



SERVIZIO SANITARIO REGIONALE
EMILIA-ROMAGNA
Azienda Ospedaliero - Universitaria di Ferrara

Dal papilledema all'atrofia ottica

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UO Clinica Neurologica

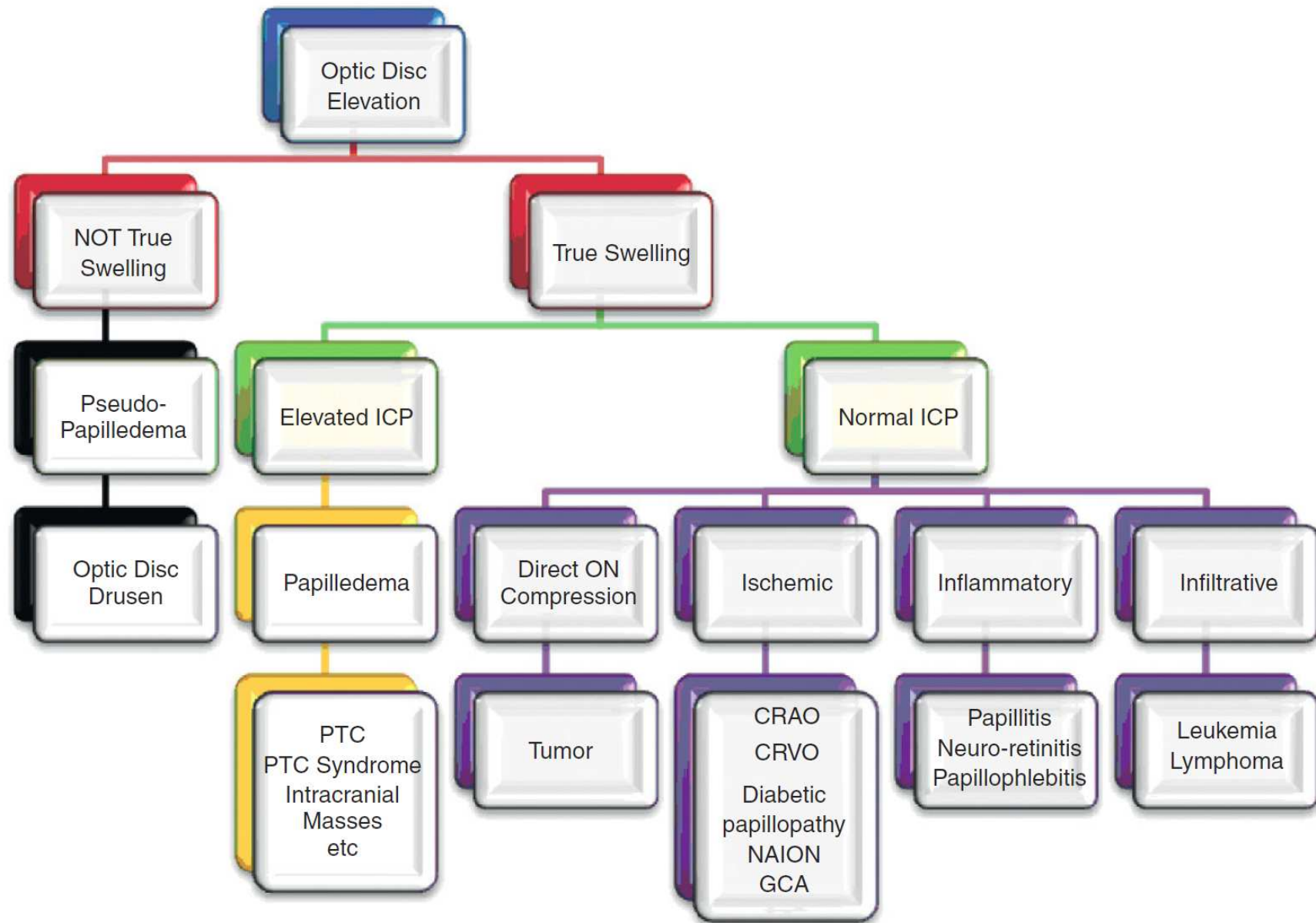
Dip. Scienze Biomediche e Chirurgico-Specialistiche

*Centro Interdipartimentale di Ricerca sulle Malattie Neuroinfiammatorie e Neurodegenerative del
Sistema Nervoso Centrale*

Università degli Studi di Ferrara

Az. Ospedaliero-Universitaria, Ferrara

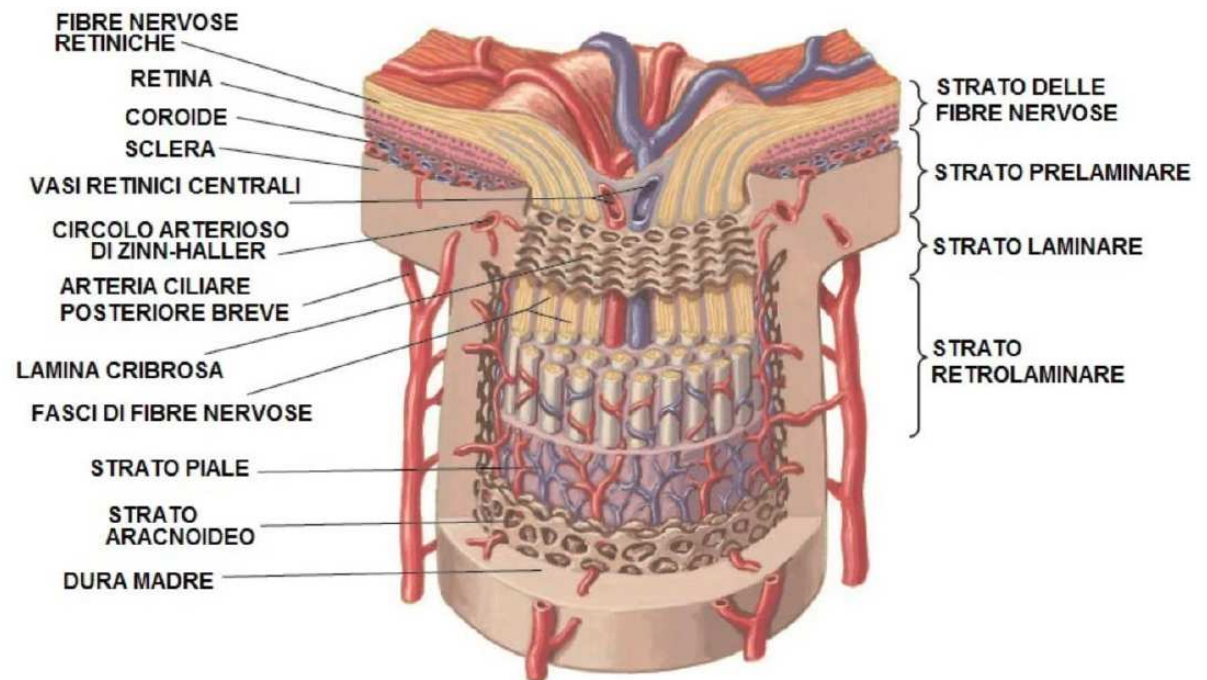
Argomenti di Neuroftalmologia – 29 Settembre 2018



ICP, intracranial pressure; PTC, pseudotumor cerebri; ON, optic nerve; CRAO, central retinal artery occlusion; CRVO, central retinal vein occlusion; NAION, nonarteritic ischemic optic neuropathy; GCA, giant cell arteritis.

- Papilledema
- Papillite
- Pseudopapilledema

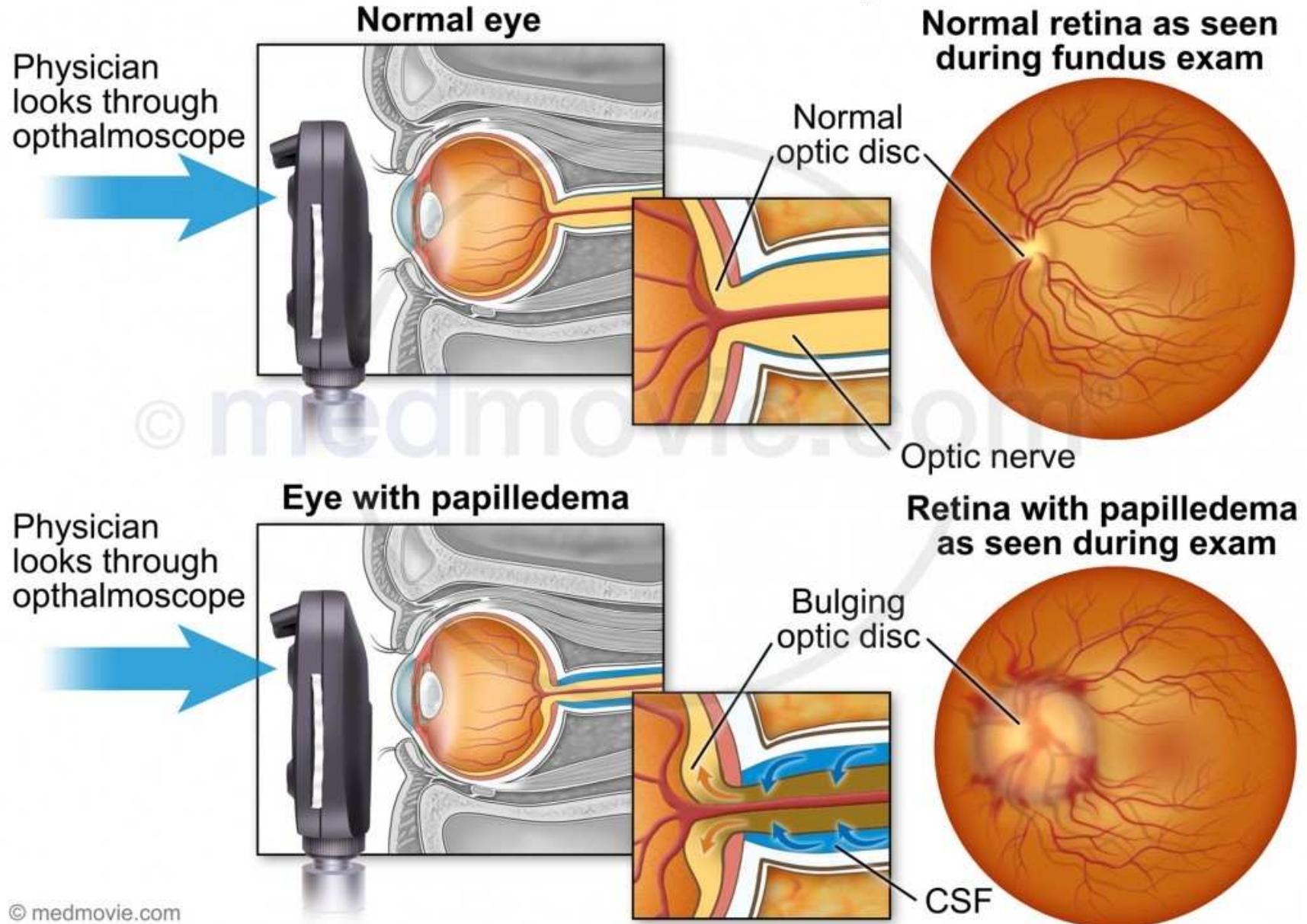
Anatomia del nervo ottico



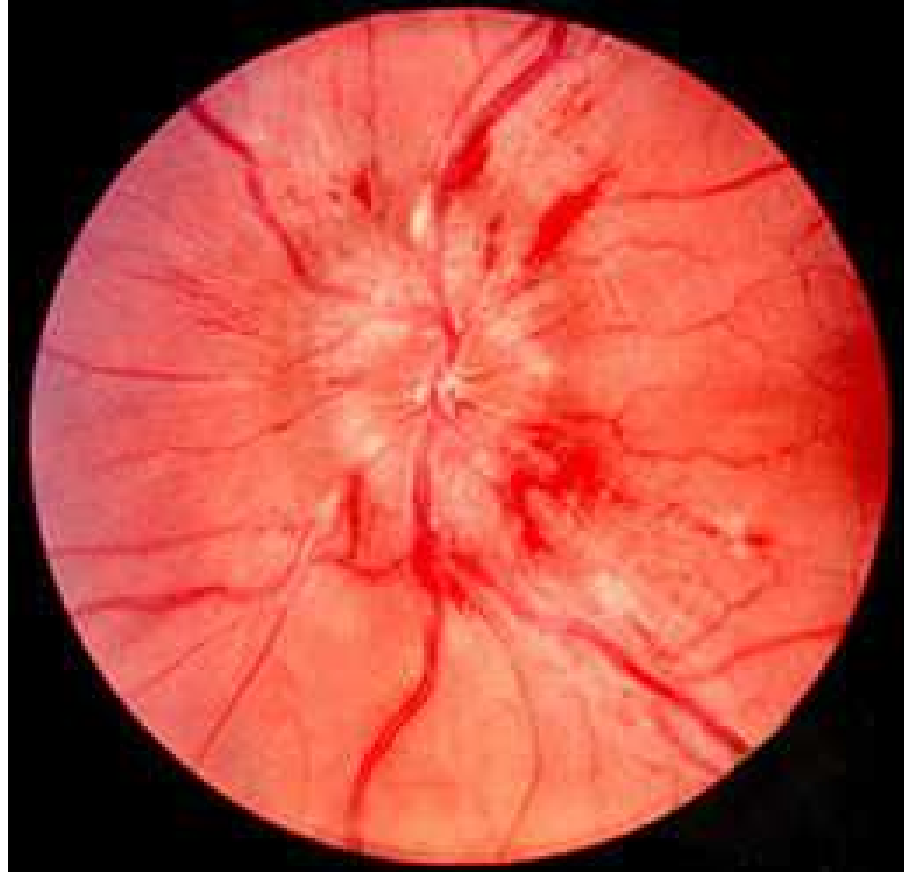
Papilledema

- **Rigonfiamento del nervo ottico:** Ostacolo al trasporto assoplasmatico e accumulo di liquido a livello della lamina cribrosa per incremento della pressione liquorale che circonda i nervi ottici nella regione retrolaminare a causa di elevata pressione intracranica
- Asimmetrico o unilaterale ma spesso bilaterale nei bambini
- Segni oftalmologici:
 - Sollevamento della papilla ottica con iperemia
 - Congestione venosa
 - Offuscamento delle vene retiniche (margini sfumati della papilla)
 - Assenza di pulsazioni venose
 - Emorragie a fiamma e macchie cotonose (segni tardivi)
 - Per lo più bilaterali
- **Cefalea** tipicamente al risveglio, offuscamento del visus transitorio (annebbiamenti di qualche secondo)
- **Diplopia orizzontale** generalmente peggiore a distanza in relazione a disfunzione del VI n. cranico
- **Normale acuità visiva** (se il papilledema florido non dà luogo a difetti della macula o se una forma cronica non ha dato luogo ad atrofia ottica).
- Difetti campimetrici visivi generalmente limitati ad un **allargamento della macchia cieca**
- **Scotoma centrale e restringimento del campo visivo** possono caratterizzare il lungo termine (mesi o anni) con associata perdita della visione per colori nell'atrofia che viene a generarsi nel papilledema cronico non trattato

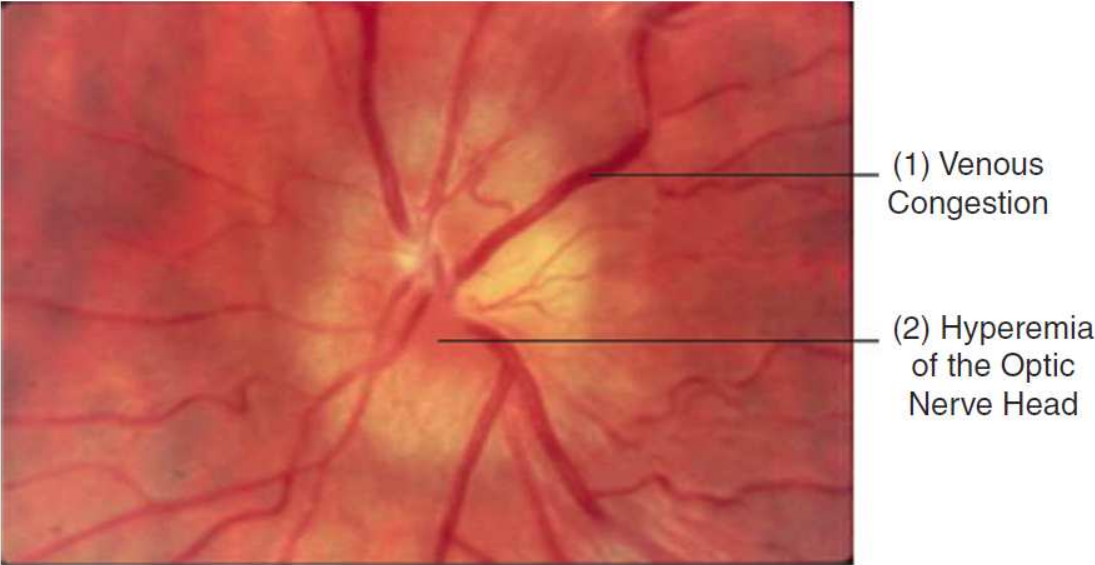
Fundus Exam - Normal vs. Papilledema



Papilledema



Papilledema



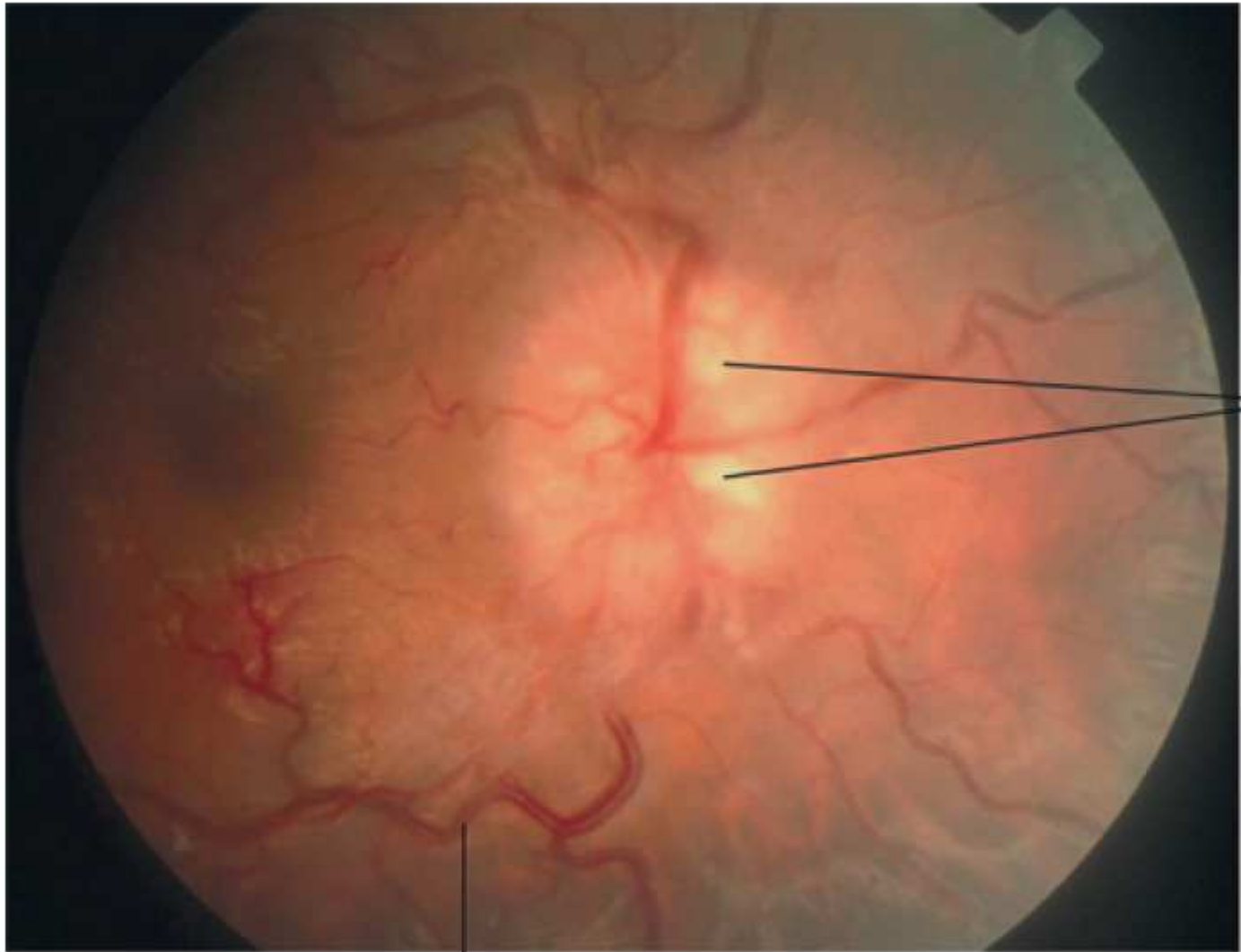
Tortuous Vein

Peripapillary Splinter / Flame Hemorrhages



Retina Striae
(Paton's Lines)

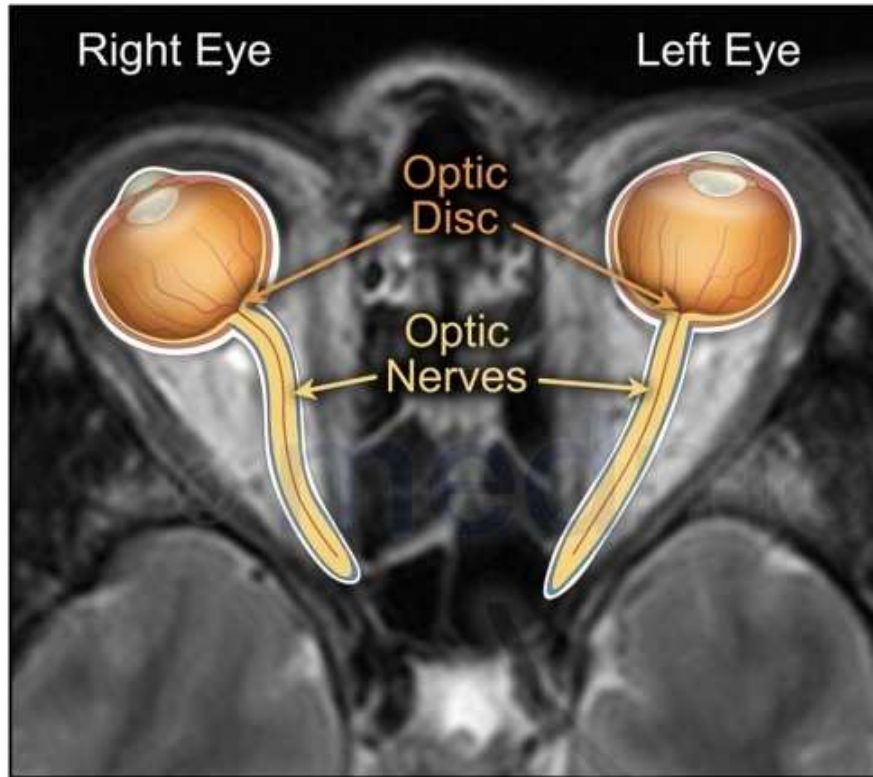
Cotton-Wool Spots



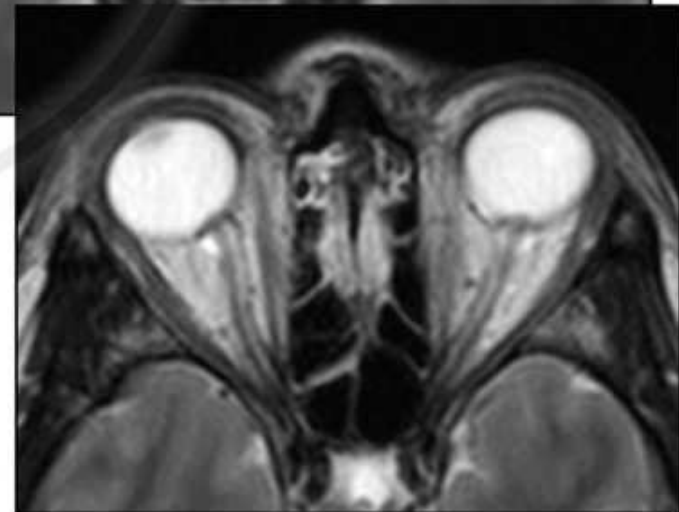
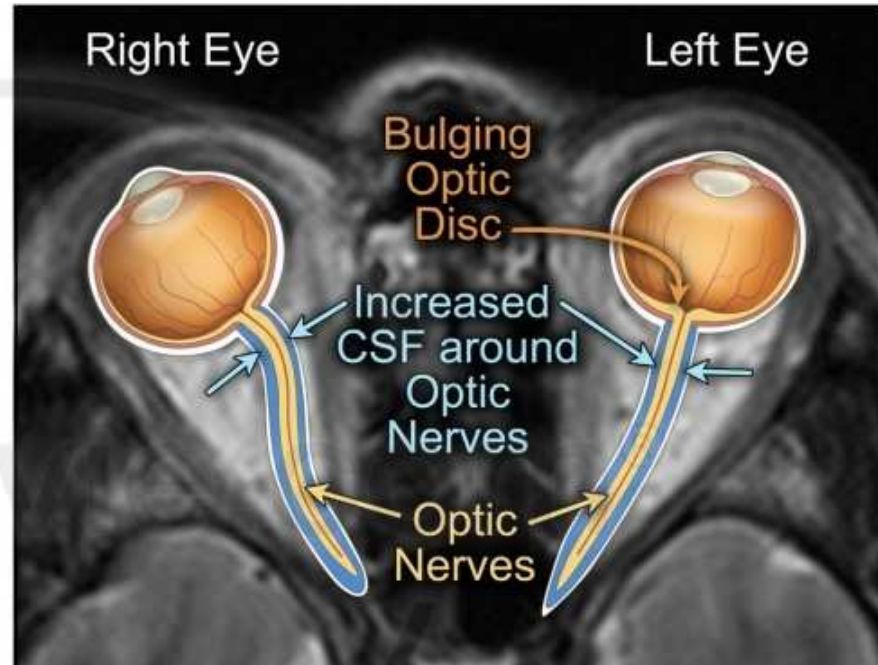
Hard Exudates
Become
Pseudodrusen

Engorged Tortuous Vein

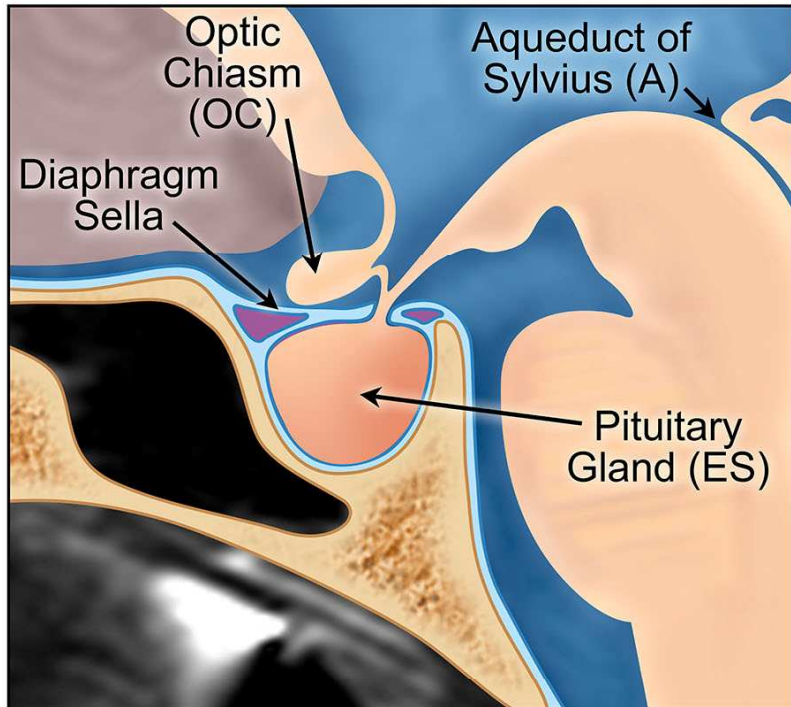
Normal Optic Nerves



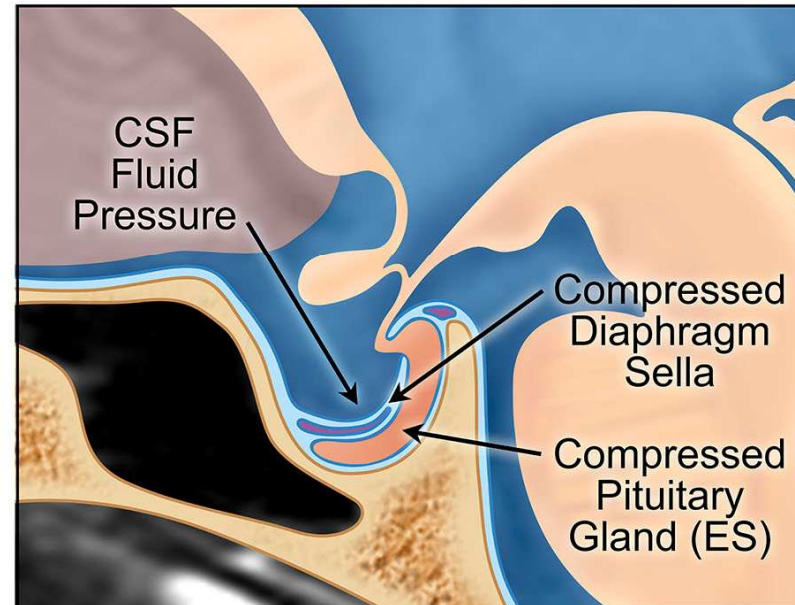
Optic Nerve with Increased CSF Pressure and Papilledema



Normal Pituitary Gland



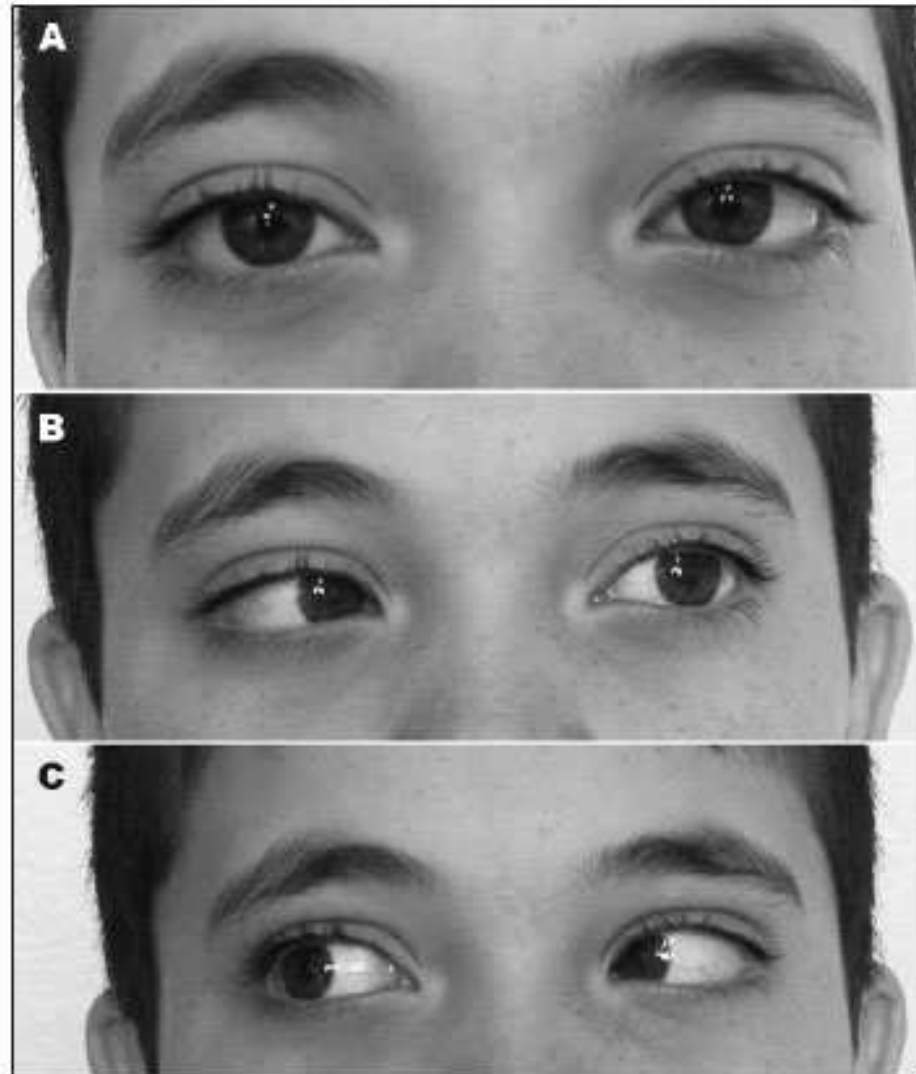
Empty Sella Pattern Indicating Lengthy Duration of High CSF Fluid Pressures



Erniazione dello spazio subaracnoideo attraverso il diaframma con spiazzamento dell'ipofisi
→ ipopituitarismo



Ipt endocranica e
paresi bilaterale
del VI n c



Ipertensione intracranica idiopatica

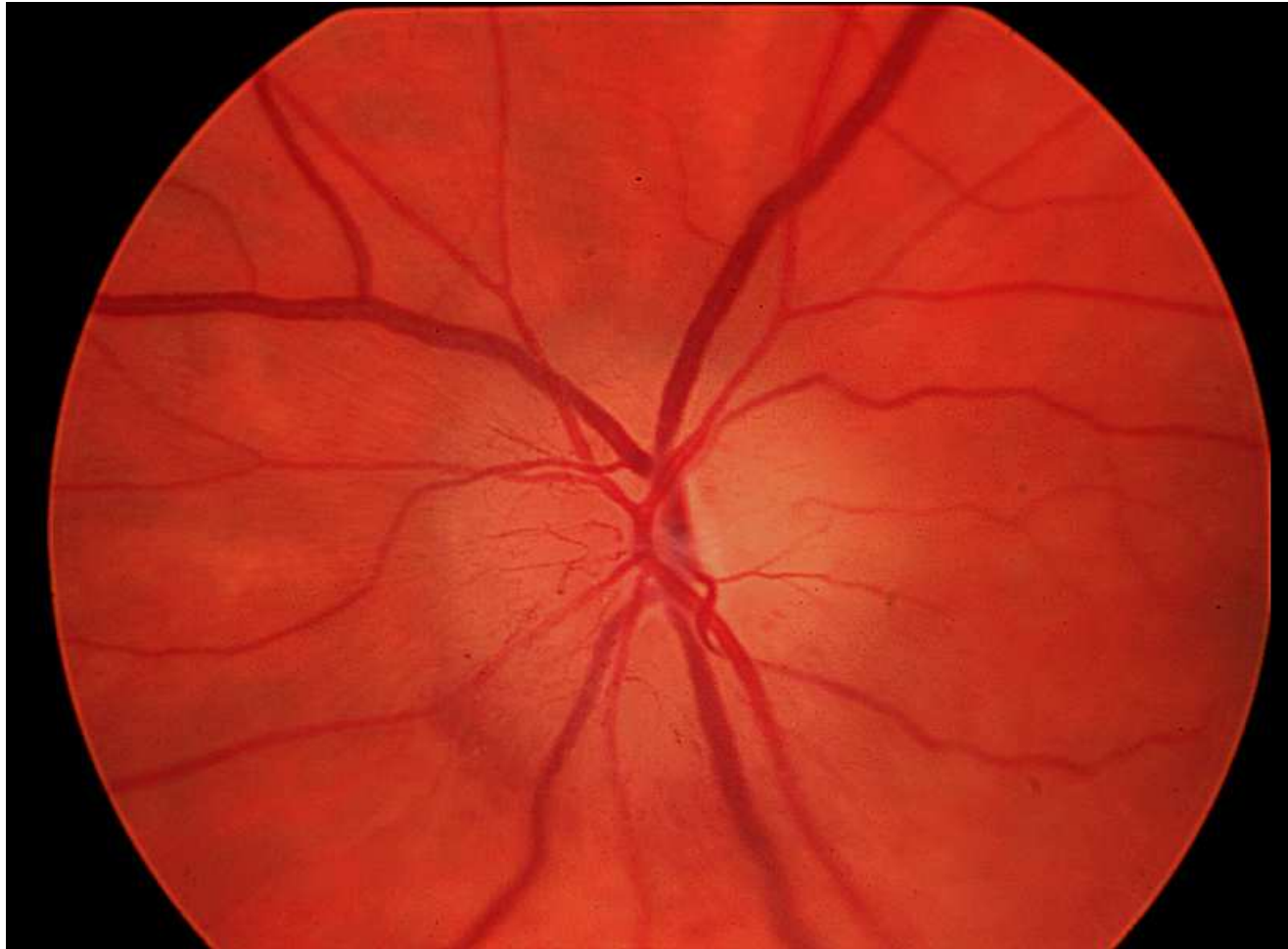
- Ipertensione Intracranica Benigna (BIH) o Pseudotumor cerebri (PTC)
- Aumento della pressione intracranica in assenza di un tumore o di altra patologia.
- Neuroimaging e rachicentesi (terapeutica)
- Trattamento farmacologico (acetazolamide) o chirurgico
- Più frequente in donne tra i 20 e i 40 anni, specialmente se obese.
- I sintomi principali sono:
 - Cefalea
 - Nausea, vomito
 - Tinnito pulsatile
 - Diplopia
 - Deficit visivi
- Terapia dell'IIH → prevenire cecità, atrofia ottica



Papillite

- Rigonfiamento della papilla ottica per infiammazione
- Rigonfiamento più comune nei bambini che negli adulti con NO
- Caratteristiche oftalmoscopiche simili alla NO, ma iperemia e emorragie più marcate
- Riduzione dell'acuità visiva e della visione per colori
- Difetto pupillare afferente presente

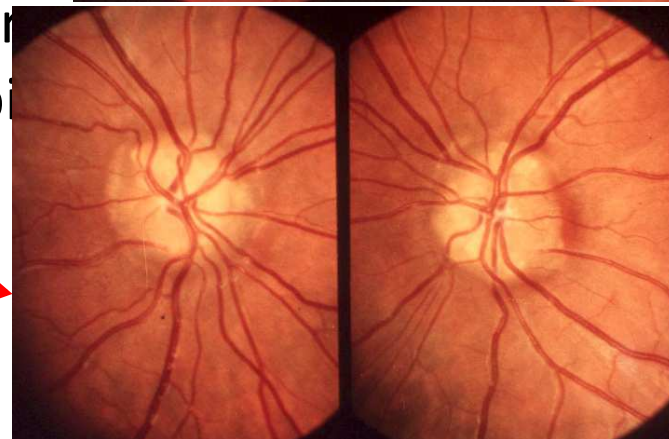
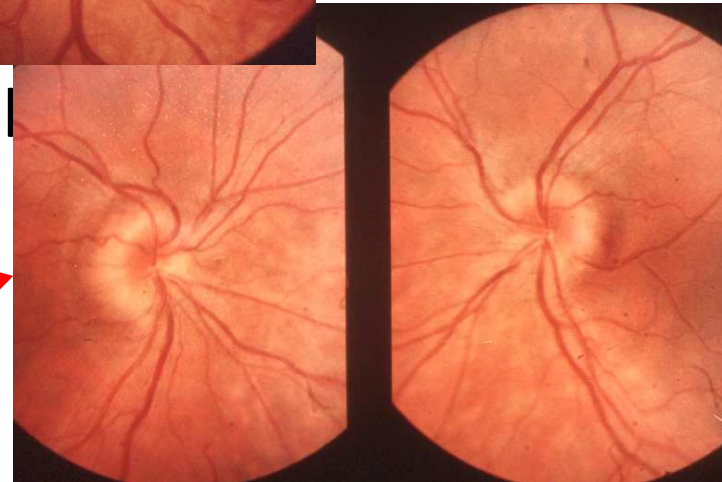
Papillite



Pseudo

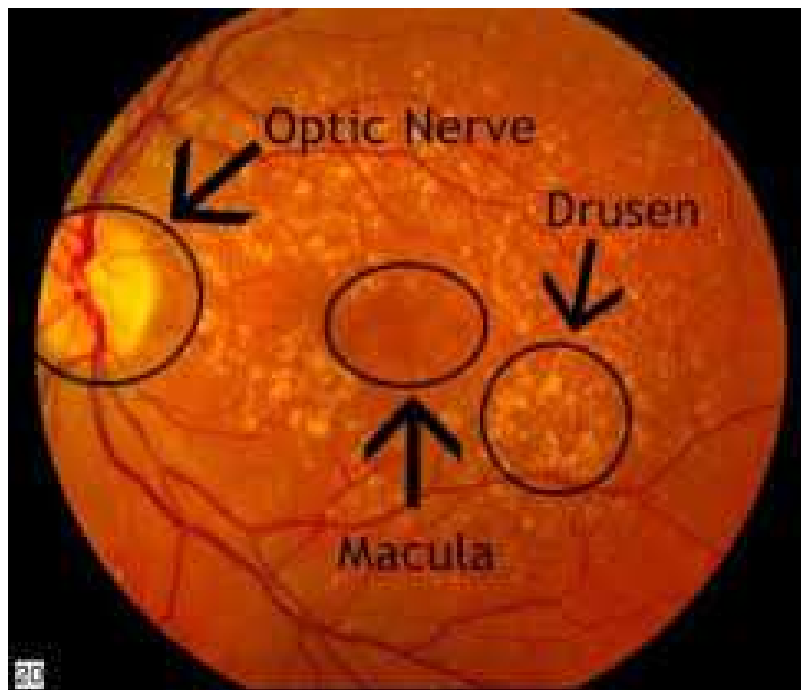
- Rigonfiamento discoidale correlati ad incremento del volume del liquido intraoculare
- Assenza di sintomi
- Diagnosi clinica con esame oftalmologico da parte di professionisti esperti
- Cause:
 - Ipermetropia
 - *Tilted disc*
 - Strato mielinico delle fibre nervose
 - Anomalie congenite della papilla ottica
 - Drusen della papilla ottica

meccanismi non uorale

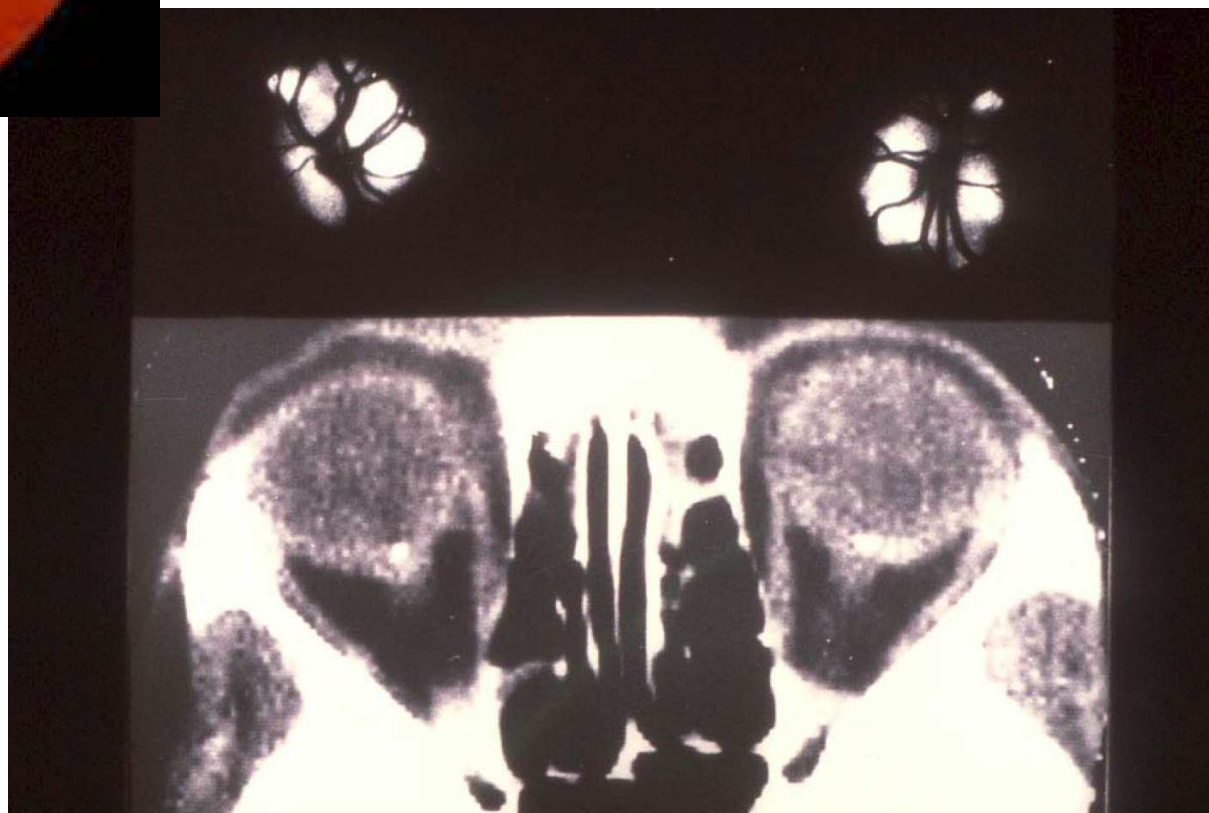


Drusen della papilla ottica

- Causa comune dello pseudopapilledema
- Piccoli aggregati di mucopolisaccaridi e materiale proteico profondamente all'interno della papilla nei bambini → riemergono in superficie calcificati ad età avanzata
- Generalmente benigni, col tempo possono generare difetti campimetrici '*wedge-shaped*'
- Eredità autosomica a penetranza variabile
- La presenza di ramificazioni vascolari e il decorso superficiale dei vasi della papilla ottica differenziano dal papilledema
- Lesione ecogenica della testa del n. ottico all'ultrasonografia
- Autofluorescenza in angiografia senza uso di fluoresceina
- Non presenta stravasi nelle fasi precoci all'angiografia
- Minor colorazione della papilla sull'angiogramma
- OCT aiuta nella DD



Drusen della papilla ottica



Characteristics of true versus pseudopapilledema

True papilledema	Pseudopapilledema
Optic disc elevation with hyperemia	Optic disc is yellow with small or absent cup
Venous engorgement	Subtle or no venous congestion
Obscuration of major retinal vessels (blurred disc margins)	Peripapillary vessels clearly seen
Absent venous pulsations	Spontaneous venous pulsations often present
Flame-shaped hemorrhages and cotton wool spots (late signs)	Anomalous vessels may be present
Majority are bilateral	May be associated with hyperopia Irregular, bulgy disc (drusen) Usually unilateral

Etiology of optic disc swelling in children

Optic disc swelling associated with neurological diseases	Optic disc swelling associated with systemic diseases	Optic disc swelling in primary ocular disease	Optic disc swelling secondary to exogenous agents
Brain tumors	Malignant hypertension	Uveitis (e.g., juvenile rheumatoid arthritis, sarcoidosis, Vogt–Koyanagi–Harada syndrome)	Corticosteroid withdrawal
Dural venous sinus thrombosis	Diabetic papillopathy	Optic neuritis	Vitamin A intoxication (or vitamin A derivative isotretinoin for acne treatment)
Dural arteriovenous malformations	Severe anemia	Malnutrition	Malnutrition
Meningitis	Cyanotic heart disease		
Encephalitis	Leukemia		
Neurofibromatosis	Sarcoidosis		
Congenital hydrocephalus	Addison’s disease		
Pseudotumor cerebri	Mucopolysaccharidosis		
Craniosynostosis	Systemic lupus erythematosus		

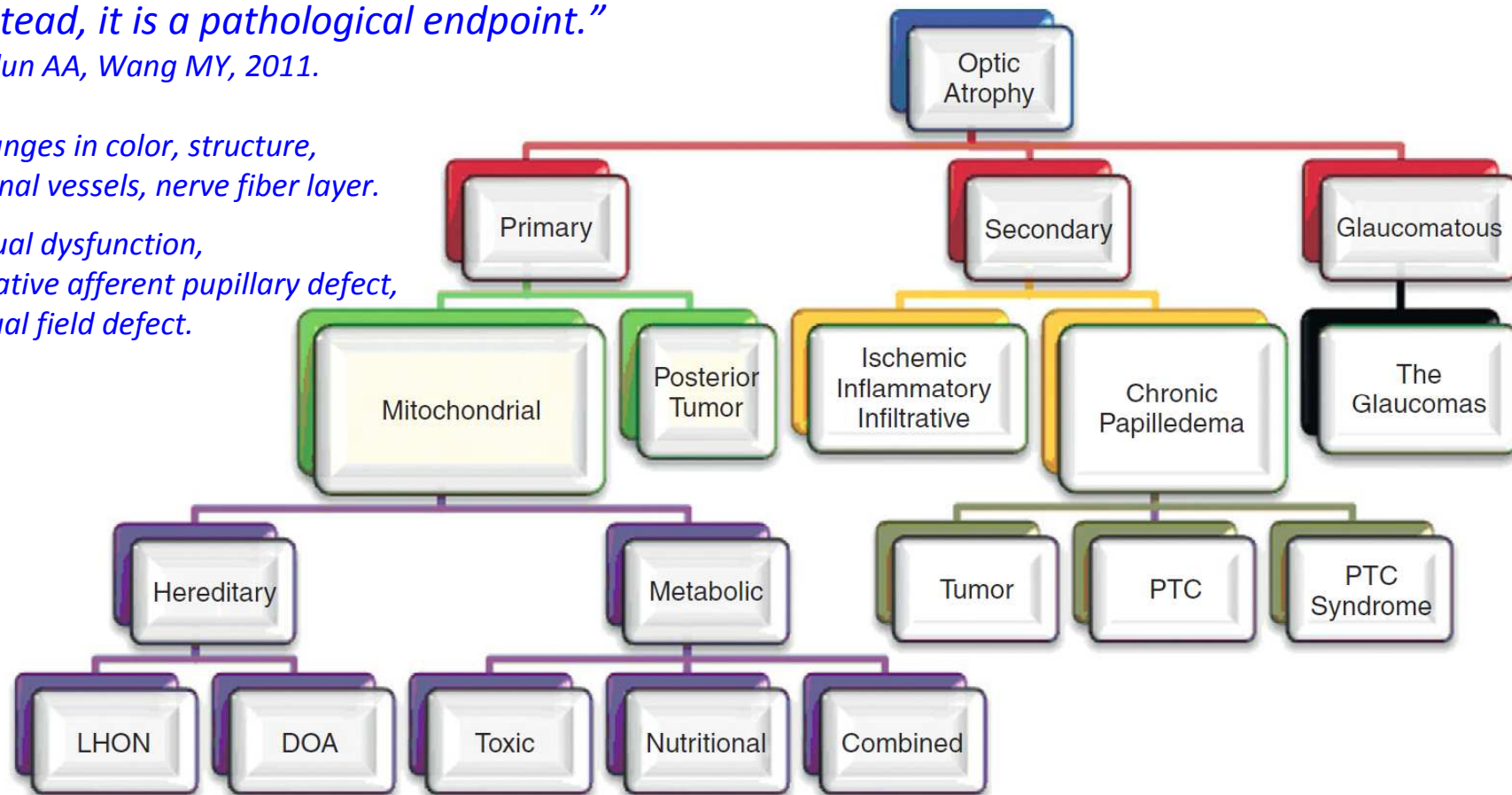
Atrofia ottica

*“Optic atrophy is not a diagnosis;
instead, it is a pathological endpoint.”*

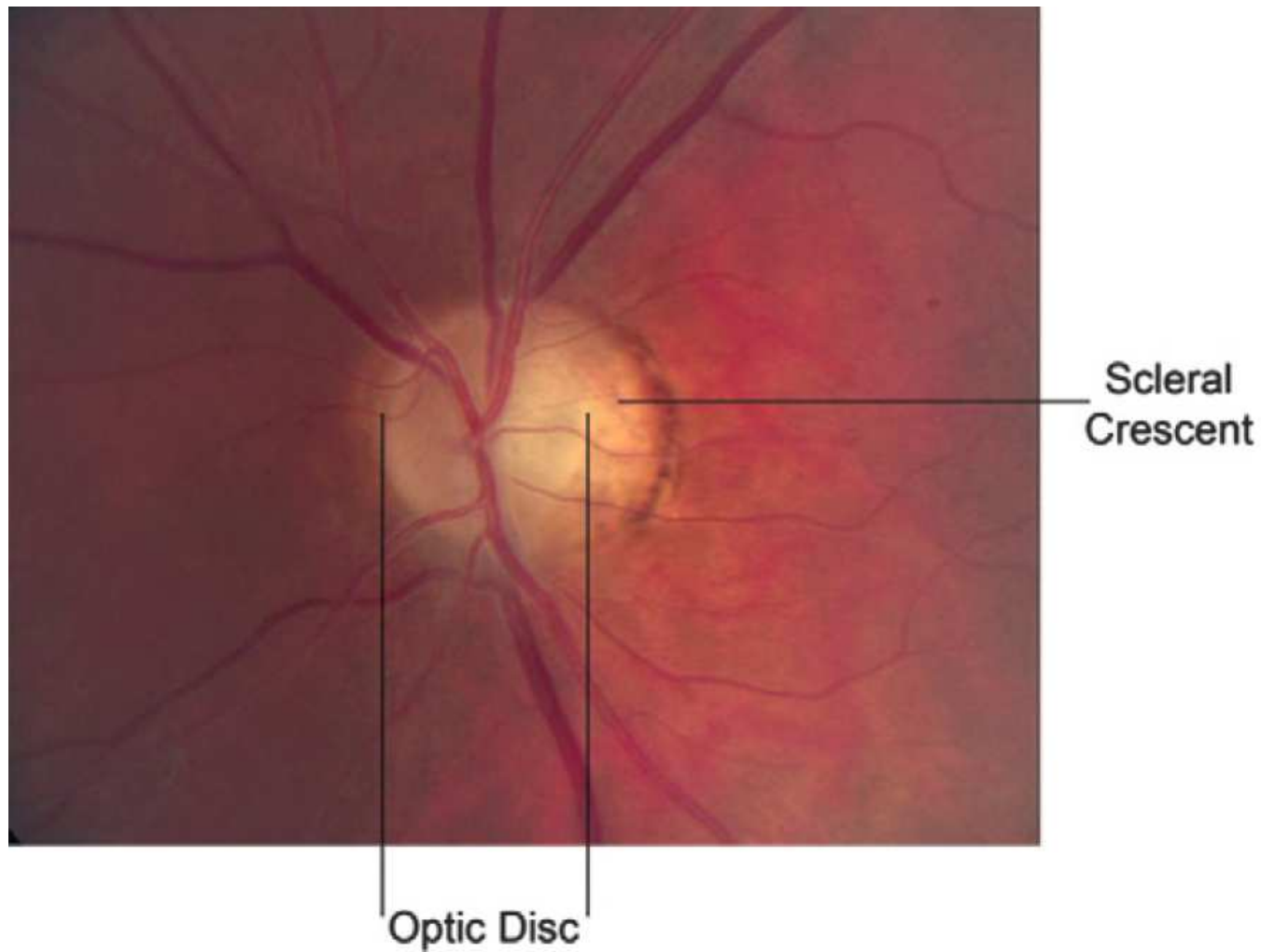
Sadun AA, Wang MY, 2011.

*Changes in color, structure,
retinal vessels, nerve fiber layer.*

*Visual dysfunction,
Relative afferent pupillary defect,
visual field defect.*

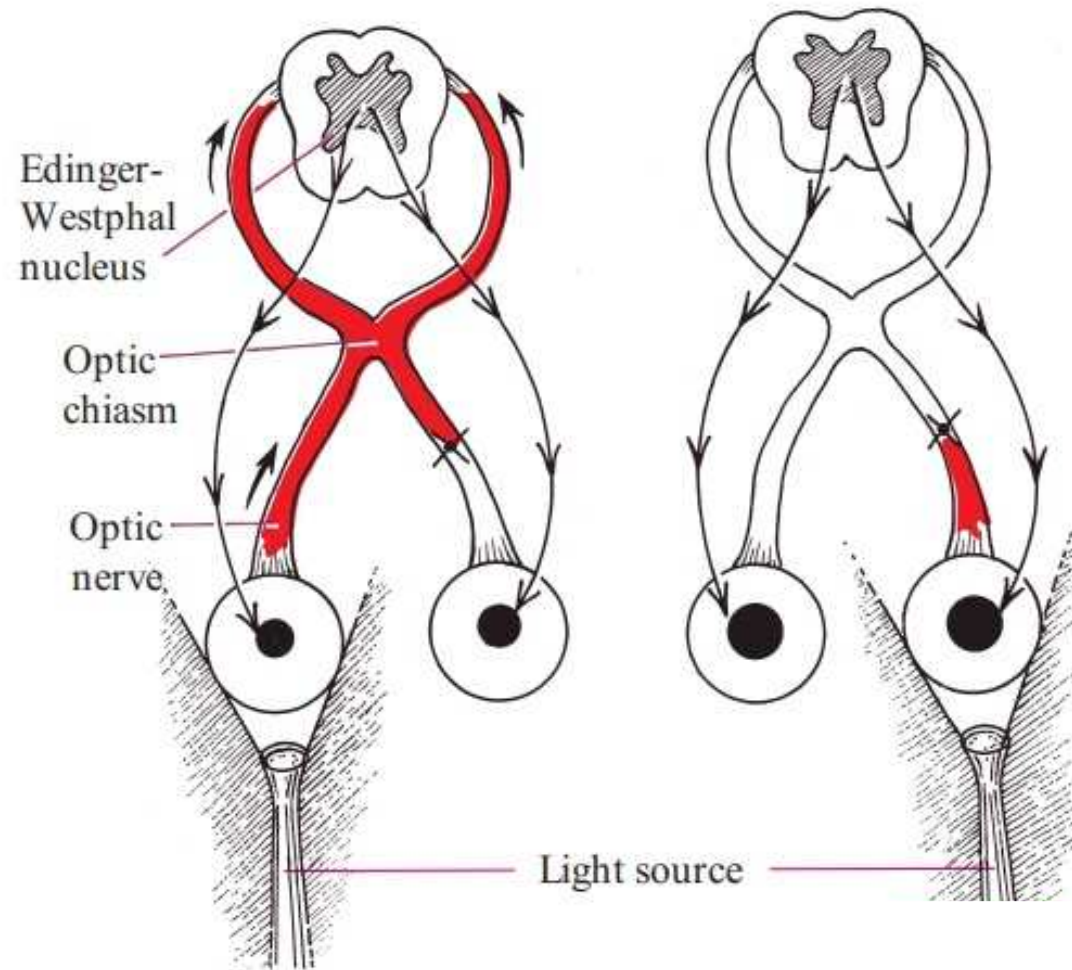


LHON, Leber’s hereditary optic neuropathy; DOA, autosomal-dominant optic atrophy; PTC, pseudotumor cerebri.



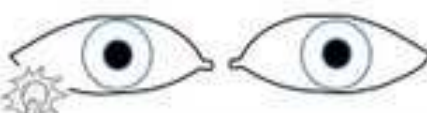
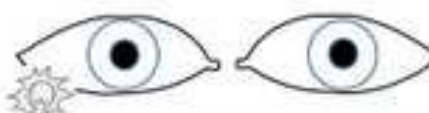
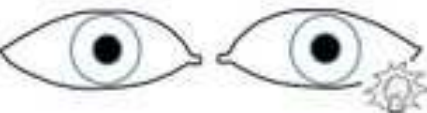
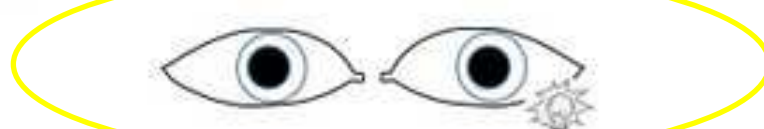
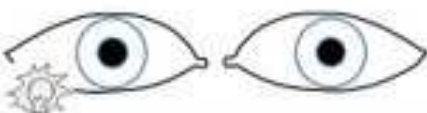
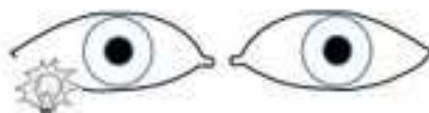


Degenerazione walleriana (assonale anterograda)
Attivazione astrociti e sostituzione assoli con tessuto gliale
Esame F.O.: luce riflette sulle cellule gliali opache, scarsa irrorazione
→ Pallore della papilla ottica

Riflesso fotomotore



Difetto pupillare afferente relativo

Normal swinging light test	Left Relative Afferent Pupillary Defect* (RAPD)
<p>(1) Begin with dark room, bright pen light, and patient fixated at distant object (to avoid a near pupil response).</p> 	<p>(1) Begin with dark room, bright pen light, and patient fixated at distant object.</p> 
<p>(2) Shine light into right (R) eye. Both pupils should constrict.</p> 	<p>(2) Shine light into right (R) eye. Both pupils constrict.</p> 
<p>(3) Swing light to left (L) eye. Both pupils remain constricted.</p> 	<p>(3) Swing light to left (L) affected eye. Instead of pupil constriction, both pupils will dilate.</p> 
<p>(4) Swing light back to right eye. Both pupils remain constricted.</p> 	<p>(4) Swing light back to right (normal) eye. Both pupils constrict.</p> 

A RAPD indicates unilateral or asymmetric optic nerve pathology (eg. asymmetric glaucoma) or retinal disease and should ALWAYS be referred to an optometrist or an ophthalmologist.

* In a right RAPD, the both pupils paradoxically dilate when light is shone in the right eye during the swinging flashlight test.