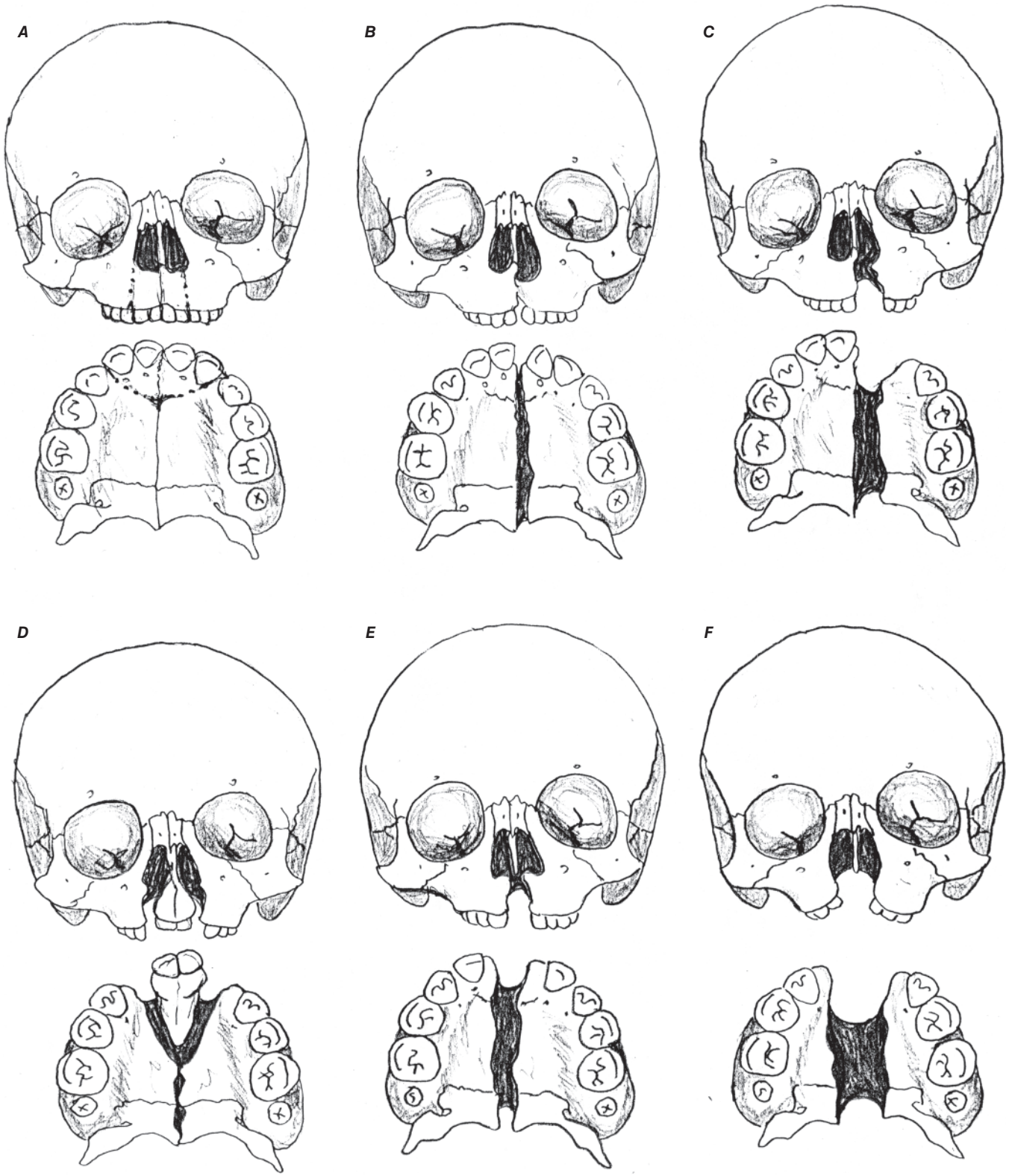

FIGURE A-2.3.2. Cleft lip (premaxilla) with cleft (maxillary) palate: (young child) (*A*) normal with dotted lines outlining the premaxilla; (*B*) incomplete left cleft lip with unilateral left cleft palate; (*C*) unilateral left cleft lip and palate; (*D*) bilateral cleft lip and palate; (*E*) midline cleft lip and palate; (*F*) agenesis of the premaxilla with wide midline cleft palate.

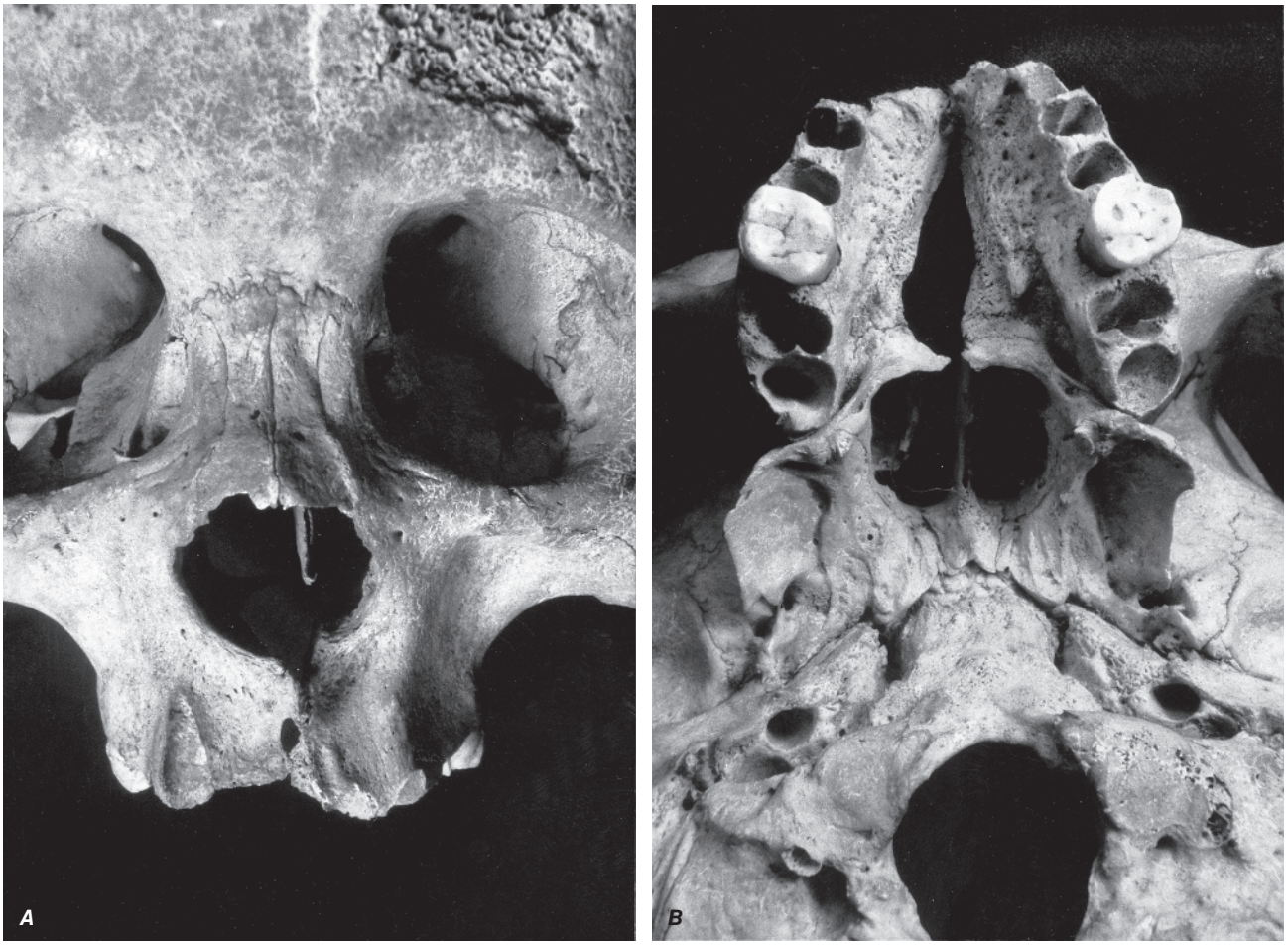

Figure A-2.3.3. Cleft lip with cleft palate: (*A*) unilateral right incomplete cleft lip with (*B*) unilateral right cleft palate, adult female (NMNH 316482), SW Colorado.

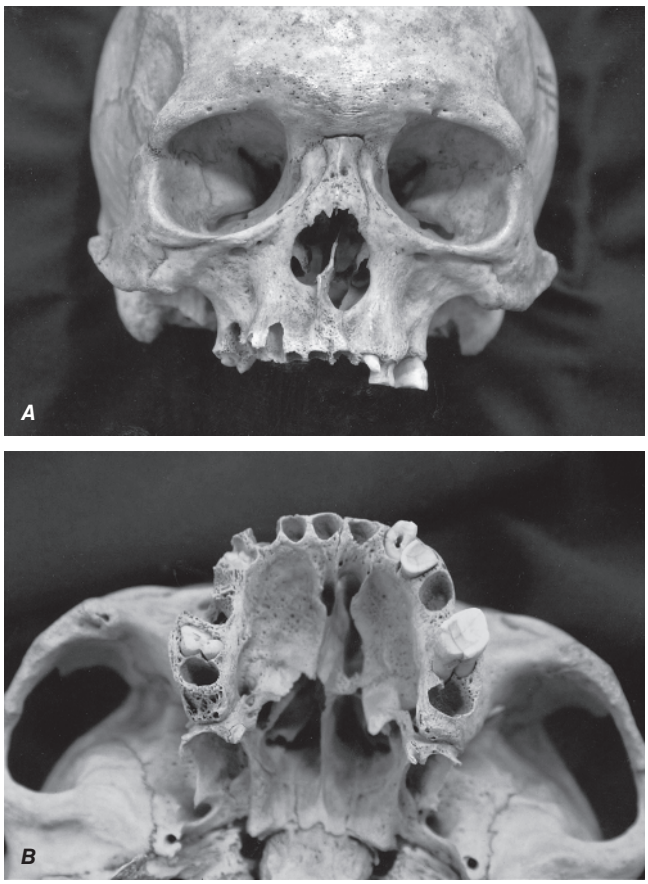

Figure A-2.3.4. Cleft lip with cleft palate: (*A*) unilateral left incomplete cleft lip with (*B*) unilateral left cleft palate, adult male (NMNH 266052), Pachacamac, Peru.

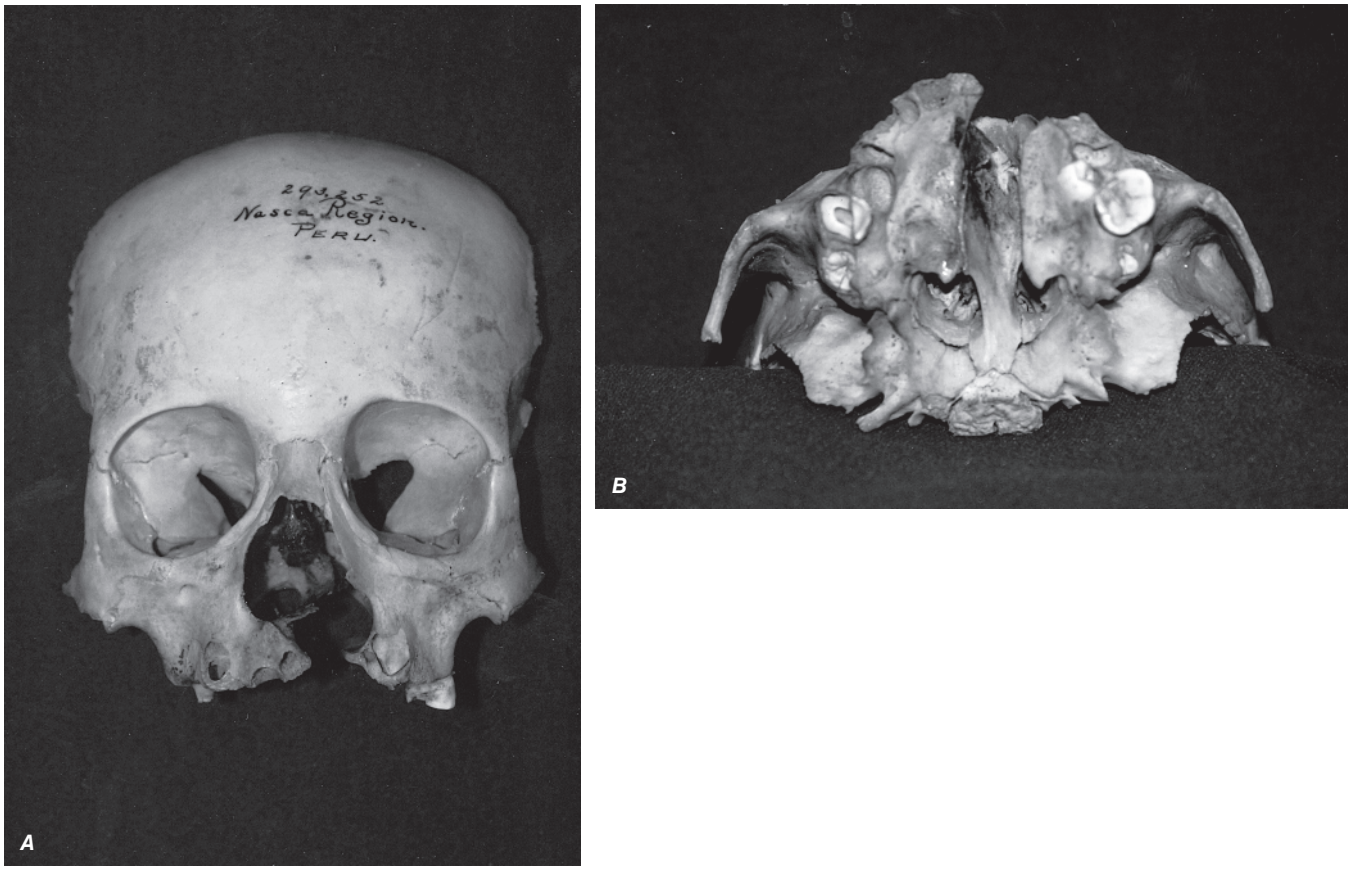

FIGURE A-2.3.5. Cleft lip with cleft palate: (*A*) unilateral left cleft lip with (*B*) unilateral cleft palate, 8- to 10-year-old child (NMNH 293262), Nasca, Peru.

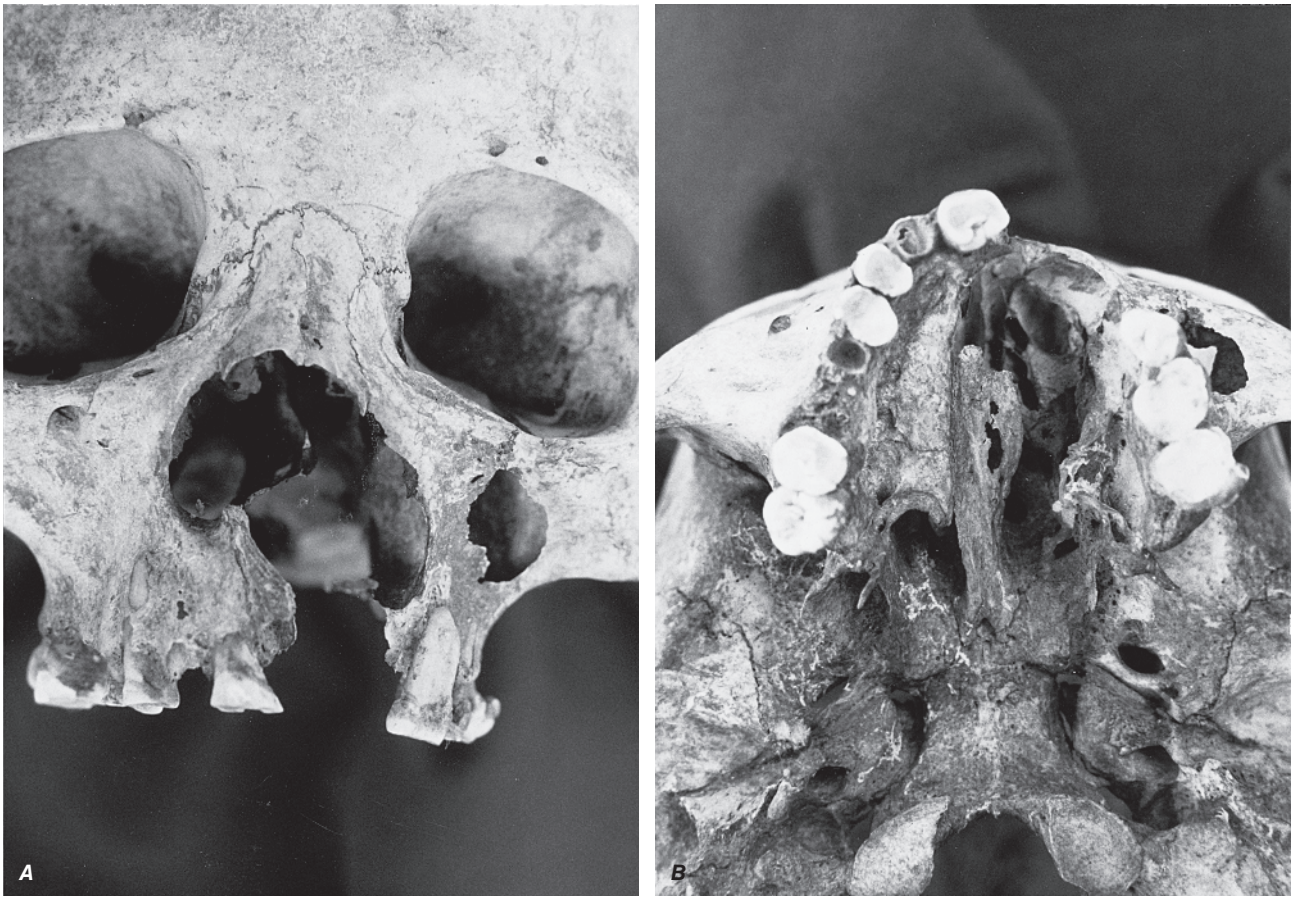

FIGURE A-2.3.6. Cleft lip with cleft palate: (*A*) unilateral left cleft lip with (*B*) unilateral cleft palate, adult female (ASU), Sandoval village, SW Colorado.

A-2.4. Cleft Palate

Isolated cleft maxillary palate evolves as a separate disturbance from cleft lip with cleft palate. Delay in the development and descent of the primitive tongue from the nasal region can interfere with the timing of the approach of the two maxillary palatine shelves toward each other as they meet first with the premaxillary primary palate and end by fusing at the dorsal border. Hypoplasia or aplasia of one or both halves is generally the cause. Cleft palate can be unilateral (the most common form), bilateral asymmetrical, or symmetrical, mild or severe (Fig. A-2.4.1). Mild forms can be expressed as notching of the dorsal border (Fig. A-2.4.2). Bilateral clefts leave the vomer unattached, while a unilateral cleft leaves one side united with the vomer. Clefting also affects the developing soft palate and uvula (Freni and Zapisek 1991).

A-2.5. Cleft Mandible

Very rarely, the two mandibular halves may fail to unite during the first year, with the two halves held together by fibrous tissue (Fig. A-2.5). Mild expressions can appear as a central notch or indentation between the central incisors (Weinberg and Van de Mark 1972).

A-2.6. Mandibular Hypoplasia

This involves the ascending ramus and dorsal portion of the mandibular body. Unilateral expressions are associated with facial asymmetry known medically as hemifacial microsomia. Bilateral expressions (usually mild) often go undetected unless asymmetrical in expression. Adjacent maxillary bone can be affected, particularly the zygomatic process. Various expressions can range from mild to severe (Fig. A-2.6.1). Type I hemifacial microsomia involves only mild/moderate mandibular hypoplasia, while type II involves a greater degree of hypoplasia with a narrow abnormally shaped ramus (Figs. A-2.6.2–A-2.6.5). Aplasia affecting most of the ramus signifies a type III expression (Kaban et al. 1981).

A-2.7. Bifid Mandibular Condyle

This develops when the fibrous tissue septa acting as scaffolding for the calcifying cartilage extending into the

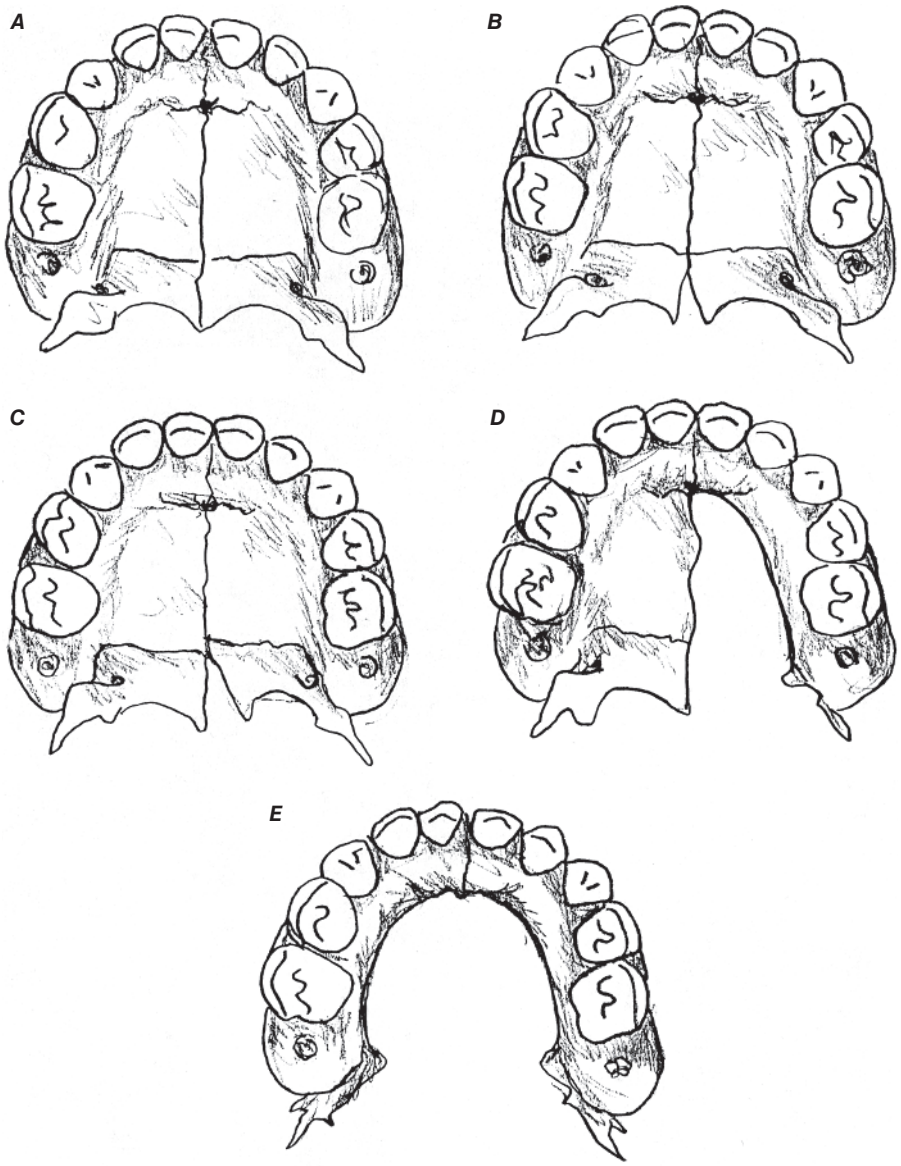

FIGURE A-2.4.1. Cleft palate: (young child) (*A*) normal palate; (*B*) bilateral notched palate; (*C*) unilateral left notched palate; (*D*) unilateral left cleft; (*E*) complete bilateral cleft.

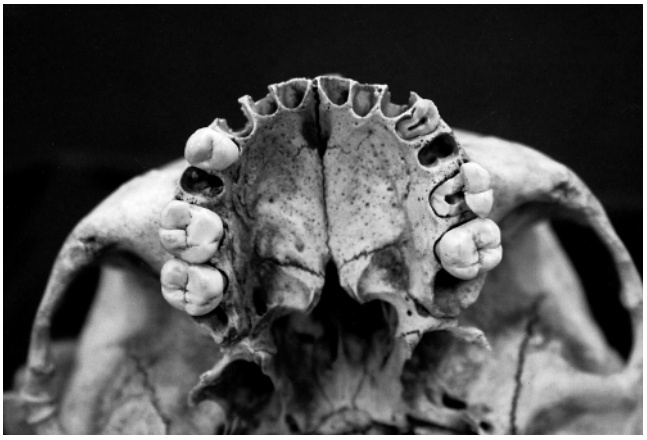

FIGURE A-2.4.2. Bilateral notched cleft palate: adult female (NMNH 264519), Chicama, Peru.

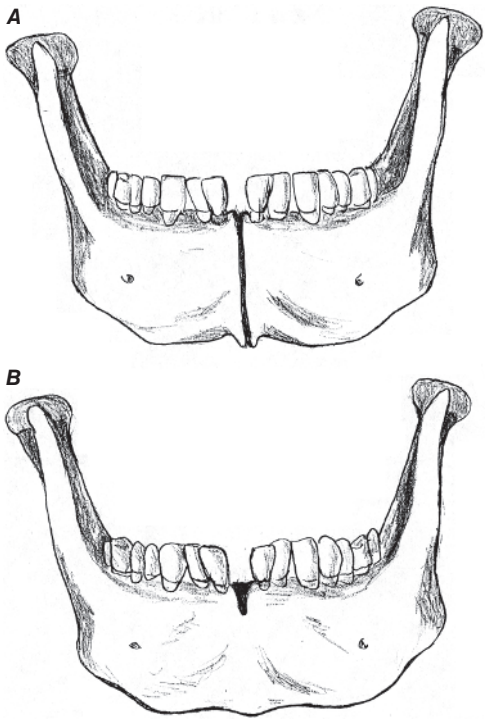

FIGURE A-2.5. Cleft mandible: (*A*) complete cleft; (*B*) notched mandible.

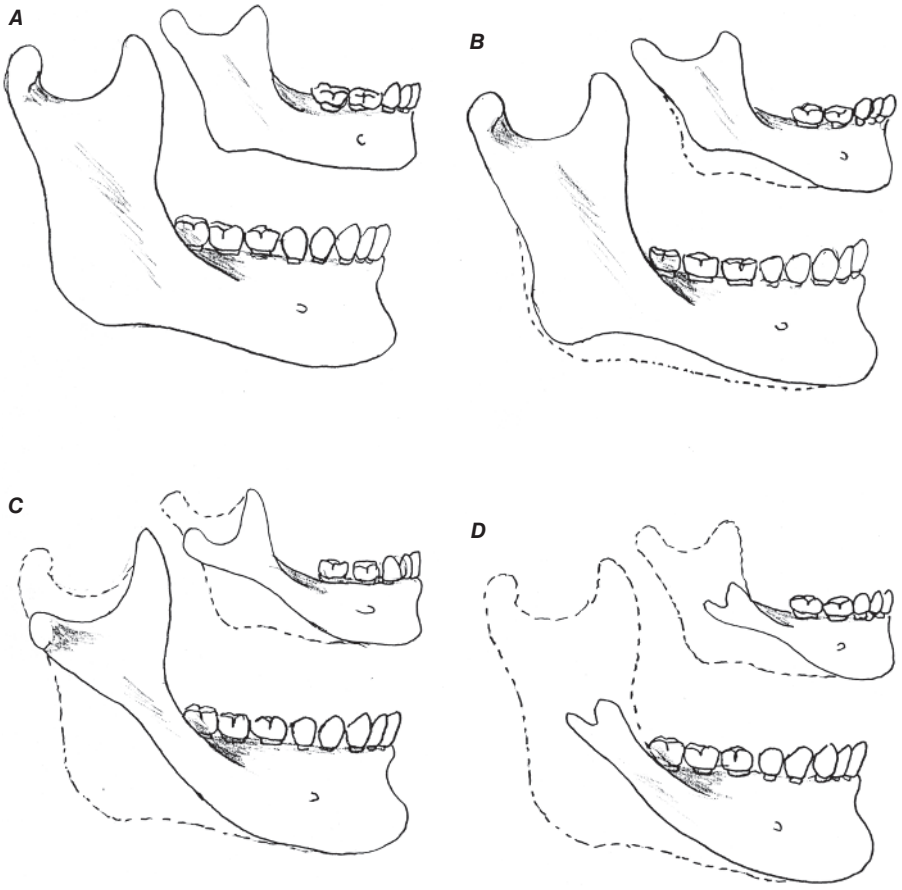

FIGURE A-2.6.1. Mandibular hypoplasia: (adult and child) (*A*) normal; (*B*) type I; (*C*) type II; (*D*) severe type III (dotted lines represent normal).

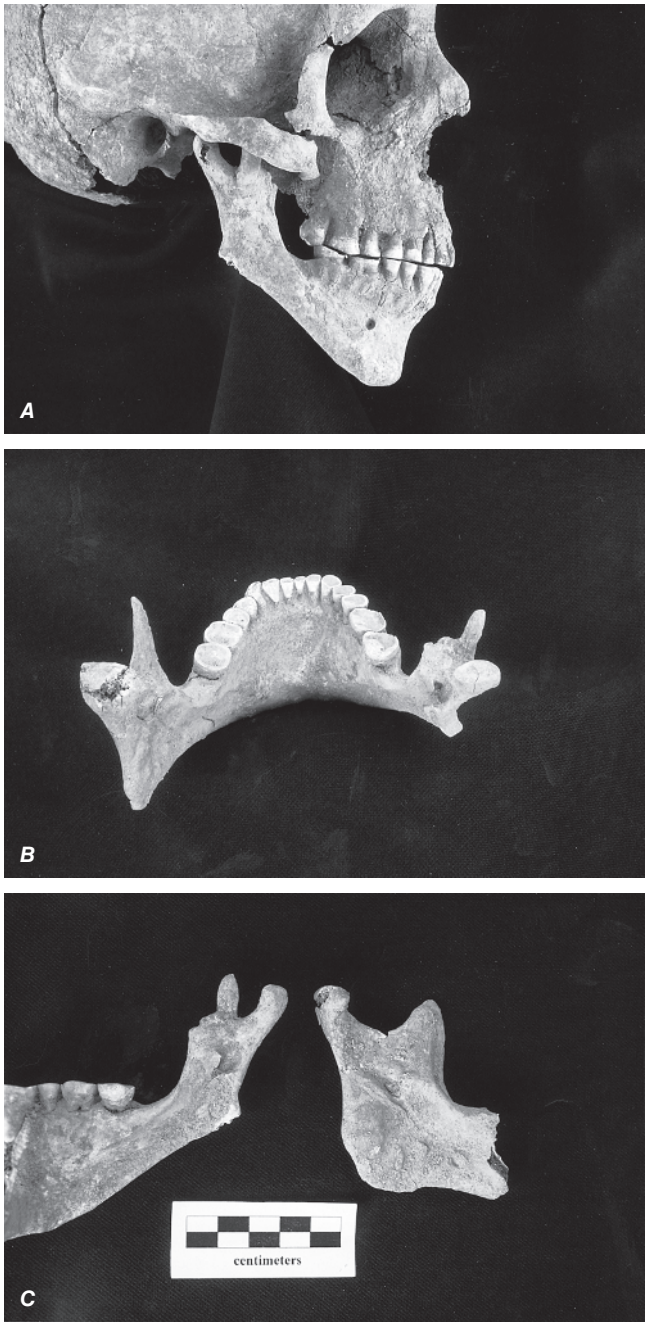

FIGURE A-2.6.2. Mandibular hypoplasia: unilateral right; (*A*) right side of the face, (*B*) dorsal view of the mandible, and (*C*) right and left sides of the mandible, adult male, La Playa, NW Mexico.

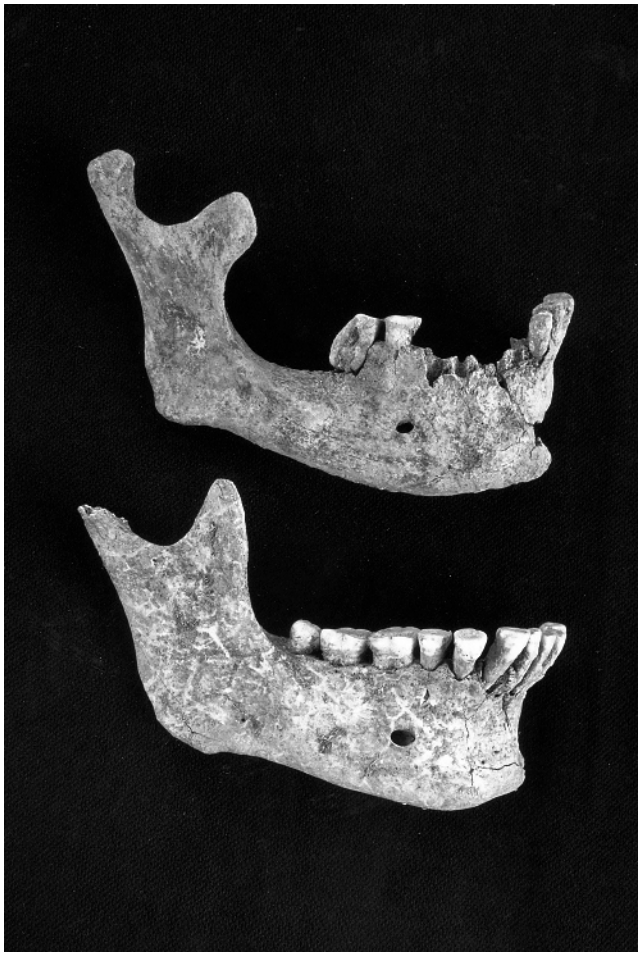

FIGURE A-2.6.3. Mandibular hypoplasia: unilateral right, adult female mandible compared with (lower) normal mandible, La Playa, NW Mexico.

mandibular head of the ramus fails to recede (Fig. A-2.7.1). Postnatal bifurcation can also occur with injury to the condylar head. Whereas the trauma-induced bifurcation is oriented anteroposteriorly, developmental bifurcation is positioned mediolaterally (Fig. A-2.7.2). Bifid condyles have a corresponding bifid articular temporal joint space. Unilateral expressions are more common than bilateral expressions (Blackwood 1957; McCormick et al. 1989).

A-2.8. Coronoid Hyperplasia

This is a genetically programmed progressive hyperdevelopment from the coronoid ossification center, culminating in enlarged bilateral coronoids by adolescence (Figs. A-2.8.1 and A-2.8.2). This prevents the mouth from fully opening as the expanded coronoid processes impinge on the posterior aspects of the zygomatics. It occurs mostly in males from the same family (Schultz and Theisen 1989).

A-2.9. Palate Inclusion (Fissural) Cyst

Overlying ectodermal tissue of the developing palate fails to retreat as membranous bones of the palate come together, leaving a pocket filled with fluid or semisolid

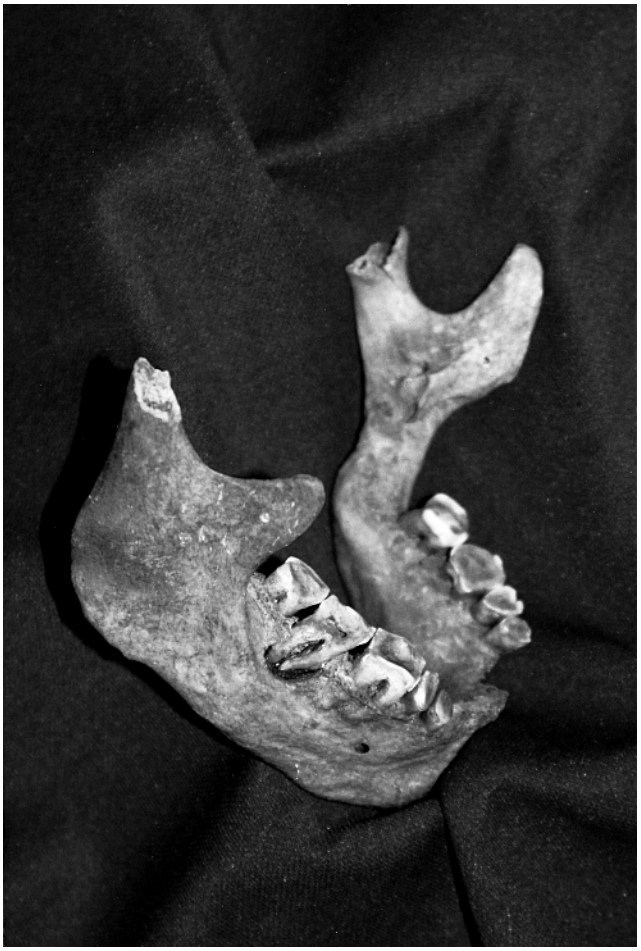

FIGURE A-2.6.4. Mandibular hypoplasia: unilateral left, adult female mandible (NMNH 242146), Dos Pueblos site, Santa Cruz Island, CA.

FIGURE A-2.6.5. Mandibular hypoplasia: unilateral right, adult female mandible, Irene Mound, Mississippian Mound Complex (contributed by Clark Spencer Larsen).

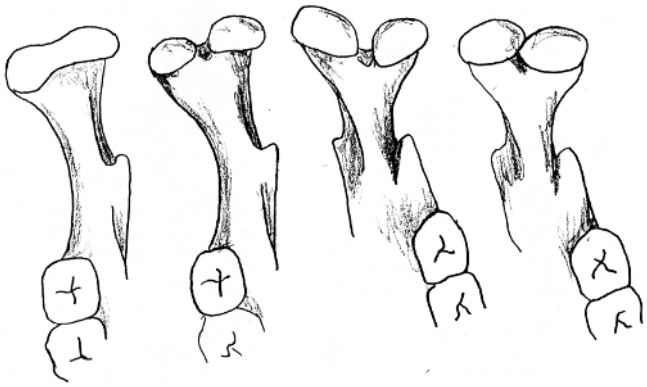

FIGURE A-2.7.1. Bifid mandibular condyle: normal (left) with examples of developmental bifurcation.

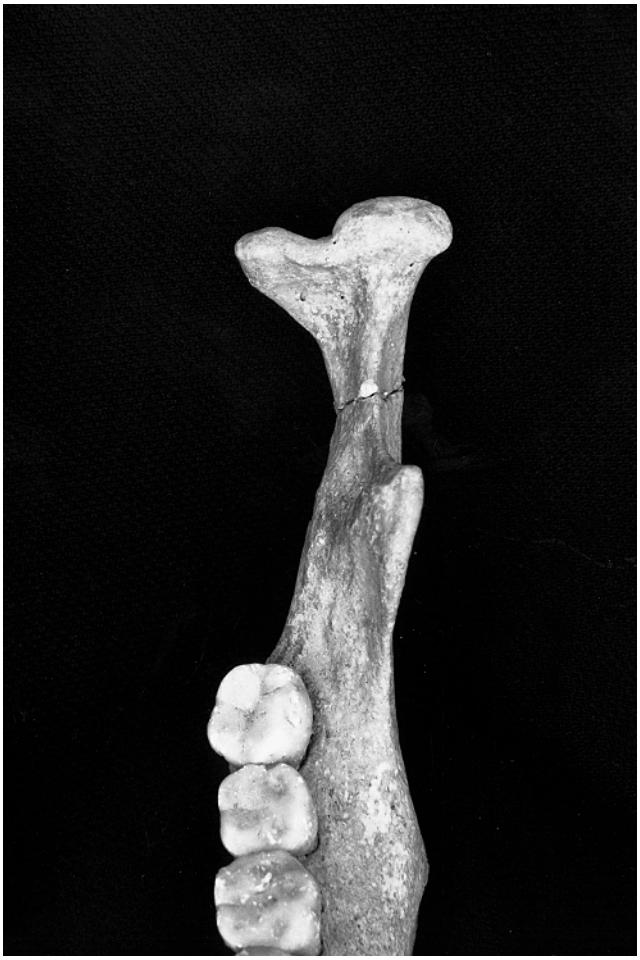

FIGURE A-2.7.2. Bifid mandibular condyle: left side of the bilateral expression, adult female, Byzantine Petras, Crete.

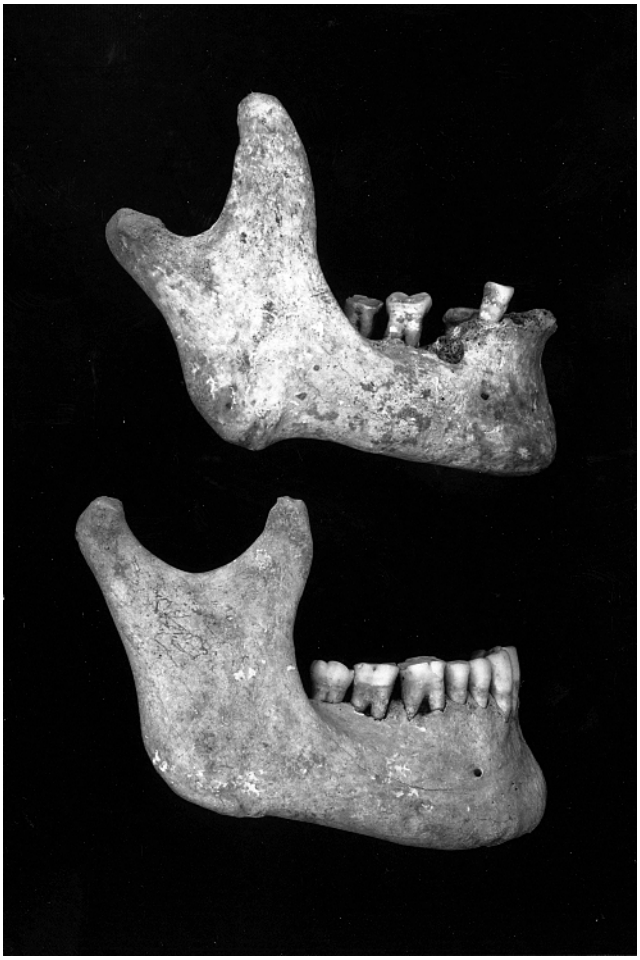

FIGURE A-2.8.1. Mandibular coronoid hyperplasia: (top) affected adult male compared with (bottom) normal adult male, Frankish Corinth, Greece.

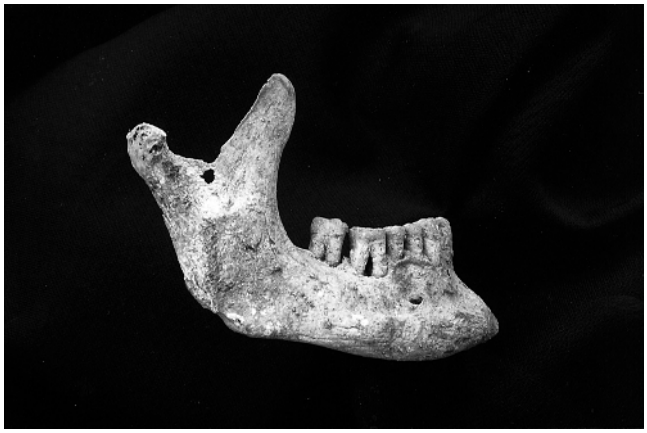

FIGURE A-2.8.2. Mandibular coronoid hyperplasia: adult male, Classical Period, Corinth, Greece.

material bounded by trapped epithelial cells within the developing bone. The incisive canal marks the junction of the premaxillary primary palate with the paired maxillary palatine shelves and is the most common site for development of an inclusion cyst—the median anterior inclusion cyst (Figs. A-2.9.1A and A-2.9.2A). The median palatal inclusion cyst develops midline between the two palatine shelves, and it can grow quite large over time (Fig. A-2.9.1B). The globulomaxillary inclusion cyst forms at the junction of the premaxilla primary palate and one of the maxillary palatine shelves (Fig. A-2.9.1C), between the roots of the lateral incisor and canine teeth (Little and Jakobsen 1973; Schafner et al. 1983).

A-2.10. Mandibular Inclusion Cyst

This is commonly referred to as the Stafne defect and takes shape as part of the primordial sublingual salivary gland bordering the submandibular fossa of the membranous bone develops prematurely, impinging upon developing membranous bone (Figs. A-2.10.1 and A-2.10.2). As the gland expands with age, it leaves a shallow or deep oval depression below the mylohyoid line near the inferior border of the inner retromolar aspect of the mandibular body (Stafne 1942; Wolf et al. 1986).

A-2.11. Mandibular Torus

This is a form of hyperplasia developing from the inner alveolar plate ossification center, resulting in one or more rounded bony protuberances below the alveolar margin on the lingual side of the mandible, generally between the canine and first molar. This is most often bilateral with more than one protuberance, varying in size and shape on each side. Sometimes the torus develops unilaterally, and they can appear as mild expressions to quite large in form (Figs. A-2.11.1–A-2.11.3).

Bony hyperplasia can also form along the median suture of the palate to form a palatine torus, but it is usually much less significant. Early development of both types of torus has been noted in infancy and early childhood, enlarging with maturity (Hauser and De Stefano 1989).

A-3. EXTERNAL AUDITORY MEATUS AND TYMPANIC PLATE DEVELOPMENT

While the first pharyngeal arch contributes to facial bones, the distal end of the ectodermal groove separating the external arch from its internal endothelial lined pouch leads to the development of the external auditory

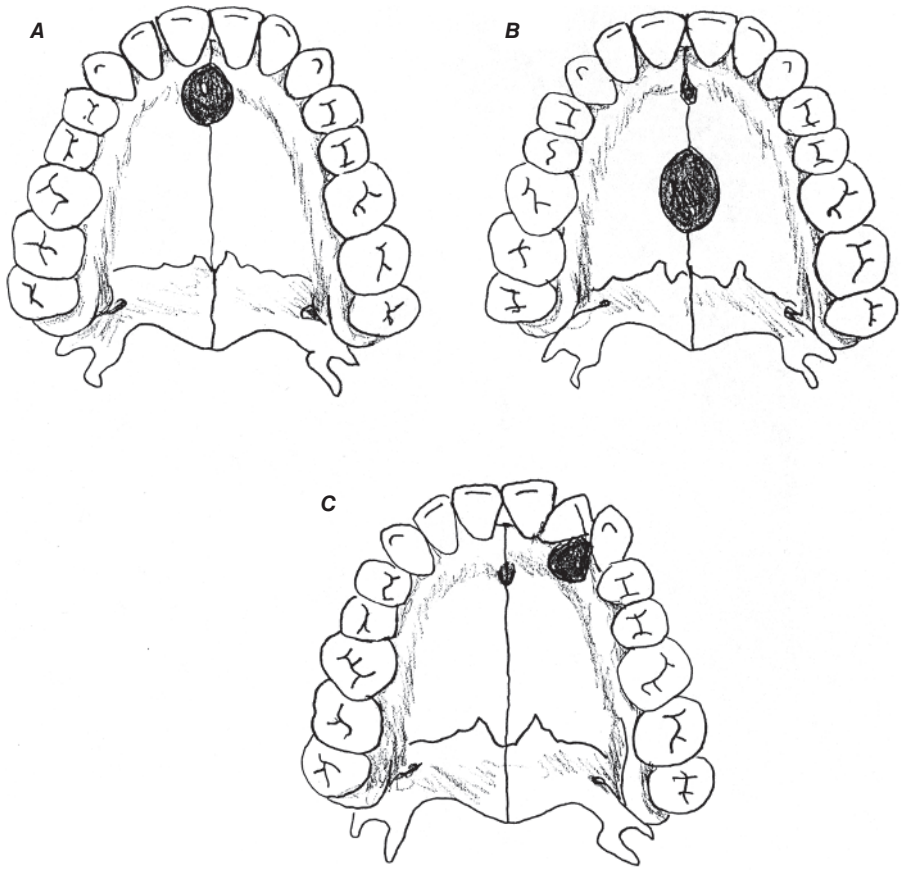

FIGURE A-2.9.1. Palatal inclusion (fissural) cysts: (*A*) median anterior within incisive canal; (*B*) median palatal between palatal plates; (*C*) globulomaxillary at the junction of premaxilla and maxilla.

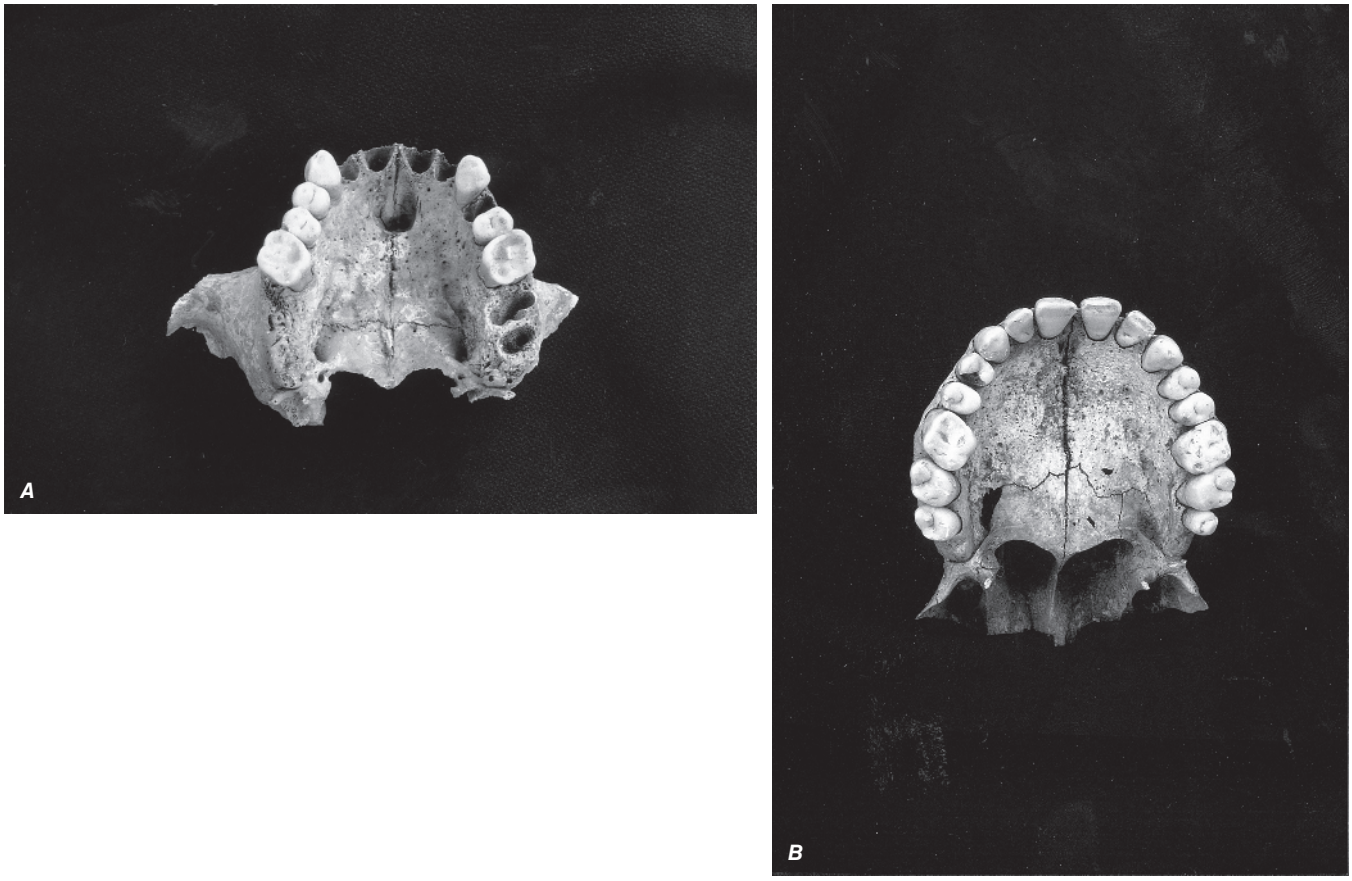

FIGURE A-2.9.2. Palatal inclusion (fissural) cyst: (*A*) adult median anterior cyst within the incisive canal, compared with (*B*) adult palate normal incisive canal (note left peg third molar), Frankish Corinth, Greece.

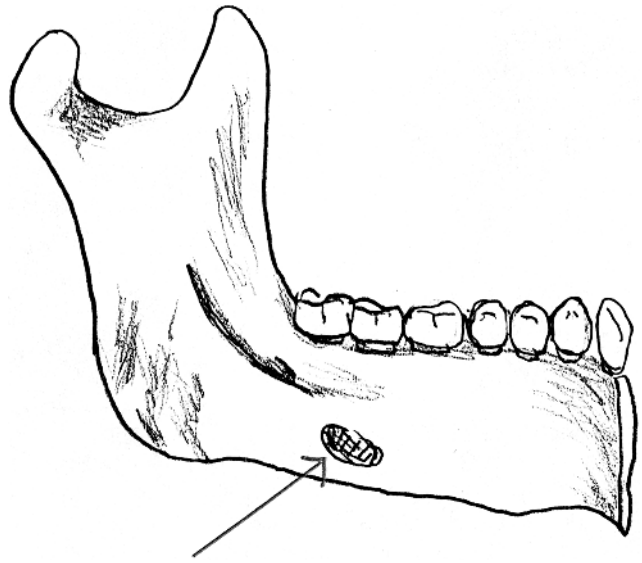

FIGURE A-2.10.1. Mandibular inclusion cyst (Stafne defect).

FIGURE A-2.10.2. Mandibular inclusion cyst (Stafne defect): unilateral right, adult male, La Playa, NW Mexico.

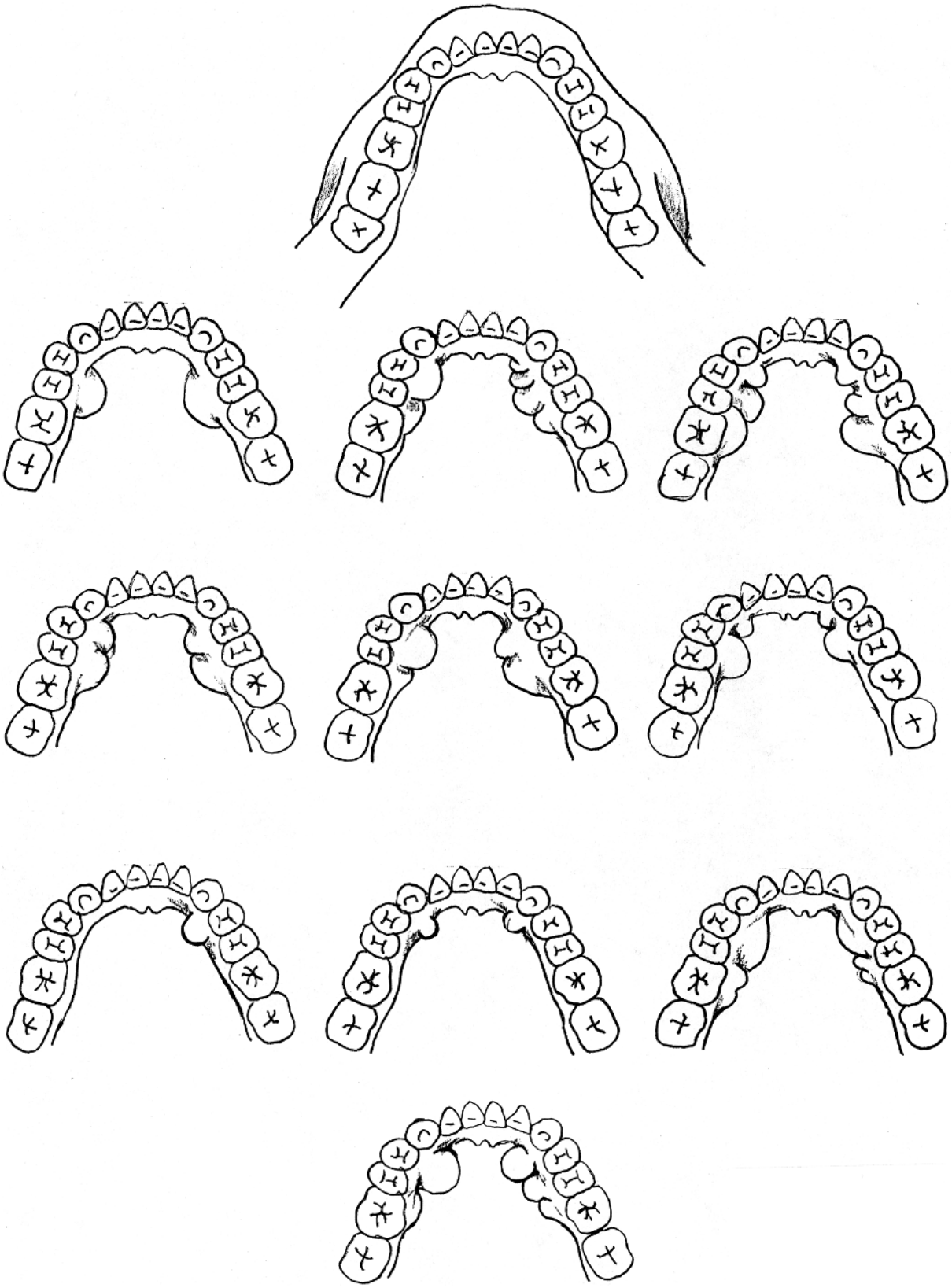

FIGURE A-2.11.1. Mandibular torus: normal mandible followed by torus variations drawn mostly from American Southwest skeletal collections.

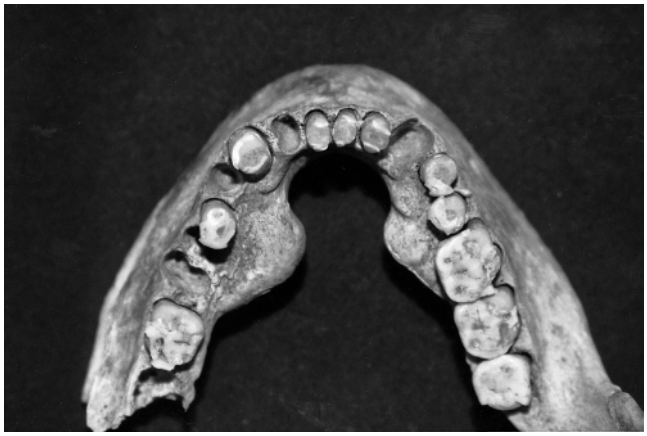

FIGURE A-2.11.2. Mandibular torus: adult male (NMNH 262945), Puye, NM.

meatus (Fig. A-3.0). The distal end of the endodermal pouch grows sack-like to form the inner ear's tympanic cavity while the proximal end remains narrow to form the eustachian tube as the external anterior portion of the ectodermal groove disappears.

The thin membrane separating the pouch and groove forms the membranous tympanic plate—the floor of the external auditory meatus—as the distal end of the ectodermal groove creates the external ear opening. By the ninth week, four small ossification centers form around the edges of the tympanic membrane, coalescing to form a U-shaped tympanic ring that eventually fuses to the squamosal part of the temporal bone by the thirty-fifth week. At birth, the bony tympanic ring frames the fibrocartilaginous tympanic plate that grows with the expansion of the auditory canal.

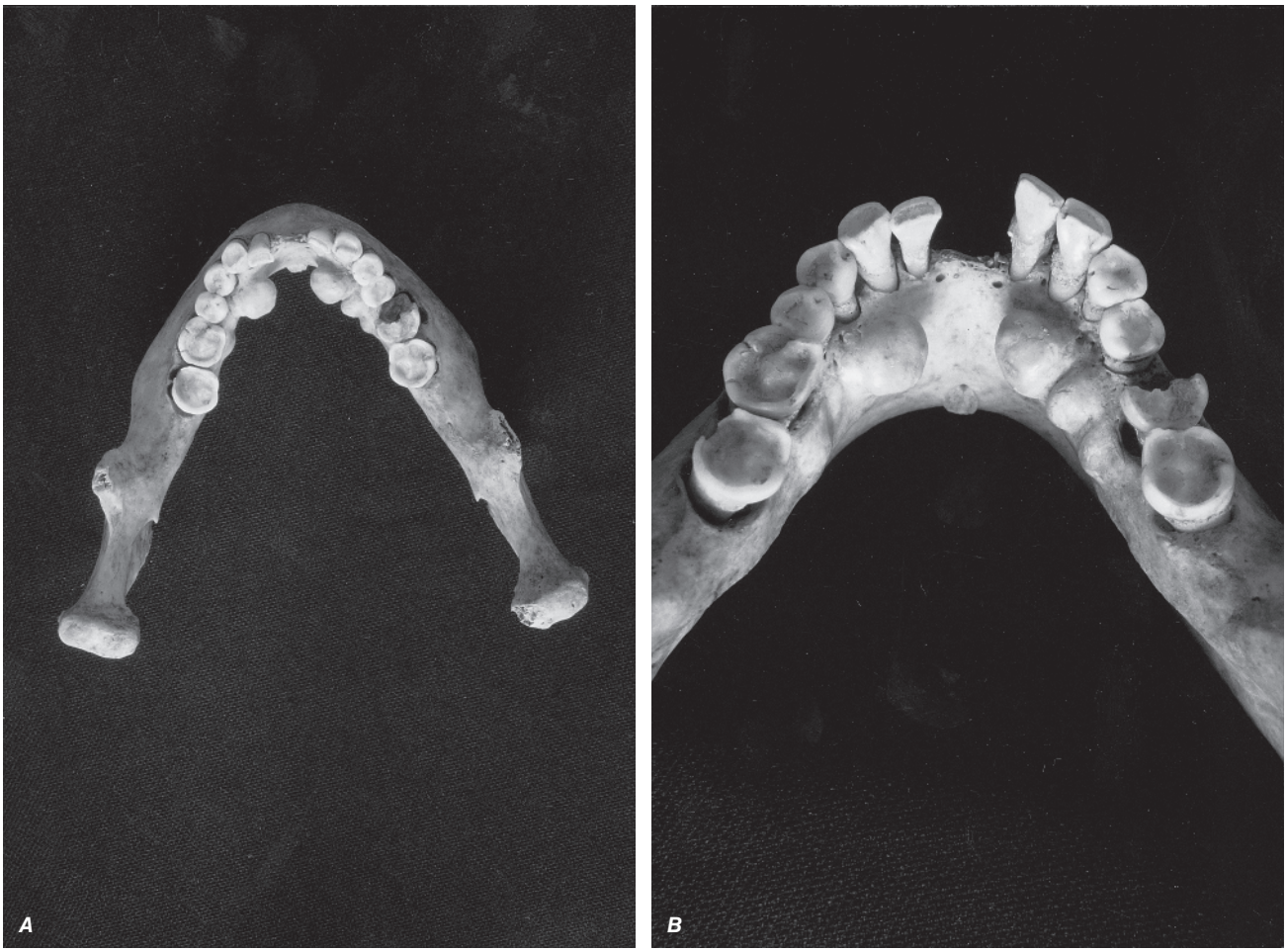

FIGURE A-2.11.3. Mandibular torus: (*A*) adult female, (*B*) close-up, Bronze Age Da Shan Qian, Inner Mongolia, PRC.

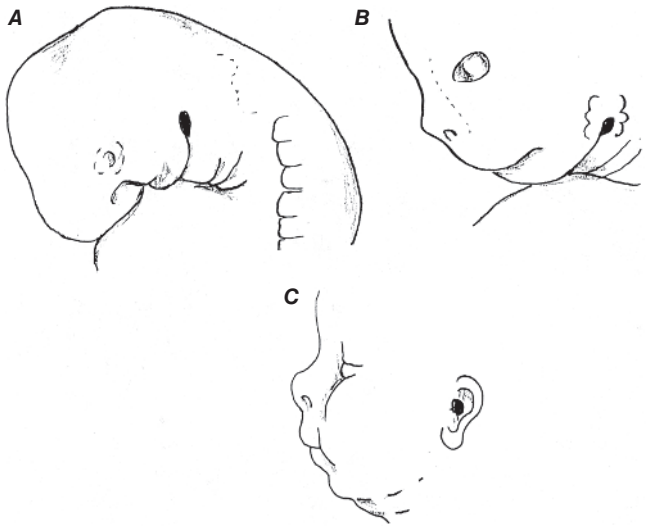

FIGURE A-3.0. External auditory meatus development: (*A*) fifth embryonic week; (*B*) seventh embryonic week; (*C*) newborn.

Ossification spreads throughout the tympanic plate rapidly from the anterior and posterior portions, but they are slow to merge, leaving an unossified area that appears as an opening in the bony tympanic plate, often referred to as foramen of Huschke. But it is not a true foramen as it essentially remains fibrocartilage until final ossifying closure by age 5 (Fig. A-3.2A–D). Bony extensions from the tympanic plate form a sheath around the developing styloid and the outer border juts out in semifolds from the external auditory meatus.

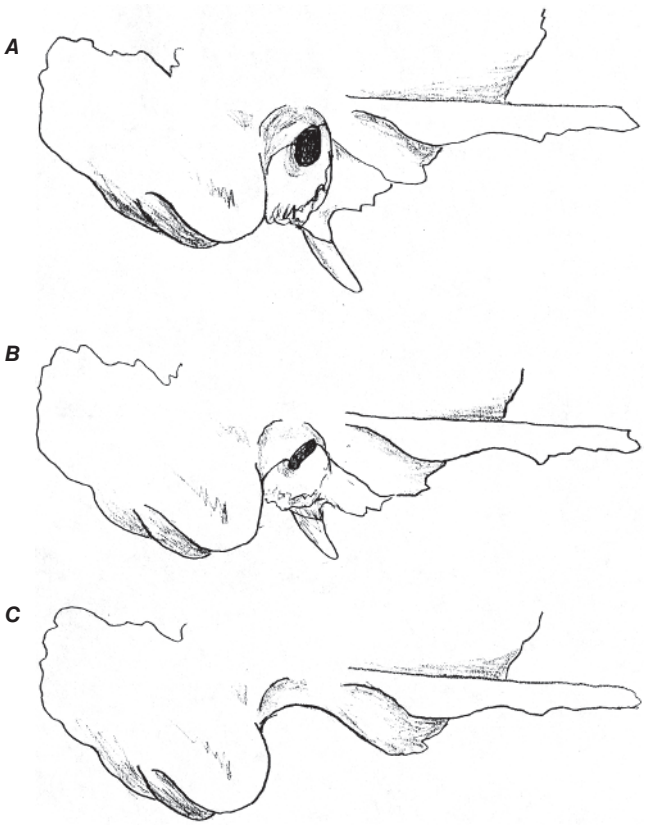

FIGURE A-3.1.1. Atresia (aplasia)/hypoplasia external auditory meatus: (*A*) normal opening; (*B*) narrow opening caused by hypoplasia; (*C*) atresia (aplasia).

EXTERNAL AUDITORY MEATUS AND TYMPANIC PLATE ANOMALIES

A-3.1. Atresia (Aplasia)/Hypoplasia External Auditory Meatus

Complete aplasia is a rare occurrence when the dorsal end of the first ectodermal groove fails to develop, usually unilaterally with the right side affected more than the left. The tympanic plate does not form and the styloid is rudimentary or absent and the petrous portion may be smaller (Hrdlicka 1933). The external soft tissue auricle of the ear is also affected. Hypoplasia can also occur with the development of the external auditory meatus producing a narrow opening with smaller tympanic plate (Figs. A-3.1.1–A-3.1.3).

A-3.2. Tympanic Aperture

When the tympanic plate fails to completely ossify from its fibrocartilaginous precursor, it leaves the bony opening to persist throughout life. This aperture can vary in size, and occasionally, the fibrocartilaginous tympanic plate fails completely to ossify, leaving a wide cleft in the floor of the external auditory meatus (Fig. A-3.2E,F).

A-3.3. External Auditory Torus

This is often referred to as external auditory exostosis but differing from pathological bony tumors (Hutchinson et al. 1997; Mann 1984). The torus forms with growth at the junction border between the tympanic plate and the temporal squamosal part of the auditory canal as a smooth bony ripple or nodule that can vary in size from slight to a large, usually as a single expression, sometimes double. Hyperplasia of one or both nodules on the primordial arms of the tympanic ring leads to this development that usually appears bilateral. Most often, it occurs at the inferior junction but can develop at the superior junction or both (Figs. A-3.3.1 and A-3.3.2).

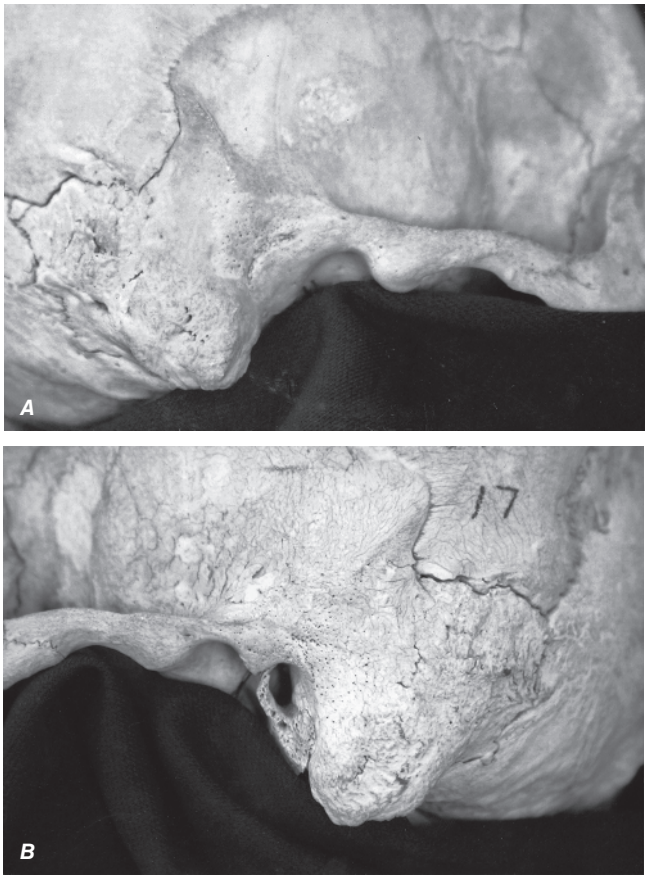

FIGURE A-3.1.2. Atresia (aplasia) external auditory meatus: (*A*) unilateral right, (*B*) normal left side, adult male (NMNH 264542), Chicama, Peru.

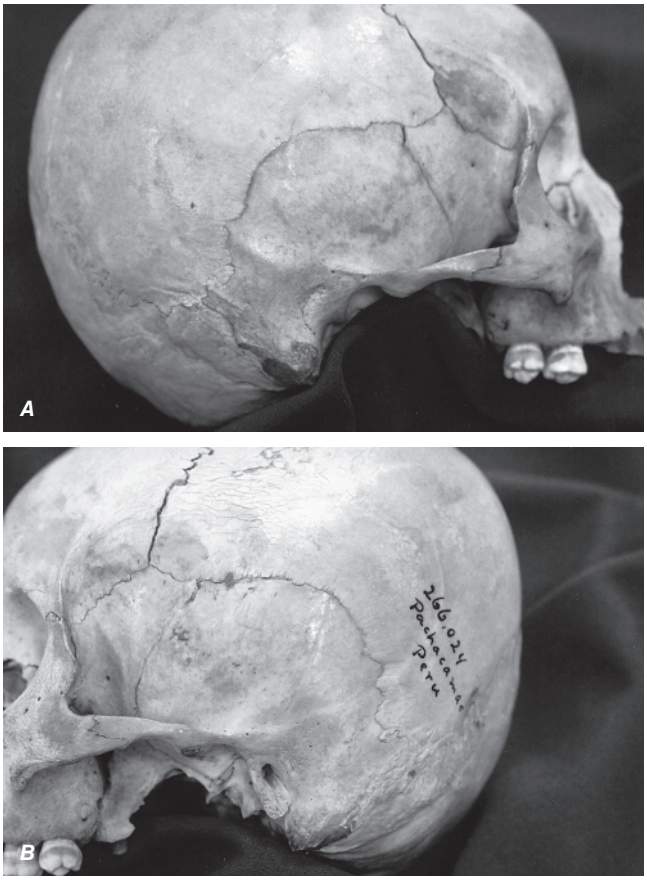

FIGURE A-3.1.3. Atresia (aplasia) external auditory meatus: (*A*) unilateral right, (*B*) normal left side, child (NMNH 266024), Pachacamac, Peru.

A-4. STYLOHYOID CHAIN DEVELOPMENT

The remaining pharyngeal arches contribute to the cartilages in the neck, while the associated pouches evolve into various glands as the ectodermal grooves between them disappear. The laryngeal cartilages—thyroid, cricoid, arytenoid, corniculate, and cuneiform cartilages—evolve from the last two pharyngeal arches, while the second and third arches provide cartilage for the stylohyoid chain. As cartilage tissue never looses the potential to ossify, sometimes trauma or genetic programming can lead to ossification of the laryngeal cartilages. Ossification of adult male thyroid cartilage is not unusual.

Paired Reichert's cartilages from the second pharyngeal arch form the bony styloid processes, stylohyoid ligaments connecting the styloids to the lesser cornua of the hyoid, and form the upper body of the hyoid as the ends of the cartilages meet midline. Paired cartilages from the third arch provide the greater cornua and lower part of the hyoid body. The dorsal end of Reichert's cartilage separates and becomes enclosed in the tympanic cavity to form the stapes.

The stylohoid chain begins with the proximal base of the styloid (tympanohyal) ensheathed by the tympanic plate. The apex of the elongated distal portion of the styloid (stylohyal) forms the attachment for the stylohyoid ligament (epihyal) that connects with the lesser cornua (hypohyal) attached to the superior portion of the hyoid at its junction with the greater cornua. The tympanohyal and stylohyal ossify from separate ossification centers, with the former present at birth and the latter appearing after birth, eventually uniting after puberty (Fig. A-4.0).

STYLOHYOID CHAIN ANOMALIES

A-4.1. Stylohyoid Chain Variations in Ossification

The stylohyal may not ossify leaving a shortened, hypoplastic (rudimentary) styloid (Fig. A-4.1.1B). Both

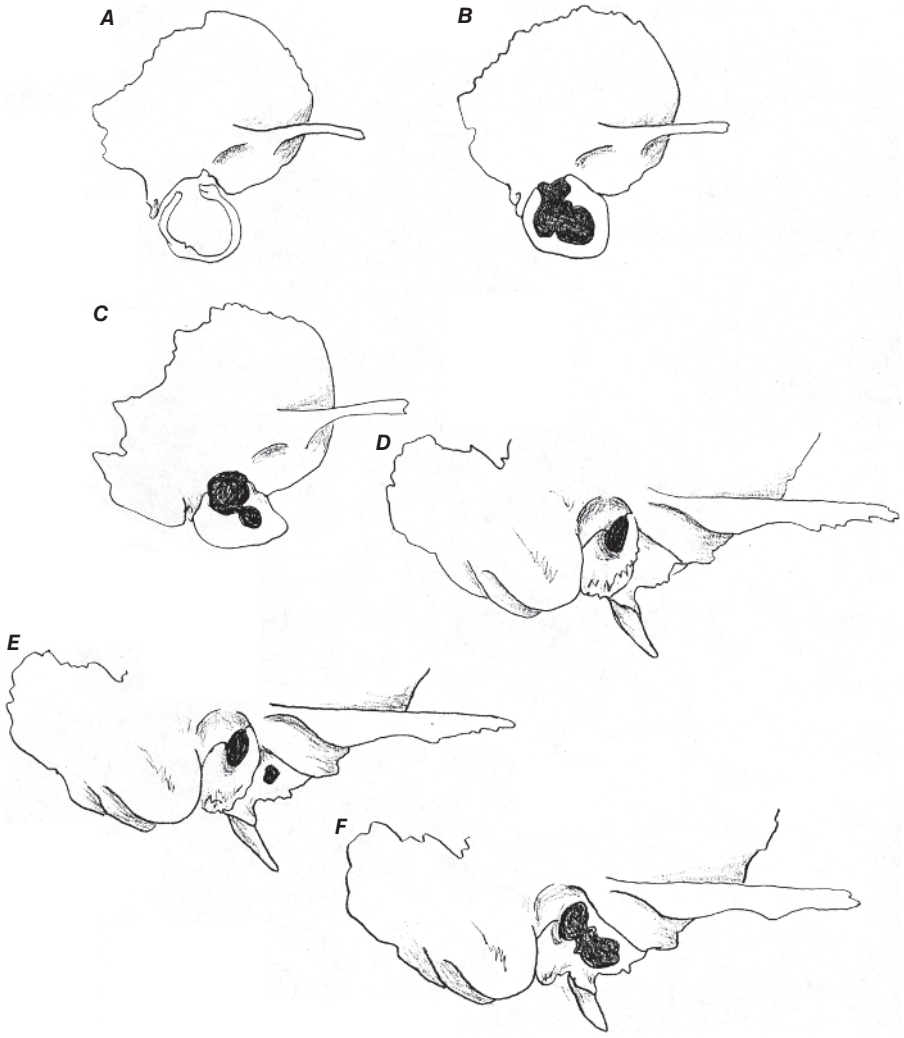

FIGURE A-3.2. Tympanic aperture: (*A*) tympanic ring at birth; (*B*) ossifying tympanic ring at 1 year; (*C*) continued ossification at 2 years; (*D*) adult complete ossification; (*E*) adult tympanic aperture; (*F*) adult tympanic cleft.

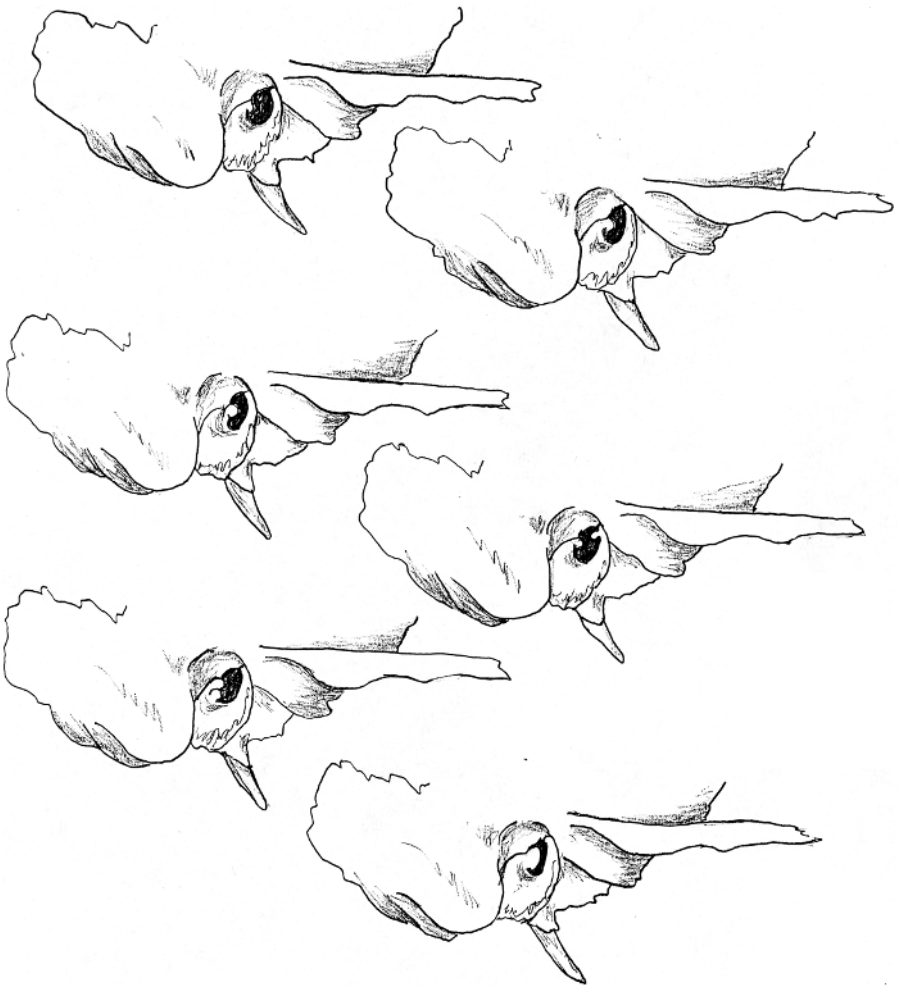

FIGURE A-3.3.1. External auditory meatus torus: variations from La Playa, NW Mexico.

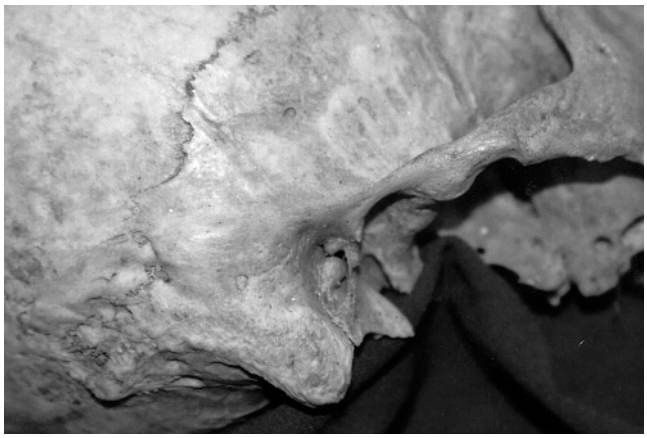

FIGURE A-3.3.2. External auditory meatus torus: adult male (NMNH 266023), Pachacamac, Peru.

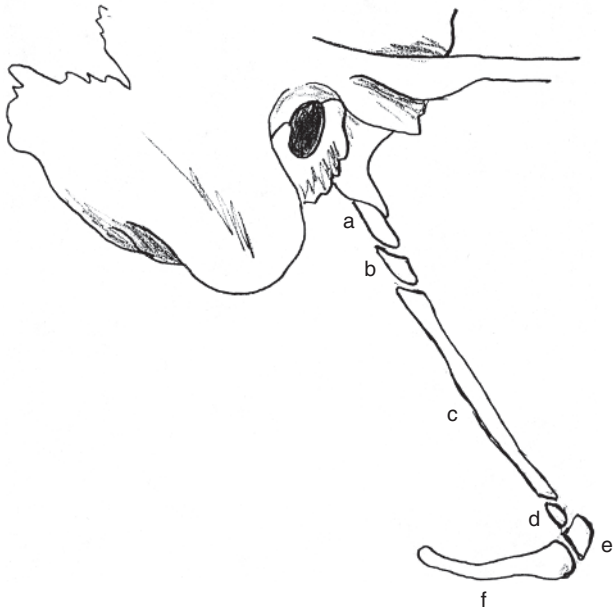

FIGURE A-4.0. Stylohyoid chain segments: (a) proximal end of the bony styloid process—tympanohyal; (b) bony distal end of the styloid process—stylohyal; (c) stylohyoid ligament—epihyal; (d) lesser cornu of the hyoid—hypohyal; (e) hyoid body; (f) greater cornua of the hyoid.

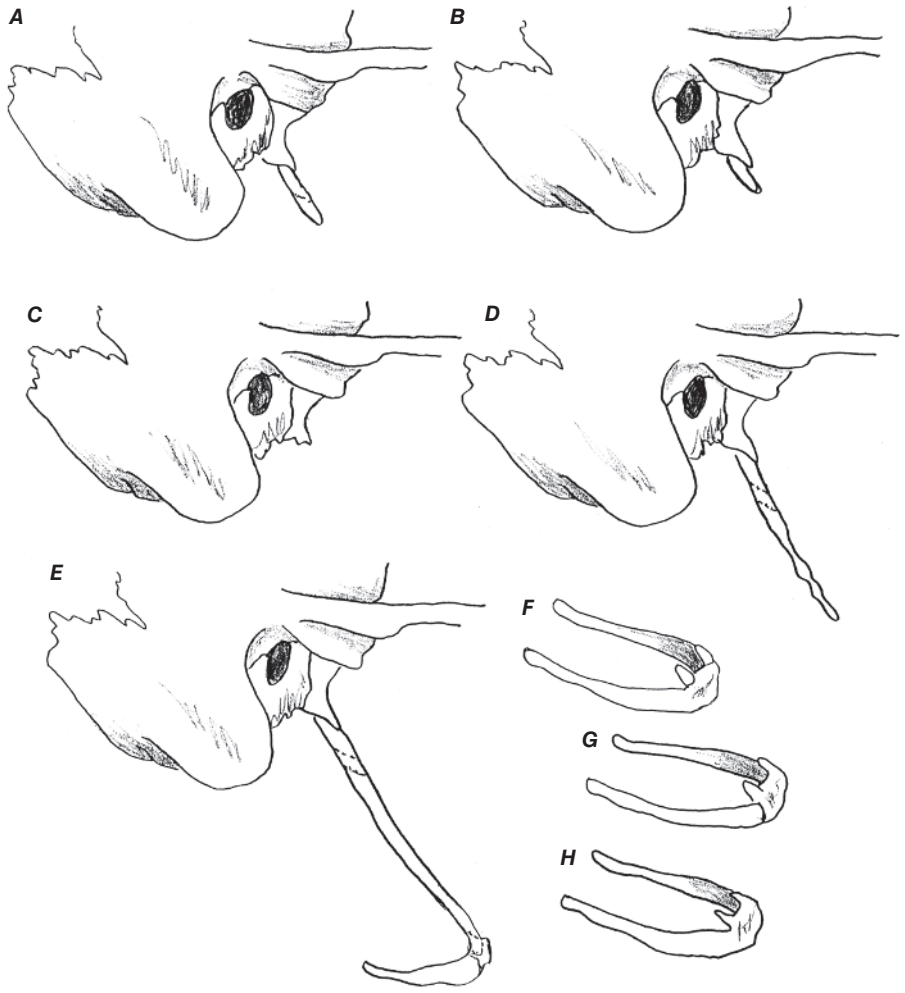

FIGURE A-4.1.1. Stylohyoid chain variations in ossification: (*A*) both parts of the styloid process ossified; (*B*) only the proximal portion of the styloid process ossified (bony hypoplasia); (*C*) neither part of the styloid process ossified (bony aplasia); (*D*) ossification of both parts of the styloid process and stylohyoid ligament (bony hyperplasia); (*E*) complete ossification of the stylohyoid chain; (*F*) separate ossified lesser cornua with greater cornua united with the hyoid body; (*G*) lesser cornua ossified and united to the hyoid body; (*H*) both ossified lesser cornua and greater cornua united with the hyoid body.

the stylohyal and tympanohyal may not ossify, leaving the bony styloid absent (Fig. A-4.1.1C). Part or all of the epihyal—the stylohyoid ligament—may ossify with the styloid to produce a greatly elongated bony styloid that may be crooked with a blunt tip (Figs. A-4.1.1D and A-4.1.2), appearing as a hyperplastic styloid process, either unilaterally or bilateral with or without asymmetry. The lesser cornua, attached to the hyoid by fibrous joints, may remain cartilaginous or ossify and sometimes uniting with the hyoid. The bony greater cornua are usually connected by fibrous joints to the hyoid body but may unite with it (Figs. A-4.1.1F and A-4.1.3) if the joint fails to develop. Rarely, both lesser and greater cornua ossify as part of the hyoid body (Figs. A-4.1.1H and A-4.1.4). Very rarely, the entire stylohyoid chain ossifies to unite all of its parts, or each segment ossifies separately with fibrous connective tissue holding them all together (Camarda et al. 1989; Gossman and Tarsitano 1977).

A-4.2. Thyroglossal Developmental Cyst

This can form anywhere along the descending pathway of the primordial thyroglossal duct as it passes from the base of the tongue to the anterior midline of the throat, past the hyoid body to its final position below the thyroid and cricoid cartilages to form the thyroid gland (Fig. A-4.2.1). Remnant thyroglossal duct tissue left behind forms a cyst that can impress upon adjacent structures such as the hyoid body (Figs. A-4.2.1B and A-4.2.2) (Moore 1985:1027; Sadler 2006:271–272).

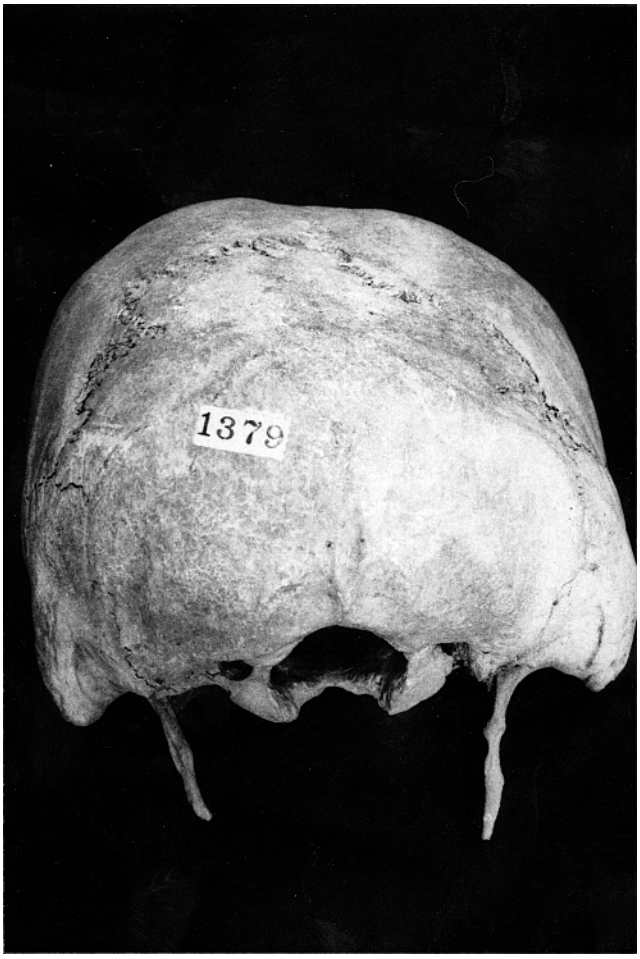

FIGURE A-4.1.2. Ossified stylohyoid ligament: bilateral, united with the bony styloid processes, adult male, Old Walpi, AZ (Field Museum).

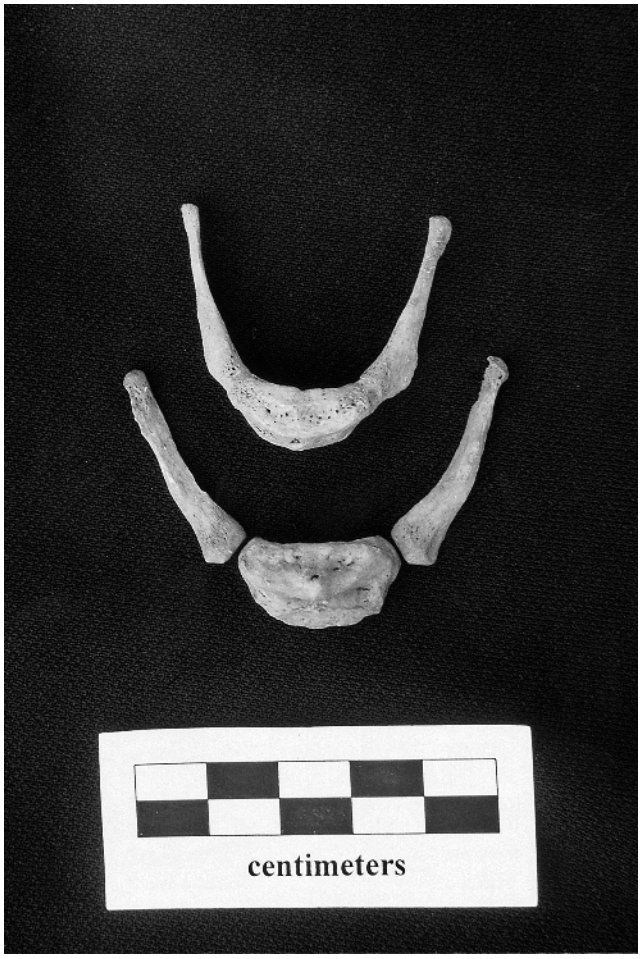

FIGURE A-4.1.3. Greater cornua united with the hyoid body: (top) adult female compared with (bottom) the normal separated cornua, adult male, Byzantine Panakton, Greece.

FIGURE A-4.1.4. Ossified lesser cornua and greater cornua united with the hyoid body: adult male (NMNH 308623), Hawikuh, NM.

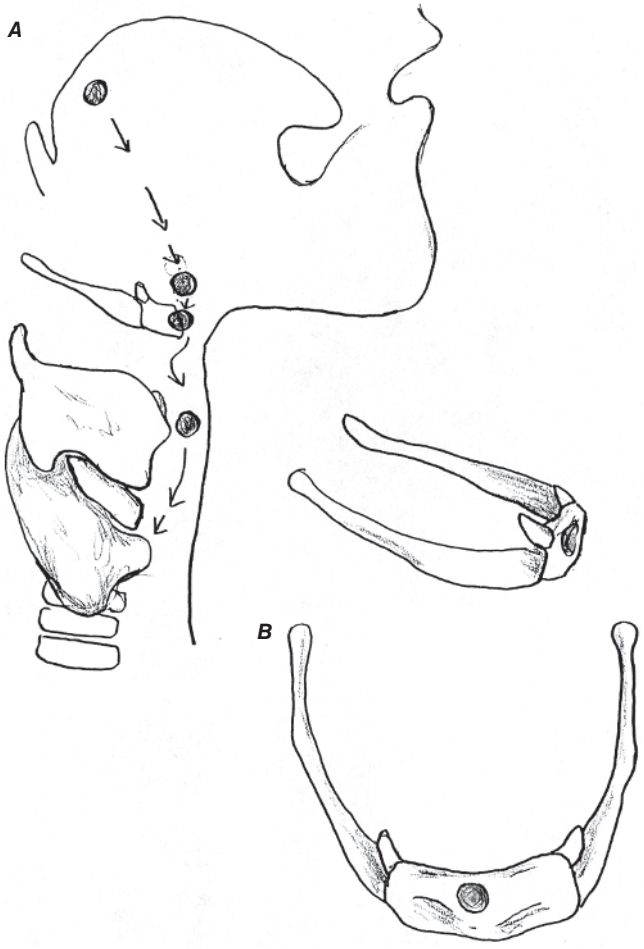

FIGURE A-4.2.1. Thyroglossal cyst development: (*A*) embryonic descent of primordial thyroglossal duct from below the tongue, past the hyoid body to the final position below the thyroid and cricoid cartilages—circles represent where cysts may form from remnant tissue; (*B*) hyoid body with imprint of the thyroglossal cyst.

FIGURE A-4.2.2. Thyroglossal cyst: (top) normal hyoid body compared with (bottom) the hyoid with thyroglossal cyst, young adult male, Frankish Corinth, Greece.

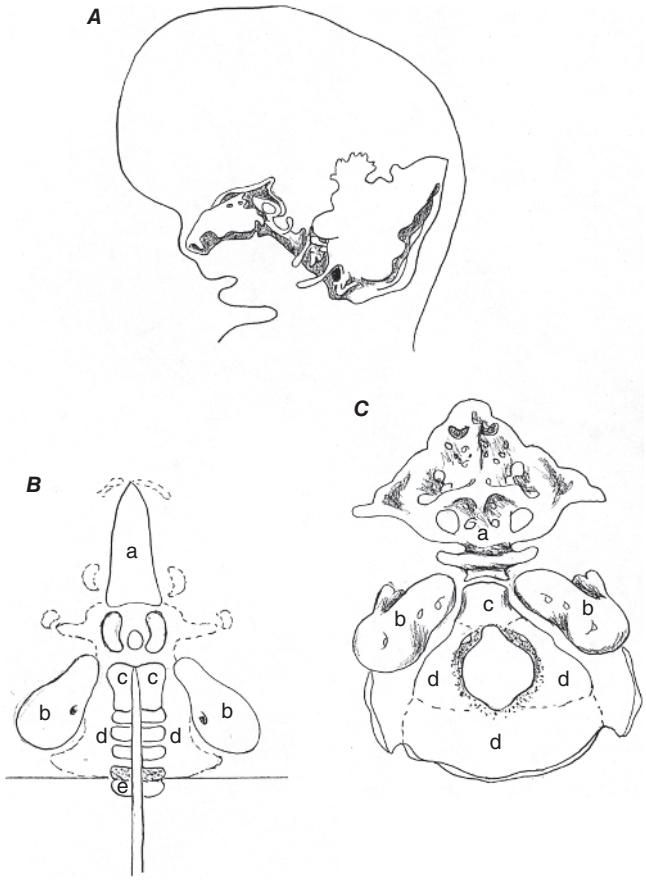

FIGURE A-5.0.1. Skull base (chondocranium) development: (*A*) the developing fetal cartilaginous plate; (*B*) developing precursor cartilages; (*C*) corresponding later development with broken lines representing contributing cranial portion of the first cervical sclerotome (a) ventral trabecular cartilages, (b) lateral otic capsules, (c) post-hypophyseal fossa parachordal cartilages surrounding the cranial end of the neural tube, (d) occipital sclerotmes, and (e) first cervical sclerotome with dotted cranial portion joining occipital ones, line divides occipital base with the atlas.

A-5. SKULL BASE DEVELOPMENT

The forerunner of the skull base—the chondocranium—forms by the end of the first month from a cartilaginous plate known as the prechordal cranial base, extending from the nasal region to surround the cranial end of the neural tube, thus cradling the developing brain. This plate stems from the expansion and fusion of three basic pairs of cartilage precursors. The ventral cartilages (trabecular cartilages) developing in the interorbital–nasal region to ultimately form the nasal cartilages, ethmoid, lesser wings, and roots of greater wings as well as the body of the sphenoid. Lateral cartilages (otic capsules) develop around the otocysts, giving rise to the petromastoids of the temporals. Paired cartilages (parachordal cartilages) just distal to the fossa for the hypophysis join together and develop into the basioccipital with small ventral portions of the occipital condyles. Abutting the parachordal cartilages, four somitomeres condense into occipital somites to form three nonsegmenting sclerotomes. The occipital sclerotomes fuse with the parachordal cartilages, while the cranial portion of the resegmenting first cervical sclerotome splits from its caudal portion to join the last occipital sclerotome in the formation of the lateral exoccipitals and supraoccipital bones (Fig. A-5.0.1). The supraoccipital quickly fuses with the occipital squamosa at the highest nuchal line (the mendosa line). The lateral exoccipitals unite with the supraoccipital by the third year and with the basioccipital by the sixth year (Fig. A-5.0.2).

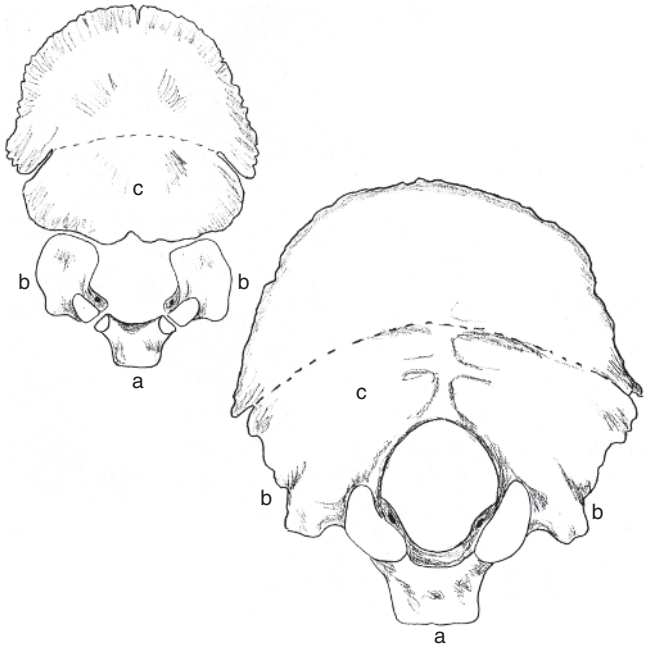

FIGURE A-5.0.2. Skull base (chondocranium): (infant and adult) (a) basioccipital; (b) lateral exoccipitals; (c) supraoccipital, fusion with occipital squamosa represented by broken lines.

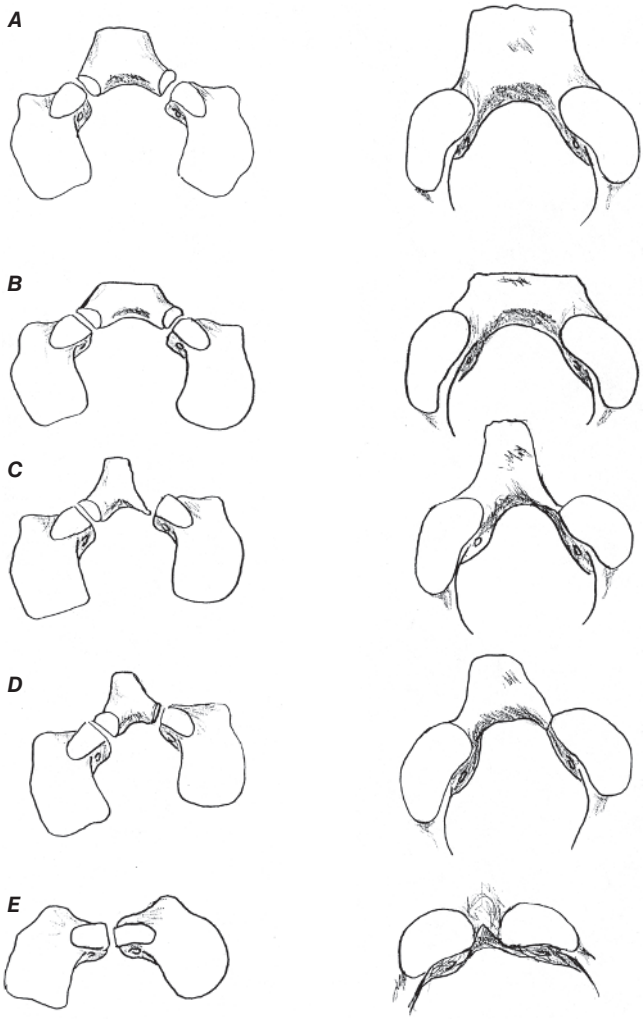

FIGURE A-5.1. Basioccipital hypoplasia/aplasia: (infant and adult) (*A*) normal basioccipital; (*B*) bilateral hypoplasia; (*C*) unilateral left aplasia; (*D*) unilateral left hypoplasia; (*E*) bilateral aplasia.

SKULL BASE ANOMALIES

A-5.1. Basioccipital Hypoplasia/Aplasia

Developmental disturbance of one or both parachordal cartilages can result in hypoplasia or aplasia of the basioccipital with or without distortion of the foramen magnum (Fig. A-5.1).

A-5.2. Basioccipital Clefts

Failure of the two parachordal cartilages to completely fuse presents as a vertical cleft (Fig. A-5.2A). Cranial shifting of the border between the atlas and occipital that moves the border upward can be variably expressed, and on rare occasions, the shift affects the developing parachordal cartilages, producing bilateral or unilateral horizontal clefts. The nonpathological clefts are filled with cartilaginous fibers in life. This kind of shift can also delete one or both ventral portions of the occipital condyles forming on the dorsal ends of the parachordal cartilages resulting in the appearance of condylar hypoplasia (Fig. A-5.2B–D).

OCCIPITAL–CERVICAL (O-C) BORDER DEVELOPMENT

The O-C border is between the occipital base, exoccipitals, axis apical dens and the atlas, axis (Fig. A-5.3.0). Note that the apical dens develops above the border in the primordial region of the proatlas of early life forms before descending below the border to join the rest of the axis as the permanent border develops (Shapiro and Robinson 1976). See the development of vertebral column under Chapter B for more details.

A-5.3. Cranial Shifting of the O-C Border

The border between the base of the occipital and atlas is moved upward (Fig. A-5.3.1), affecting the last

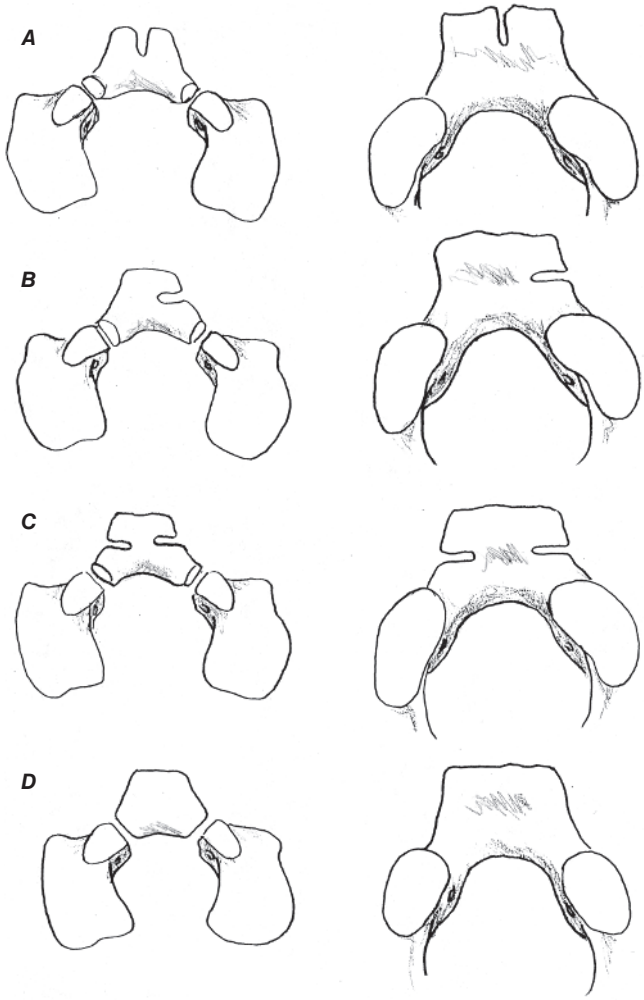

FIGURE A-5.2. Basioccipital clefts: (infant and adult) (*A*) vertical cleft from failure of parachordal cartridges to completely fuse; (*B*) unilateral left horizontal cleft caused by attempted cranial border shift; (*C*) bilateral horizontal cleft from attempted cranial border shift; (*D*) bilateral agenesis of ventral portions of occipital condyles from attempted cranial border shift (hypoplasia of occipital condyles).

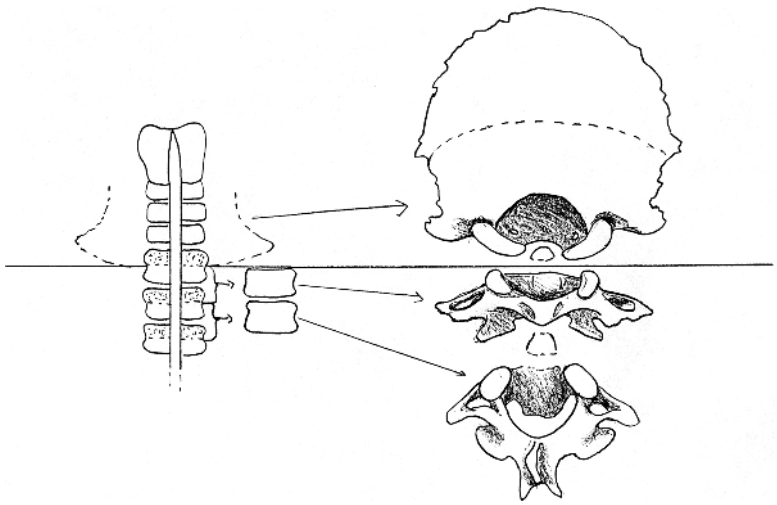

FIGURE A-5.3.0. Occipital–cervical border: represented by a solid line with basioccipital and apical dens forming above the O-C border, atlas, dens, and axis body developing below the O-C border.

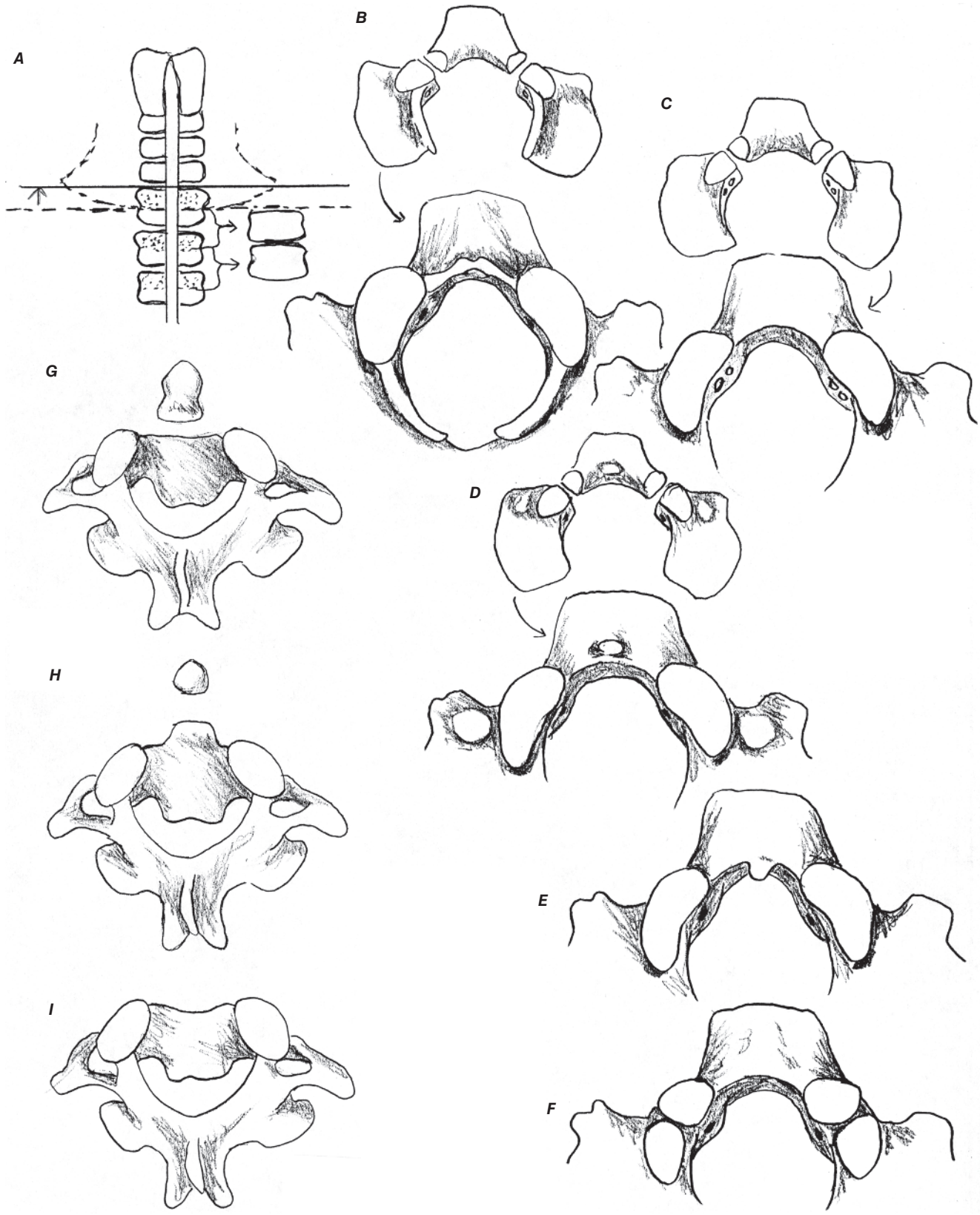

FIGURE A-5.3.1. Cranial shifts at the occipital–cervical (O-C) border: (*A*) schematic drawing shows the border (solid line) moving upward from the normal border represented by a dashed line; (newborn and adult) basioccipital; (*B*) complete shift occipital vertebra expression; (*C*) mild shift double hypoglossal canals; (*D*) precondylar and paracondylar (bony protrusions) processes; (*E*) precondylar extended tubercle from the anterior rim of the foramen magnum; (*F*) bifurcated occipital condyles; (*G*) separate dens (type I dens defect—os odontoideum); (*H*) separate dens' tip (type II dens defect—ossiculum terminale); (*I*) agenesis of dens, both apical tip and base (type V dens defect).

FIGURE A-5.3.2. O-C cranial border shift precondylar tubercle: adult female and adult male, Warring States Period, Da Shan Qian, Inner Mongolia, PRC.

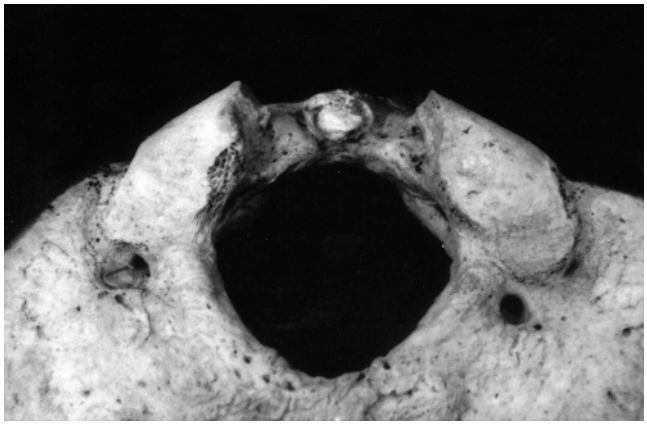

FIGURE A-5.3.3. O-C cranial border shift separated the apical dens' tip attached to the foramen magnum rim: adult male (NMNH 271905), Amoxiumqua, NM.

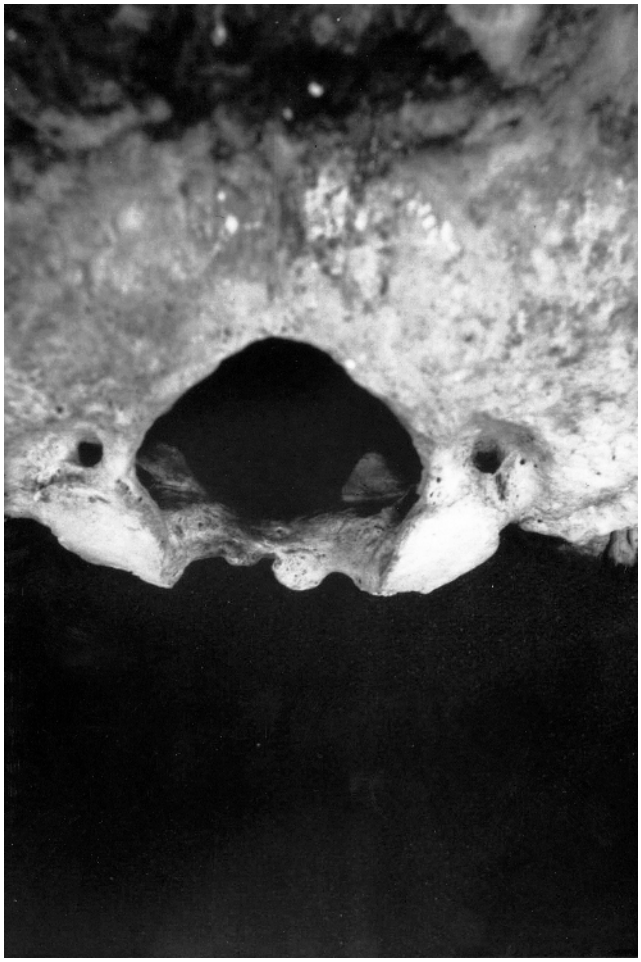

FIGURE A-5.3.4. O-C cranial border shift precondylar process: adult male (NMNH 308602), Hawikuh, NM.

occipital sclerotome as it coalesces with the cranial portion of the resegmenting first cervical sclerotome (Fig. A-5.3.1A). The upward shift forces this segment into attempted separation from the base of the skull to form a substitute proatlas or occipital vertebral segment. Complete separation from the base of the skull is not possible, generally leaving only variable expressions of an occipital vertebra arising from the exoccipitals, represented by attempts to form vertebral arching around the foramen magnum. Usually, there is some form of anterior arch with an incomplete posterior arch protruding around the foramen magnum that often appears distorted by the unusual projections.

Minor forms of cranial border shifting include a small bony tubercle extending from the anterior rim of the foramen magnum as if simulating a pseudo-apical dens tip (Figs. A-5.3.1E and A-5.3.2). Raised bony distortions around the foramen magnum may appear, including the precondylar process (sometimes double in form), and other forms of raised bony processes in the paracondylar spaces, bilaterally or unilaterally (Figs. A-5.3.1D, A-5.3.4, and A-5.3.5). Paramastoid processes near the notch for the jugular foramen may also be part of this phenomenon. These bony processes can be small, or large enough to articulate with the atlas. The hypoglossal canals commonly bifurcate with mild cranial border shifts (Fig. A-5.3.1C), and the occipital condyles may also bifurcate (Fig. A-5.3.1F).

Cranial shifting at the O-C border can sometimes, though rarely, affect the cartilaginous precursor apical dens (odontoid) of the axis vertebra developing within the region of the primordial foramen magnum (Dawson and Smith 1979). With a stable developing O-C border, the dens' apical tip descends from the area of the anterior rim of the foramen magnum to unite with the base of the dens as it moves downward to join the body of the axis vertebra (Fig. A-5.3.0). Shifting upward of the

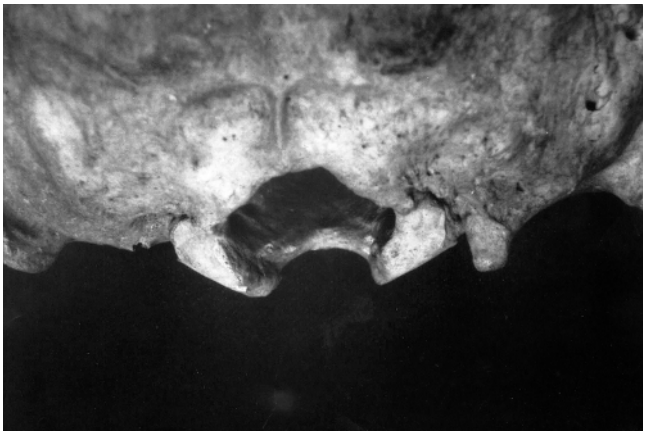

FIGURE A-5.3.5. O-C cranial border shift paracondylar process: adult female (NMNH 271804), Amoxiumqua, NM.

O-C border can delay and prevent the apical tip of the dens from uniting with its base (type II dens defect: ossiculum terminale), leaving it a separate ossicle (Figs. A-5.3.1H and A-5.3.6) that may attach to the anterior of the atlas or anterior rim of the foramen magnum (Fig. A-5.3.3). The entire dens may not join with the axis body (type I dens defect: os odontoideum) (Fig. A-5.3.1G). Agenesis of the tip (type IV dens defect) leaves the dens appearing short and blunt (Fig. A-5.3.7). The base of the dens may fail to ossify within the precursor cartilage, leaving the apical segment as a separated ossicle (type III dens defect) or the entire dens, both base and apical tip, may fail to ossify (type V dens defect) (Fig. A-5.3.1I). With agenesis of the dens or dens apical tip, there will be no articulating facet for it on the atlas (Fig. A-5.3.7). Severe dens defects can lead to serious neurological problems and often involve

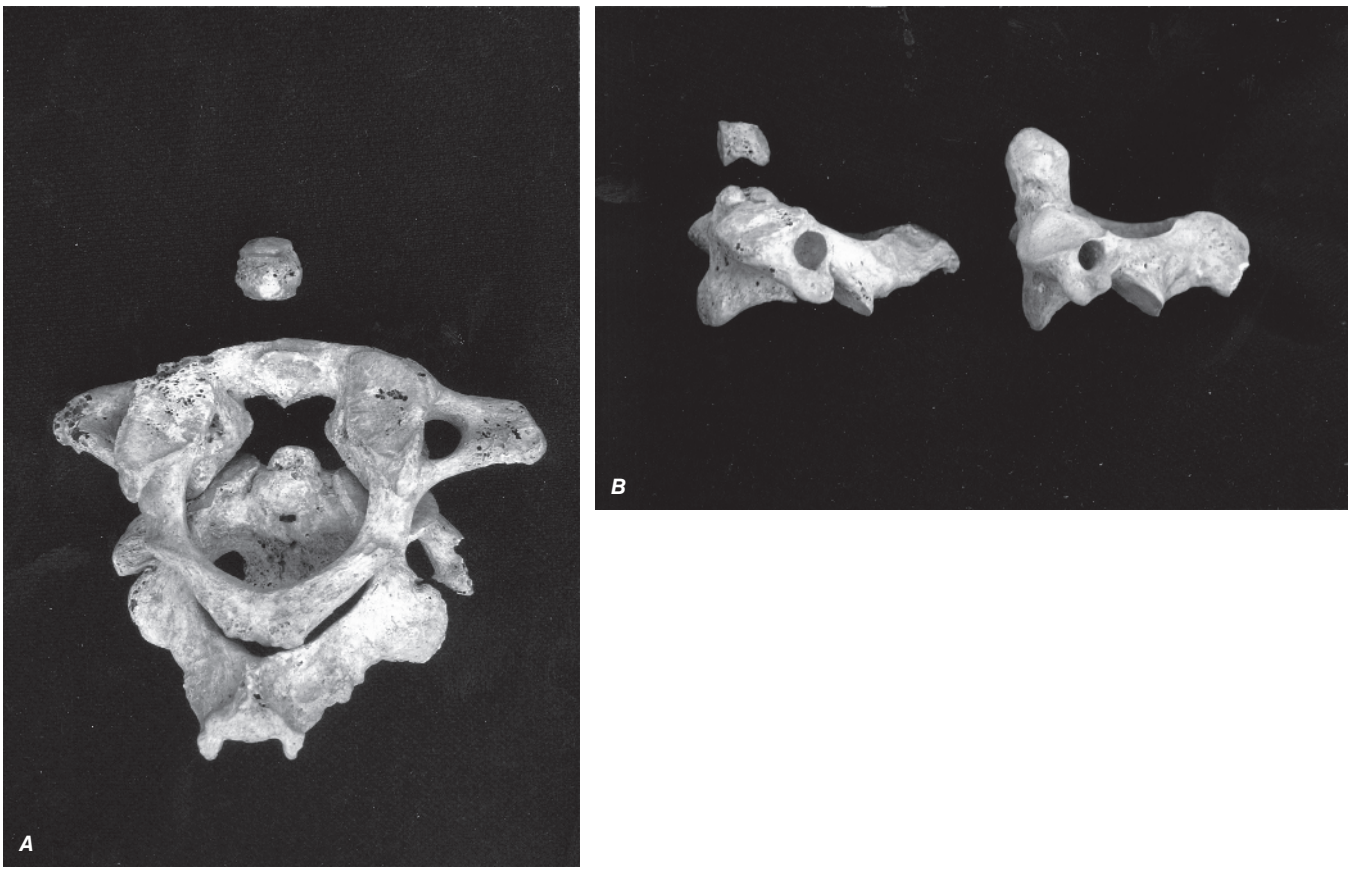

FIGURE A-5.3.6. O-C cranial border shift separated the apical dens' tip: (*A*) atlas with facet for dens and axis with separated apical dens' tip; (*B*) separated apical dens' tip compared with the normal axis dens, adult male, Frankish Corinth, Greece.

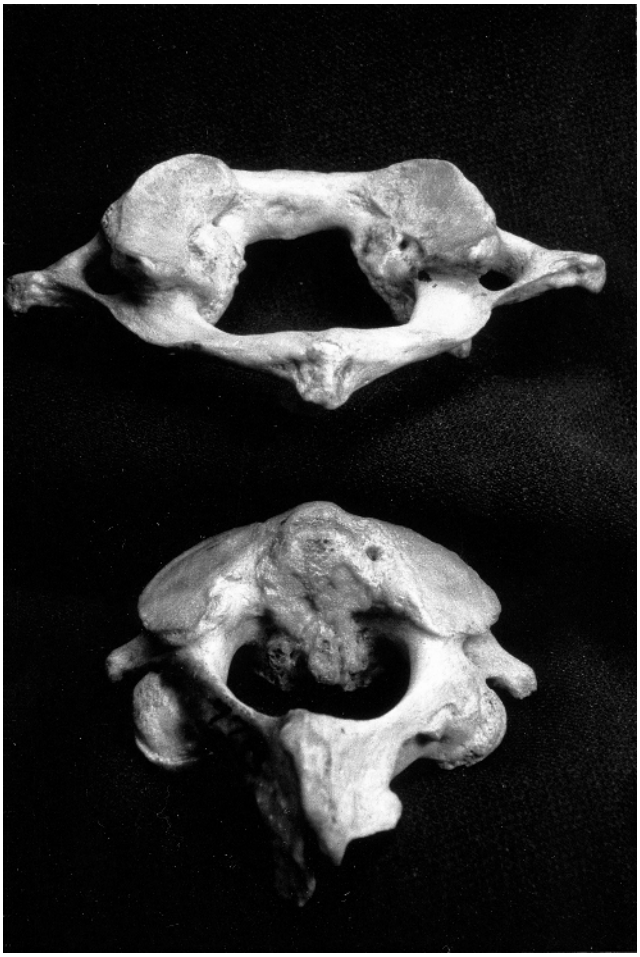

FIGURE A-5.3.7. O-C cranial border shift with agenesis of the apical dens' tip: atlas without facet for dens and axis with agenesis of the apical dens' tip, adult male (778), NMNH Terry collection.

basilar impression—indentation of the base of the skull. Neurological symptoms are often triggered by trauma to the neck that produces subluxation of the O-C (atlanto–axial) junction.

A-5.4. Caudal Shifting of the O-C Border

As the border moves downward between the atlas and axis vertebrae (Fig. A-5.4.1), the atlas becomes distorted as it is assimilated into the skull base (Figs. A-5.4.1B and A-5.4.2). Caudal shifting of the O-C border is far more common than cranial shifting with bilateral or unilateral expressions. The atlas tries to become part of the exoccipitals but fails to loose its vertebral identity. It often displays transverse foramina, whereas the occipital vertebra never does. The anterior arch is usually complete, but the posterior arch is often incomplete and distorted. Distortion of the foramen magnum is often present.

Mild expressions include condylar hypoplasia (Fig. A-5.4.1C) when caudal shifting attempts to obliterate the development of the major portion of the articulating condyles on the exoccipitals. Another form of caudal shifting of this border can affect the apical tip of the axis' dens, forcing it to articulate with the anterior rim of the foramen magnum. A precondylar facet appears on the anterior rim of the foramen magnum with a matching facet on the dens' apical tip (Fig. A-5.4.1D). The adjacent superior anterior arch of the atlas can also be affected with an additional matching facet (Fig. A-5.4.3). Caudal shifting interferes with the timing of the descent of the apical tip of the dens during morphogenesis, allowing it to protrude onto the anterior rim of the foramen magnum.

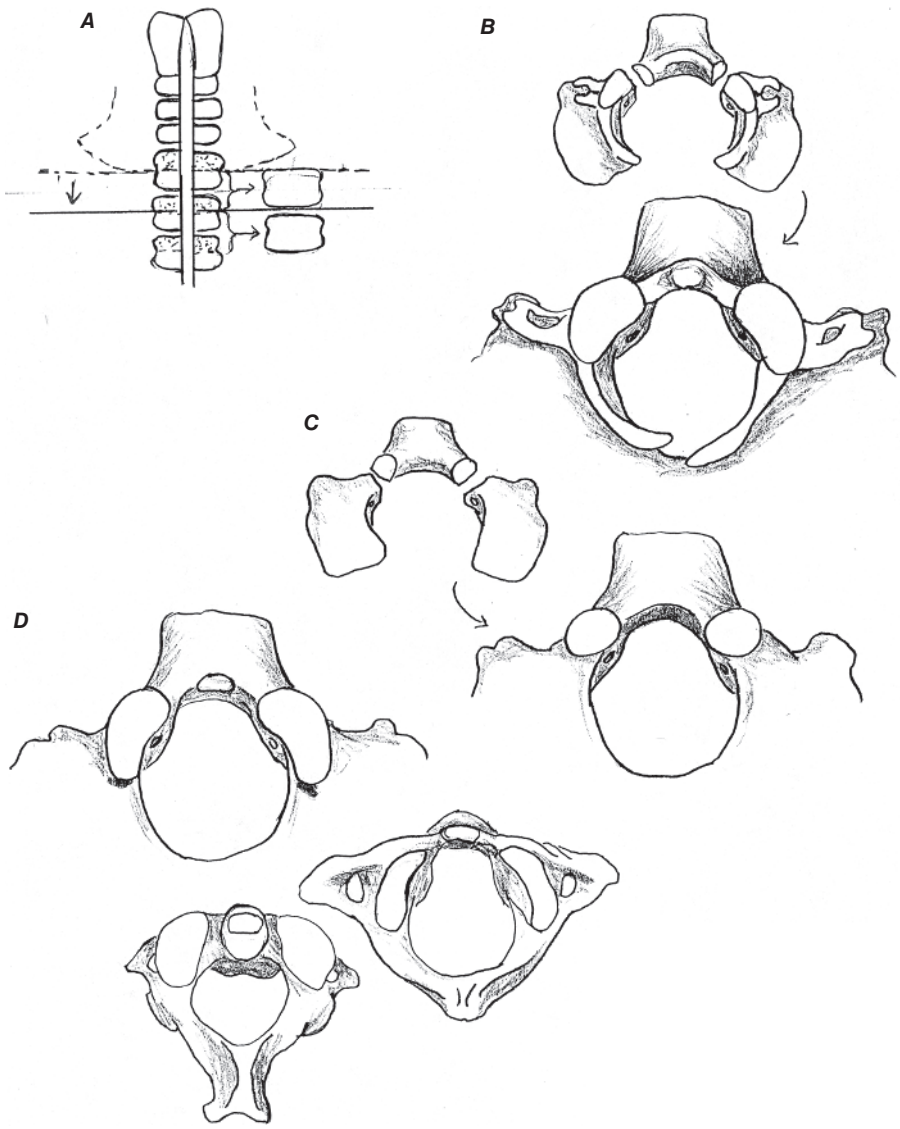

FIGURE A-5.4.1. Caudal shifts at the occipital–cervical (O-C) border: (*A*) schematic drawing shows the border (solid line) moving downward from the normal border represented by a dashed line; (newborn and adult) basioccipital; (*B*) complete shift occipitalized atlas; (*C*) mild shift condylar hypoplasia; (*D*) greater shift with matching articular facets on the anterior rim of the basioccipital, atlas, and dens of axis (not seen on newborn).

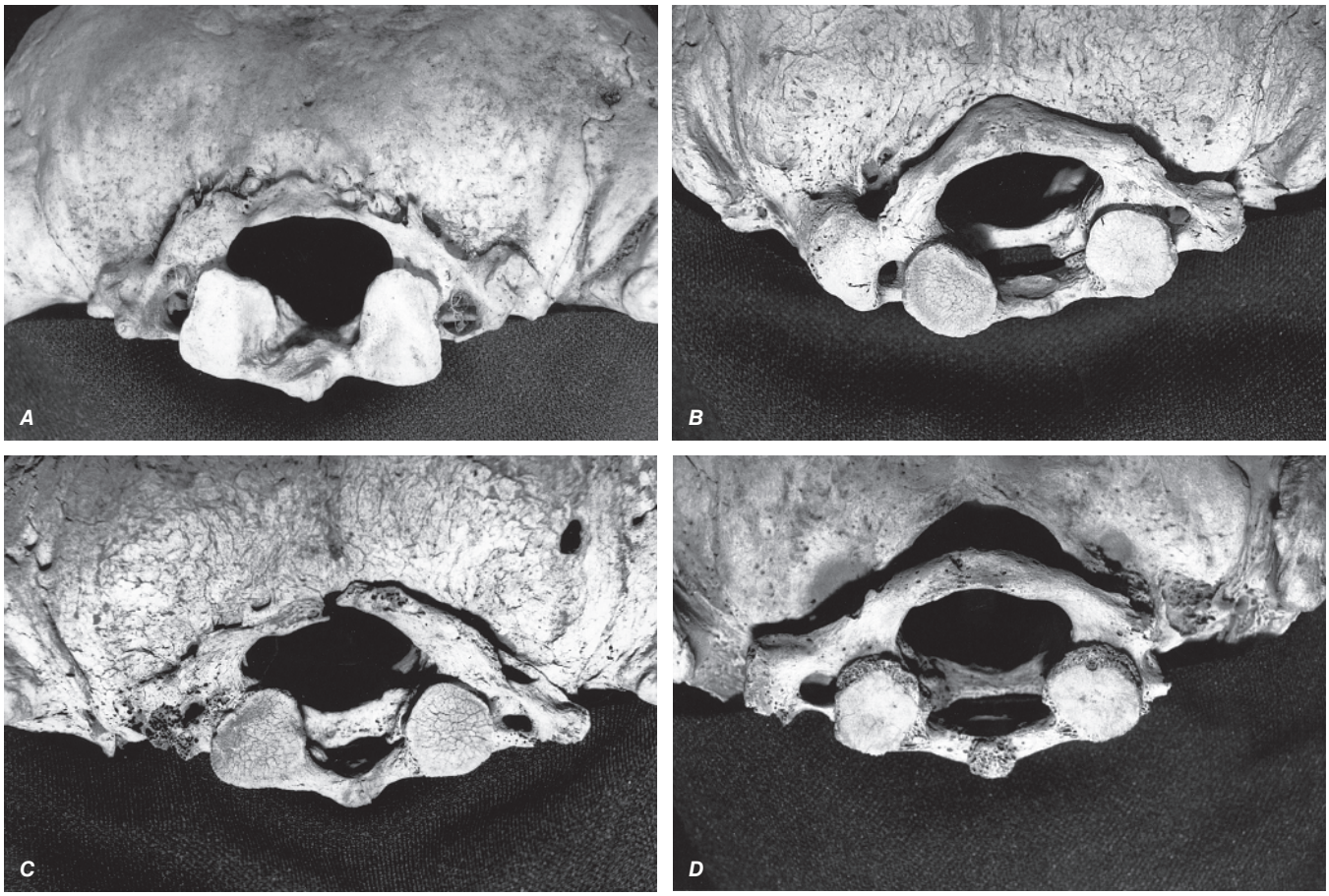

FIGURE A-5.4.2. O-C caudal border shift occipitalized atlas: (*A*) adult male (NMNH 157693), Chaviz Pass, AZ; (*B*) adult female (NMNH 264644), Chicama, Peru; (*C*) adult female (NMNH 264615), Chicama, Peru; (*D*) adult male (NMNH 266361), Pachacamac, Peru.

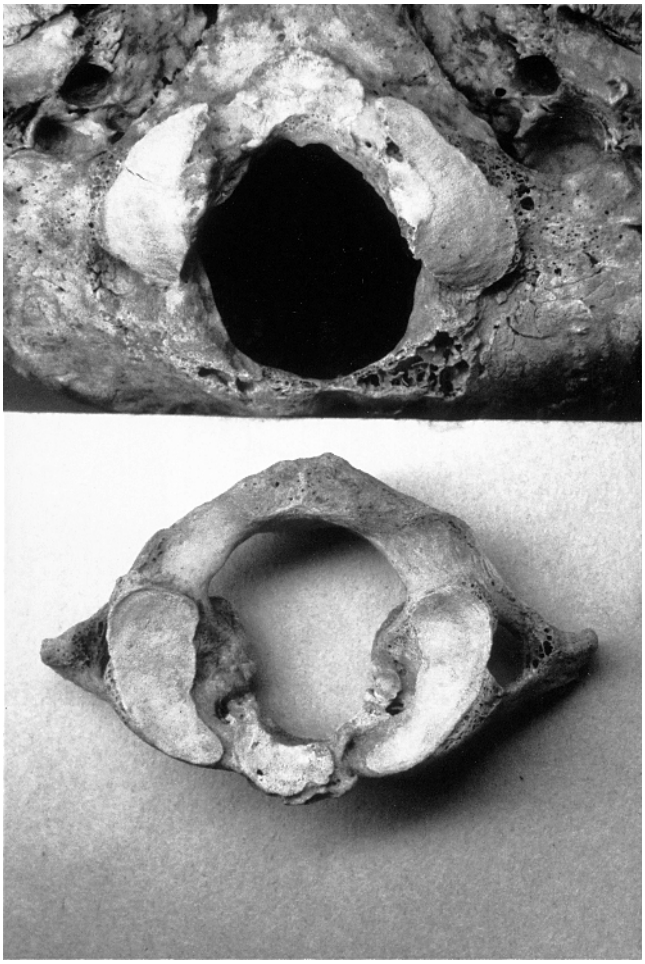

FIGURE A-5.4.3. O-C caudal border shift facets: matching articulating facets anterior rim of the foramen magnum and anterior arch of the atlas, adult female (NMNH 269267), Puye, NM.