

THE END-STAGE EYE

Phthisis Bulbi

A morphologic approach to ocular atrophy and disorganization

Dr Abu Suraih Sakhri

MBBS, MD (Pathology)

www.abusuraisakhri.com

Learning objectives

- Define phthisis bulbi and distinguish it from congenital microphthalmia
- Explain the major initiating conditions and the structural changes they produce
- Recognize the defining findings of the end-stage globe and apply them to cases

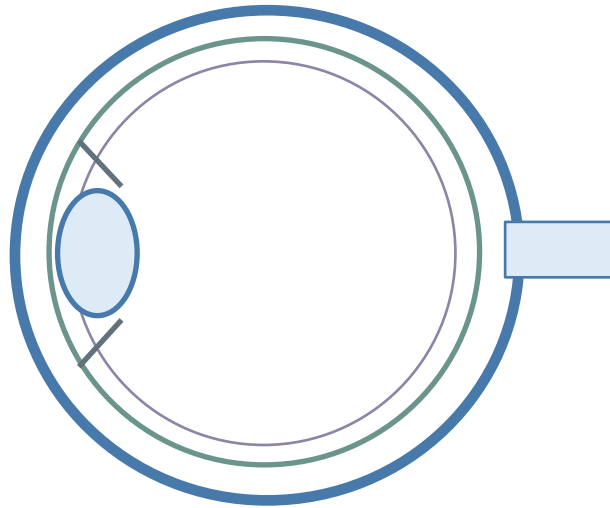
A morphologic diagnosis—not one etiologic disease

- Phthisis bulbi describes a small, atrophic eye with disorganized intraocular contents
- Several different antecedent disorders converge on a similar end-stage phenotype
- The morphologic diagnosis does not, by itself, establish the initiating cause

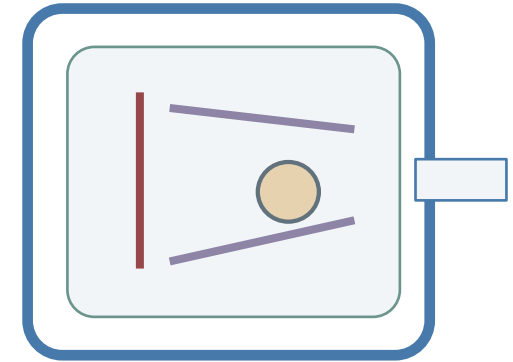
The diagnosis integrates globe size with architecture.

The defining combination

- An abnormally small globe is necessary but not sufficient to infer phthisis
- Loss of normal internal ocular organization is the crucial associated feature
- Both structural features should be assessed together



Organized anatomy

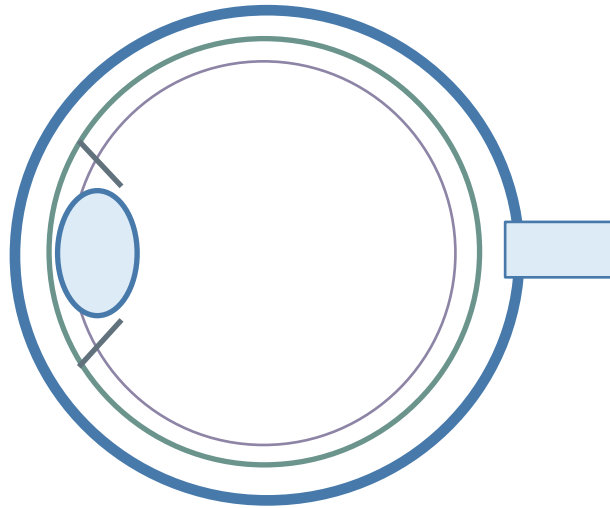


Atrophy + disorganization

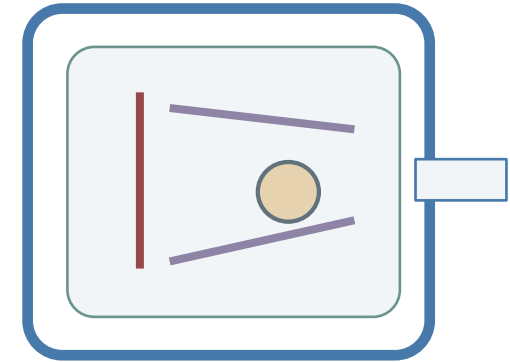
Original, editable schematics

The alternative: Congenital microphthalmia

- Hypoplastic or microphthalmic eyes are small from development
- That these are generally not internally disorganized
- A small, relatively organized eye is not equivalent to a phthisical eye



Organized anatomy

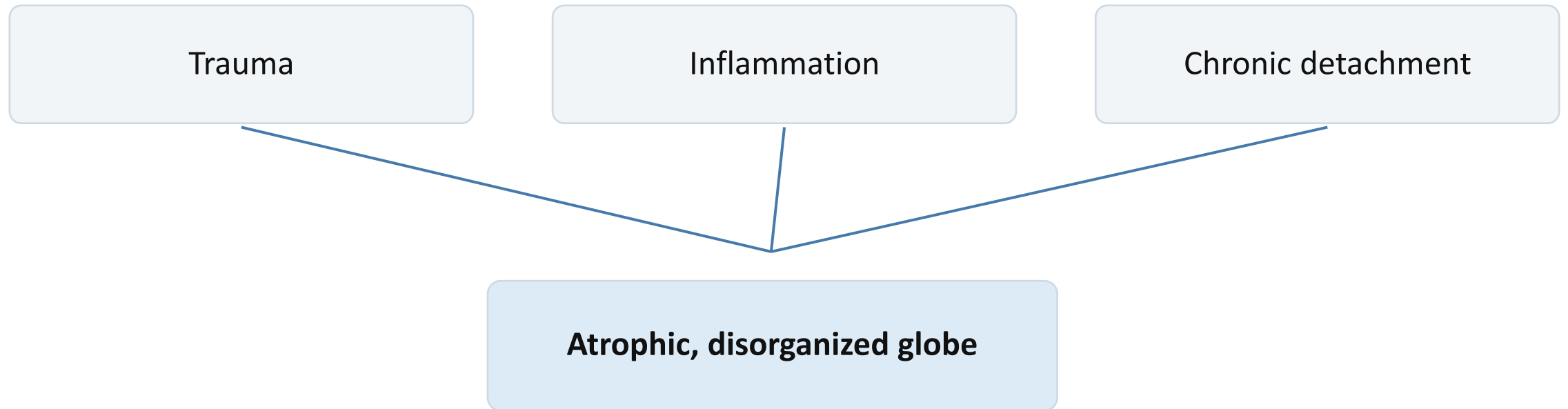


Atrophy + disorganization

Original, editable schematics

Convergent causes of phthisis

- Trauma, intraocular inflammation and chronic retinal detachment are explicitly named
- Other severe ocular insults can culminate in the same atrophic phenotype
- End-stage morphology can obscure the original cause



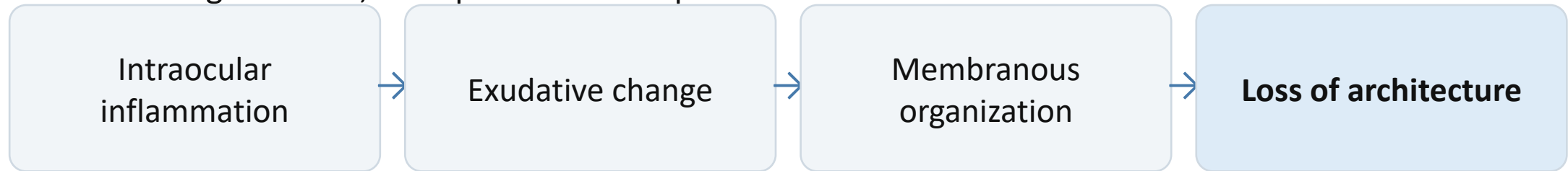
Alternative causes—not a chronological sequence

Trauma as an initiating insult

- Trauma is a named antecedent
- Evaluate the end-stage eye for global disorganization, not an isolated old injury
- A trauma history supports etiology but is not itself the morphologic criterion

Persistent intraocular inflammation

- Intraocular inflammation is another explicitly recognized antecedent
- Exudative fluid and an organizing cyclitic membrane may coexist in advanced cases
- In end-stage disease, multiple ocular compartments can be affected

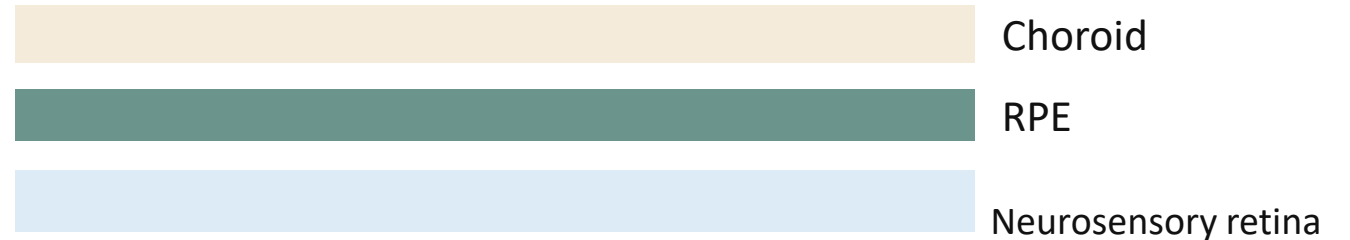


Schematic sequence: Teaching synthesis

Chronic retinal detachment

- Chronic retinal detachment is both a potential antecedent and a typical finding
- The detached neurosensory retina no longer contacts its normal RPE support
- The whole globe may become atrophic and disorganized in advanced disease

Apposed interface



Detached interface



Original layered schematic

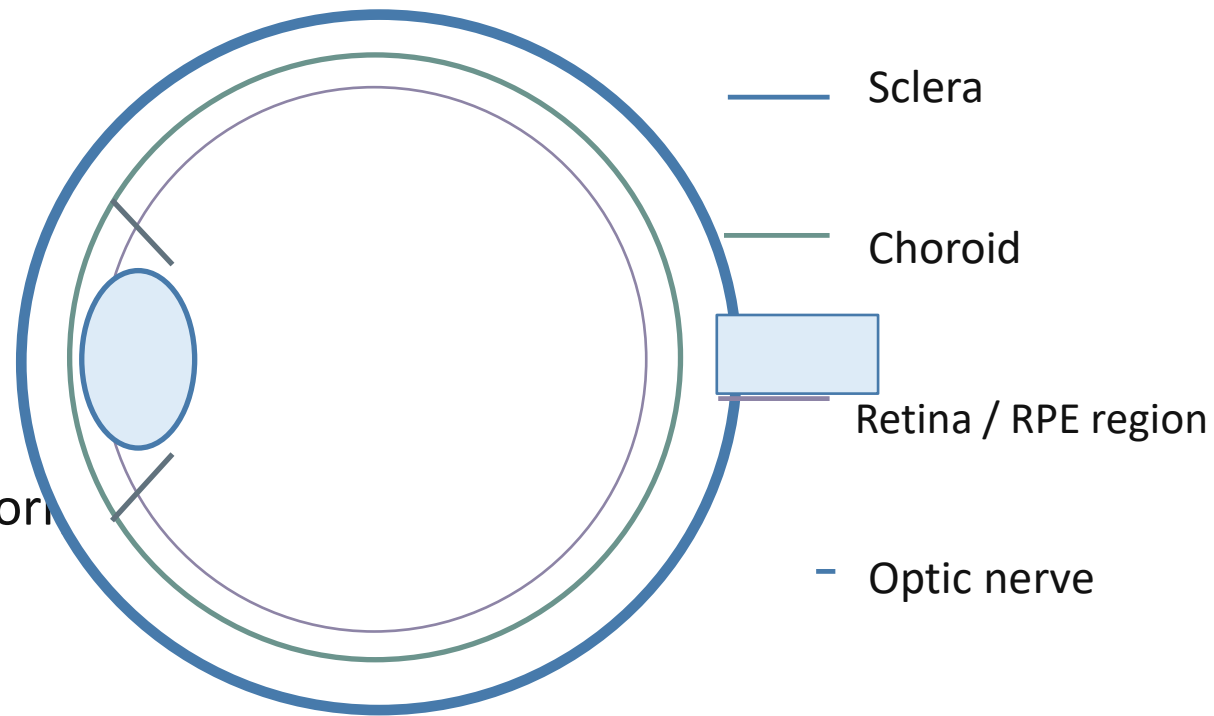
One eye; Several coexisting lesions

- Phthisical eyes may contain effusion, a cyclitic membrane and retinal detachment
- Optic nerve atrophy, intraocular bone and scleral thickening may also be present
- These findings are a constellation; A rigid number-of-features cutoff is not supplied

Anatomic compartment	Associated finding
Ciliary body / choroid	Ciliochoroidal effusion
Across ciliary regions	Cyclitic membrane
Neurosensory retina / RPE	Chronic retinal detachment
Optic nerve	Atrophy
Inside the globe	Possible intraocular bone
Posterior sclera	Thickening

Anatomic orientation to the globe

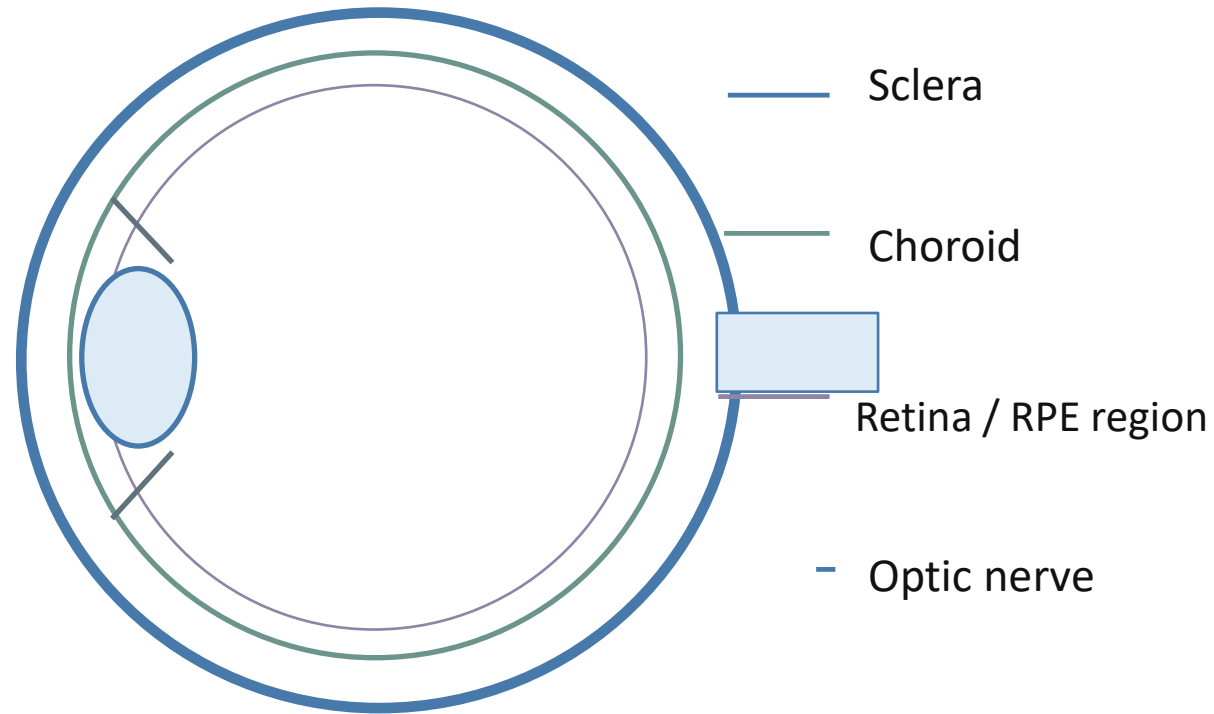
- Outer coat: Sclera; Vascular middle layer: Uvea; Internal sensory layer: Retina
- The ciliary body and choroid provide landmarks for ciliochoroidal effusion
- The optic nerve exits posteriorly. The retina is internal to the choroid



Schematic: Organized globe

What internal organization normally preserves

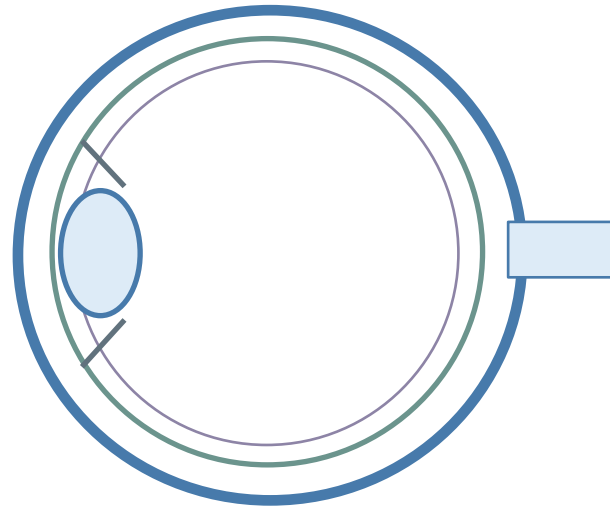
- The neurosensory retina lies adjacent to the RPE
- The ciliary body forms part of the anterior uveal complex
- The sclera defines the external coat and overall globe contour



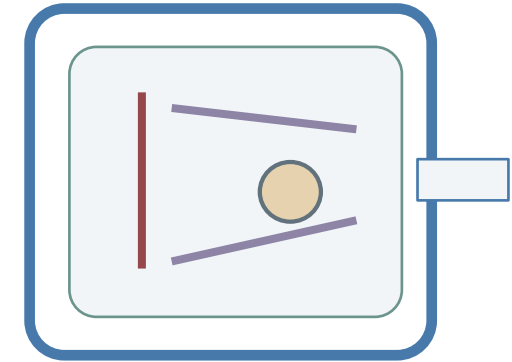
Schematic: Organized globe

The end-stage architecture

- Shrinking of the globe accompanies major displacement or disruption of its contents
- Normal tissue relationships may be difficult to trace
- The endpoint is qualitatively more than a single compartment lesion



Organized anatomy



Atrophy + disorganization

Original, editable schematics

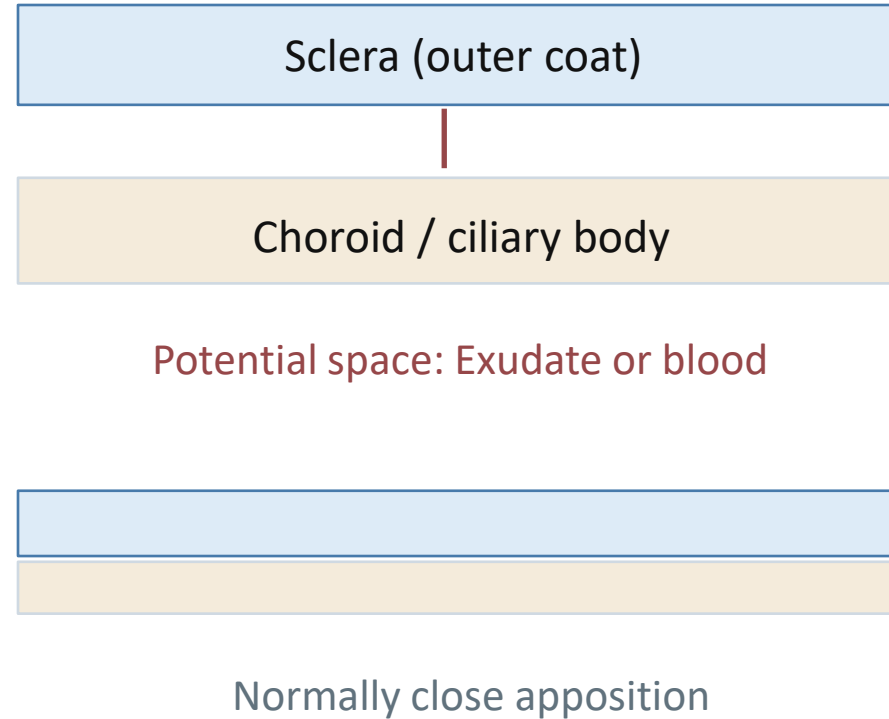
A map of the characteristic changes

- Ciliary body/choroid:
Ciliochoroidal effusion
- Vitreous cavity: Cyclitic
membrane; Retina: Chronic
detachment
- Posterior structures: Optic nerve
atrophy and thickened sclera;
Intraocular bone may occur

Anatomic compartment	Associated finding
Ciliary body / choroid	Ciliochoroidal effusion
Across ciliary regions	Cyclitic membrane
Neurosensory retina / RPE	Chronic retinal detachment
Optic nerve	Atrophy
Inside the globe	Possible intraocular bone
Posterior sclera	Thickening

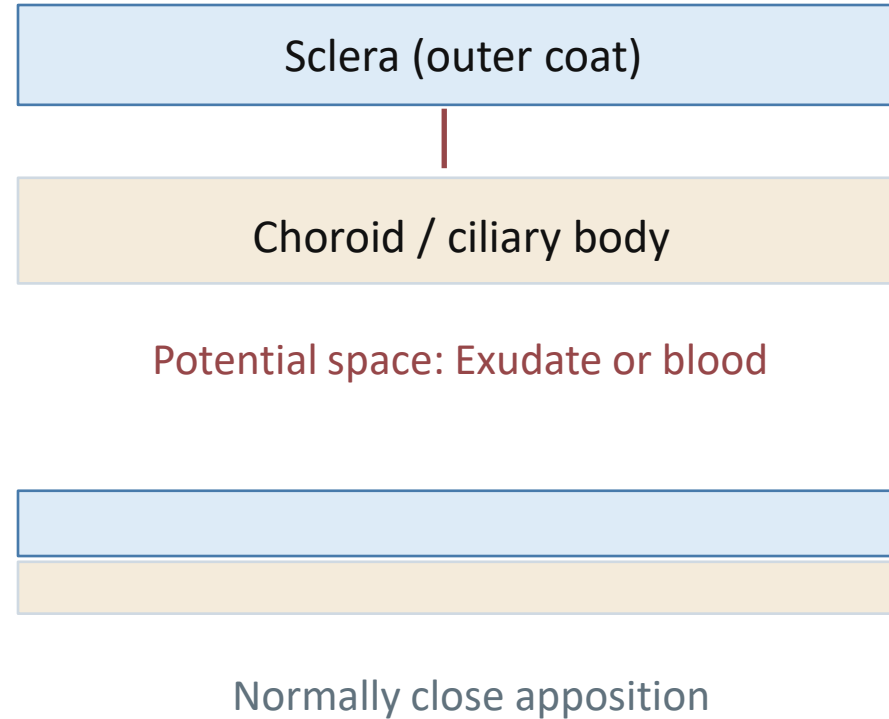
Ciliochoroidal effusion: Definition

- Exudate or blood accumulates between ciliary body and sclera
- Similar fluid may occur between choroid and sclera
- The descriptor concerns a specific tissue plane, not generalized vitreous hemorrhage



Effusion is a potential-space lesion

- The outer uveal tissues and sclera are normally closely apposed
- Fluid separates these tissue interfaces in ciliochoroidal effusion
- Identify the correct anatomic interval before using this term



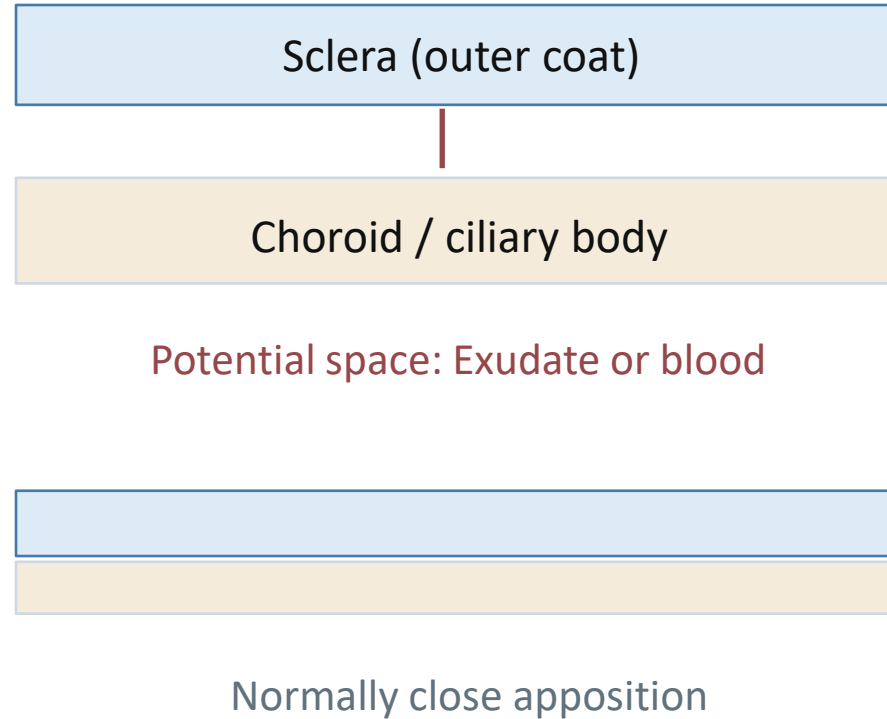
Exudate versus blood in the effusion

Feature	Exudate	Blood
Material	Proteinaceous / inflammatory fluid	Hemorrhagic material
Core location	Ciliary body–sclera or choroid–sclera	Same potential spaces
Scope of inference	Presence does not uniquely identify cause	Presence does not uniquely identify cause

Robbins explicitly allows exudate or blood in the ciliochoroidal plane

Where ciliochoroidal effusion is found

- Anterior component: Between ciliary body and sclera
- Posterior component: Between choroid and sclera
- The shared diagnostic feature is separation along the uveoscleral interface

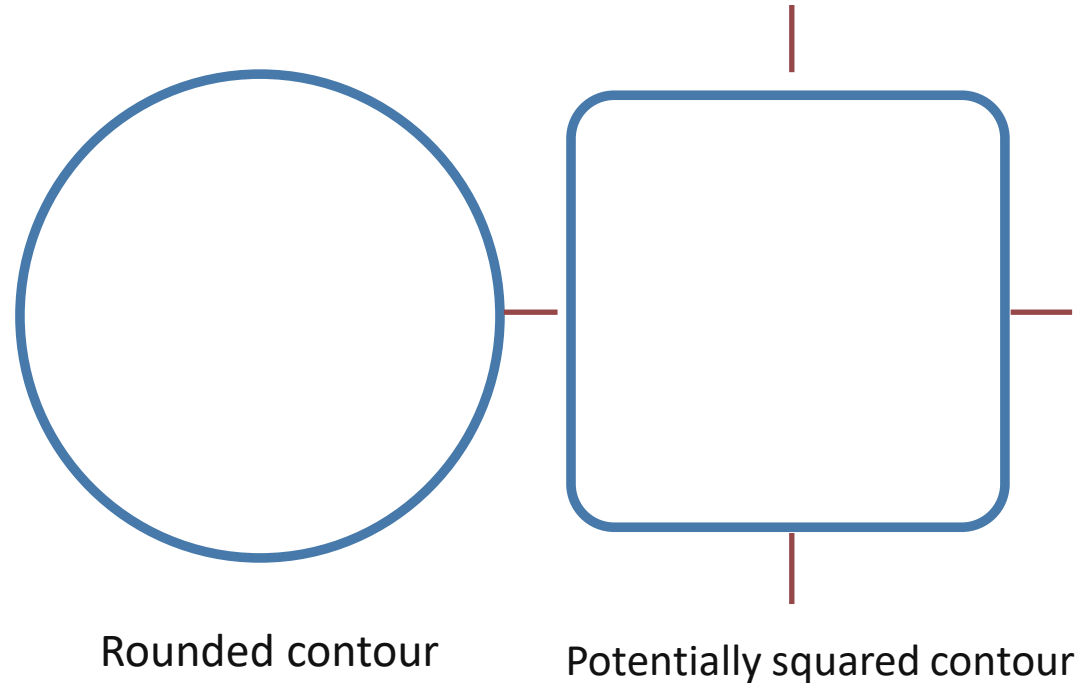


Hypotony in the phthisical eye

- Ciliochoroidal effusion is typically associated with low intraocular pressure
- The physiologic state is ocular hypotony
- An association; It does not provide a numerical pressure threshold

Why the contour can appear square

- Normal extraocular muscle forces continue to act on the globe
- A hypotonic eye resists those forces differently than a normally pressurized globe
- The net result may be a squared rather than rounded external contour



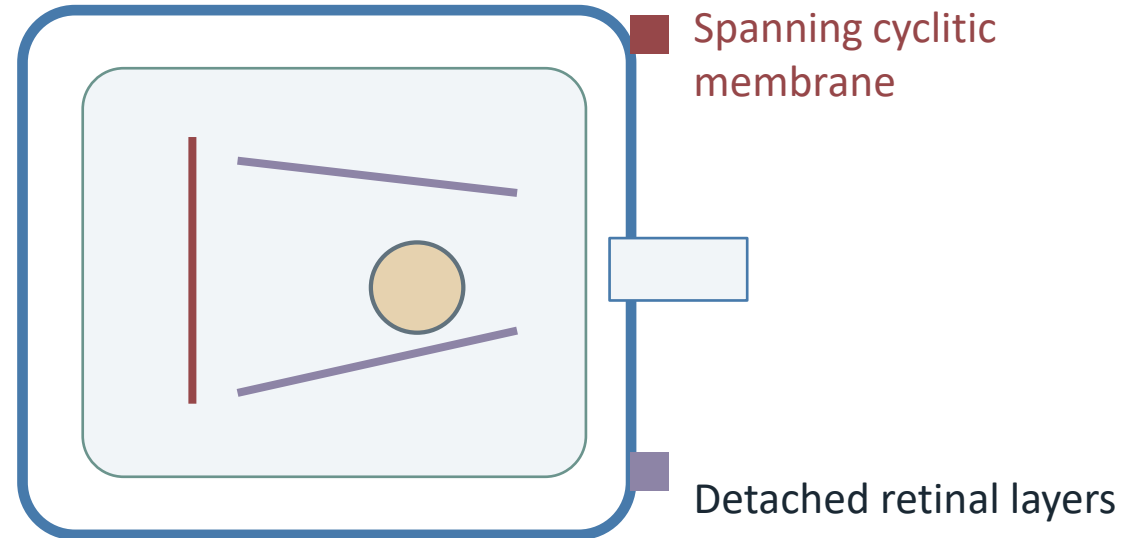
Low pressure is not the entire diagnosis

Dimension	Question to ask
Size	Is the globe atrophic/small?
Architecture	Are internal ocular structures disorganized?
Physiology	Is low intraocular pressure present?
Context	Is there evidence of a severe antecedent insult?

Hypotony is a physiologic correlate, not a synonym for phthisis

The ciliary body in an end-stage globe

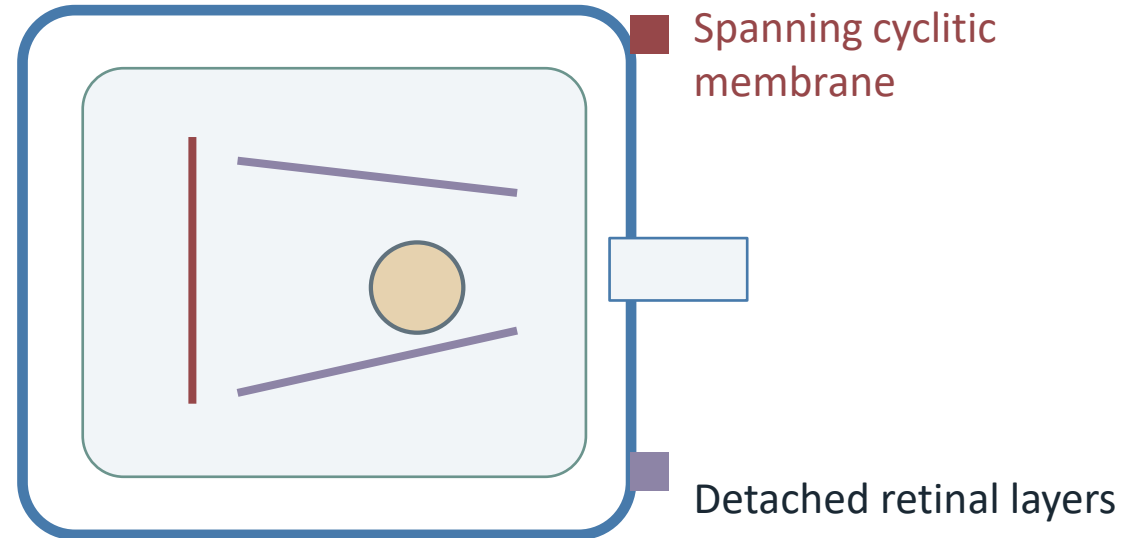
- The ciliary body is an anterior uveal structure
- Effusion may separate it from the sclera
- A cyclitic membrane can extend across the globe between opposite ciliary regions



Diagrammatic ciliary-body attachments

Cyclitic membrane: Defining geometry

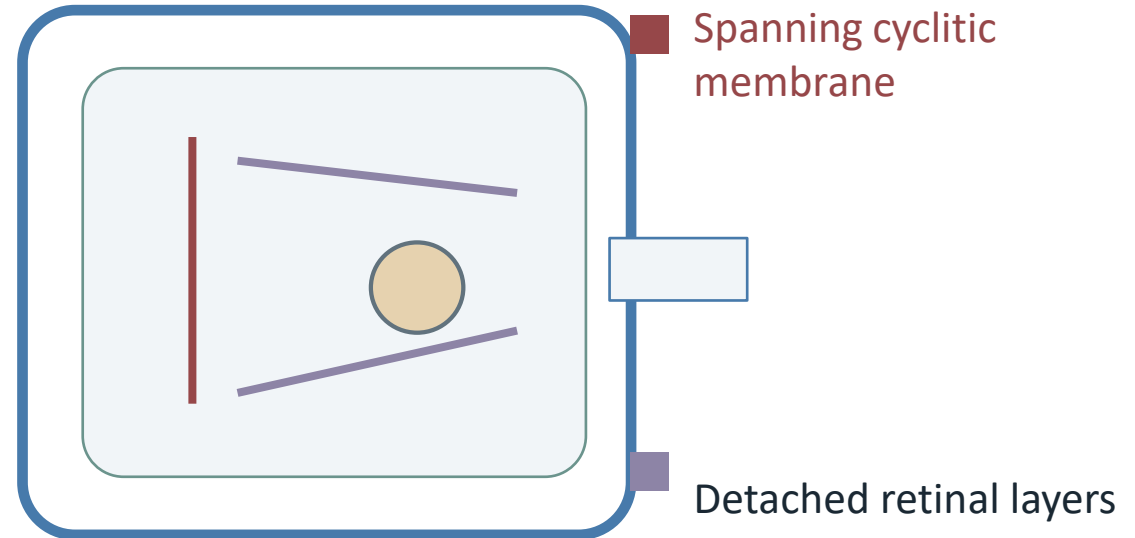
- A membrane extends from one aspect of the ciliary body across the eye
- Its opposite attachment reaches another aspect of the ciliary body
- The spanning configuration distinguishes it from an isolated retinal fold



Diagrammatic ciliary-body attachments

Understanding cyclitic membrane location

- The membrane traverses the internal ocular cavity
- It is defined by its relation to the ciliary body—not the scleral wall
- Diagrammatic depictions should show a spanning membrane rather than an annular coat



Diagrammatic ciliary-body attachments

Cyclitic membrane versus detached retina

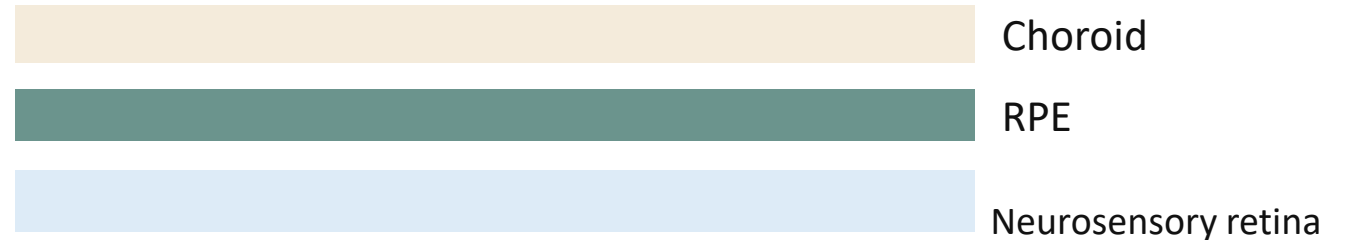
Feature	Cyclitic membrane	Retinal detachment
Structure	Spanning intraocular membrane	Neurosensory retina
Anatomic anchor	Ciliary body to ciliary body	Interface with RPE
Appearance in phthisis	Typical associated lesion	Typical associated lesion

A cyclitic membrane spans between ciliary-body regions

The retina–RPE interface

- The retinal pigment epithelium lies directly external to the neurosensory retina
- Detachment means separation of these two layers
- Understanding that interface prevents confusion with a ciliochoroidal effusion

Apposed interface



Detached interface



Original layered schematic

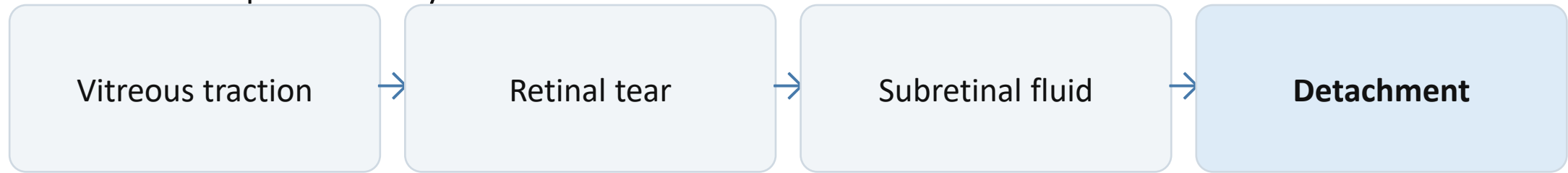
Retinal detachment: Two broad mechanisms

	Rhegmatogenous	Nonrhegmatogenous
Retinal break	Present	Absent
Fluid source/path	Vitreous passes through retinal tear	Exudative or other non-break mechanisms
End-stage link	May persist chronically	May persist chronically

Rhegmatogenous detachment involves a full-thickness retinal break

Rhegmatogenous detachment: Structural sequence

- Vitreoretinal traction can create a full-thickness tear
- Liquefied vitreous accesses the subretinal potential space
- Persistent separation may become chronic

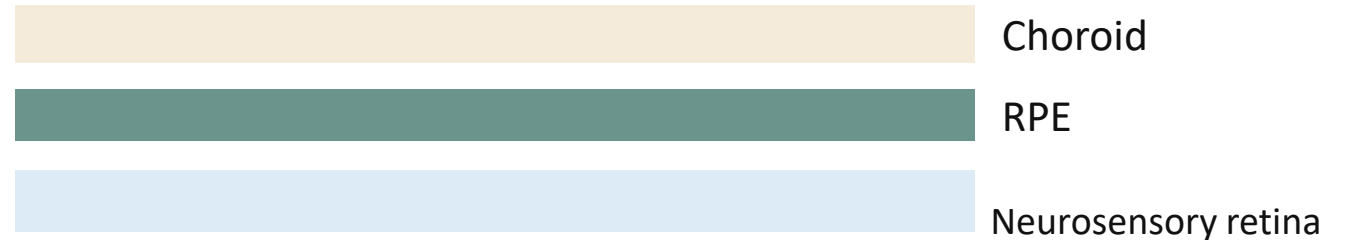


Schematic sequence: Teaching synthesis

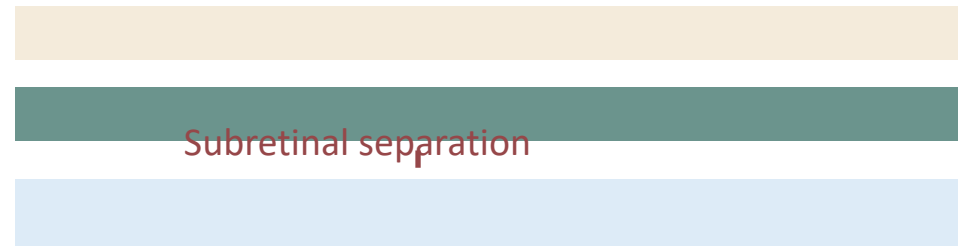
Nonrhegmatogenous detachment

- Retinal separation occurs without a full-thickness retinal defect
- The chapter includes exudative mechanisms and lesions that impair RPE barrier function
- The anatomic endpoint remains separation of neurosensory retina from RPE

Apposed interface



Detached interface

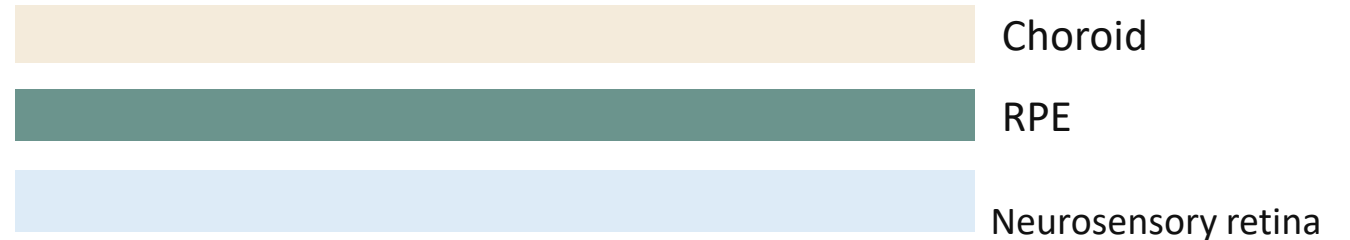


Original layered schematic

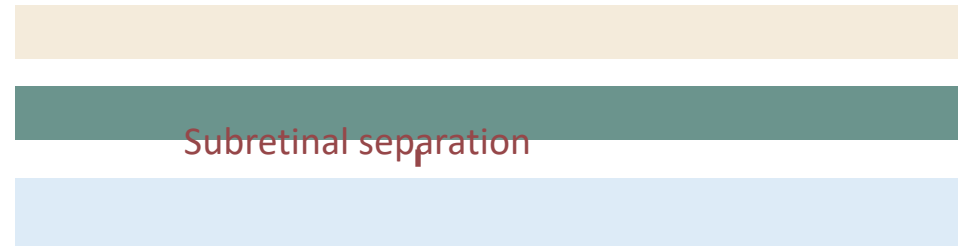
Why chronicity matters in retinal detachment

- The chapter links absent photoreceptor outer segments with chronic detachment
- Long-standing separation represents more than a transient change in retinal position
- Phthisis can contain a chronically detached and structurally abnormal retina

Apposed interface



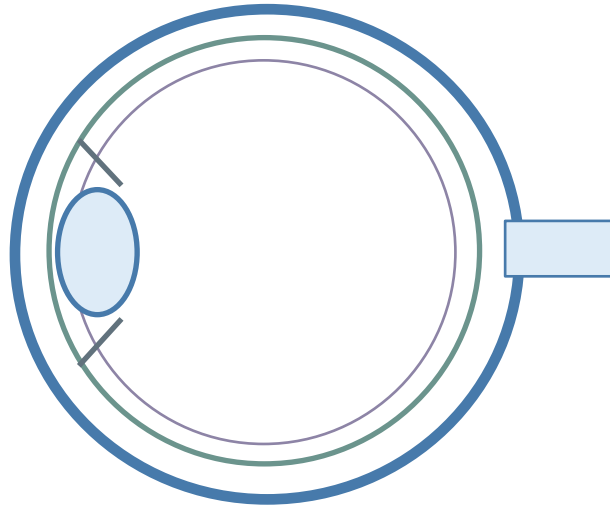
Detached interface



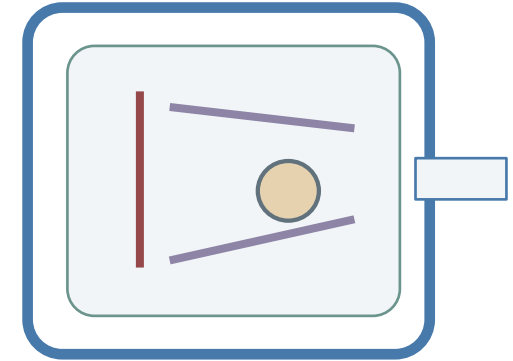
Original layered schematic

Retinal detachment is not sufficient alone

- A detached retina can exist in a globe that retains normal overall size
- Phthisis adds gross atrophy and severe loss of internal organization
- Do not assign the end-stage diagnosis from detachment in isolation



Organized anatomy

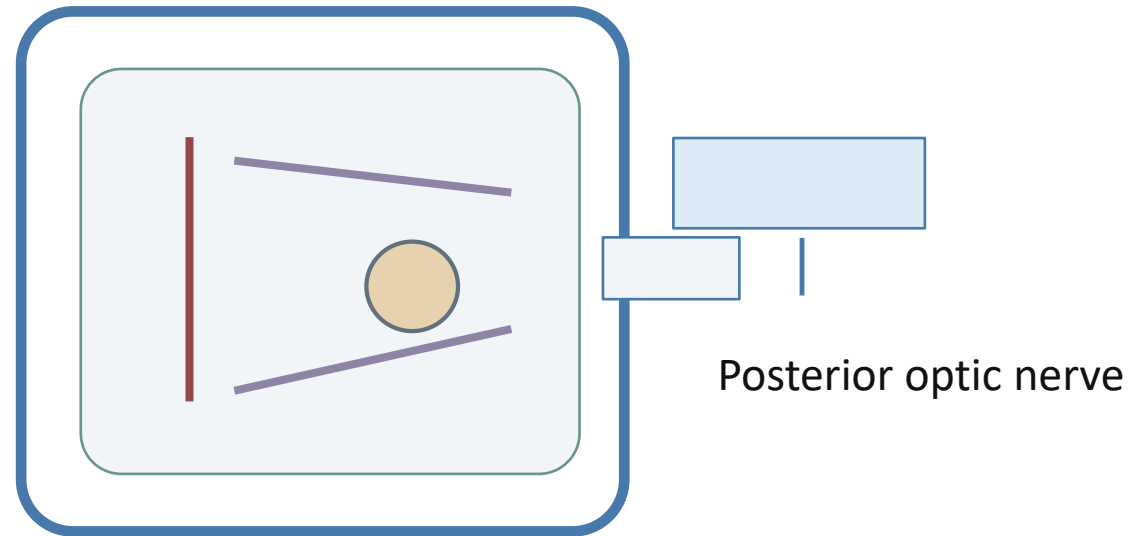


Atrophy + disorganization

Original, editable schematics

Optic nerve atrophy in phthisis

- Optic nerve atrophy is among the typical associated findings
- The nerve is evaluated as a posterior consequence of ocular injury
- Atrophy does not independently determine which initiating disease caused phthisis



Atrophy is an associated lesion

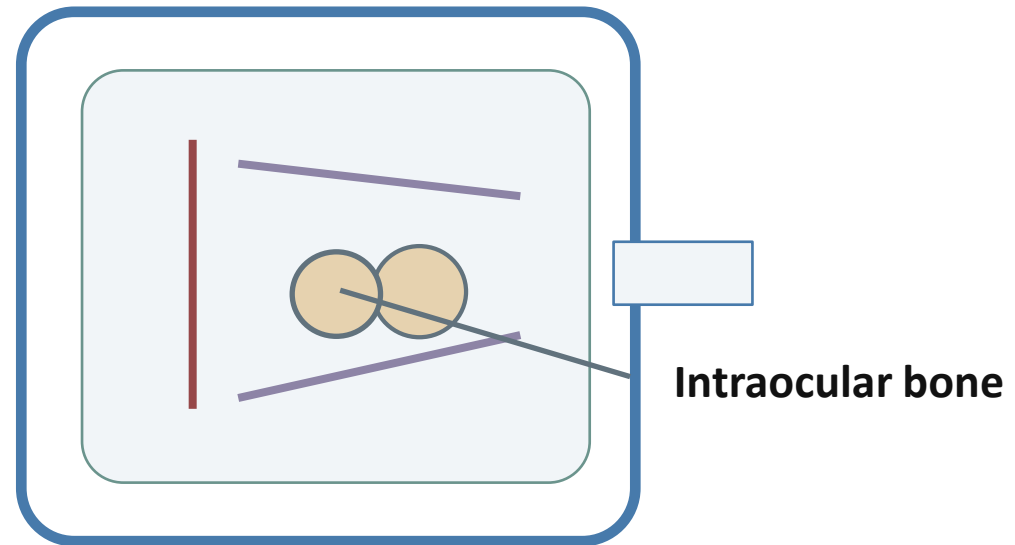
Optic nerve damage: A parent-context comparison

	Advanced glaucoma	Phthisis bulbi
Primary morphologic emphasis	Cupped, atrophic optic nerve	Small, disorganized globe
Other tissues	Globe may remain organized	Multiple ocular compartments affected
Relationship	Distinct diagnostic process	Optic nerve atrophy may coexist

Robbins also describes optic nerve atrophy with advanced glaucoma

Intraocular bone: A striking late change

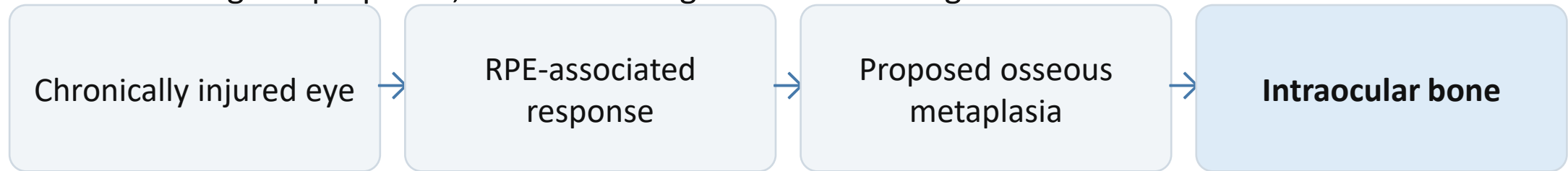
- True intraocular bone among characteristic features
- The proposed origin is osseous metaplasia of the retinal pigment epithelium
- “Thought to originate” is a qualified mechanistic statement, not proof in every case



Proposed RPE osseous metaplasia

Osseous metaplasia: What the term means

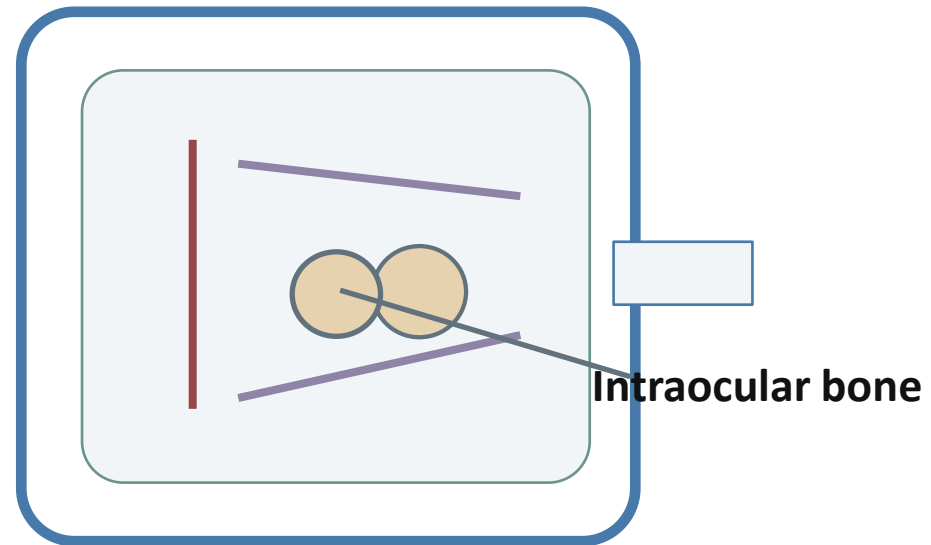
- Metaplasia denotes replacement by a differentiated tissue type inappropriate for the site
- Here the described product is bone arising within the injured eye
- The RPE origin is proposed; Avoid asserting a universal lineage



Schematic sequence: Teaching synthesis

Bone location is an internal finding

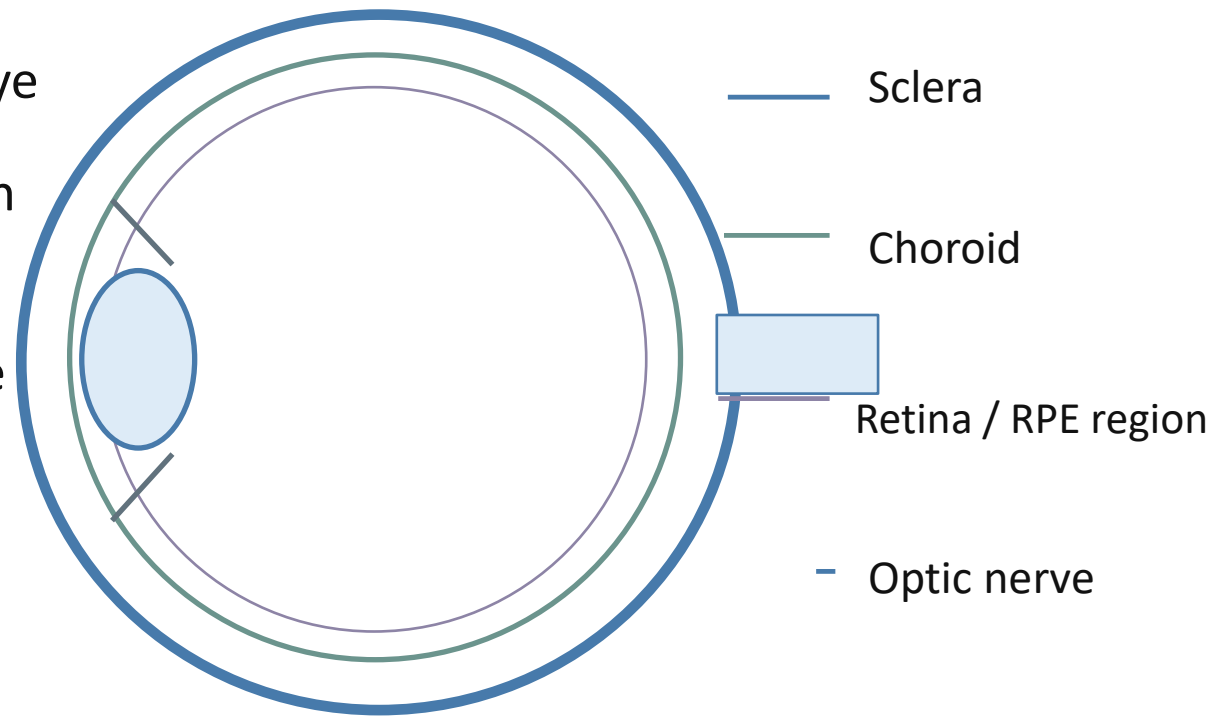
- An intraocular bone focus lies within the contents of the globe
- It should not be confused with the surrounding osseous orbit
- A simple drawing can establish location; Diagnostic tissue requires genuine histology



Proposed RPE osseous metaplasia

Scleral thickening

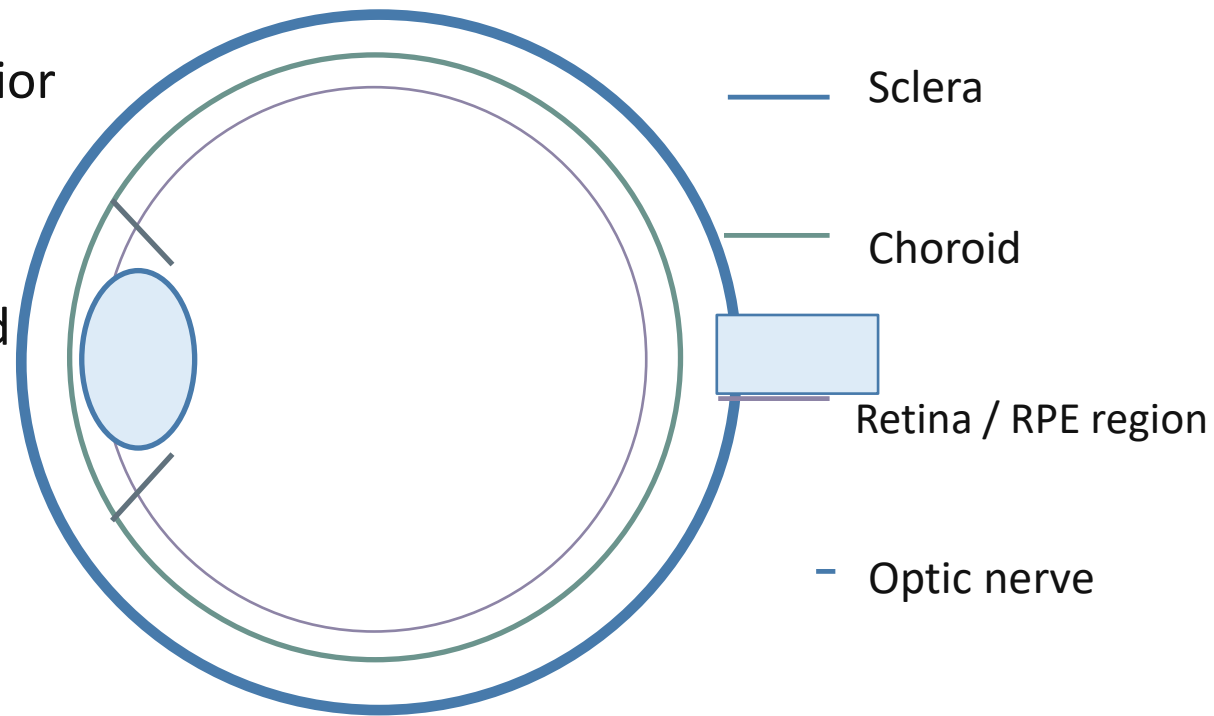
- The sclera may become thickened in the phthisical eye
- Posterior scleral thickening in particular
- This change occurs alongside internal disorganization and globe atrophy



Schematic: Organized globe

Posterior sclera: Site-specific observation

- When assessing the wall, examine anterior and posterior portions separately
- Posterior-predominant thickening is a text-described tendency
- Do not assume that every phthisical specimen shows uniform wall thickness



Schematic: Organized globe

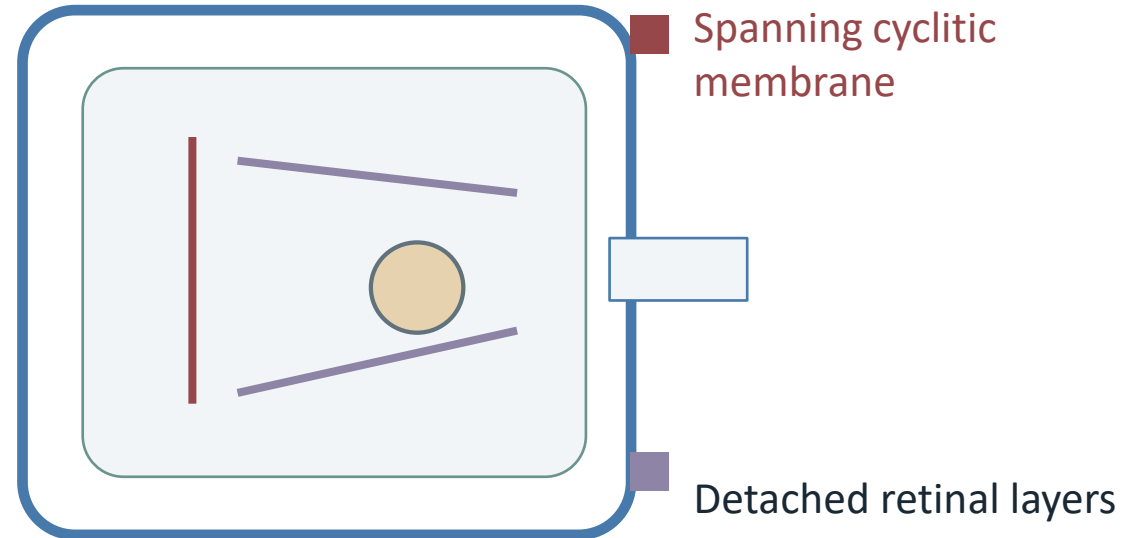
Integrating the posterior findings

- Chronic retinal detachment changes the retina–RPE interface
- Optic nerve atrophy concerns the nerve leaving the globe
- Posterior scleral thickening concerns the outer ocular coat

Anatomic compartment	Associated finding
Ciliary body / choroid	Ciliochoroidal effusion
Across ciliary regions	Cyclitic membrane
Neurosensory retina / RPE	Chronic retinal detachment
Optic nerve	Atrophy
Inside the globe	Possible intraocular bone
Posterior sclera	Thickening

Integrating the anterior findings

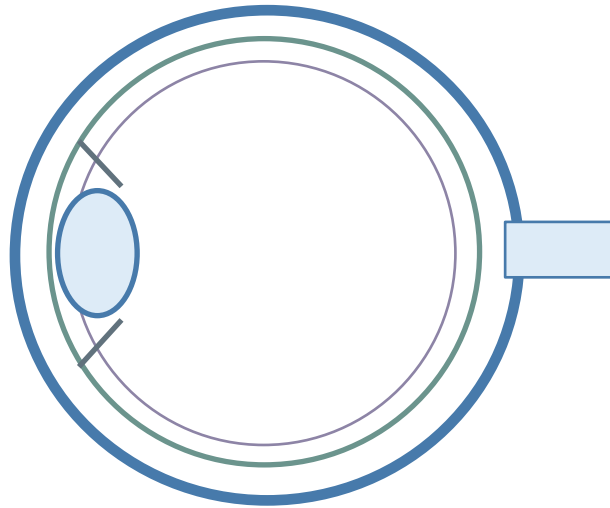
- Ciliary-body separation contributes to ciliochoroidal effusion
- Cyclitic membrane relates to the ciliary-body regions
- Together these demonstrate architectural disruption beyond the posterior retina



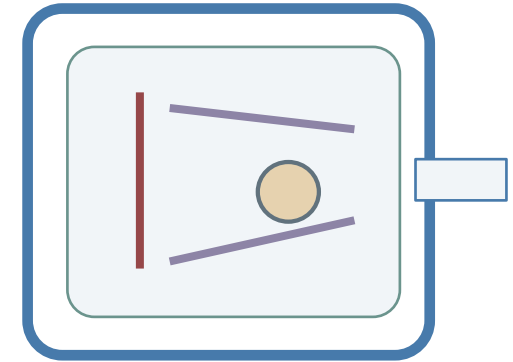
Diagrammatic ciliary-body attachments

Normal versus phthisical globe: One schematic

- Normal: Preserved ocular boundaries and internal relationships
- Phthisical: Reduced size, contour deformation and altered internal planes
- The diagram is an editable schematic, not a histologic specimen



Organized anatomy



Atrophy + disorganization

Original, editable schematics

Gross impression versus internal pathology

Inspection level	Core features
External	Small, atrophic; possibly squared contour
Internal	Disorganized planes; membrane; detachment
Posterior	Optic nerve atrophy; posterior scleral thickening
Distinctive additional finding	Intraocular bone

External appearance may suggest globe shrinkage or contour deformation

A tissue-by-tissue recognition checklist

- Sclera: Thickness, especially posteriorly
- Ciliary body/choroid: Effusion planes; Vitreous cavity: Spanning membrane
- Retina–RPE: Chronic separation; Nerve: Atrophy; Interior: Bone

Anatomic compartment

Associated finding

Ciliary body / choroid

Ciliochoroidal effusion

Across ciliary regions

Cyclitic membrane

Neurosensory retina / RPE

Chronic retinal detachment

Optic nerve

Atrophy

Inside the globe

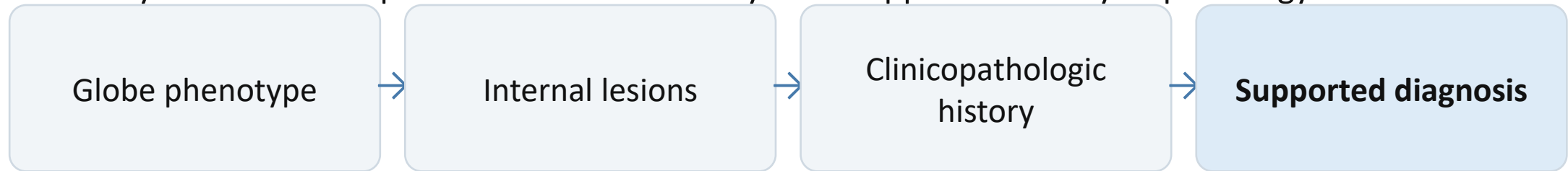
Possible intraocular bone

Posterior sclera

Thickening

How to use the constellation diagnostically

- First establish the small/disorganized-globe phenotype
- Then document which characteristic changes are actually present
- Finally infer the most plausible antecedent only from supported history or pathology



Schematic sequence: Teaching synthesis

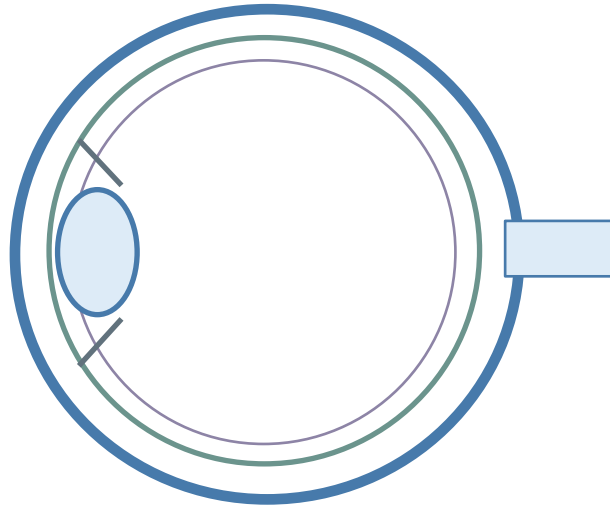
A structured morphology description

Compartment	Target observation
Globe	Size, contour, internal architecture
Uvea	Ciliochoroidal separation / contents
Retina / cavity	Detachment / cyclitic membrane
Posterior wall	Sclera; optic nerve
Additional	Possible intraocular bone

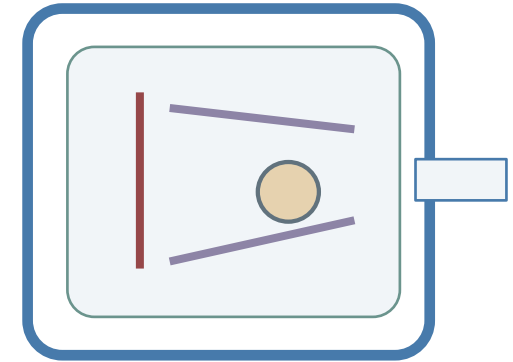
Describe overall globe size and degree of architectural loss

Do not equate small eyes with phthisis

- Congenital developmental small eyes can maintain internal organization
- Acquired severe ocular injury may produce both shrinkage and disorder
- Internal architecture provides the key discriminating feature



Organized anatomy



Atrophy + disorganization

Original, editable schematics

Do not infer mechanism from a single sign

Isolated finding	Inference to avoid
Effusion	“It must have been traumatic”
Optic nerve atrophy	“It must have been glaucoma”
Small globe	“It must be phthisis”
Bone	“RPE origin is proven in this case”

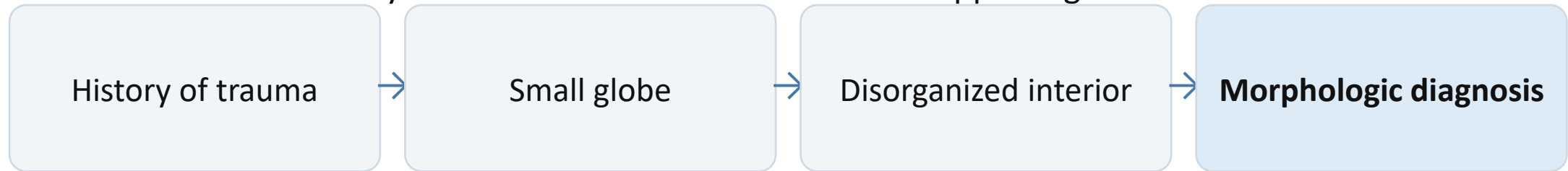
Effusion does not uniquely identify inflammation, hemorrhage or trauma

Case 1: A small globe after trauma

- History: Remote severe ocular trauma
- Current pathology: Markedly atrophic globe with retinal detachment and a cyclitic membrane
- Question: Which end-stage diagnosis best integrates these findings?

Case 1: Clinicopathologic reasoning

- Trauma provides a recognized initiating context
- Small size plus disorganized architecture establishes the phenotype
- Detached retina and cyclitic membrane add characteristic supporting observations



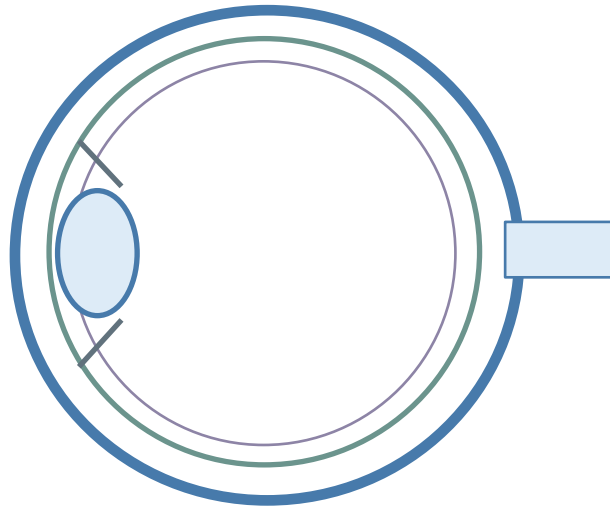
Schematic sequence: Teaching synthesis

Case 2: A congenitally small eye

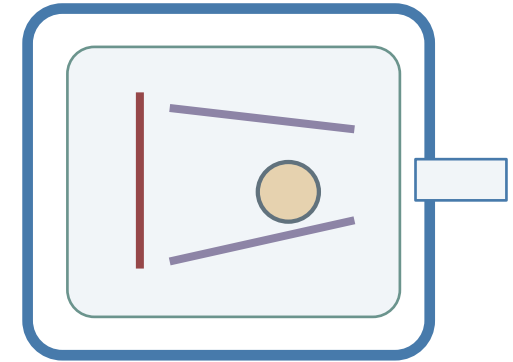
- History: Eye known to be small since birth
- Internal architecture is largely preserved on assessment
- Question: Does small size alone justify the diagnosis phthisis bulbi?

Case 2: The decisive distinction

- Developmental smallness and acquired end-stage atrophy are not interchangeable
- A preserved internal arrangement weighs against phthisis
- A clear history and anatomic assessment prevent false equivalence



Organized anatomy



Atrophy + disorganization

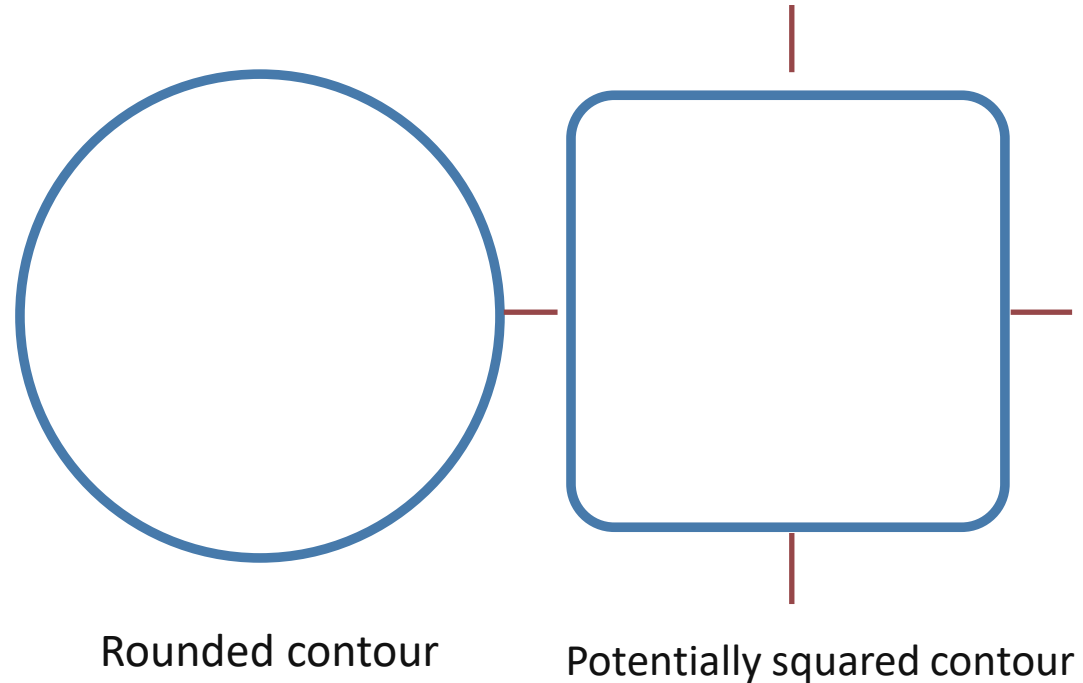
Original, editable schematics

Case 3: Low pressure with effusion

- A small eye shows ciliochoroidal effusion and reduced intraocular pressure
- It also contains a detached retina and posterior scleral thickening
- Question: How do physiologic and structural evidence complement each other?

Case 3: Integrating pressure and morphology

- Hypotony is typically associated with ciliochoroidal effusion
- Reduced wall tension permits altered action of normal extraocular muscle pull
- Definitive morphologic description still rests on the disorganized, atrophic globe

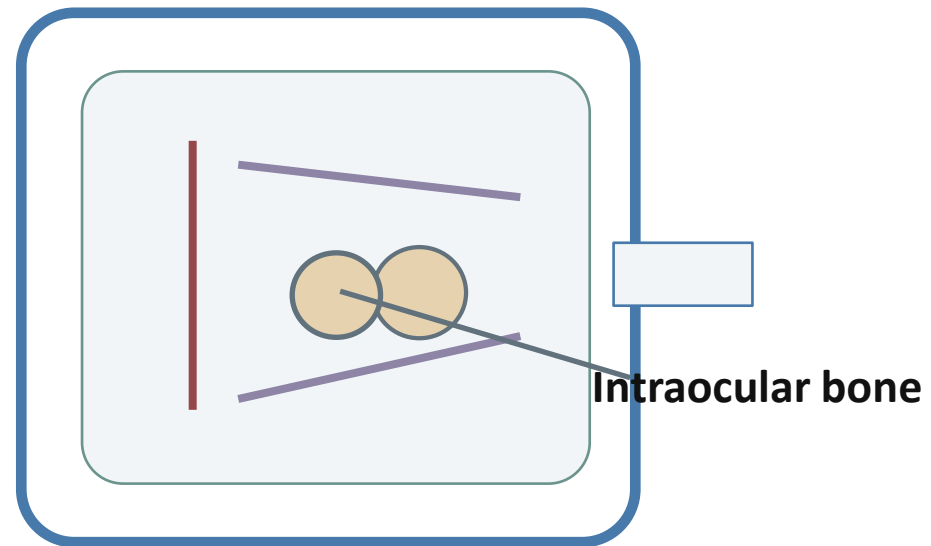


Case 4: Unexpected intraocular bone

- A chronically damaged globe contains a focus of intraocular bone
- The globe is small, with internal disorganization
- Question: What tissue of origin does Robbins propose?

Case 4: Appropriate level of certainty

- Intraocular bone is a described associated finding
- The text proposes the RPE as its metaplastic source
- Present the origin as a hypothesis, not a validated lineage in every specimen



Proposed RPE osseous metaplasia

Final synthesis: Multiple compartments fail

- Etiologic triggers differ; The endpoint is an atrophic, disorganized eye
- Ciliochoroidal effusion, cyclitic membrane and chronic detachment localize internal damage
- Optic nerve atrophy, bone and posterior scleral thickening complete the morphologic pattern

Anatomic compartment	Associated finding
Ciliary body / choroid	Ciliochoroidal effusion
Across ciliary regions	Cyclitic membrane
Neurosensory retina / RPE	Chronic retinal detachment
Optic nerve	Atrophy
Inside the globe	Possible intraocular bone
Posterior sclera	Thickening

Summary: Morphology that matters

- Small globe + internal disorganization = central diagnostic combination
- Recognize effusion, spanning membrane, detachment and posterior changes
- Distinguish congenital microphthalmia and avoid overclaiming etiology or RPE origin

The diagnosis integrates globe size with architecture.

Source and visual provenance

- Primary: Robbins, Cotran & Kumar, Pathologic Basis of Disease, 11th ed., Ch. 29, p. 1209
- Context: same chapter, retinal detachment and optic nerve sections
- All ocular diagrams in this presentation are original, editable teaching schematics—not tissue images