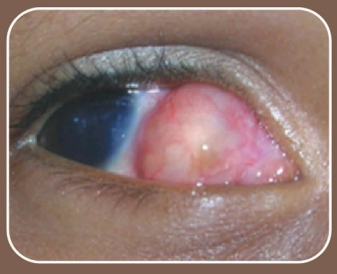
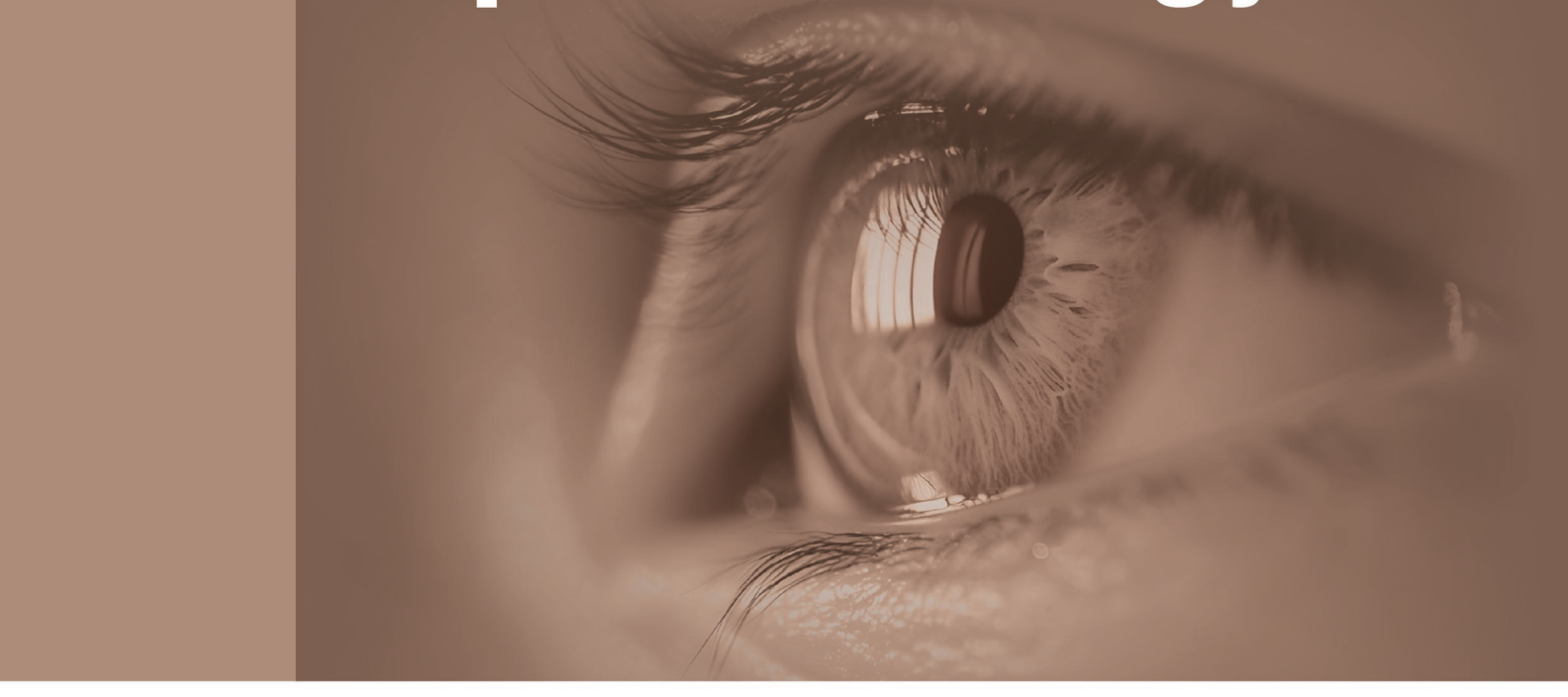
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Comprehensive Pediatric Ophthalmology



PK Mukherjee

JAYPEE

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CHAPTER

16 Disorders of the Sclera in Children

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Scleral disorders are relatively rare at all ages, but they are still rarer under 20.

The sclera is the outer coat of the eyeball, constituting almost 85% of the total surface, and is opaque; the rest of the globe is formed by a transparent cornea. It is stronger than the cornea when considered to be a protective part; it does not have any optical function, while the cornea is predominantly optical in function. Besides being protective, the other functions of the sclera are to anchor extraocular muscles, keep the interior of the eyeball dark, and let intraocular blood vessels and nerves pass through the sclera without supplying blood to it, making it avascular all over except 1 mm round the cornea at the limbus. Its sensations are duller than those of the cornea, retaining a moderate pain sensation. The sclera maintains the shape of the eyeball.

A fully developed sclera constitutes a hollow sphere of 22 mm in diameter with two openings, one large anterior and one small posterior. The first is referred to as the anterior scleral, while the later is called the posterior scleral foramen. The anterior foramen makes the limbus and has a diameter of 11.00 mm; the posterior foramen is 2.00 mm in diameter and transmits the optic nerve along with its covering and blood vessels. The posterior foramen is covered by a thin fibrous sheet of tissue with perforation in it; called lamina cribrosa, it is not flat but is depressed backward. Nothing passes through the lamina cribrosa; the exact purpose of the foramen is not well understood. The lamina is not as rigid as the wall of the sclera; it bulges forward in papilledema and bends back in glaucoma.

The foramina other than the two mentioned earlier are meant for the transmission of:

- Long and short ciliary vessels and nerves
- Exit of venae vorticosae
- Passage of anterior ciliary vessels.
- These foramina are situated round the entry of the optic nerve, in two groups as per their distance from the optic nerve. The long and short ciliary nerves and vessels pass through them.
- These apertures are situated more anteriorly, four in number, 4–7 mm behind the equator.
- Through the anterior aperture, pass the anterior ciliary arteries. They enter the sclera just in front of the insertion of the recti, and the anterior ciliary veins make their way through these openings **(Fig. 16.1)**.

The color of the adult sclera is marble white; it has a blue tinge at birth, which changes to white by 1 year. The anterior part of the sclera is visible round the cornea in the interpalpebral aperture and is referred to as the

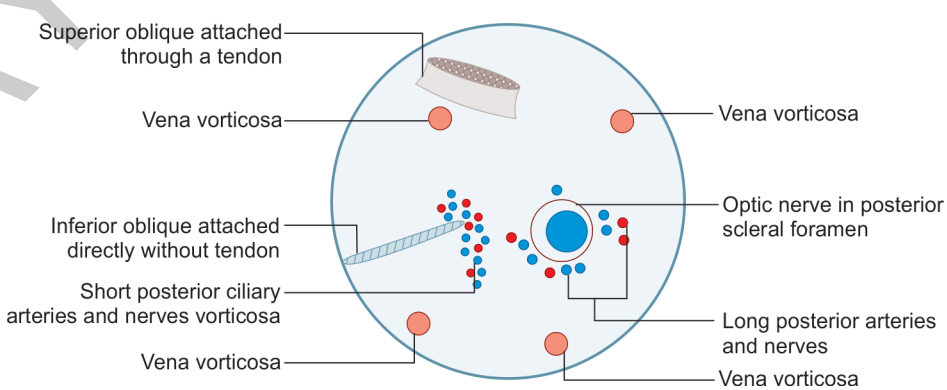


Fig. 16.1: Structures at the posterior sclera.

white of the eye in contrast to the cornea iris complex, which is called the black of the eye. The sclera is covered by transparent bulbar conjunctiva. The conjunctiva can be moved over the sclera and lifted up with ease. The second structure that covers the sclera is Tenon's capsule, which extends from the optic nerve to the limbus, not reaching the corneal margin; it terminates about 1–2 mm away from the limbus. The Tenon's capsule encompasses the globe snugly, with a potential space between the two. The sub-Tenon's space is used to place sub-Tenon drugs. The potential space is traversed by fine fibrils that enhance the attachment of the Tenon's capsule to the sclera. Posteriorly, the Tenon's capsule terminates round the optic nerve, merging with its covering. The sclera is not uniformly thick throughout; it is thicker at the limbus and round the optic nerve; it is 1 mm round the optic nerve and 0.75 mm round the limbus; and it is thinnest at the insertion of the recti.

ATTACHMENT OF EXTRAOCULAR MUSCLES

No Extraocular Muscle Originates from the Sclera

The attachment of the extraocular muscles to the sclera has a great bearing on the movement of the globe. Knowledge of the position of attachments is of great importance in squint surgery. The attachment of extraocular muscles to the sclera can be put into two groups: those attached anterior to the equator and those attached behind the equator **(Fig. 16.2)**.

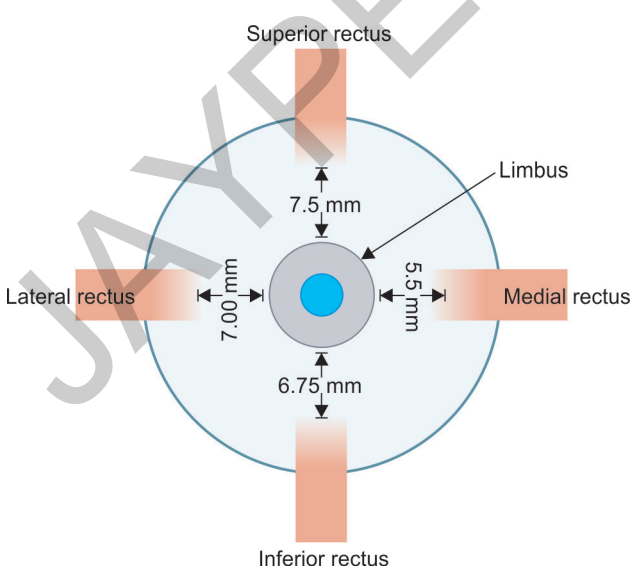


Fig. 16.2: Attachment recti anterior to equator.

The first group consists of four recti; the second group consists of two obliques. During squint surgery, the distance of attachment is measured in millimeters from the limbus, which is not equal for all muscles. The distance of insertion of the obliques from the limbus is far more that of the recti. The width of attachment of individual muscles is variable. The inferior oblique is the only muscle that attaches itself directly to the sclera. The other extraocular muscles are attached by various lengths of tendons. The sclera is thinnest at the insertion of the muscles; the muscles blend intimately with the sclera; hence, precaution must be taken while detaching muscles in squint surgery to avoid inadvertently nicking the sclera.

Anatomically Weak Spots in Sclera (Fig. 16.3)

The weak spots on the sclera are:

- Thin sclera at attachment of recti muscles.
- Thinned sclera following trauma and inflammation.
- The third group consists of various foramina through which the vessels and nerves pass.
- The other weak spots are:
  - Corneoscleral junction
  - Equator
  - Sclera over the ciliary body
  - Posterior pole

Development and Congenital Anomalies of Sclera

The sclera is fully mesodermal; it develops from the paraaxial mesenchyme round the developing optic nerve, which has two parts: the anterior and posterior parts. The anterior part grows earlier and faster than its posterior counterpart. The anterior part of mesoderm not only gives rise to sclera but also to the attachment of recti that grow backward. In the initial stage, the limbus is far posterior to its final position on the developing ciliary body. The developing mesoderm reaches the future posterior pole by the 3rd month to grow round the developing optic nerve and give rise to the lamina cribrosa. The scleral spur, which is located relatively anteriorly, becomes evident by the 16th week; by the 20th week, the sclera is almost fully developed.

Disorder of Sclera

Disorders of the sclera can be:

- Congenital
- Acquired

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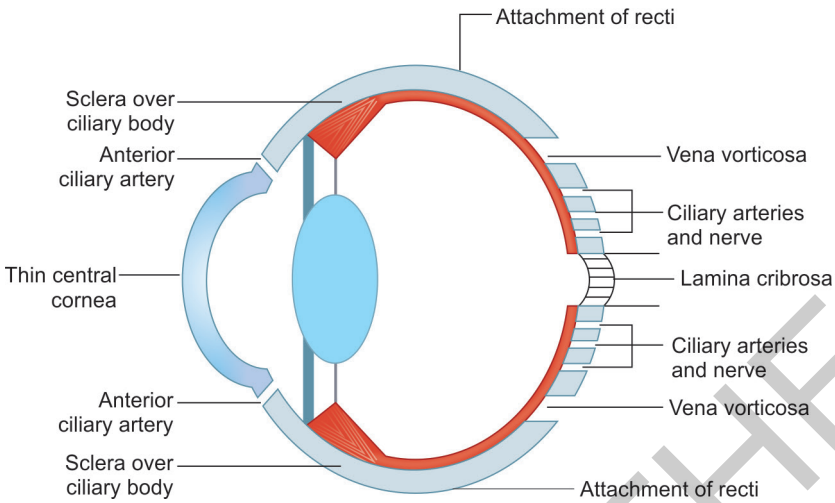


Fig. 16.3: Weak spots on the sclera and cornea.

Congenital

The congenital anomalies may be any of the following in isolation or in combination; most of the congenital anomalies of the sclera are bilateral and mostly symmetric.

They are caused by:

- Failure of the development of the optic cup.
- Retarded growth of the optic cup.
- Failure of mesoderm round the optic vesicle.

There are four types of common congenital anomalies:

1. Colobomas are generally associated with colobomas of the uvea.
2. Small eyeballs
   i. Anophthalmos
   ii. Microphthalmos
   iii. Nanophthalmos
3. Large eyeballs, congenital scleral ectasia
   i. Posterior staphyloma
   ii. Idiopathic scleral staphyloma
4. Other congenital anomalies of the sclera are:
   i. Blue sclera
   ii. Scleral melanosis
   iii. Intrascleral nerve loop
   iv. Sclerocornea

   Anomalies 1, 2, and 3 do not effect vision.

Acquired

The acquired disorders are:

- Ectasia
- Inflammation
- Trauma
- Infection (rare)

SCLERAL ECTASIA

The ectasia of the sclera are types of staphylomas involving the sclera.

> A staphyloma is defined as—ectatic cicatrization of outer coat of the eyeball with the uvea incarcerated.

The basic mechanisms of scleral staphyloma are the thinning and disorganization of scleral fibers with raised intraocular pressure; it may happen even with normal intraocular tension, depending on the thinning of the sclera.

The staphylomas can involve:

- Cornea only
  - Partial
  - Complete
- Sclera only
- Combination

As per location, scleral staphylomas are ***(Fig. 16.4)****:*

- Intercalary
- Ciliary
- Equatorial
- Posterior
- Combination

As per duration:

- Congenital
- Acquired

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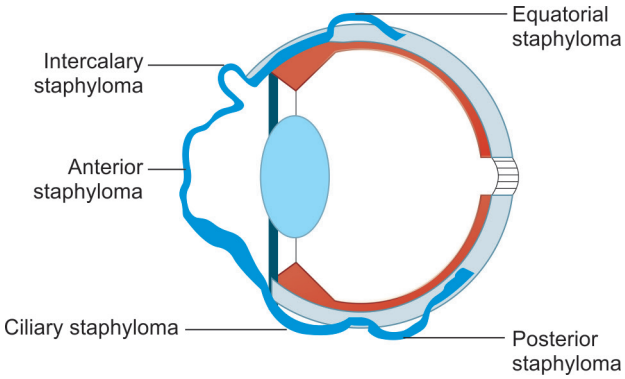


Fig. 16.4: Various types of corneoscleral staphylomas.

Flowchart 16.1: Types of staphylomas.

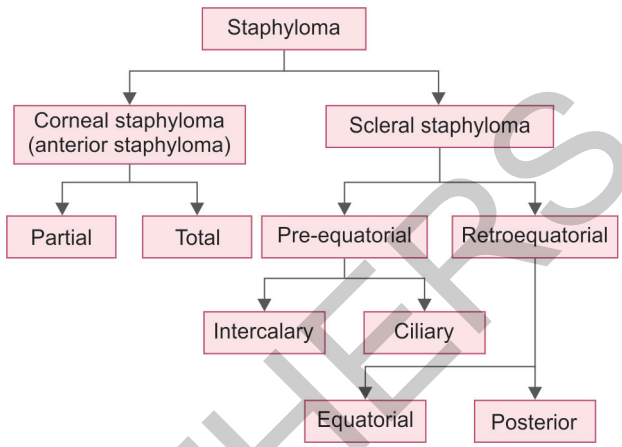


TABLE 16.1: Staphylomas and their predisposing factors.

S. No.	*Staphyloma*	*Outer coat involved*	*Anatomical defect*	*Uvea involved*
1.	Corneal	Center of the cornea	Thin central cornea	Iris
2.	Intercalary	Limbus	Entry of anterior ciliary vessels	Root of the iris
3.	Ciliary	Anterior sclera	Nonspecific	Ciliary body
4.	Equatorial	Equator of the sclera	The thinnest part of the sclera	Choroid
5.	Posterior	Posterior pole	Congenital weak	Choroid

Congenital Scleral Staphyloma

About 6% of eyes have some degree of ectasia; all of them are hidden either under the extraocular muscle or in a deeper situation behind the equator. They are symptomless, becoming evident at the time of either squint or retinal detachment surgery. Some authors also put posterior staphyloma in the category of congenital anomalies. Topographically, congenital staphylomas are equatorial or posterior.

Acquired Staphyloma of Sclera

Predisposing factors for producing scleral staphylomas are:

- Anatomical
  - Natural perforation passes through the following vessels and nerves:
    - Anterior ciliary arteries
    - Long and short ciliary arteries and nerves
    - Vertex vein
  - Lamina cribrosa
- Pathological
  - Buphthalmos—primary or secondary
  - Trauma
  - Severe scleritis
  - Necrotizing scleritis
  - Scleromalacia

  The last three are not seen in children.
- Infection—oculosporidiosis in endemic areas.

Flowchart 16.1 shows various types of staphylomas of the eye.

The relationship of staphylomas to the predisposing factors is shown in **Table 16.1**.

PATHOGENESIS OF VARIOUS TYPES OF SCLERAL STAPHYLOMA

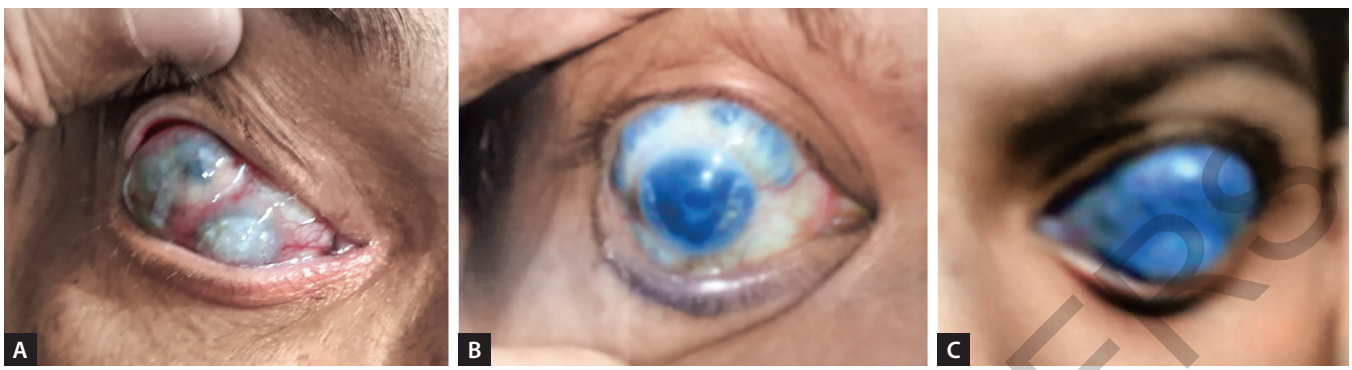
Intercalary Staphyloma

Intercalary staphyloma generally develops at the upper limbus, mostly following a limbal incision for classical lens extraction with an incarcerated root of the iris.

Ciliary Staphyloma

The prerequisite for ciliary staphyloma is thinning of the sclera due to stretching, as seen in buphthalmos or following scleritis, scleromalacia, and oculosporidiosis. In 5–6% of cases, no cause can be found. The effect of

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Figs. 16.5A to C: Extensive ciliary staphylomas and annular staphyloma since childhood.

intraocular pressure depends on the thickness of the sclera. A very thin sclera may bulge even in the presence of normal intraocular pressure; a relatively thick sclera may stand even with a high intraocular pressure (IOP) **(Figs. 16.5A to C)**.

The number of ciliary staphylomas varies from 1 to >1 in various places of various sizes; however, they may be numerous enough to encircle the limbus, causing an annular staphyloma. Generally, these eyes are painful without vision.

Equatorial Staphyloma

The most common cause of equatorial staphylomas is congenital idiopathic weakness. They amount to 6–7% of all staphylomas. Unlike intercalary and ciliary staphylomas, they are not visible and were discovered during squint and retinal detachment surgery.

Posterior Staphyloma

Posterior staphylomas, found exclusively in pathological myopia, may produce a slight bulging of the involved eye. On retinoscopy, a posterior staphyloma gives two readings, one at the rim of the staphyloma and the other at the bottom of the staphyloma. The difference between the two readings gives the depth of the staphyloma, i.e., measured by adding minus lenses to the initial reading. The staphylomas may be large enough to engulf the optic nerve. The posterior staphylomas are best visualized on indirect ophthalmoscopy. They are confirmed by ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), and optical coherence tomography (OCT). The best results are seen by wide-field OCT and 3D MRI. Like all staphylomas, posterior staphylomas, too, do not respond well to treatment. Posterior scleral support has been tried without much success.

Symptoms

- Small staphylomas under the extraocular muscles and very posteriorly placed may be asymptomatic and go undetected unless the extraocular muscle is detached for any reason.
- Anteriorly placed congenital staphylomas are visible as black, lobulated protrusions under the conjunctiva, which is generally attached to the staphyloma.
- The presence of irregular astigmatism is common with intercalary and ciliary staphylomas.
- High myopia is the rule in posterior staphyloma.
- Visual loss depends on the position and size of the staphyloma.
- Unmanageable glaucoma is a common feature.

INFECTION OF SCLERA IN CHILDREN

Avascularity and the absence of epithelium exposed to the exterior make the sclera immune to infections. However, a severe infection of the peripheral cornea may spill over to the sclera at the limbus. An infected penetrating wound or an infected foreign body may cause a scleral abscess.

INFLAMMATION OF SCLERA

Inflammations of the substance of the sclera, which are referred to as scleritis, are not seen in children. Older children in late teens may suffer from episcleritis, i.e., an inflammation of the superficial sclera. It is mostly an autoimmune disease seen in rheumatoid arthritis; in many cases, no cause can be pinpointed. Episcleritis is a self-limited condition that is often mistaken for phlycten. The main differentiating points are that episcleritis is seen in late teens away from the limbus, is more diffuse than phlycten, may be tender, and does not involve the cornea. The common factor among the two is their positive

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response to local steroids, both of which are known to recur.

Other congenital anomalies of the sclera are:

Blue Sclera

A fully developed sclera is marble white with streaks of blood vessels traversing the surface and occasionally associated with melanosis without any bulge. In contrast to this, the thin sclera of newborns is more translucent, with a blue tinge imparted by the underlying choroid. Within 1 year, the sclera of an infant gets the white color of the adult sclera. Sometimes, the sclera remains bluish throughout life without clinical significance.

The systemic conditions associated with blue sclera are:

- Osteogenesis imperfect (blue sclera-brittle bone syndrome)
- Ehlers–Danlos syndrome (adults)
- The two local conditions with a black tinge are buphthalmos and microphthalmos.

Scleral Melanosis

Scleral melanosis is not uncommon in children; it may be present at birth or may make its presence felt in the first year and progress until the end of the first decade to become stationary. It is basically melanocytic hyperpigmentation. Generally bilateral, it is asymptomatic and does not involve vision. It may be associated with melanosis of the conjunctiva. The conjunctiva can be moved freely over pigmentation. A differential diagnosis consists of conjunctival nevi and acquired melanosis. It does not require any treatment **(Fig. 16.6)**.



Fig. 16.6: Scleral melanosis along with melanosis of the skin.

Intrascleral Nerve Loop

The nerves and vessels destined for the uvea traverse through the substance of the sclera without serving it. In rare instances, a small bunch of long ciliary nerves change their intrascleral course to appear on the surface of the sclera under the transparent conjunctiva as small nodules, generally on the nasal side. The cause of the condition is not known. It does not require treatment.

Sclerocornea

The normal sclera stops at the limbus, leaving a transparent cornea. On rare instances, the sclera engulfs the cornea from periphery, converting the transparent cornea as opaque as the sclera. The corneoscleral step at the limbus is obliterated, and the cornea is generally flat.

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Printed in India

Available at all medical bookstores or buy online at www.ejaypee.com

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Medical Publishers (P) Ltd.
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ISBN 978-93-6616-156-3