Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve...

[Two-words] Disc

Optic...

Myelinated...

Optic Nerve...

Megalo-...

Optic Nerve...

[Two-words] Syndrome

Morning-Glory Disc
These three are all secondary to abnormal closure of the embryonic optic fissure

- **Optic Nerve Coloboma**
- **Optic Pit/Hole**
- **Morning-Glory Disc**

**Megalopapilla**

**Myelinated RNFL**

**Optic Nerve Hypoplasia**

**Tilted-Disc Syndrome**

**Developmental Abnormalities of the Optic Nerve Head**

**Tilted-Disc Syndrome**

1. Superior pole appears elevated, inferior recessed
2. Associated with situs inversus of retinal vessels
3. Fundus abnormality produces myopic astigmatism
4. VF testing reveals bitemporal hemianopia that doesn’t respect the vertical and may resolve with refraction
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble…
2)
3)

Optic Pit/Hole

Morning-Glory Disc

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
1) May resemble... deep cupping
2)
3)

These three are all secondary to abnormal closure of the embryonic optic fissure

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Morning-Glory Disc

Optic Pit/Hole

Tilted-Disc Syndrome
Optic Nerve Coloboma
1) May resemble... deep cupping
2) Can be... [laterality]
3)
These three are all secondary to abnormal closure of the embryonic optic fissure

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Morning-Glory Disc

Tilted-Disc Syndrome

Optic Pit/Hole

Developmental Abnormalities of the Optic Nerve Head
Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
1) May resemble... **deep cupping**
2) Can be... **bilateral, asymmetric**
3) Part of the **CHARGE association**

These three are all secondary to abnormal closure of the embryonic optic fissure

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Morning-Glory Disc

Tilted-Disc Syndrome
These three are all secondary to abnormal closure of the embryonic optic fissure

**Optic Nerve Coloboma**
1) May resemble **deep cupping**
2) Can be **bilateral, asymmetric**
3) Part of the **CHARGE association**

**Megalopapilla**

**Optic Pit/Hole**

**Myelinated RNFL**
-- Myelin normally starts at LGN, ends at lamina cribrosa
-- Can be patchy, discontinuous
-- Corresponding VF has absolute scotoma

**Optic Nerve Hypoplasia**
-- Abnormally low number of axons
-- DFE: Small pale disc with double ring sign
-- VA **20/20<->NLP**
-- VF defects invariably present
-- Remember the 4 D's (more on this shortly)

**Morning-Glory Disc**
-- DFE reveals:
1) Funnel-shaped
2) Number of vessels crossing the rim seems abnormally high
-- Tissue is contractile, so cup seems to open and close (like its namesake, the morning-glory flower)
-- VA usually **20/200**, but can be **20/20<->NLP**
-- 1/3 develop serous RD

**Tilted-Disc Syndrome**
1) Superior pole appears elevated, inferior recessed
2) Associated with **situs inversus** of retinal vessels
3) Fundus abnormality produces myopic astigmatism
4) VF testing reveals bitemporal hemianopia that doesn't respect the vertical and may resolve with refraction
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

**Optic Nerve Coloboma**
1) May resemble...**deep cupping**
2) Can be...**bilateral, asymmetric**
3) Part of the...**CHARGE association**

**Optic Pit/Hole**

**Morning-Glory Disc**
1) Superior pole appears...**elevated**
2) Inferior...**recessed**
3) Associated with...**situs inversus** of retinal vessels
4) Fundus abnormality produces...**myopic astigmatism**
5) VF testing reveals...**bitemporal hemianopia** that doesn't...**resolve with refraction**

**Megalopapilla**

**Myelinated RNFL**
--Myelin normally starts at LGN, ends at lamina cribrosa
--Can be...**patchy, discontinuous**
--Corresponding VF has absolute scotoma

**Optic Nerve Hypoplasia**
--Abnormally...**low number of axons**
--DFE: Small pale disc with...**double ring sign**
--VA...**20/20<->NLP**
--VF defects invariably present
--Remember the...**4 D's**

**Optic Pit/Hole**
--Associated with...**serous RD** in adulthood

**Morning-Glory Disc**
1) Funnel-shaped...2) Number of vessels crossing the rim seems abnormally...**high**
3) Tissue is...**contractile**, so cup seems...to...**open and close** (like its namesake, the morning-glory flower)
4) VA usually...**20/200**, but can be...**20/20<->NLP**
5) 1/3 develop...**serous RD**
Developmental Abnormalities of the Optic Nerve Head

- Myelinated RNFL
  - Myelin normally starts at LGN, ends at lamina cribrosa
  - Can be patchy, discontinuous
  - Corresponding VF has absolute scotoma

- Megalopapilla
  - Abnormally large diameter of disc and cup
  - VF testing may reveal an enlarged blind spot

- Optic Nerve Hypoplasia
  - Abnormally low number of axons
  - DFE: Small pale disc with double ring sign
  - VA 20/20→NLP
  - VF defects invariably present
  - Remember the 4 D’s (more on this shortly)

- Optic Pit/Hole
  - Associated with serous RD in adulthood

- Morning-Glory Disc
  - DFE reveals:
    1) Funnel-shaped
    2) Number of vessels crossing the rim seems abnormally high
  - Tissue is contractile, so cup seems to open and close (like its namesake, the morning-glory flower)
  - VA usually 20/200, but can be 20/20→NLP
  - 1/3 develop serous RD

- These three are all secondary to abnormal closure of the embryonic optic fissure

What is the CHARGE association?
- Coloboma
- Heart abnormalities
- Choanal atresia
- Retardation
- Genitourinary abnormalities
- Ear abnormalities

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Pit/Hole

Morning-Glory Disc

Tilted-Disc Syndrome

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Myelinated RNFL
-- Myelin normally starts at LGN, ends at lamina cribrosa
-- Can be patchy, discontinuous
-- Corresponding VF has absolute scotoma

Megalopapilla
-- Abnormally large diameter of disc and cup
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Optic Nerve Hypoplasia
-- Abnormally low number of axons
-- DFE: Small pale disc with double ring sign
-- VA 20/20–NLP
-- VF defects invariably present
-- Remember the 4 D's (more on this shortly)

Optic Pit/Hole
-- Associated with serous RD in adulthood

Morning-Glory Disc
-- DFE reveals: 1) Funnel-shaped…2) Number of vessels crossing the rim seems abnormally… high
-- Tissue is… contractile, so cup seems to… open and close (like its namesake, the morning-glory flower)
-- VA usually… 20/200, but can be… 20/20–NLP
-- 1/3 develop… serous RD

These three are all secondary to abnormal closure of the embryonic optic fissure

What is the CHARGE association?
- Coloboma
- Heart abnormalities
- Choanal Atresia
- Retardation
- Genitourinary abnormalities
- Ear abnormalities

Optic Nerve Coloboma
1) May resemble… deep cupping
2) Can be… bilateral, asymmetric
3) Part of the… CHARGE association

Optic Pit/Hole

Morning-Glory Disc

Tilted-Disc Syndrome
1) Superior pole appears… elevated, inferior… recessed
2) Associated with… situs inversus of retinal vessels
3) Fundus abnormality produces… myopic astigmatism
4) VF testing reveals… bitemporal hemianopia that doesn't… respect the vertical and may… resolve with refraction

Developmental Abnormalities of the Optic Nerve Head

Coloboma
Heart abnormalities
Choanal Atresia
Retardation
Genitourinary abnormalities
Ear abnormalities
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble…deep cupping
2) Can be…bilateral, asymmetric
3) Part of the…CHARGE association

Optic Pit/Hole
--Associated with…
[retinal condition, and age]

Morning-Glory Disc

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome

1/3 develop…serous RD
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Pit/Hole
--Associated with...serous RD in adulthood

Morning-Glory Disc

Megalopapilla

Myelinated RNFL

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Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Pit/Hole
--Associated with...serous RD in adulthood

Morning-Glory Disc

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

- **Optic Nerve Coloboma**
  1) May resemble... **deep cupping**
  2) Can be... **bilateral, asymmetric**
  3) Part of the... **CHARGE association**

- **Optic Pit/Hole**
  --Associated with... **serous RD in adulthood**

- **Morning-Glory Disc**
  --DFE reveals:
  1) A funnel-shaped... **[classic term]**
  2)

- **Megalopapilla**

- **Myelinated RNFL**

- **Optic Nerve Hypoplasia**

- **Tilted-Disc Syndrome**
  1) Superior pole appears... **elevated**
  2) Associated with... **situs inversus of retinal vessels**
  3) Fundus abnormality produces... **myopic astigmat**
  4) VF testing reveals... **bitemporal hemianopia** that doesn't... resolve with refraction
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble... *deep cupping*
2) Can be... *bilateral, asymmetric*
3) Part of the... *CHARGE association*

Optic Pit/Hole
--Associated with... *serous RD in adulthood*

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped... *excavation*
  2) 

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble…deep cupping
2) Can be…bilateral, asymmetric
3) Part of the…CHARGE association

Optic Pit/Hole
--Associated with…serous RD in adulthood

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped…excavation
2) Number of vessels crossing the rim seems abnormally… [high v low]

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome

1/3 develop…serous RD [high v low]
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble… **deep cupping**
2) Can be… **bilateral, asymmetric**
3) Part of the… **CHARGE association**

Optic Pit/Hole
--Associated with… **serous RD in adulthood**

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped… **excavation**
  2) Number of vessels crossing the rim seems abnormally… **high**

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

Optic Pit/Hole
--Associated with...serous RD in adulthood

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Nerve Hypoplasia
--Abnormally low number of axons
--DFE: Small pale disc with double ring sign
--VA: 20/20<->NLP
--VF defects invariably present
--Remember the 4 D's (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears...elevated, inferior...recessed
2) Associated with...situs inversus of retinal vessels
3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn't respect the vertical and may resolve with refraction

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...[descriptor], so cup seems to...[type of change] (like its namesake, the morning-glory flower)

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Pit/Hole
--Associated with...serous RD in adulthood

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble **deep cupping**
2) Can be **bilateral, asymmetric**
3) Part of the **CHARGE association**

Optic Pit/Hole
--Associated with **serous RD in adulthood**

Morning-Glory Disc
--DFE reveals:
   1) A funnel-shaped **excavation**
   2) Number of vessels crossing the rim seems abnormally **high**
   --Tissue is **contractile**, so cup seems to **open and close** (like its namesake, the morning-glory flower)
   --VA usually... **[range]**, but can be... **[range]**

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

Optic Pit/Hole
--Associated with…serous RD in adulthood

Optic Nerve Coloboma
1) May resemble…**deep cupping**
2) Can be…**bilateral, asymmetric**
3) Part of the…**CHARGE association**

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped…**excavation**
  2) Number of vessels crossing the rim seems abnormally…**high**
--Tissue is…**contractile**, so cup seems to…**open and close** (like its namesake, the morning-glory flower)
--VA usually…**20/200**, but can be…**20/20<->NLP**

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome

*These three are all secondary to abnormal closure of the embryonic optic fissure*
Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
1) May resemble... **deep cupping**
2) Can be... **bilateral, asymmetric**
3) Part of the... **CHARGE association**

Optic Pit/Hole
--Associated with... **serous RD in adulthood**

Morning-Glory Disc
--DFE reveals:
  1) A funnel-shaped... **excavation**
  2) Number of vessels crossing the rim seems abnormally... **high**
  --Tissue is... **contractile**, so cup seems to... **open and close** (like its namesake, the morning-glory flower)
  --VA usually... **20/200**, but can be... **20/20<>NLP**
  --1/3 develop... **[retinal condition]**

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped...excavation
2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
--VA usually...20/200, but can be...
20/20<->NLP
--1/3 develop...serous RD

Optic Pit/Hole
--Associated with...serous RD in adulthood

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Megalopapilla

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped... excavation
2) Number of vessels crossing the rim seems abnormally... high
--Tissue is... contractile, so cup seems to... open and close (like its namesake, the morning-glory flower)
--VA usually... 20/200, but can be... 20/20<->NLP
--1/3 develop... serous RD

Optic Nerve Coloboma
1) May resemble... deep cupping
2) Can be... bilateral, asymmetric
3) Part of the... CHARGE association

Optic Pit/Hole
--Associated with... serous RD in adulthood

Optic Nerve Hypoplasia
--Abnormally low number of axons
--DFE: Small pale disc with... double ring sign
--VA... 20/20<->NLP
--VF defects invariably present
--Remember the 4 D's (more on this shortly)

Megalopapilla
1) Abnormally large diameter of... [? and ?] up
2) VF testing may reveal an... enlarged blind spot

Myelinated RNFL
--Myelin normally starts at LGN, ends at lamina cribrosa
--Can be patchy, discontinuous -- Corresponding VF has absolute scotoma

Tilted-Disc Syndrome
1) Superior pole appears... elevated, inferior... recessed
2) Associated with... situs inversus of retinal vessels
3) Fundus abnormality produces... myopic astigmatism
4) VF testing reveals... bitemporal hemianopia that doesn’t... resolve with refraction

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped…excavation
2) Number of vessels crossing the rim seems abnormally…high
--Tissue is…contractile, so cup seems to…open and close (like its namesake, the morning-glory flower)
--VA usually…20/200, but can be…20/20<->NLP
--1/3 develop…serous RD

Optic Nerve Coloboma
1) May resemble…deep cupping
2) Can be…bilateral, asymmetric
3) Part of the…CHARGE association

Optic Pit/Hole
--Associated with…serous RD in adulthood

Optic Nerve Hypoplasia
--Abnormally low number of axons
--DFE: Small pale disc with double ring sign
--VA 20/20<->NLP
--VF defects invariably present
--Remember the 4 D's (more on this shortly)

Megalopapilla
1) Abnormally large diameter of…disc and cup
2) VF testing may reveal an…enlarged blind spot

Myelinated RNFL
--Myelin normally starts at LGN, ends at lamina cribrosa
--Can be patchy, discontinuous--Corresponding VF has absolute scotoma

Tilted-Disc Syndrome
1) Superior pole appears…elevated
2) Inferior…recessed
3) Fundus abnormality produces…myopic astigmatism
4) VF testing reveals…bitemporal hemianopia that doesn’t…respect the vertical and may…resolve with refraction

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped excavation
2) Number of vessels crossing the rim seems abnormally high
--Tissue is contractile, so cup seems to open and close (like its namesake, the morning-glory flower)
--VA usually 20/200, but can be 20/20<->NLP
--1/3 develop serous RD

Optic Nerve Coloboma
1) May resemble deep cupping
2) Can be bilateral, asymmetric
3) Part of the CHARGE association

Optic Pit/Hole
--Associated with serous RD in adulthood

Optic Nerve Hypoplasia
--Abnormally low number of axons
--DFE: Small pale disc with double ring sign
--VA 20/20<->NLP
--VF defects invariably present
--Remember the 4 D's (more on this shortly)

Megalopapilla
1) Abnormally large diameter of disc and cup
2) VF testing may reveal an [specific VF finding]

Myelinated RNFL
--Myelin normally starts at LGN, ends at lamina cribrosa
--Can be patchy, discontinuous
--Corresponding VF has absolute scotoma

Tilted-Disc Syndrome
1) Superior pole appears elevated, inferior recessed
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These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble... **deep cupping**
2) Can be... **bilateral, asymmetric**
3) Part of the... **CHARGE association**

Optic Pit/Hole
--Associated with... **serous RD in adulthood**

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped... **excavation**
2) Number of vessels crossing the rim seems abnormally... **high**
--Tissue is... **contractile**, so cup seems to... **open and close** (like its namesake, the morning-glory flower)
--VA usually... **20/200**, but can be... **20/20<->NLP**
--1/3 develop... **serous RD**

Megalopapilla
1) Abnormally large diameter of... **disc and cup**
2) VF testing may reveal an... **enlarged blind spot**

Myelinated RNFL

Optic Nerve Hypoplasia

Tilted-Disc Syndrome
1) Superior pole appears... **elevated**
2) Inferior... **recessed**
3) Fundus abnormality produces... **myopic astigmatism**
4) VF testing reveals... **bitemporal hemianopia** that doesn’t...
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

An abnormally large cup in a preemie with cerebral palsy is suggestive of what condition?

Megalopapilla
1) Abnormally large diameter of...disc and cup
2) VF testing may reveal an...enlarged blind spot

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

Optic Pit/Hole
--Associated with...serous RD in adulthood

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped...excavation
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--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
--VA usually...20/200, but can be...
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Tilted-Disc Syndrome
1) Superior pole appears...elevated
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Developmental Abnormalities of the Optic Nerve Head

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**Optic Nerve Coloboma**
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

**Optic Pit/Hole**
--Associated with...serous RD in adulthood

**Morning-Glory Disc**
--DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
--VA usually...20/200, but can be...
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--1/3 develop...serous RD

**Megalopapilla**
1) Abnormally large diameter of...disc and cup
2) VF testing may reveal an...enlarged blind spot

**Tilted-Disc Syndrome**
1) Superior pole appears...elevated
2) Inferior...recessed
3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn’t...resolve with refraction

An abnormally large cup in a preemie with cerebral palsy is suggestive of what condition?
Periventricular leukomalacia

An abnormally large cup in a preemie with cerebral palsy is suggestive of what condition? Periventricular leukomalacia

Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

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1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

**Optic Pit/Hole**
--Associated with...serous RD in adulthood

**Morning-Glory Disc**
--DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
--VA usually...20/200, but can be...
  20/20<->NLP
--1/3 develop...serous RD

**Megalopapilla**
1) Abnormally large diameter of...disc and cup
2) VF testing may reveal an...enlarged blind spot

**Tilted-Disc Syndrome**
1) Superior pole appears...elevated
2) Inferior...recessed
3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn’t...resolve with refraction

An abnormally large cup in a preemie with cerebral palsy is suggestive of what condition?
Periventricular leukomalacia

Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

**Optic Nerve Coloboma**
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
3) Part of the...CHARGE association

**Optic Pit/Hole**
--Associated with...serous RD in adulthood

**Morning-Glory Disc**
--DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
--Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
--VA usually...20/200, but can be...
  20/20<->NLP
--1/3 develop...serous RD

**Megalopapilla**
1) Abnormally large diameter of...disc and cup
2) VF testing may reveal an...enlarged blind spot

**Tilted-Disc Syndrome**
1) Superior pole appears...elevated
2) Inferior...recessed
3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn’t...resolve with refraction

An abnormally large cup in a preemie with cerebral palsy is suggestive of what condition?
Periventricular leukomalacia
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

Optic Nerve Coloboma
1) May resemble...deep cupping
2) Can be...bilateral, asymmetric
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What is the causative event, and when does it occur?

Periventricular leukomalacia

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Via retrograde trans-synaptic degeneration of ganglion cell axons

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And it (almost) goes without saying...What condition must be considered in any individual with an enlarged cup?

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**Where does myelination normally begin?**

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Where does myelination normally begin?
At the lateral geniculate nucleus

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- **Where does myelination normally begin?**
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(Note: Because we're talking about the axons of retinal ganglion cells, it's probably better to say that myelination begins at the lamina cribrosa and ends at the LGN.)
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A hypoplastic nerve with double-ring sign can easily be mistaken for what?
A normal sized optic nerve head and cup (the outer edge of the ring is interpreted as the edge of the optic rim)

Tilted Disc Syndrome

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**Morning-Glory Disc**
--DFE reveals:
1) A funnel-shaped...**excavation**
2) Number of vessels crossing the rim seems abnormally...**high**
--Tissue is...**contractile**, so cup seems to...**open and close** (like its namesake, the morning-glory flower)
--VA usually...**20/200**, but can be...
20/20<->NLP
--1/3 develop...**serous RD**

**Megalopapilla**
1) Abnormally large diameter of...**disc and cup**
2) VF testing may reveal an...**enlarged blind spot**

**Myelinated RNFL**
1) Myelin normally ends at the...**lamina cribrosa**
2) Can be...**patchy and discontinuous**
3) Part of the...**CHARGE association**

**Optic Nerve Hypoplasia**
1) Abnormally low number of...**axons**
2) DFE: Small pale disc with...**double ring** sign
3) Corresponding VF has an...**absolute** scotoma

**Tilted-Disc Syndrome**
1) Superior pole appears...**elevated**
2) Associated with...**situs inversus** of retinal vessels
3) Fundus abnormality produces...**myopic astigmatism**
4) VF testing reveals...**bitemporal hemianopia** that doesn't...
   respect the vertical and may...
   resolve with refraction
Developmental Abnormalities of the Optic Nerve Head

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--DFE reveals:
  1) A funnel-shaped excavation
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--Tissue is contractile, so cup seems to open and close (like its namesake, the morning-glory flower)
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Optic Pit/Hole
--Associated with serous RD in adulthood

Optic Nerve Coloboma
1) May resemble deep cupping
2) Can be bilateral, asymmetric
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Optic Nerve Hypoplasia
1) Abnormally low number of axons
2) DFE: Small pale disc with double ring sign
3) VA 20/15<->NLP
4) VF defects invariably present
5) Remember the 4 D’s (more on this shortly)

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Megalopapilla
1) Abnormally large diameter of disc and cup
2) VF testing may reveal an enlarged blind spot

Tilted-Disc Syndrome
1) Superior pole appears elevated, inferior recessed
2) Associated with situs inversus of retinal vessels
3) Fundus abnormality produces myopic astigmatism
4) VF testing reveals bitemporal hemianopia that doesn’t respect the vertical and may resolve with refraction

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

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--Associated with serous RD in adulthood

Optic Nerve Hypoplasia
1) Abnormally low number of axons
2) DFE: Small pale disc with double ring sign
3) VA 20/15<->NLP
4) VF defects...
5) [how likely?]

Megalopapilla
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Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

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5) Remember the...**4 D's** (more on this shortly)

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5) Remember the...**4 D's** (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears...**elevated**
2) Inferior...**recessed**
3) Fundus abnormality produces...**myopic astigmatism**
4) VF testing reveals...**bitemporal hemianopia** that doesn't...with refract
Developmental Abnormalities of the Optic Nerve Head

Tilted-Disc Syndrome
1) Superior pole appears elevated, inferior recessed
2) Associated with situs inversus of retinal vessels
3) Fundus abnormality produces myopic astigmatism
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Optic Pit/Hole
--Associated with serous RD in adulthood

Is the VF loss associated with optic nerve hypoplasia progressive, or nonprogressive?
Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
1) May resemble...deep cupping
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Optic Pit/Hole
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2) DFE: Small pale disc with...double ring sign
3) VA...20/15<->NLP
4) VF defects...invariably present
5) 1/3 develop...serous RD

Is the VF loss associated with optic nerve hypoplasia progressive, or nonprogressive? Nonprogressive
Developmental Abnormalities of the Optic Nerve Head

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--DFE reveals:
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Optic Nerve Hypoplasia
1) Abnormally low number of...axons
2) DFE: Small pale disc with...double ring sign
3) VA...20/15<->NLP
4) VF defects...invariably present
5) Remember the...mnemonic

Tilted-Disc Syndrome
1) Superior pole appears...elevated
2) Associated with...situs inversus of retinal vessels
3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn't...respect the vertical and may...resolve with refraction
5) Myelinated RNFL
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Mnemonic 56
Tilted-Disc Syndrome
Developmental Abnormalities of the Optic Nerve Head

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-- DFE reveals:
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2) Number of vessels crossing the rim seems abnormally... high
-- Tissue is... contractile, so cup seems to... open and close the more... like its namesake, the morning-glory flower
-- VA usually... 20/200, but can be... 20/20<->NLP
-- 1/3 develop... serous RD

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Tilted-Disc Syndrome

You see a child with optic nerve hypoplasia. What should you do for her?

Get MRI brain (various CNS abnormalities are associated with optic-nerve hypoplasia), and refer for endocrine evaluation (multiple endocrine deficiencies are associated, especially pituitary-related)

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Optic Nerve Coloboma
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Tilted-Disc Syndrome
1) Superior pole appears...[elevated v recessed], inferior...[elevated v recessed]
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
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Tilted-Disc Syndrome
1) Superior pole appears...elevated, inferior...recessed
2)
3)
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Developmental Abnormalities of the Optic Nerve Head

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1) Abnormally low number of… axons
2) DFE: Small pale disc with… double ring sign
3) VA… 20/15<->NLP
4) VF defects… invariably present
5) Remember the… 4 D’s (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears… elevated, inferior… recessed
2) Associated with… [two words] of retinal vessels
3)
4)
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped... excavation
2) Number of vessels crossing the rim seems abnormally... high
--Tissue is... contractile, so cup seems to... open and close (like its namesake, the morning-glory flower)
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5) Remember the... 4 D's (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears... elevated, inferior... recessed
2) Associated with... situs inversus of retinal vessels
3) 4)

These three are all secondary to abnormal closure of the embryonic optic fissure
Developmental Abnormalities of the Optic Nerve Head

Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped…excavation
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--Tissue is…contractile, so cup seems to…open and close (like its namesake, the morning-glory flower)
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5) Remember the…4 D’s (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears…elevated, inferior…recessed
2) Associated with…situs inversus of retinal vessels
3) Fundus abnormality produces… [refractive error]
4)

These three are all secondary to abnormal closure of the embryonic optic fissure
Morning-Glory Disc
--DFE reveals:
1) A funnel-shaped…excavation
2) Number of vessels crossing the rim seems abnormally…high
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Tilted-Disc Syndrome
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4) These three are all secondary to abnormal closure of the embryonic optic fissure
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5) Remember the…4 D's (more on this shortly)

Tilted-Disc Syndrome
1) Superior pole appears…elevated, inferior…recessed
2) Associated with…situs inversus of retinal vessels
3) Fundus abnormality produces…myopic astigmatism
4) VF testing reveals…[specific VF finding] that doesn’t respect the vertical, and may resolve with…[simple clinical maneuver]
Developmental Abnormalities of the Optic Nerve Head

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--DFE reveals:
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Tilted-Disc Syndrome
1) Superior pole appears…elevated, inferior…recessed
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3) Fundus abnormality produces…myopic astigmatism
4) VF testing reveals…bitemporal hemianopia that doesn’t respect the vertical, and may resolve with…refraction
Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure

**Optic Nerve Coloboma**
1) May resemble...deep cupping
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1) Superior pole appears...elevated, inferior...recessed
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3) Fundus abnormality produces...myopic astigmatism
4) VF testing reveals...bitemporal hemianopia that doesn't respect the vertical, and may resolve with...refraction

If a patient has tilted discs + difficulty with night vision, what diagnosis should be considered?

Congenital stationary night blindness (CSNB)

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Developmental Abnormalities of the Optic Nerve Head

These three are all secondary to abnormal closure of the embryonic optic fissure:

- **Optic Nerve Coloboma**
  1) May resemble...deep cupping
  2) Can be...bilateral, asymmetric
  3) Part of the...CHARGE association

- **Optic Pit/Hole**
  --Associated with...serous RD in adulthood

- **Morning-Glory Disc**
  --DFE reveals:
  1) A funnel-shaped...excavation
  2) Number of vessels crossing the rim seems abnormally...high
  --Tissue is...contractile, so cup seems to...open and close (like its namesake, the morning-glory flower)
  --VA usually...20/200, but can be...
  20/20<->NLP
  --1/3 develop...serous RD

- **Megalopapilla**
  1) Abnormally large diameter of...disc and cup
  2) VF testing may reveal an...enlarged blind spot

- **Myelinated RNFL**
  1) Myelin normally ends at the...lamina cribrosa
  2) Can be...patchy and discontinuous
  3) Corresponding VF has an...absolute scotoma

- **Optic Nerve Hypoplasia**
  1) Abnormally low number of...axons
  2) DFE: Small pale disc with...double ring sign
  3) VA...
  20/20<->NLP
  4) VF defects...invariably present
  5) Remember the...4 D’s (more on this shortly)

- **Tilted-Disc Syndrome**
  1) Superior pole appears...elevated, inferior...recessed
  2) Associated with...situs inversus of retinal vessels
  3) Fundus abnormality produces...myopic astigmatism
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As for the four D's of optic nerve hypoplasia...

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These three are all secondary to abnormal closure of the embryonic optic fissure
What are the **4 D’s** of optic nerve hypoplasia?

-- **D**

-- **D**

-- **D**

*Hints forthcoming…*

-- **D**

-- **D**
What are the 4 D’s of optic nerve hypoplasia?

- **D**: Drink (heavy EtOH consumption during pregnancy)
- **D**: Diabetes
- **D**: Drugs (especially Dilantin or other seizure meds)
- **D**: De Morsier syndrome

Concern mom’s life while she is pregnant w/ the child who will have ON hypoplasia

Hints forthcoming…

A congenital condition with significant CNS findings
Developmental Abnormalities of the Optic Nerve Head

What are the **4 D’s** of optic nerve hypoplasia?

- **Drink** (ie, heavy EtOH consumption during pregnancy)
- **Diabetes**
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- **De Morsier syndrome**

(5 D’s if you count this one)

(should be lower-case, but it looked funny)
What are the **4 D’s** of optic nerve hypoplasia?

--**Drink** (ie, heavy EtOH consumption during pregnancy)

In other words, optic-nerve hypoplasia is part of the [fetal alcohol syndrome](https://en.wikipedia.org/wiki/Fetal_alcohol_syndrome)

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What triad constitutes de Morsier syndrome?
Developmental Abnormalities of the Optic Nerve Head

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What triad constitutes de Morsier syndrome?

--Optic nerve hypoplasia (duh)

--Absence of the

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What is the noneponymous name of de Morsier syndrome?

Septo-optic dysplasia
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What triad constitutes de Morsier syndrome?

--Optic nerve hypoplasia (duh)
--Absence of the septum pellucidum
--Agenesis of the corpus callosum
--Pituitary dwarfism

Note: The listed triad is from the BCSC Peds book. Per the Neuro book, the triad includes pituitary dwarfism, not corpus callosum agenesis (although it states that “the corpus callosum may be thinned or absent”)

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Of the four, which is the most common cause of optic nerve hypoplasia?

Maternal diabetes is notorious for causing a specific pattern of optic nerve hypoplasia--what is it?

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**invariably present**

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There is another optic-nerve condition that presents with nonprogressive bitemporal **superior** loss that doesn’t respect the vertical midline. What is it?

**Fuch’s coloboma**
Developmental Abnormalities of the Optic Nerve Head

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Pituitary adenoma compressing the optic chiasm causes bitemporal superior VF. Would that be an acceptable alternative answer?

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Not really, for two reasons:
--As alluded to in the question, it is an optic chiasm condition, not an optic nerve condition; and
--VF loss secondary to chiasmal compression is progressive--as the tumor enlarges, so too do the VF cuts

There is another optic-nerve condition that presents with nonprogressive bitemporal superior loss that doesn't respect the vertical midline. What is it?

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What is the basic pathology in Fuch’s coloboma?
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OK, but why on earth do these pts get a VF defect?

It's actually pretty simple. As indicated below, these eyes are staphyloma-ish inferonasally. Thus, the ‘axial length’ inferonasally is longer than it is in other regions. Because of this extra axial length, the refractive correction used during the performance of a visual-field test—a correction based on the non-staphylomatous fovea—is not myopic enough for the inferonasal retina. The subsequent uncorrected refractive error produces a refractive scotoma in the superotemporal VF.

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OK, but why on earth do these pts get a VF defect?

It's actually pretty simple. As indicated below, these eyes are staphyloma-ish inferonasally. Thus, the 'axial length' inferonasally is longer than it is in other regions. Because of this extra axial length, the refractive correction used during the performance of a visual-field test--a correction based on the non-staphylomatous fovea--is not myopic enough for the inferonasal retina. The subsequent uncorrected refractive error produces a refractive scotoma in the superotemporal VF.

This implies that the VF defect will resolve if the 'proper' refractive correction is employed. Does it?

What pattern of VF loss is associated with SSONH? Bitemporal superior loss that doesn’t respect the vertical midline

There is another optic-nerve condition that presents with nonprogressive bitemporal superior loss that doesn’t respect the vertical midline. What is it? Fuch's coloboma

What is the basic pathology in Fuch's coloboma?
The optic nerve head is tilted such that it is deeply depressed inferonasally--almost staphyloma-ish
What are the 4 D's of optic nerve hypoplasia?

- Drink (ie, heavy EtOH consumption during pregnancy)
- Diabetes
- Drugs (especially Dilantin or other seizure meds)
- De Morsier syndrome

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