ANATOMY OF OPTIC NERVE

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Embryology of optic nerve
Introduction
Parts of optic nerve
Blood supply
Clinical significance
3rd week of gestation: the first evidence of primitive eye formation occurs.

Neural plate destined to form prosencephalon.

Depression

Optic sulcus formation
Optic sulcus deepens & the walls of prosencephalon bulge out

Optic vesicle formation

Proximal part of optic vesicle become constricted & elongated

4th week

Optic stalk formation

- Optic stalk is the original connection between the optic vesicle & the forebrain
Embryology of Optic Nerve

- Develops in the framework of the optic stalk
- Optic Nerve Head
  - Formed as the optic stalk encloses the hyaloid artery (8th week)
  - From the hyaloid artery, the vascular bud develops within Bergmeister’s papilla
  - Hyaloid artery disappears before birth
  - Bergmeister’s papilla becomes atrophic & the physiologic cup develops (at 15th week)
Optic Nerve

Axons

- Develops from the embryonic optic stalk
- Stalk lumen is progressively occupied by the axons growing from the ganglion cells (the 7th week)
- Axons fully occupy the stalk, reach the brain and a rudimentary optic chiasm is formed (the 8th week)
- Myelination starts near chiasm and stops at lamina cribrosa
Optic nerve sheaths:

- Form from the mesenchymal cells
- Begin to appear at the end of the 7th week

Glial element:

- Develops from the neuroectodermal cells forming the outer wall of the optic stalk
- Differentiates into astrocytes and oligodendrocytes
Vasculature

- Development of capillaries in the optic nerve and the CNS is similar.

- Vessels and connective tissue from the pia mater begin to enter the proximal optic nerve (at the 11th week).

- Capillaries are separated by astrocyte sheet and perivascular space.

- Vascularization is completed in the 18th week.
<table>
<thead>
<tr>
<th>Weeks of Gestation</th>
<th>Length (mm)</th>
<th>Developing Events</th>
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</thead>
<tbody>
<tr>
<td>4</td>
<td>2.5-6 mm</td>
<td>Short optic stalk</td>
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<tr>
<td>5</td>
<td>5-9 mm</td>
<td>Development of hyaloid vasculature</td>
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<tr>
<td>6</td>
<td>8-14 mm</td>
<td>Embryonic cleft closes</td>
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<tr>
<td>7</td>
<td>13-18 mm</td>
<td>Growth of axons</td>
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<td>8</td>
<td>18-31 mm</td>
<td>Stalk fully occupied by axons</td>
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<td>Axons of optic nerves reach the brain</td>
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<td>Rudimentary optic chiasm established</td>
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<td>Optic nerve vascularization starts to form</td>
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<tr>
<td>Week of Gestation</td>
<td>Length (mm)</td>
<td>Developing Events</td>
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<tr>
<td>11</td>
<td>65-73 mm</td>
<td>Vascular-connective septa invade the nerve</td>
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<tr>
<td>12</td>
<td>80 mm</td>
<td>Pia mater, arachnoid &amp; dura mater distinguishable</td>
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<td>Glial filaments appear</td>
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<td>14</td>
<td>105 mm</td>
<td>Subarachnoid space appears</td>
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<tr>
<td>15</td>
<td>117-123 mm</td>
<td>Physiologic cup starts to form</td>
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<tr>
<td>18</td>
<td>160 mm</td>
<td>Vascularization of the optic nerve completed</td>
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<tr>
<td>23</td>
<td>220 mm</td>
<td>Myelination starts</td>
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Optic Nerve

- 2\textsuperscript{nd} cranial nerve
- Starts from optic disc, extends upto optic chiasma
- Backward continuation of nerve fiber layer of retina (axons of ganglion cells)
- Also contains afferent fibers of light reflex and some centrifugal fibers
An outgrowth of brain
Not covered by neurilemma: **does not regenerate when cut**
Fibers of optic nerve are very fine (2-10 µm in diameter) & are millions in number
Surrounded by meninges unlike other peripheral nerves
Both primary & secondary neurons are in retina
About 47-50 mm in length

Divided into 4 parts:

- Intraocular (1 mm)
- Intraorbital (30 mm)
- Intracanalicular (6-9 mm)
- Intracranial (10 mm)
Intraocular Part

- Passes through sclera, choroid & appears in eye as optic disc
- 1.5 mm in diameter
- Expands to 3 mm behind sclera due to presence of myelin sheath
- Divided into 4 portions (from anterior to posterior):

  ![Diagram showing Intraocular Part](image)

  - SNFL
  - Prelaminar
  - Lamina Cribrosa
  - Retrolaminar
Intra Ocular Part
(optic papillae or optic disc):

- Retina
- Choroid
- Sclera

**OPTIC NERVE HEAD**

- Surface nerve fibre layer
- Prelaminar region
- Lamina cribrosa region
- Retrolaminar region
Surface Nerve Fiber Layer

- Composed of axonal bundles (94% nerve fibers of retina + 5% astrocytes)
- Optic disc is covered by thin layer of astrocytes, **ILM of Elschnig**: separates it from vitreous
- When central portion of membrane gets thickened: **Central meniscus of Kuhnt**
- Near the optic nerve, all layers of retina (except NFL) are separated from it by: **Intermediate tissue of Kuhnt**
Prelaminar Region

- Predominant structures: neurons and increased quantity of astroglial tissue
  - **Border tissue of Jacoby** (a cuff of astrocytes) separates the nerve from the choroid
  - The loose glial tissue does not bind the axon bundles together as do the Muller cells of the retina

**SO**

the disc swells so easily in papilloedema while the adjacent retina does not
Lamina Cribrosa

- Fibrillar sieve-like structure
- Composed of fenestrated sheets of scleral connective tissue lined by glial tissue
- Bundles of ON fibers leave the eye through LC
- **Border tissue of Elsching:**
  - rim of collagenous tissue with few glial cells
  - intervenes b/w the choroid and sclera & ON fibers
Retrolaminar Region

- Characterized by decrease in astrocytes & acquisition of myelin supplied by oligodendrocytes
- Addition of myelin sheath doubles the diameter of ON (from 1.5 to 3.0 mm) as it passes through the sclera
- Axonal bundles are surrounded by connective tissue septa
- The posterior extent of the retrolaminar region is not clearly defined
1a-Internal limiting membrane of retina
1b-Inner limiting membrane of Elschnig
2-Central meniscus of Kuhnt
3- Spur of collagenous tissue separating the anterior lamina cribrosa (6) from the choroid
4-Border tissue of Jacoby
5- Intermediary tissue of Kuhnt
6- Central meniscus of Kuhnt
7-Posterior lamina cribrosa
Ophthalmoscopic Features of Optic Nerve Head

- **Optic Disc**: part of nerve head visible with ophthalmoscope
- Intra papillary parts:
  - optic cup & neuroretinal rim
  - separated by *scleral ring of Elsching*
Why the normal disc is Pink?

- Light entering the disc diffuses among adjacent columns of glial cells and capillaries
- Acquires the pink color of the capillaries
- Light rays that exit through the tissue via the nerve fiber bundles are pink

&

give the disc its characteristic color
Nerve fiber loss in chronic glaucoma:
- leads to increased exposure of the lamina as axons are lost
- its pores become more visible as the cup enlarges,
- there is increased white reflex at disc
Disc shape
- Usually oval
- Vertical diameter being on average 9% longer than horizontal diameter
- The cup is 8% wider in the horizontal

Disc size
- Normal disc area ranges widely from 0.86 mm$^2$ to 5.54 mm$^2$
- **Macrodiscs:** area > 4.09 mm$^2$
- **Microdiscs:** area < 1.29 mm$^2$
Primary macrodiscs: associated with condition such as pits of the optic nerve ‘Morning glory syndrome’

Secondary macrodisc: associated with high myopia and buphthalmos

NAION is common in smaller ON heads due to problems of vascular perfusion and of limited space

Same is true for optic nerve head drusen due to blockage of orthograde axoplasmic flow
- **Pseudo papilloedema** is encountered with smaller optic nerve head - particularly in highly hypermetropic eye
- Susceptibility of the superior & inferior disc regions to damage: due to higher pore-to-disc area
▪ Funnel shaped depression
  - varies in form & size
  - usually off-centre towards the temporal side
▪ Cup correlates with disc:
  - large in large discs
  - small in small discs (may be absent)
▪ 3D measurement of cup shape: using confocal microscopy or stereoscopic techniques
Neuroretinal Rim

- Tissue outside the cup
- Contains the retinal nerve axons as they enter the nerve head
- ISNT rule (inferior- thickest)
- Greater axonal mass and vascularity in the inferotemporal region
In primary open angle glaucoma:
- progressive loss of retinal ganglion cells
- leading to enlargement of cup, particularly at upper & lower poles of disc
- leading to vertically oval cup

But: Horizontally oval cup-normal

Occurrence of flame shaped haemorrhages on rim, usually at inferior or superior temporal margin: early sign of glaucoma
Cup/Disc Ratio

- Ratio of cup & disc width
- Measured in same meridian, usually vertical or horizontal
- Doesn’t differ by more than 0.2 in 99% subjects

Applied

- Asymmetry of greater than 0.2 is of diagnostic importance in glaucoma
Parapapillary Chorioretinal Atrophy

- Crescentric region of chorioretinal atrophy, found temporally in normal disc
- May be exaggerated in chronic glaucoma or high myopia
- Two zones of PPCRA:
  - Zone alpha/choroidal crescent
  - more peripheral zone & is an irregular hypo- or hyper pigmented region
  - Corresponds to RPE that failed to extend to the disc margin
Zone beta or Scleral Crescent

- Related to disc centrally or zone alpha peripherally
- Consists of marked atrophy of pigment epithelium & choriocapillaries, with good visibility of larger choroidal vessels

Applied

- The zones are larger in total area & individually in the presence of chronic glaucoma
Retinal Vessels

- Emerge on medial side of cup, slightly decentered superonasally
- Temporal arteries take an arcuate course as they leave the disc
- Nasal arteries take more direct, though curved course
- Course of arteries and veins is similar but not identical

This avoids excessive shadowing of rods & cones
Venous pulsation:
- observed at disc in 15-90% of normal subjects
- due to pulsatile collapse of the veins as ocular pressure rises with arterial inflow into uvea

Arterial pulsation:
- rare, usually pathological
  Eg. High ocular pressure or aortic incompetence
Intraorbital Part

- Extends from back of the eyeball to the optic foramina
- Sinuous course to give play for the eye movements
- Covered by dura, arachnoid and pia
- The pial sheath contains capillaries and sends septa to divide nerve into fasciculi
- The SAS containing CSF ends blindly at the sclera but continues intracranially
- Central retinal artery, accompanying vein enter SAS inferomedially about 10 mm from the eyeball
- Posteriorly, near optic foramina, the ON is closely surrounded by annulus of Zinn & origin of four rectus muscles

- Some fibers of SR & MR are adherent to its sheath
- Account for the painful ocular movements seen in retrobulbar neuritis
The long & short ciliary nerves & arteries surround the ON before these enter the eyeball.

B/w ON & LR muscle are situated the ciliary ganglion, divisions of the oculomotor nerve, the nasociliary nerve, the sympathetic & the abducent nerve.

The ophthalmic artery, superior ophthalmic vein & the nasociliary nerve cross the ON superiorly from the lateral to medial side.
Relation of intraorbital part of optic nerve
Intracanalicular Part

- Closely related to ophthalmic artery
- OA crosses the nerve inferiorly from medial to lateral side in the dural sheath
- Leaves the sheath at the orbital end of the canal
- Sphenoid and post ethmoidal sinuses lie medial to it and are separated by a thin bony lamina

Applied

- This relation accounts for retrobulbar neuritis following infection of the sinuses
Intracranial Part

- Lies above the cavernous sinus & converges with its fellow to form the chiasm
- Ensheaths in pia mater
- Receives arachnoid & dural sheaths at the point of its entry into the optic canal
- Internal carotid artery runs, at first below & then lateral to it
- Medial root of the olfactory tract & the anterior cerebral artery lie above it
Arrangements of nerve fibers in optic nerve

- Exactly same as in retina
- Fibers from the peripheral part of the retina:
  - lie deep in the retina
  - occupy the most peripheral part of the optic disc
- Fibers originating closer to the optic nerve head:
  - lie superficially in the retina
  - occupy a more central portion of the disc
OPTIC NERVE HEAD:

- Peripheral fibers → deep in Retina → superficially in optic nerve
- Fibers close to optic nerve head → superficial in retina → central in optic nerve
In the distal region

- Exactly as in retina
  - i.e. upper temporal & lower temporal fibers are situated on the temporal half of the optic nerve
  - separated from each other by a wedge shaped area occupied by the Pmb
- The upper nasal and lower nasal fibers are situated on the nasal side

In the proximal region

- The macular fibers are centrally placed
Arrangement of fibres in ON head & distal region

Arrangement of fibres in proximal region of ON
Blood supply of optic nerve

Intraocular part

1. Surface nerve fiber layer

Supplied by:

i. Peripapillary arterioles of CRA origin
ii. Epipapillary arterioles of CRA origin
iii. Rich anastomoses with prelaminar region
iv. Occasional anastomoses with choriocapillaries
v. Precapillary branches from cilioretinal arteries when present
Prelaminar and laminar region

Derive from short posterior ciliary arteries

Arterial circle of Zinn-Haller

Retrolaminar region

- Receives its supply mainly from arteries & arterioles of pial sheath of neighbouring leptomeninges
Optic disc edema occurs as prelaminar axons swell from orthogonal axoplasmic flow at level of lamina cribrosa.

Insufficient blood flow through posterior ciliary arteries due to thrombosis, hypotension, vascular occlusion cause ON head infarction.
Venous drainage of optic nerve head

- In each zone:
  - venules drain into central retinal vein
  - or when present into a duplicated vein (an embryonic persistence of hyaloid veins)
- Occasionally septal veins in retrolaminar region drain into pial veins
- Some small venules from prelaminar region or from SNFL (opticociliary veins) drain into choroid
Optiociliary veins may enlarge in optic nerve sheath meningiomas
Intraorbital Part

- Supplied by 2 systems of vessels: a periaxial and an axial
- Periaxial consists of 6 branches of internal carotid artery:
  - Ophthalmic artery
  - Long posterior ciliary arteries
  - Short posterior ciliary arteries
  - Lacrimal artery
  - Central artery of retina before it enters ON
  - Circle of Zinn
The axial system consists of:
- Intraneural branches of central retinal artery
- Central collateral arteries which come off from CRA before it pierces the nerve
- Central artery of ON
Blood Supply Of Optic Nerve
Intracanalicular Part

- Ophthalmic artery is sole supply to this portion, except occasional branch from CRA on its inf. aspect
- Branches from ophthalmic artery arises within the canal or in the orbit
- Pial network is poor in this region, because arteries reach pia along connective tissue bands binding the nerve to surrounding dural sheath
This supply is vulnerable to shearing injury in skull fracture
Intracranial part

- **Perichiasmal artery**: branch of superior hypophyseal branch of ICA
  - Runs back along the medial side of the ON, joins its fellow of the opposite side along the anterior border of the chiasm & supplies both
  - It is probably the largest supply to intracranial part
- **Ophthalmic artery** gives number of small collateral arteries running backwards along inferior surface of nerve, winding round its margin in superior aspect
- Additional branches from anterior cerebral artery and anterior communicating artery also supply the same
Carotid artery aneurysms, displacement of carotid artery can compress ON
Blood Supply Of Optic Nerve Head

Blood Supply Of The Various Parts Of The Optic Nerve
• Chiefly by central retinal vein & to lesser extent via pial venous system
• Both system drain into the ophthalmic venous system in the orbit & less commonly directly into cavernous sinus
Blood brain barrier at the optic nerve

- The capillaries of ON head, the retina and the CNS, have non-fenestrated endothelial linings with tight junctions b/w adjacent endothelial cells
- These are responsible for blood tissue barrier to the diffusion of small molecules across capillaries
- However it is incomplete as a result of continuity b/w the extracellular spaces of choroid and ON head at level of choroid (in prelaminar region)
- There is no barrier to diffusion across the highly fenestrated capillaries of the choroid
Signs of Optic Nerve Dysfunction

- Reduced VA
- Afferent pupillary defects
- Visual field defects
- Dischromatopsia
Diminished light sensitivity

Diminished contrast sensitivity

- Optic disc changes on fundoscopy include:
  - Disc edema
  - Hyperemia
  - Paleness
  - Atrophy
Lesions of the visual pathway

Lesions of the optic nerve

- Complete blindness on the affected side
- Abolition of direct light reflex on ipsilateral side & consensual on contralateral side
- Near (accommodation) reflex is present
- Causes- optic atrophy
  - Traumatic avulsion of optic nerve
  - Indirect optic neuropathy
  - Acute optic neuritis
Lesion through proximal part of optic nerve

- Ipsilateral blindness
- Contralateral hemianopia
- Abolition of direct light reflex on affected side & consensual on contralateral side
- Near reflex is intact
Dia Showing Defects Of Visual Pathway
Hyperopic Optic Disc

- Disc usually lacking physiological cup
- Have crowded appearance mimicking papilledema

Myopic Optic Disc

- Disc is larger
- Surrounded by white crescent of bare sclera, on the temporal side
- CDR is bigger mimicking glaucomatous cupping
Congenital Anomalies of Optic Nerve

**Without systemic association**
- Tilted optic disc
- Optic disc drusen
- Optic disc pit
- Myelinated nerve fiber

**With systemic association**
- Optic disc coloboma
- Morning glory syndrome
- Optic nerve hypoplasia
- Aicardi syndrome
- Megalopapilla
- Peripapillary staphyloma
- Optic disc dysplasia
Optic Disc Coloboma

- Results from an incomplete closure of the embryonic fissure
- Defect of the inferior aspect of ON
- White mass: glial tissue fills the defect
- Inferior NRR: thin or absent, superior NRR: relatively normal
Myelinated nerve fiber

- White, feathery patches that follow NFL Bundles
- Peripheral edges fanned out
- Simulated disc edema
Optic disc pit

- Round or oval, gray or white depression in the optic disc
- Commonly found temporally
Tilted optic disc

- Occurs when nerve exits the eye at an oblique angle
- Superotemporal disc: raised, simulating disc swelling
- Inferotemporal disc: flat or depressed
- Resulting in an oval-shaped disc with long axis at an oblique angle
Optic disc drusen

- Globules of mucoproteins & mucopolysaccharides that progressively calcify in the optic disc
- Thought to be the remnants of the axonal transport system of degenerated retinal ganglion cells
Morning Glory Disc

- Congenital funnel shaped excavation of the posterior pole
- White tuff of glial tissue covers central portion of cup
- Blood vessels appear to be increased in no. & emanate from the edge of disc
Optic nerve hypoplasia

- Optic nerve head appears abnormally small due to a low no. of axons
- Gray or pale disc surrounded by light-colored peripapillary halo
- Double ring sign
Aicardi syndrome

- Rare genetic disorder in which corpus callosum is partly or completely missing
Megalopapilla

- Abnormally large disc with large cup to disc ratio
- Area > 2.5 mm²
- Pale NRR
Peripapillary staphyloma

- Area around disc is deeply excavated, with atrophic changes in RPE
- Generally unilateral
Papilloedema

- Swelling of ON head secondary to raised intracranial pressure
Optic Atrophy

Primary Optic Atrophy

Secondary Optic Atrophy
Optic Neuropathy

Arteritic anterior ischaemic optic neuropathy

Non-arteritic anterior ischaemic optic neuropathy
References

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