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## Imaging of developmental anomalies of the eye and the orbit.

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*AJNR Am J Neuroradiol* 1992, 13 (2) 793-803

<http://www.ajnr.org/content/13/2/793.citation>

This information is current as  
of May 9, 2025.

# Imaging of Developmental Anomalies of the Eye and the Orbit

Larissa T. Bilaniuk<sup>1</sup> and Martha Farber

From the Children's Hospital of Philadelphia, Philadelphia, PA (LTB) and the Scheie Eye Institute, University of Pennsylvania, Philadelphia, PA (MF)

The development of the eye and the orbit encompasses a series of stages that take place during the embryonic and fetal periods (1–4). These stages overlap each other temporally, and are interrelated. Multiple diverse genetic and acquired conditions may derange this normal pattern of development. The specific type of anomaly that results, however, depends more heavily on the time that the insult or the developmental arrest occurred than on the specific etiology of the derangement (5). Familiarity with the sequence of developmental events is a prerequisite for understanding the various anomalies of the visual organ and for correct interpretation of diagnostic studies. Imaging modalities now allow precise delineation of the ocular and orbital (6) anomalies and of any associated central nervous system, facial (7), and systemic abnormalities. Such information may be critical in planning the treatment and management of the patient, as well as in establishing a prognosis.

## Normal Development

The eye develops from the neuroectoderm (neural plate), the surface ectoderm, and neural crest cells (1, 3, 4). Recent experimental embryologic studies have disproved the long held theory that mesoderm contributes significantly to the structures of the eye and the orbit. However, it is believed that the paraxial mesoderm that lies beneath the neuroectoderm induces it to become the eye-forming tissue, and also nourishes and

supports growth of the eye (2, 4). The mesenchymal tissue that gives rise to the eye and to the adnexal structures originates from neural crest cells that themselves arise from ectoderm located at the junction of surface ectoderm and neuroectoderm. Neural crest cells also give rise to the bony orbit, the fat, and nerve sheaths. At a specific time during the fetal development, the crest cells migrate to surround the optic cup and optic stalk (8). The development of the globe and the development of the orbit are independent of each other. In the developing eye and orbit, the paraxial mesoderm contributes only to the vascular endothelium and to the striated extraocular muscles. The origin, belly, and insertion of the extraocular muscles develop concurrently, and each individual muscle develops at the same time (9).

During the period of embryogenesis (first 3 weeks after conception), the neuroectoderm forms the neural plate with a central groove and paired neural folds that flank the groove on each side. During this period, a group of cells within the neural ectoderm is induced by the underlying paraxial mesoderm to become the eye-forming tissue. The neural groove deepens and begins to close. At the 22nd day, before the anterior neuropore closes, the optic primordium can be identified at either side of the still-open anterior neural groove. Eventually, the neural folds fuse to form a tube; the eye and the chiasm form from that portion of the neural tube destined to become the diencephalon.

By the 24th day, while the neural tube is still open anteriorly, an ocular pit forms in each of the paired neural folds (Fig. 1A) and extends laterally to touch the surface ectoderm on its own side. As the optic pits deepen into sulci, neural crest cells insinuate themselves between the surface ectoderm and the neural ectoderm of the optic vesicles.

<sup>1</sup> Address reprint requests to Dr Bilaniuk, Children's Hospital of Philadelphia, 34th St. and Center Blvd., Philadelphia, PA 19104.

**Index terms:** Eyes, abnormalities and anomalies; Orbits, abnormalities and anomalies; Pediatric neuroradiology

AJNR 13:793–803, Mar/Apr 1992 0195-6108/92/1302-0793

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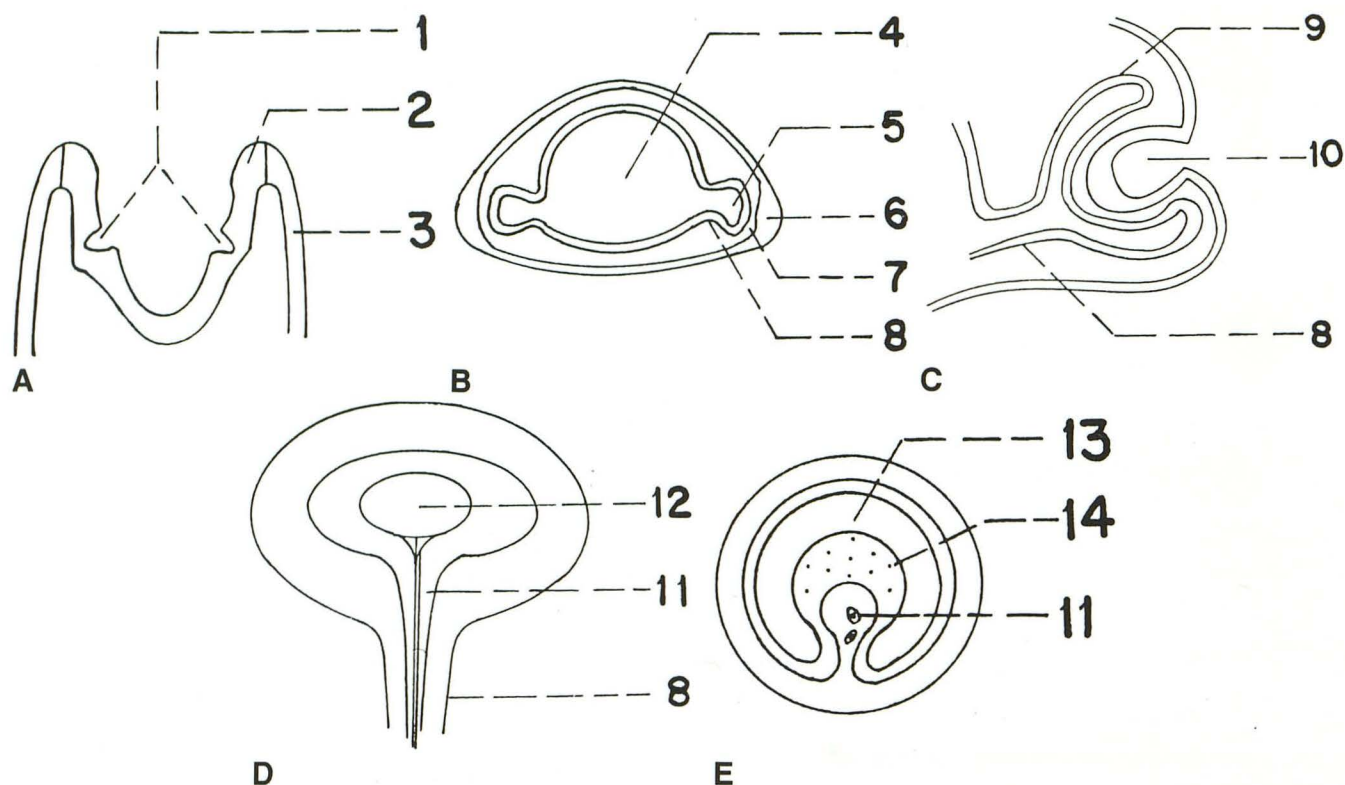


Fig. 1. A-D, Schematic drawings of early stages of eye development. E, Cross section of optic stalk with optic fissure. 1, optic pits; 2, neural ectoderm; 3, surface ectoderm; 4, forebrain cavity; 5, optic vesicle; 6, lens placode; 7, neural crest; 8, optic stalk; 9, optic cup; 10, lens vesicle; 11, optic fissure with hyaloid vessels; 12, lens; 13, optic stalk lumen; 14, axons from ganglion cells of the retina.

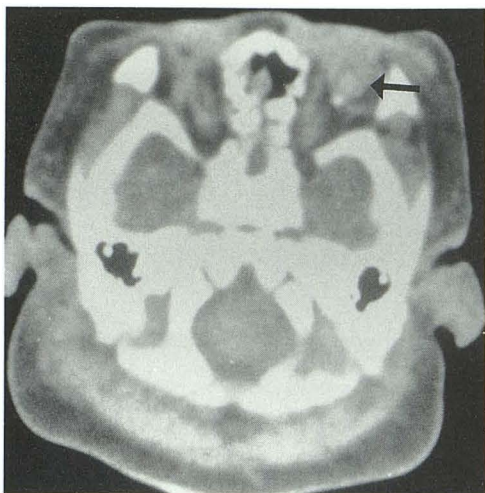
By the 26th day, the neural tube is closed at the anterior neuropore. The optic pits have deepened and expanded to form prominent outpouchings, the optic vesicles, that now project from the neural tube on short necks (the primitive optic stalks) (Fig. 1B). The surface ectoderm overlying the vesicles thickens and forms a lenticular placode which then invaginates and, through multiple steps, eventually forms the lens. At the same time, the optic vesicle invaginates to form a cup with an outer layer of cells destined to become the retinal pigment epithelium, an inner layer destined to form the neurosensory retina, and an intervening cavity called the subretinal space. This process can be compared to a hollow rubber ball where one side has been pushed in against the opposite side (Fig. 1C). However, the optic cup is not completely continuous; ventrally it contains a fetal (embryonic) fissure that permits mesenchymal and vascular tissues (the hyaloid artery) to enter the globe (Fig. 1D). This fissure extends along the optic stalk. In normal development, the fissure is eventually closed at its margins by multipotential cells.

During its development, the eye has a transi-

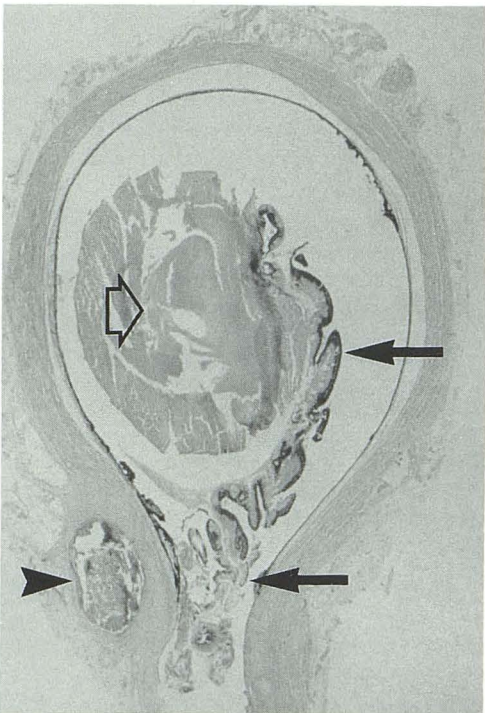
tory vascular system (the hyaloid artery and branches) (Fig. 1D) that forms the primary vitreous and nourishes the structures of the eye. This primitive vascular system involutes by the 35th week of gestation. The vascular primary vitreous also atrophies, and secondary vitreous is formed.

The optic nerve develops in the optic stalk, which connects the optic vesicle to the forebrain cavity (Fig. 1B). A single layer of undifferentiated neuroectodermal cells that lines the cavity of the forebrain is continuous with the retinal pigment epithelium of the outer cup (4). During the invagination of the optic vesicle, there is also invagination of the optic stalk (Fig. 1E). At the sixth week, axons from ganglion cells of the retina grow posteriorly into the optic stalk, toward the central nervous system. There is also proliferation of glial cells to form a supporting framework and mantle for the optic nerve. During normal embryogenesis, an excess of ganglion cells is generated. All these ganglion cells send axons into the optic stalk toward the brain. Those axons that do not make appropriate connections or do not reach their target centrally undergo degeneration. Thus, *normally* there is massive retrograde degenera-





A



B

Fig. 2. Right secondary anophthalmia and left congenital cystic eye.

A, Axial CT shows absence of the right globe. Somewhat thickened rectus muscles lying in the center of the orbit extend to irregular soft tissue that does not have any characteristics of a globe. The left orbit contains a congenital cystic eye (arrow) with a centrally located malformed lens and a small calcific density at its posterior aspect.

B, Histologic section of the congenitally cystic eye demonstrates the abnormal, centrally located lens (open arrow), disorganized retinal tissue (closed arrows) that extends into the optic stalk, and bony tissue (arrowhead) adjacent to the cystic eye.

tion of optic nerve fibers and retinal ganglion cells (10).

Once the lumen of the stalk is obliterated by

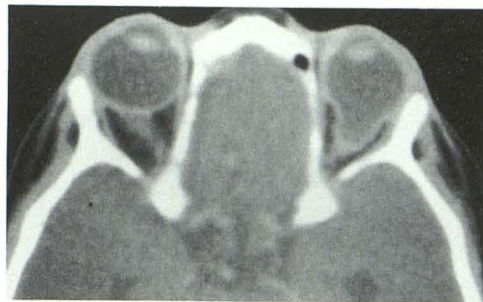


Fig. 3. Coloboma of the left posterior globe (morning glory syndrome). Axial CT demonstrates a smaller left globe with prominent outpouching posteriorly. (Case courtesy of Thomas P. Naidich, MD, Miami, FL.)

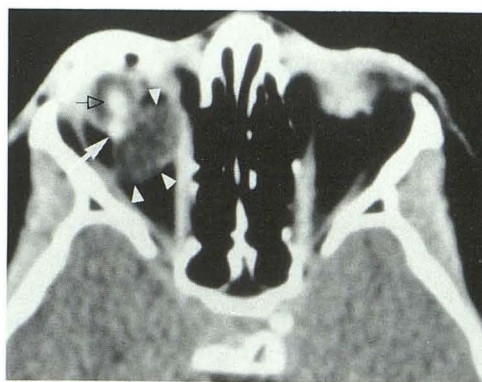


Fig. 4. Microphthalmia with cyst. Axial CT shows a small deformed right globe with a centrally located lens (open arrow) and calcified or ossified tissue (arrow) at the junction of the globe with the cyst (arrowheads). Extreme microphthalmia is present on the opposite (left) side.

the process of invagination and by the subsequent growth of axons, the direct communication between the cavity of the optic vesicle and the third ventricle is lost. The optic recess on the floor of the third ventricle represents the site of the original connection (4) and is the only residual of the previous direct communication between the third ventricle and the eye.

Initially, the chiasm is formed only from crossed optic nerve fibers. Later, between the 11th and 13th weeks, uncrossed fibers enter the chiasm. Myelination begins posteriorly in the region of the geniculate bodies at about the fifth month and reaches the globe by the eighth month (4).

### Anophthalmia and Microphthalmia

Anophthalmia results when there is failure of the optic pit to form a vesicle (primary anophthalmia) or when the optic vesicle forms and then degenerates (secondary anophthalmia) (Fig. 2A).



Complete failure of eye development is extremely rare. Only histologic examination can differentiate between true anophthalmia and severe microphthalmia. In cases of microphthalmia, ocular structures are identified, whereas they are absent in primary anophthalmia.

Microphthalmos is considered simple or pure when the eye is small but anatomically correct, and complex when the eye is malformed (11). Some use the term nanophthalmos to refer to the simple microphthalmos, but others reserve this term for those cases where there is marked decrease in the axial length of the eye (less than 18 mm), microcornea, and absence of systemic abnormalities (12). Microphthalmos, whether simple or complex, can be unilateral or bilateral; it can be the only abnormality or it can be associated with systemic disorders that may be due to genetic causes (eg, microphthalmos is present in 75% of Trisomy 13 patients), environmental causes (viral prenatal infections), or unknown causes (13). Weiss et al (11, 12) proposed that the pathogenetic mechanism for complex microphthalmos is inadequate production of secondary vitreous. They also stated that small optic cup, low intraocular pressure, altered vitreous proteoglycans, and abnormal release of growth factors play a role in the development of simple microphthalmos. The complex microphthalmos is further subdivided into two categories: noncolobomatous and colobomatous forms.

### Congenital Cystic Eye

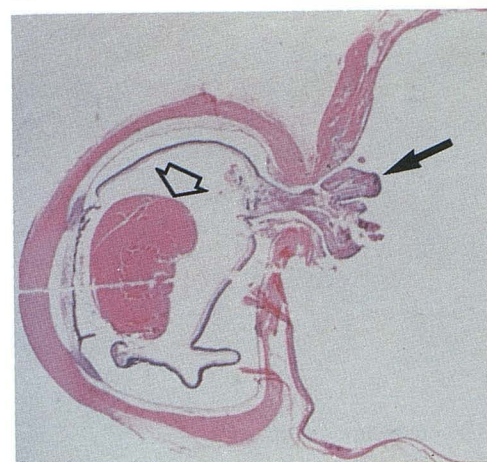
If the invagination of the optic vesicle does not occur or is incomplete, then a congenital cystic eye results (Fig. 2). This is an extremely rare condition. Microscopic examination reveals the cyst to be lined by malformed retina that shows an inverted arrangement of its layers, because there has been no invagination. That is, the outer layer lies on the side towards the cavity of the cyst (14) (Fig. 2B). Tendons of extraocular muscles insert into the outer fibrous wall of the cystic eye and there is an abnormal lens (Fig. 2). Imaging studies may not be able to differentiate this entity from a very disorganized severe microphthalmia.

### Colobomas

The term coloboma is of Greek origin meaning curtailment and is used in ophthalmology to refer to defects in ocular structures (14). Those colobomas that result from failure of the embryonic



A



B

Fig. 5. Microphthalmia with cyst.

A, Photograph of child with right microphthalmia and a huge cyst that obscures the small eye.

B, Histologic section from another case demonstrates a smaller malformed globe with an abnormal, centrally located lens (*open arrow*) and neuroectodermal tissue (*solid arrow*) prolapsing through a coloboma into a much larger cyst. (Case courtesy of Dr Morton Smith, Washington University, Seattle, WA.)

choroidal fissure to close properly are classified as typical colobomas (15). Colobomas are common malformations, constituting about 2% of all congenital abnormalities. They are usually bilateral (16).

A variant of typical coloboma of the disc is designated the morning glory syndrome, because in this malformation the optic disc is posteriorly displaced at the apex of a funnel-shaped staphylomatous excavation (Fig. 3). This configuration is thought to resemble the flower (17). Apple and colleagues (17) explain this on the basis of the fact that, during development, just as the cells at the margins of the embryonic fissure remain undifferentiated for a longer time and thus are more sensitive to insults, so do the cells of the future optic nerve head that may be considered to be one huge embryonic fissure.



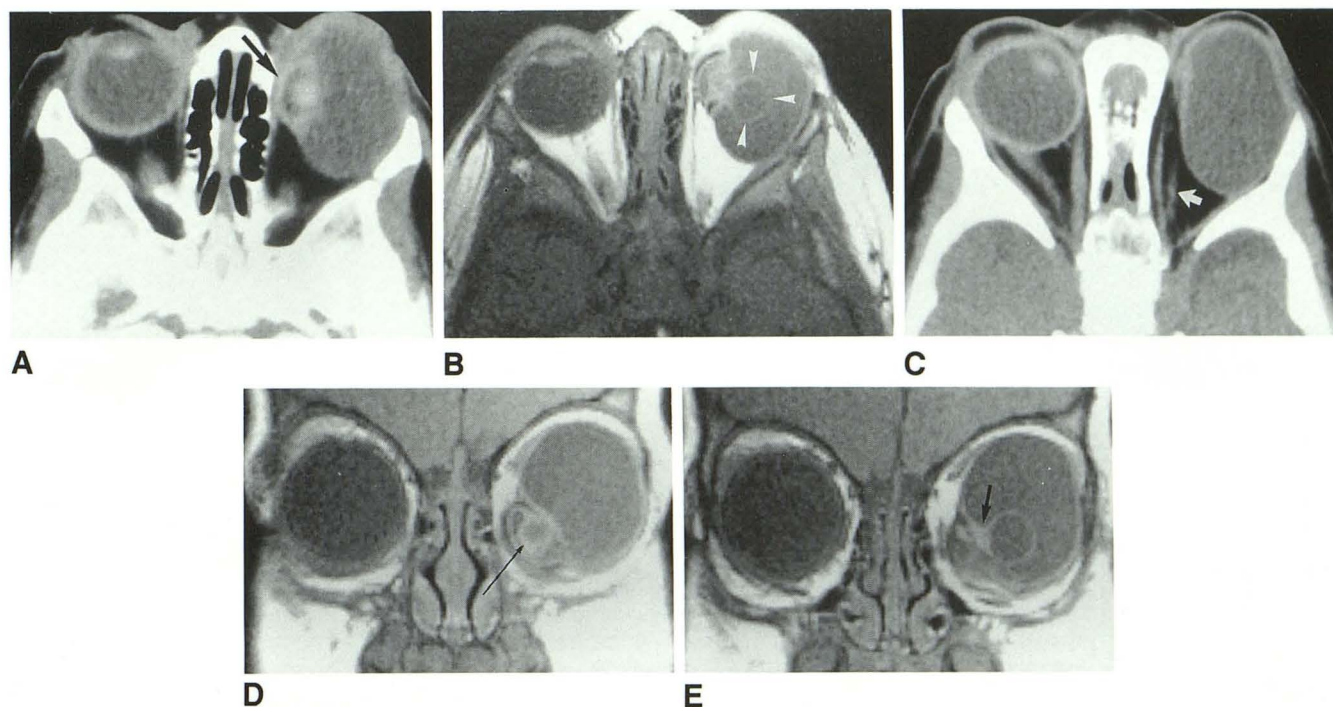


Fig. 6. Microphthalmos with a cyst.

A, Axial CT reveals a very small malformed globe (*arrow*) with centrally located lens lying medial to a huge cyst which has expanded the bony orbit.

B, Axial MR at the same level reveals a small deformed medially located globe and a huge cyst that contains a smaller cyst (*arrowheads*) adjacent to the lateral aspect of the abnormal globe. The second centrally located cyst was not identified on CT.

C, Axial CT section slightly superior to A reveals extraocular muscles and an optic nerve sheath complex (*arrow*) that extends to the abnormally small globe.

D, Coronal MR demonstrates the small deformed globe containing a lens (*arrow*). These lie inferomedial to the huge cyst.

E, Coronal MR slightly posterior to D reveals the smaller centrally located cyst whose wall is continuous with abnormal tissue (*arrow*) that is continuous with the abnormally small globe. (Case courtesy of Richard Boyer, MD, Primary Children's Medical Center, Salt Lake City, UT.)



Fig. 7. Aplasia of the right optic nerve. Axial T1 MR shows only the intracranial segment of the left optic nerve (*arrow*). No right optic nerve could be identified intracranially or within the right orbit.

### Microphthalmia with Cyst

Microphthalmos with cyst is a severe colobomatous malformation that results when neuroectodermal tissue (the inner layer of the retina) proliferates at the margins of the unclosed fetal fissure and protrudes outward forming a cyst or cysts (14, 18). The sizes of the eye and the cyst vary: the globe may be near normal size or small and the cyst small (Fig. 4), or there may be a tiny malformed eye with a huge cyst (Figs. 5 and 6). This entity may be associated with numerous systemic abnormalities.

Imaging modalities play an important role in the diagnosis of microphthalmos with a cyst for they demonstrate and characterize the eye and the cyst, and frequently demonstrate continuity between the globe and the cyst (19) (Figs. 4 and 6). Thus, this entity can be differentiated from other eye anomalies. In addition, computed tomography (CT) or magnetic resonance (MR) can demonstrate associated brain abnormalities.



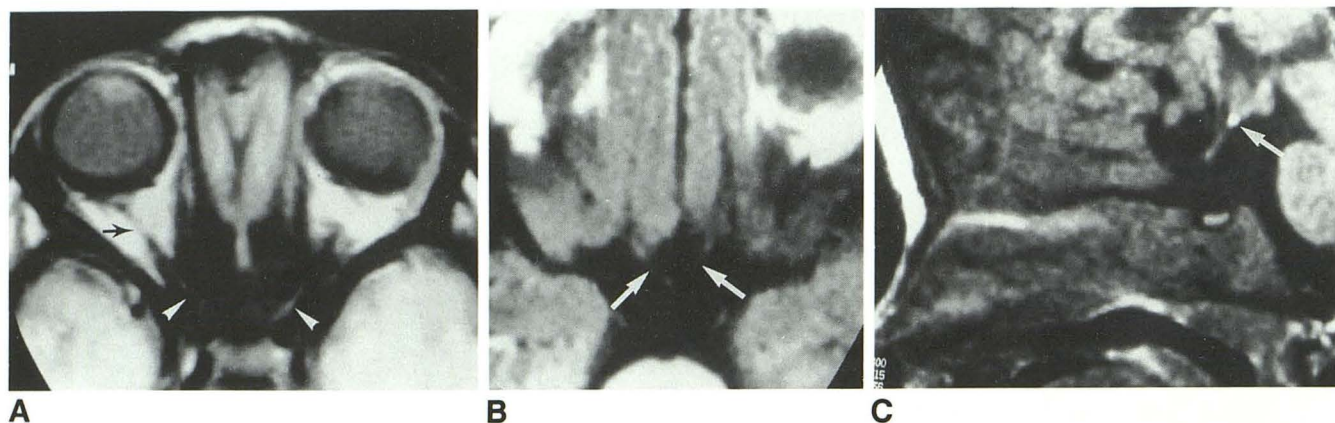


Fig. 8. Marked bilateral optic nerve hypoplasia with pituitary anomaly.

A, Proton density MR demonstrates tiny optic canals (arrowheads). The right optic nerve sheath complex (arrow) is very small.

B, Slightly higher MR section demonstrates extremely small intracranial optic nerves (arrows).

C, Sagittal MR shows absence of the pituitary stalk. There is abnormally high signal intensity (arrow) in the inferior hypothalamus at the site where the pituitary stalk would be expected to have arisen.

### Optic Nerve Aplasia and Hypoplasia

Optic nerve aplasia is due to failure of the embryonic fissure to reach the optic stalk. It is a sporadic, unilateral condition and is not associated with any systemic abnormalities (Fig. 7) (3).

Optic nerve hypoplasia (Fig. 8) is a subnormal number of axons. It is a frequent congenital anomaly that may be isolated or may be associated with ocular, facial, endocrine, or central nervous system anomalies (10). Among such entities are septo-optic dysplasia (Fig. 9) and encephalocele. One prospective study of 93 children with optic nerve hypoplasia found that those children who had bilateral optic nerve hypoplasia and poor vision had the highest incidence of associated nonocular congenital abnormalities and medical problems (10). In this study, developmental delay was the most frequently associated abnormality, and hypothyroidism was the most frequent endocrine disturbance. Those children that had bilateral optic nerve hypoplasia with good vision or those that had unilateral optic nerve hypoplasia fared much better.

The proposed pathogenesis of optic nerve hypoplasia is excessive degeneration of optic nerve axons and retinal ganglion cells during development of the eye and visual pathways (much more than the normal amount) (10). Thus, one cause of optic hypoplasia can be major midline and hemispheric anomalies that prevent the axons of the retinal ganglion cells from making connections at their intracranial target sites. However, there are cases where the optic nerve hypoplasia is the only abnormality. In these instances, the

pathogenesis may be primary agenesis of retinal ganglion cells.

In children with optic nerve hypoplasia, imaging studies are of great value to exclude any associated intracranial abnormality. The imaging studies demonstrate small optic nerves and chiasm, display the morphology of the pituitary gland (Fig. 8) and depict associated brain anomalies (Fig. 9). Slight symmetrical decrease in the size of the optic nerves may be difficult to appreciate on imaging, just as it is difficult to appreciate clinically. Marked decrease in the size of the optic nerves or unilateral involvement is much more easily detected.

### Persistent Hyperplastic Primary Vitreous

Persistent hyperplastic primary vitreous (PHPV) (Fig. 10) results when the embryonic

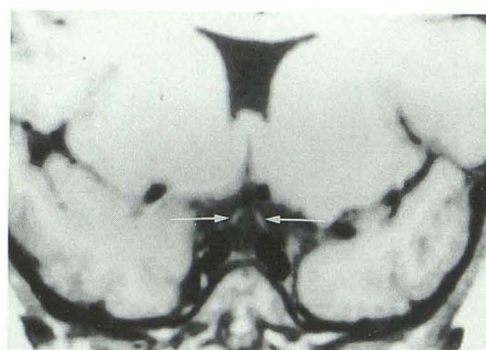


Fig. 9. Septo-optic dysplasia. Coronal T1 MR shows absence of the septum pellucidum and extremely small intracranial optic nerves (arrows). A more posterior section (data not shown) demonstrated an anomaly of the posterior pituitary gland.



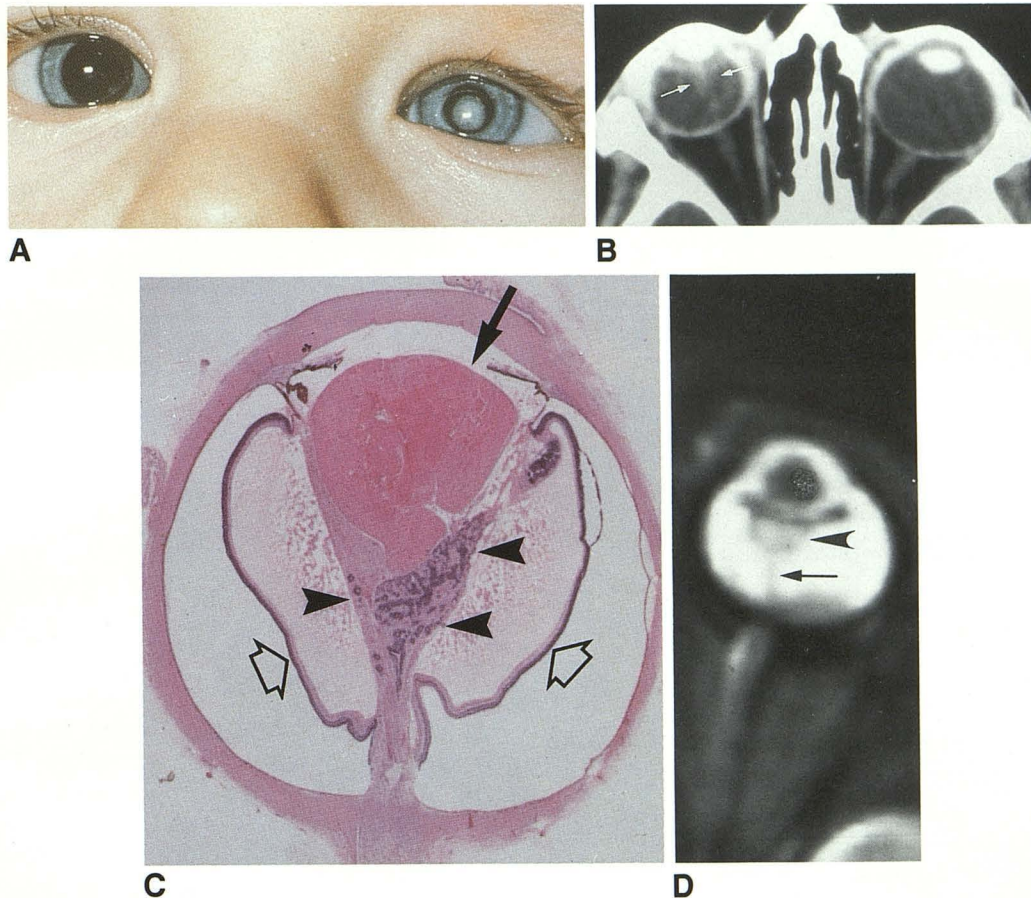


Fig. 10. Persistent hyperplastic primary vitreous. Four different cases.

A, Photograph of a child with left leukocoria due to persistent hyperplastic primary vitreous.

B, Histologic section of persistent hyperplastic primary vitreous shows the abnormal lens (arrow) and triangular fibrovascular material (arrowheads) extending from it toward the optic nerve head. The retina (open arrows) is detached.

C, Axial CT reveals a smaller right globe with tissue of increased attenuation (arrows) surrounding the posterior aspect of the right lens and extending posteriorly toward the right optic nerve head.

D, Axial T2 MR demonstrates a deformed lens with prominent retrolental tissue (arrowhead) that has a thinner band of tissue (arrow) extending from it toward the posterior aspect of the globe.

intraocular vascular system (hyaloid artery with its branches) and the fibrovascular tissue of the primary vitreous fail to regress normally (20). It occurs in normal full-term infants, is usually unilateral and is usually associated with microphthalmos. The most typical clinical findings are leukocoria (white pupil), microphthalmia, and cataract (21). The presence of microphthalmia and absence of calcification are important factors differentiating PHPV from retinoblastoma, since retinoblastoma also presents with leukocoria but typically shows calcifications and a globe that is normal in size or enlarged. Glaucoma is a relatively frequent complication of PHPV (26% in one series) and thus the affected eye may be buphthalmic (21). Other complications that may occur

include recurrent spontaneous hemorrhages, retinal detachment, and phthisis bulbi.

In PHPV, histologic examination reveals a hyaloid artery that extends from the optic disc to a fibrovascular retrolental tissue (22) (Fig. 10B). Imaging studies, CT and MR, demonstrate a smaller globe with increased attenuation or hyperintensity on T1-weighted images and tissue that extends from the posterior aspect of the lens to the back of the globe in a band-like or triangular configuration (20, 23, 24).

### Congenital Glaucoma

Glaucoma is a condition that results from abnormal elevation of intraocular pressure. The elevated intraocular pressure is most often due to



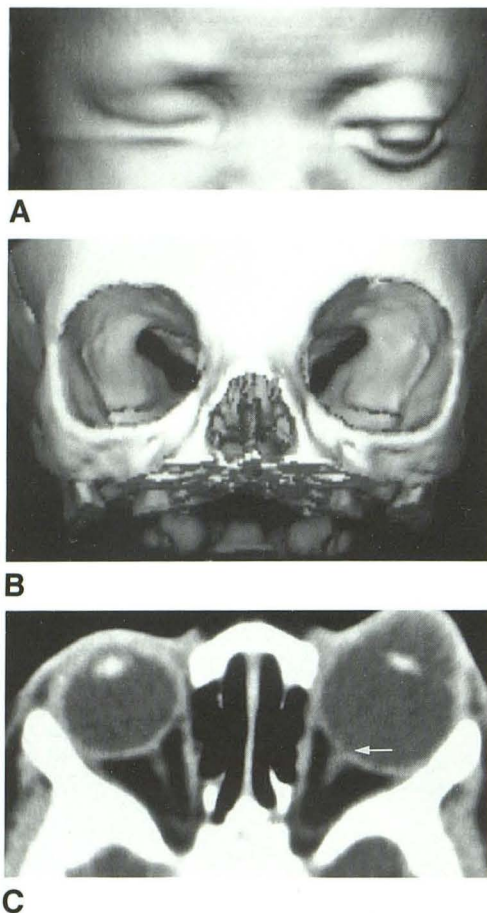


Fig. 11. Severe primary congenital glaucoma of the left eye.

A, Three-dimensional soft tissue CT shows marked enlargement of the left eye that protrudes between the lids.

B, Three-dimensional bone CT shows enlargement of the bony left orbit.

C, Axial CT demonstrates prominent diffuse enlargement of the left eye, particularly the anterior chamber. The left lens is flattened. There is slight indentation (*arrow*) at the optic nerve head consistent with cupping.

increased resistance to normal outflow of aqueous humor from the eye. Primary congenital glaucoma is most frequently due to abnormal development of the trabecular meshwork. Anderson's investigations (25) suggest that the primary defect of congenital glaucoma may be premature or excessive formation of collagenous beams within the trabecular meshwork. These beams are thought to generate traction forces that cause compaction of the trabecular sheets with consequent obstruction to the outflow of aqueous humor (25).

Congenital glaucoma is bilateral in up to 80% of cases (26). It may be the only abnormality, or

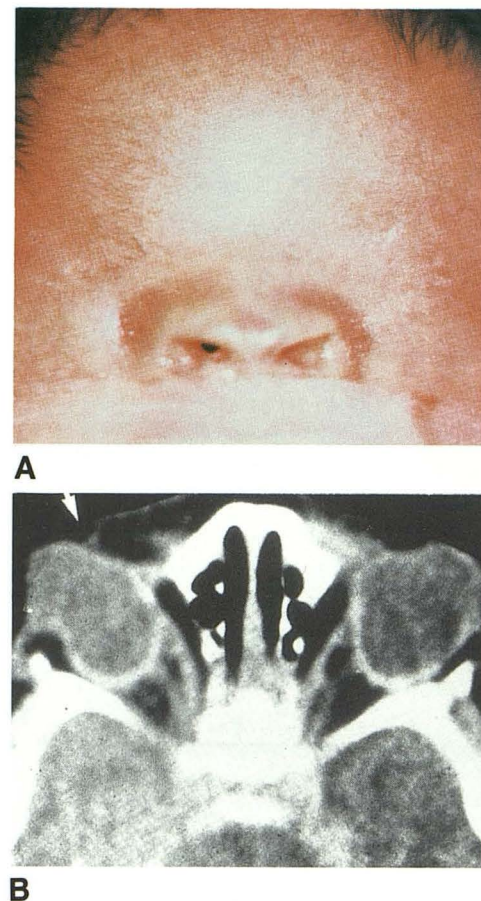


Fig. 12. Cryptophthalmia.

A, Photograph of the infant shows total absence of the eyelids and continuous skin from forehead across the orbits to the cheeks. (Case courtesy of Dr. James Katowitz, The Children's Hospital of Philadelphia, PA.)

B, Axial CT shows malformed anterior structures of each globe and continuity of the skin (*arrow*) with the tissue of the orbits. There is absence of the cornea, anterior chamber, and lens on each side. (Reprinted with permission from Levine et al (28).)

it may occur in association with other developmental disorders including phakomatoses. The glaucoma may manifest itself at birth or during the first few years of life. Increase in intraocular pressure causes enlargement of the entire eye (Fig. 11).

Secondary congenital glaucoma refers to those cases of glaucoma that result from intrauterine eye inflammation (eg, rubella), trauma, or ocular tumor (eg, retinoblastoma). Insults or maldevelopments that result in glaucoma occur anywhere from the fourth lunar month of gestation onward, at a time when the anterior structures of the eye are developing.



## Cryptophthalmos

Cryptophthalmos ("hidden eye") results when the lid folds fail to develop. Cryptophthalmos is considered typical when there is complete absence of the lids (complete ablepharia) and the skin is continuous from the forehead, across the orbits, to the cheeks (27) (Fig. 12). The skin over the globes is shown pathologically to be corneal epithelium that has undergone metaplastic change (2). There is absence of eyelashes, meibomian glands, lacrimal glands, lacrimal puncta, and usually also eyebrows (27). Cryptophthalmos is not an isolated anomaly but is part of a systemic syndrome that often includes anomalies of face, cranium, extremities, and urogenital system

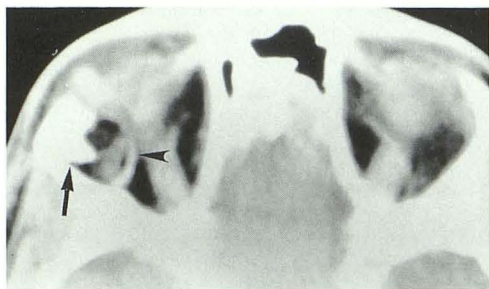


Fig. 13. Dermoid cyst. Axial CT demonstrates a heterogeneous mass with a wall of higher attenuation (*arrowhead*). The content of the mass is heterogeneous with some fatty tissue (low attenuation). In addition, the dermoid extends into the frontal zygomatic suture (*arrow*).

(28). Imaging studies are needed to demonstrate the globes and the rest of the intraorbital structures prior to surgical intervention.

## Obstruction of the Nasal Lacrimal Duct

The nasal lacrimal duct forms in the invaginated surface ectoderm between the inner canthus and the inferior turbinate. Obstruction of this duct is a frequent anomaly and is usually due to an imperforate membrane at the lower end of the duct (29). The membrane consists of epithelium of the lacrimal duct and the mucosa of the nose. The obstruction is manifested by overflow of tears (epiphora), or less commonly by a dacryocystocele. Imaging studies are helpful in differentiating this entity from other masses that may present in the newborn.

## Congenital Tumors of the Orbit

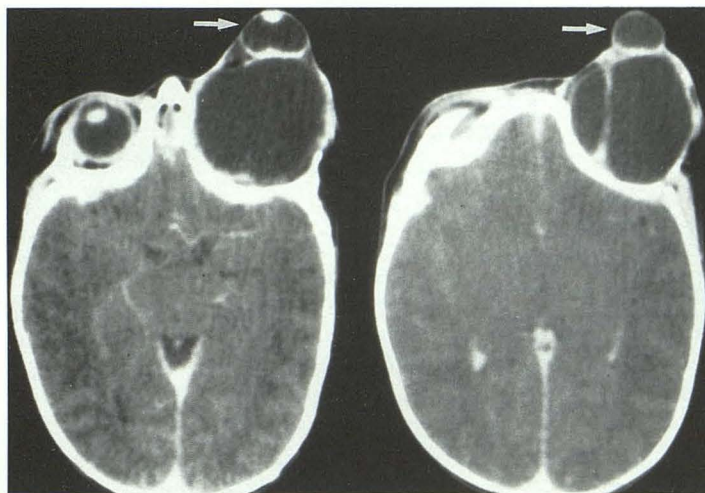
Congenital tumors of the orbit may be derived from one germ cell layer, such as dermoid cysts, from two germ cell layers (teratoid tumors), or from three germ cell layers (teratoma) (30). These congenital tumors are not true neoplasms but developmental choristomas—that is, malformed tissue found at a site that normally does not contain that tissue.

## Dermoid and Epidermoid Cysts

Dermoid and epidermoid cysts originate from sequestered embryonic surface ectoderm and oc-



A



B

C

Fig. 14. Left orbital teratoma.

A, Photograph of a child shows marked protrusion of the globe by a large posteriorly located mass that has expanded the orbit.

B, and C, Contrast-enhanced axial CT scans demonstrate the large cystic lesion that markedly expands the bony orbit and displaces the globe (*arrows*) anteriorly. (Case courtesy of Thomas P. Naidich, MD, Miami, FL.)



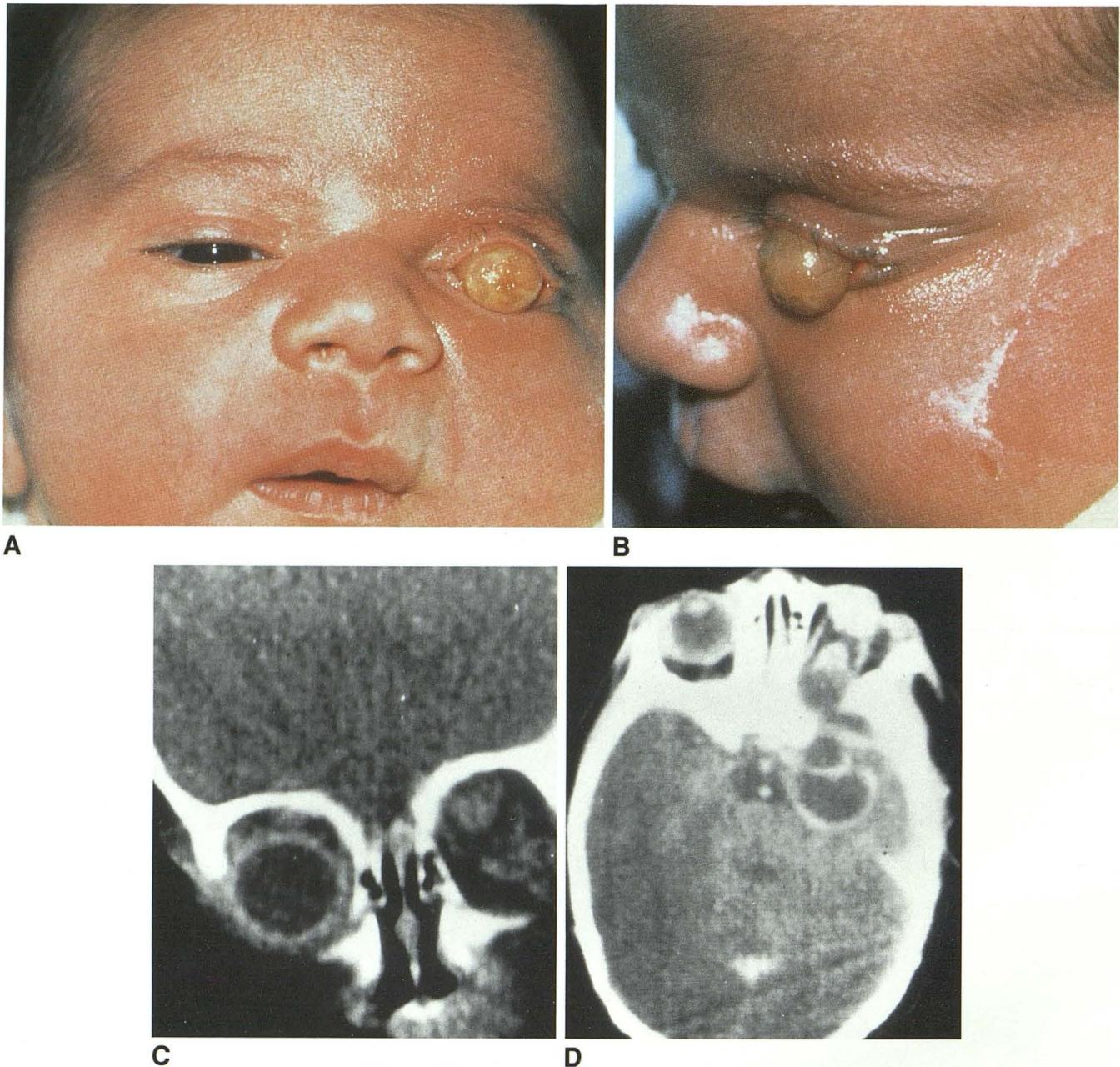


Fig. 15. Orbital teratoma.

A, Frontal and B, Left lateral views of the child show protrusion of the markedly deformed globe anteriorly through the palpebral fissure.

C, Coronal CT shows heterogeneous irregular tissue within the left orbit.

D, Contrast-enhanced axial CT reveals multiloculated cystic tissue that extends from the orbit into the left temporal fossa. At surgery, the teratoma lay entirely anterior to the temporal dura. Histologic evaluation of the tumor revealed tissues with pulmonary, gastrointestinal, muscular, and renal characteristics. Histologic examination of the globe revealed severely dysplastic microphthalmia with absent lens, iris, and anterior chamber segments; persistent hyperplastic primary vitreous; and retinal dysplasia. (Case courtesy of Lucy Rorke, MD, Philadelphia, PA.)

cur frequently in the orbit. They are typically located at the site of sutures, particularly the zygomaticofrontal and frontoethmoidal sutures. Dermoid cysts are the most common pediatric

orbital tumor. They contain adnexal structures, such as hair and sebaceous glands and, therefore, their contents are frequently oily due to the secretion of sebum. Epidermoid cysts do not con-



sist of adnexal structures but may contain cholesterol in addition to epithelial debris. Imaging studies reveal the cysts as well-defined masses that may have thin walls. The cysts may be homogeneous or heterogeneous (Fig. 13) containing tissue ranging from muscle to fat in radio-density and signal intensity. Some lesions may contain a fat-fluid level or may be diffusely fatty. Larger lesions may produce scalloping of the adjacent bony wall or even expansion of the entire orbit.

### Congenital Orbital Teratoma

Orbital teratomas are rare congenital tumors that contain tissue derived from all three germ cell layers and are usually benign (30). In most instances, the tissue is disorganized, but some teratomas may actually have fetal features. One theory of pathogenesis of the teratomas is lack of normal response of tissue to inducers (31, 32).

The most common presentation of a teratoma is extreme unilateral proptosis in an otherwise healthy full-term infant (30) (Fig. 14A). The tumor usually grows rapidly after birth. Most typically, the tumor is multicystic (Figs. 14 and 15), and may be mixed cystic-solid or completely solid. Usually the bony orbit is markedly expanded and is two to three times its normal size (Fig. 14). Rarely the teratoma may extend intracranially (33) (Fig. 15). Imaging studies permit delineation and characterization of the lesions, which leads to correct diagnosis, even when the clinical findings are confusing. Teratomas need to be differentiated from other congenital lesions, such as microphthalmos with cyst, dermoid cyst, congenital cystic eye, hemangioma, and meningoencephalocele.

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