Q

most vs least

common pediatric space-occupying lesion of orbit

Orbital Dermoid Cysts
• **Most** common pediatric space-occupying lesion of orbit

*Orbital Dermoid Cysts*
Q

- **Most** common pediatric space-occupying lesion of orbit
- Arise from \text{non-specific histo term} trapped in \text{two words}

\textit{Orbital Dermoid Cysts}
 Orbital Dermoid Cysts

- **Most** common pediatric space-occupying lesion of orbit
- Arise from **epithelial nests** trapped in **bony sutures**
Q

**Orbital Dermoid Cysts**

- Most common pediatric space-occupying lesion of orbit
- Arise from **epithelial nests trapped in bony sutures**

*During what general developmental period does this occur?*
Orbital Dermoid Cysts

- Most common pediatric space-occupying lesion of orbit
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During what general developmental period does this occur?
Embryogenesis
Q

orbital dermoid cysts

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During what general developmental period does this occur?
Embryogenesis

Does this mean dermoid cysts are congenital lesions?
Most common pediatric space-occupying lesion of orbit
Arise from **epithelial nests** trapped in **bony sutures**

**Orbital Dermoid Cysts**

*During what general developmental period does this occur?*
Embryogenesis

*Does this mean dermoid cysts are congenital lesions?*
Yes and no
Orbital Dermoid Cysts

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- Arise from *epithelial nests* trapped in *bony sutures*

*During what general developmental period does this occur?*  
Embryogenesis

*Does this mean dermoid cysts are congenital lesions?*  
*Yes and no*  
*C’mon brah--either they’re congenital, or they’re not. What’s up with this?*
**Orbital Dermoid Cysts**

- Most common pediatric space-occupying lesion of orbit
- Arise from **epithelial nests trapped in bony sutures**

*During what general developmental period does this occur?*
Embryogenesis

*Does this mean dermoid cysts are congenital lesions?*
Yes and no

*C’mon brah--either they’re congenital, or they’re not. What’s up with this?*
They’re congenital in the sense that they’re always present at birth. However, depending upon their location, they aren’t always clinically apparent at birth--and may not become so until adulthood.*
**Orbital Dermoid Cysts**

- **Most** common pediatric space-occupying lesion of orbit
- Arise from **epithelial nests** trapped in **bony sutures**
- Can be located **not nasal/temporal** (most common) or **ditto**
**Orbital Dermoid Cysts**

- Most common pediatric space-occupying lesion of orbit
- Arise from **epithelial nests** trapped in **bony sutures**
- Can be located **pre-septal** (most common) or **deep**
Orbital Dermoid Cysts

- Most common pediatric space-occupying lesion of orbit
- Arise from epithelial nests trapped in bony sutures
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**Orbital Dermoid Cysts**
Most common pediatric space-occupying lesion of orbit

Arise from epithelial nests trapped in bony sutures

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Presentation: Palpable mass that is painless, smooth and mobile
A

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*Can they be affixed to the underlying periosteum?*
Orbital Dermoid Cysts

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Can they be affixed to the underlying periosteum?
Yes, and this may render them immobile
**Orbital Dermoid Cysts**

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*Can they be affixed to the underlying periosteum?*
Yes, and this may render them immobile

*Can they be affixed to the overlying skin?*
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**Can they be affixed to the underlying periosteum?**
Yes, and this may render them immobile

**Can they be affixed to the overlying skin?**
Only if the cyst has partially ruptured, leading to scarring
Most common pediatric space-occupying lesion of orbit
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What about cysts located deep in the orbit--how do they present?
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What about cysts located deep in the orbit—how do they present?
With progressive proptosis +/- off-axial globe displacement
**Orbital Dermoid Cysts**

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- Can be located pre-septal (most common) or deep
- Pre-septal cysts typically superotemporal >> superonasal
- Presentation: Palpable mass that is painless, smooth and mobile of dermoid cysts located deep in the orbit

*What about cysts located deep in the orbit--how do they present?*
With progressive proptosis +/- off-axial globe displacement

*At what age does this occur?*
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What about cysts located deep in the orbit--how do they present?
With progressive proptosis +/- off-axial globe displacement

At what age does this occur?
Adulthood
**Orbital Dermoid Cysts**

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- Presentation: Palpable mass that is **painless**, **smooth** and **mobile**
  - Natural history: **progressive enlargement vs spontaneous involution**
**Orbital Dermoid Cysts**

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  - Natural history: **Progressive enlargement**
- CT finding: **well vs poorly** circumscribed lesion with **low vs high density lumen**
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Is it necessary to CT suspected dermoid cysts?
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*Is it necessary to CT suspected dermoid cysts?*
In most cases, no--the dx can be made clinically
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- CT finding: **Well** circumscribed lesion with **low** density lumen
- Histology:
  - Cyst lined with **epithelium**
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*What three things constitute dermal appendages/adnexal structures?*

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**Q/A**

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*What three things constitute dermal appendages/adnexal structures?*

- Hair
- Sebaceous glands
- Sweat glands
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A closely-related clinical entity presents similarly to dermoid cysts, but has a somewhat different histology--it’s lined with nonkeratinized squamous epithelium, has no dermal appendages, and contains no oil. What is this entity called?
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A closely-related clinical entity presents similarly to dermoid cysts, but has a somewhat different histology--it’s lined with nonkeratinized squamous epithelium, has no dermal appendages, and contains no oil. What is this entity called?

Epidermoid cyst (aka simple epithelial cyst)
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- Histology:
  - Cyst lined with **keratinized** epithelium
  - Dermal appendages (aka *adnexal structures*) are present
  - Cyst filled with **oil** and **keratin**
- Management: **conservative vs surgical**
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- Management: Excision
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- Management: Excision

At what age should excision be performed?

Opinions differ, but many authorities would recommend ~1 year

Because at around this age, newly-mobile toddlers are at increased risk of cyst rupture from accidental trauma (eg, from falling and bonking their face)
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- Management: **Excision**

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**Why this age?**
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- **Management:** Excision

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- **Management:** **Excision**
  - Effort should be made to remove the cyst without
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- Management: **Excision**
  - Effort should be made to remove the cyst without spilling its contents
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Why is this important?
Because if released into the orbit and/or subQ space, the cyst's contents will provoke a strong inflammatory response, with subsequent scarring.
Orbital Dermoid Cysts

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**Orbital Dermoid Cysts**

= epibulbar dermoid?

BTW: Are epibulbar dermoids related to dermoid cysts?
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Orbital Dermoid Cysts
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Orbital Dermoid Cysts

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**BTW: Are epibulbar dermoids related to dermoid cysts?**
No, they are completely unrelated entities

**Where are epibulbar dermoids located?**
On the ocular surface

- At the limbus

**With what syndrome are epibulbar dermoids associated?**
Goldenhar

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On the ocular surface

Where on the ocular surface are they typically located?
On the limbus

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CT finding: Well circumscribed lesion with low density lumen
Histology:
- Cyst lined with keratinized epithelium
- Dermal appendages (aka adnexal structures) are present
- Cyst filled with oil and keratin
Management: Excision
Effort should be made to remove the cyst without spilling its contents
Orbital Dermoid Cysts

= epibulbar dermoid? No

Most common pediatric space-occupying lesion of orbit
Arise from epithelial nests trapped in bony sutures
Can be located pre-septal (most common) or deep
Pre-septal cysts typically superotemporal or superonasal
Presentation: Palpable mass that is painless, smooth, and mobile
Natural history: Progressive enlargement
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- Cyst lined with keratinized epithelium
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- Cyst filled with oil and keratin
Management: Excision
- Effort should be made to remove the cyst without spilling its contents

BTW: Are epibulbar dermoids related to dermoid cysts? No, they are completely unrelated entities

Where are epibulbar dermoids located?
On the ocular surface

Where on the ocular surface are they typically located?
At the limbus

By what other name are epibulbar dermoids commonly known?
Limbal dermoids

Orbital Dermoid Cysts = epibulbar dermoid? No
A

Most common pediatric space-occupying lesion of orbit

Arise from epithelial nests trapped in bony sutures

Can be located pre-septal (most common) or deep

Pre-septal cysts typically superotemporal or superonasal

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Orbital Dermoid Cysts

= epibulbar dermoid? No

BTW: Are epibulbar dermoids related to dermoid cysts? No, they are completely unrelated entities

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Where on the ocular surface are they typically located? At the limbus

By what other name are epibulbar dermoids commonly known? Limbal dermoids
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  - Cyst lined with keratinized epithelium
  - Dermal appendages (aka adnexal structures) are present
  - Cyst filled with oil and keratin
- Management: Excision
  - Effort should be made to remove the cyst without spilling its contents

**Early Dermoid Cysts**

- Epibulbar dermoid?
  - No

**BTW: Are epibulbar dermoids related to dermoid cysts?**
  - No, they are completely unrelated entities

**Where are epibulbar dermoids located?**
  - On the ocular surface

**Where on the ocular surface are they typically located?**
  - At the limbus

**With what syndrome are epibulbar dermoids associated?**
  - Goldenhar
Most common pediatric space-occupying lesion of orbit
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**Orbital Dermoid Cysts**

= epibulbar dermoid? No

**BTW: Are epibulbar dermoids related to dermoid cysts?**
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**Where are epibulbar dermoids located?**
On the ocular surface

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**With what syndrome are epibulbar dermoids associated?**
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Most common pediatric space-occupying lesion of orbit

- Arise from epithelial nests trapped in bony sutures
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---

**Orbital Dermoid Cysts**

= epibulbar dermoid? *No*

BTW: Are epibulbar dermoids related to dermoid cysts?
No, they are completely unrelated entities

Where are epibulbar dermoids located?
On the ocular surface

Where on the ocular surface are they typically located?
At the limbus

With what syndrome are epibulbar dermoids associated?
**Goldenhar**

In two words, what sort of condition is Goldenhar?
It's a **craniofacial malformation**

Dermal appendages (aka adnexal structures) are present

Cyst filled with oil and keratin
**Orbital Dermoid Cysts**

- Most common pediatric space-occupying lesion of orbit
- Arise from epithelial nests trapped in bony sutures
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- Management: Excision
  - Effort should be made to remove the cyst without spilling its contents

---

**Q/A**

- **= epibulbar dermoid? No**
- **BTW: Are epibulbar dermoids related to dermoid cysts?**
  - No, they are completely unrelated entities
- **Where are epibulbar dermoids located?**
  - On the ocular surface
- **Where on the ocular surface are they typically located?**
  - At the limbus
- **With what syndrome are epibulbar dermoids associated?**
  - Goldenhar
- **In two words, what sort of condition is Goldenhar?**
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Most common pediatric space-occupying lesion of orbit
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**Orbital Dermoid Cysts**

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No, they are completely unrelated entities

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Where on the ocular surface are they typically located?  
At the limbus

With what syndrome are epibulbar dermoids associated?  
**Goldenhar**

In two words, what sort of condition is Goldenhar?  
It’s a craniofacial malformation

Epibulbar dermoids are completely unrelated to dermoid cysts. They occur on the ocular surface, typically at the limbus, and are associated with the Goldenhar syndrome, which is a craniofacial malformation.
**Orbital Dermoid Cysts**

= epibulbar dermoid? No

- Most common pediatric space-occupying lesion of orbit
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- Management: Excision
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**BTW: Are epibulbar dermoids related to dermoid cysts?**
No, they are completely unrelated entities

**Where are epibulbar dermoids located?**
On the ocular surface

**Where on the ocular surface are they typically located?**
At the limbus

**With what syndrome are epibulbar dermoids associated?**
Goldenhar

In two words, what sort of condition is Goldenhar?
It's a craniofacial malformation

Dermal appendages (aka adnexal structures) are present

Craniofacial malformations are divided into two categories based on etiology—what are they?
Most common pediatric space-occupying lesion of orbit

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**Orbital Dermoid Cysts**

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Where are epibulbar dermoids located?
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Where on the ocular surface are they typically located?
At the limbus

With what syndrome are epibulbar dermoids associated?
Goldenhar

In two words, what sort of condition is Goldenhar?
It's a craniofacial malformation

Craniofacial malformations are divided into two categories based on etiology—what are they?
Those due to , and those not due to .
Orbital Dermoid Cysts

= epibulbar dermoid? No

BTW: Are epibulbar dermoids related to dermoid cysts?
No, they are completely unrelated entities

Where are epibulbar dermoids located?
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In two words, what sort of condition is Goldenhar?
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Craniofacial malformations are divided into two categories based on etiology—what are they?
Those due to craniosynostosis, and those not due to craniosynostosis
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**Orbital Dermoid Cysts**

*epibulbar dermoid? No*

**BTW:** Are epibulbar dermoids related to dermoid cysts?
No, they are completely unrelated entities

**Where are epibulbar dermoids located?**
On the ocular surface

**Where on the ocular surface are they typically located?**
At the limbus

**With what syndrome are epibulbar dermoids associated?**
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*In two words, what sort of condition is Goldenhar?*
It's a craniofacial malformation

*Craniofacial malformations are divided into two categories based on etiology—what are they?*
Those due to craniosynostosis, and those not due to craniosynostosis

*To what process does craniosynostosis refer?*
**Orbital Dermoid Cysts**

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Dermal appendages (aka adnexal structures) are present

Craniofacial malformations are divided into two categories based on etiology—what are they?
Those due to craniosynostosis, and those not due to craniosynostosis

To what process does craniosynostosis refer?
To the premature closing of the bony sutures of the cranium
Most common pediatric space-occupying lesion of orbit

- Arise from epithelial nests trapped in bony sutures
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- No, they are completely unrelated entities

Where are epibulbar dermoids located?
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Where on the ocular surface are they typically located?
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With what syndrome are epibulbar dermoids associated?

Goldenhar

In two words, what sort of condition is Goldenhar?
- It's a craniofacial malformation

Dermal appendages (aka adnexal structures) are present

Craniofacial malformations are divided into two categories based on etiology—what are they?
- Those due to craniosynostosis, and those not due to craniosynostosis

To which category does Goldenhar belong?
A Orbital Dermoid Cysts

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BTW: Are epibulbar dermoids related to dermoid cysts?
No, they are completely unrelated entities

Where are epibulbar dermoids located?
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Where on the ocular surface are they typically located?
At the limbus

With what syndrome are epibulbar dermoids associated?
Goldenhar

In two words, what sort of condition is Goldenhar?
It's a craniofacial malformation

Craniofacial malformations are divided into two categories based on etiology—what are they?
Those due to craniosynostosis, and those not due to craniosynostosis

To which category does Goldenhar belong?
It is of the non-craniosynostosis sort
OK, confession time. To my frustration, I discovered that I inadvertently revised this topic on separate occasions, and thus am ‘stuck’ with two reviews of the same subject. That said, the two differ in a number of ways—so much so it’s worth keeping both. The second, same-but-different review commences with the next slide.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma
Orbital Dermoid Cysts

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

**choristoma**

What is a choristoma?
**Q/A**

**Orbital Dermoid Cysts**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A **choristoma**

*What is a choristoma?*

A tumor composed of histologically normal vs abnormal cells found in an abnormal location
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

What is a choristoma?
A tumor composed of histologically normal cells found in an abnormal location
What is a choristoma?
A tumor composed of histologically normal cells found in an abnormal location.

What is the name of the reverse clinical entity, i.e., one with abnormal cells found in their normal location?

A hamartoma
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma? A choristoma is a tumor composed of histologically normal cells found in an abnormal location.

What is the name of the reverse clinical entity, i.e., one with abnormal cells found in their normal location? A hamartoma.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma?

A choristoma is a tumor composed of histologically normal cells found in an abnormal location.

What is the name of the reverse clinical entity, ie, one with abnormal cells found in their normal location?

A hamartoma

What is a hamartoma?

A lesion that is a choristoma (or hamartoma) indicates what about its onset?

That, by definition, it is congenital and benign.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma?

A choristoma is a tumor composed of histologically normal cells found in an abnormal location.

What is the name of the reverse clinical entity, i.e., one with abnormal cells found in their normal location?

A hamartoma

That a lesion is a choristoma (or hamartoma) indicates what about its onset?

That, by definition, it is…[congenital vs acquired]
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma?

A choristoma is a tumor composed of histologically normal cells found in an abnormal location.

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A lesion with abnormal cells found in their normal location.

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It is congenital.

Are all choristomas/hamartomas congenital?
Yes.

Are all congenital lesions choristomas/hamartomas?
No.
What is a choristoma?
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A tumor composed of histologically normal cells found in an abnormal location.

That a lesion is a choristoma (or hamartoma) indicates what about its onset?
It is... congenital.

Are all choristomas/hamartomas congenital?
Yes.

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is an orbital dermoid cyst?
 choristoma
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What is a choristoma?

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That a lesion is a choristoma (or hamartoma) indicates what about its onset?

That, by definition, it is...congenital

Are all choristomas/hamartomas congenital?

Yes

Are all congenital lesions choristomas/hamartomas?

No
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is

A choristoma

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That a lesion is a choristoma (or hamartoma) indicates what about its onset? It is congenital.

Are all choristomas/hamartomas congenital? Yes

Are all congenital lesions choristomas/hamartomas? No.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma?

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What is a hamartoma?

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That a lesion is a choristoma (or hamartoma) indicates what about its onset?

That, by definition, it is… congenital

That a lesion is a choristoma (or hamartoma) indicates what about its status vis a vis malignancy?

That a lesion is a choristoma (or hamartoma) indicates that it is… benign
What is a choristoma?
A tumor composed of histologically normal cells found in an abnormal location.

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That a lesion is a choristoma (or hamartoma) indicates what about its onset?
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That, by definition, it is... [benign vs malignant]
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a choristoma?

A choristoma

What is a choristoma?

A tumor composed of histologically normal cells found in an abnormal location.

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That, by definition, it is... benign
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
#1
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
#1

What embryologic event results in the eventual development of a dermoid cyst?
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
#1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
#1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?
Epidermoid cysts
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
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What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?
**Epidermoid cysts** *aka...*

*The BCSC Path book uses a different name for this lesion—what is it?*
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?
#1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?
Epidermoid cysts aka... simple epithelial cyst

The BCSC Path book uses a different name for this lesion—what is it?
Simple epithelial cyst
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
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What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

Epidermoid cysts vs Dermoid cysts

<table>
<thead>
<tr>
<th>Epidermoid cysts</th>
<th>vs</th>
<th>Dermoid cysts</th>
</tr>
</thead>
<tbody>
<tr>
<td>?</td>
<td></td>
<td>Yes</td>
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</table>

Is a choristoma?
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition? Epidermoid cysts vs Dermoid cysts

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<th>Dermoid cysts</th>
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<tbody>
<tr>
<td>Is a choristoma?</td>
<td></td>
<td></td>
<td></td>
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Yes

Yes
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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<td>Is a choristoma?</td>
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<tr>
<td>?</td>
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<td>Contains dermal appendages?</td>
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---

**Orbital Dermoid Cysts**
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?
A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst?
The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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What three ‘dermal appendages’ being referred to here?

Contains dermal appendages?
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What three ‘dermal appendages’ being referred to here?
--Hair follicles
--Sebaceous glands
--Sweat glands

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<td>Keratin</td>
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Can epi/dermoid cysts appear in the nasal orbit?

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Can epi/dermoid cysts appear in the nasal orbit?
Yes, but it is uncommon and unexpected
**Orbital Dermoid Cysts**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma.

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The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?

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The differential for a preseptal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it? It's whether the mass is above or below the **canthus**.
In very general terms (think in terms of the *Path* and/or *Fundamentals* book), what sort of lesion is a dermoid cyst? A choristoma.

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

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### Differential for a preseptal mass in the nasal orbit of an infant

**The differential for a preseptal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?** It’s whether the mass is above or below the medial canthus.

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What is the primary condition of concern for a mass **above** the nasal canthus in an infant?

A meningocele (or encephalocele, or meningoencephalocele)

What is the primary condition of concern for a mass **below** the nasal canthus in an infant?

A meningocele (or encephalocele, or meningoencephalocele)

A preseptal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it? It’s whether the mass is above or below the medial canthus.
Orbital Dermoid Cysts

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma.


What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures.

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition? Epi-dermoid cysts.

In general terms, dermoid/epidermoid cysts presents in one of two manners. We'll get to what the two presentations are shortly, but first: What factor determines the manner in which it presents? The lesion’s location.

A pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it? It’s whether the mass is above or below the medial canthus.

What is the primary condition of concern for a mass above the nasal canthus in an infant? A meningocele (or encephalocele, or meningoencephalocele).

What is the primary condition of concern for a mass below the nasal canthus in an infant? Meningocele, etc.

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In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

- A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

- #1

What embryologic event results in the eventual development of a dermoid cyst?

- The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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In general terms, dermoid/epidermoid cysts presents in one of two manners. We’ll get to what the two presentations are shortly, but first: What factor determines the manner in which it presents?

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The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?

- It’s whether the mass is above or below the medial canthus

What two conditions top the DDx for a mass below the nasal canthus in an infant?

- Congenital nasolacrimal duct obstruction (CNLDO), and dacryocele

In general terms, dermoid/epidermoid cysts presents in one of two manners. We’ll get to what the two presentations are shortly, but first: What factor determines the manner in which it presents?

- The lesion’s location
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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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The lesion’s location

A pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?

It’s whether the mass is above or below the medial canthus

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The differential for a mass below the nasal canthus in an infant?

Congenital nasolacrimal duct obstruction (CNLDO), and dacryocele

What two conditions top the DDx for a mass below the nasal canthus in an infant?

Congenital nasolacrimal duct obstruction (CNLDO), and dacryocele

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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

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The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it? It’s whether the mass is above or below the medial canthus.

In a nutshell, what is the pathophysiology underlying CNLDO? Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

What is the classic sign of CNLDO? The presence of epiphora with sticky mucopurulent discharge. Further, the lacrimal sac often swells, resulting in an erythematous mass below the nasal canthus.

How is CNLDO managed? Initially conservatively, with lacrimal-sac massage + topical abx-steroid drop; if this fails, with probing at around age 1 yr.
**Q/A**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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The lesion’s location

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**CNLDO**

In a nutshell, what is the pathophysiology underlying CNLDO?

Obstruction at the proximal vs distal end of the nasolacrimal duct leads to stasis of fluid within the nasal sac, which subsequently becomes infected.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma.


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**In a nutshell, what is the pathophysiology underlying CNLDO?** Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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- The lesion’s location

Where does it present?

- Nasal vs temporal aspect of orbit

  - Nasal
  - Temporal

Age it becomes clinically apparent

- Late infancy/early toddlerhood
- Adult

What is the classic sign of CNLDO?

- The presence of epiphora with sticky mucopurulent discharge. Further, the lacrimal sac often swells, resulting in an erythematous mass below the nasal canthus.

How is CNLDO managed?

- Initially conservatively, with lacrimal-sac massage + topical abx-steroid drop; if this fails, with probing at around age 1 yr

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Where does dermoid cyst typically present?

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In a nutshell, what is the pathophysiology underlying CNLDO? Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

What is the classic sign of CNLDO? The presence of epiphora with sticky mucopurulent discharge. Further, the lacrimal sac often swells, resulting in an erythematous mass below the nasal canthus.

What blinding condition must always, always, ALWAYS be ruled out in an infant with epiphora? Glaucoma. Epiphora is part of the 'classic triad' in the presentation of congenital glaucoma.
**Orbital Dermoid Cysts**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

- A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

- #1

What embryologic event results in the eventual development of a dermoid cyst?

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A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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**In a nutshell, what is the pathophysiology underlying CNLDO?**

Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

**What is the classic sign of CNLDO?**

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**What blinding condition must always, always, ALWAYS be ruled out in an infant with epiphora?**

- Glaucoma. Epiphora is part of the ‘classic triad’ in the presentation of congenital glaucoma.

**What is CNLDO?**

- Congenital nasolacrimal duct obstruction (CNLDO)

- CNLDO is typically not a primary problem, but rather an indication of underlying disease processes.

- Epiphora is the classic sign.

- The lacrimal sac becomes infected with pus.

- The lacrimal sac will often swell, resulting in a mass below the nasal canthus.

- Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

- Epiphora is part of the ‘classic triad’ in the presentation of congenital glaucoma.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

#1

What embryologic event results in the eventual development of a dermoid cyst?

- The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

- Epi-dermoid cysts

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In a nutshell, what is the pathophysiology underlying CNLDO?

Obstruction at the distal end of the nasolacrimal duct leads to stasis of fluid within the lacrimal sac, which subsequently becomes infected.

What is the classic sign of CNLDO?

- The presence of epiphora, with sticky mucopurulent discharge. Further, the lacrimal sac often swells, resulting in an erythematous mass below the nasal canthus.

What blinding condition must always, always, ALWAYS be ruled out in an infant with epiphora?

- Glaucoma. Epiphora is part of the ‘classic triad’ in the presentation of congenital glaucoma.

What are the other two findings in the classic triad?

- Epiphora

---

In general terms, dermoid/epidermoid cysts presents in one of two manners.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

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Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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Epidermoid cyst

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The lesion's location

In a nutshell, what is the pathophysiology underlying CNLDO?

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Epi-dermoid cyst

In general terms, dermoid/epidermoid cysts presents in one of two manners. We'll get to what the two presentations are shortly, but first: What factor determines the manner in which it presents?

The lesion’s location

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Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO?

CNLDO

Pre-septal mass in nasal orbit

Above canthus

Anatomic issue: Is the mass...

Below canthus

Meningocele

Dacryocele

Dacryocele

At birth, with a bluish swelling below the nasal canthus

Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO?

No. In fact, they generally don't have much epiphora at all.

What is the key anatomic difference between CNLDO and dacryocele?

CNLDO involves obstruction at only one site ('below' the lacrimal sac), whereas a dacryocele involves obstruction at two— one below the lacrimal sac (as in CNLDO), but also one 'above' it (usually at the common canaliculus)
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

#1

What embryologic event results in the eventual development of a dermoid cyst?

The sequestration of dermoid elements within bony sutures

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The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?

It's whether the mass is above or below the medial canthus

Above canthus

Meningocele

Below canthus

Anatomic issue: Is the mass…

CNLDO

Dacryocele

Epidermoid cysts

In nasal orbit

Dacryocele

CNLDO

Meningocele

Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO?

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When and how does dacryocele present?

At birth, with a bluish swelling below the nasal canthus

What is the key anatomic difference between CNLDO and dacryocele?

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For more about dacryoceles, see slide-set O8
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Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO?

No. In fact, they generally don't have much epiphora at all.

When and how does dacryocèle present?

At birth, with a classic swelling below the nasal canthus.
**Orbital Dermoid Cysts**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition? Epi-dermoid cysts

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Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO? No. In fact, they generally don’t have much epiphora at all.

When and how does dacryoccele present? At birth, with a bluish swelling below the nasal canthus

A closely-related entity, differing only with regard to histology, presents in a manner identical to that of dermoid cysts. What is this condition? Epi-dermoid cysts

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures

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In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids? #1

What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition? Epi-dermoid cysts

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Do dacryoceles present with sticky, mucopurulent epiphora a la congenital NLDO? No. In fact, they generally don’t have much epiphora at all.

When and how does dacryoceles present? At birth, with a bluish swelling below the nasal canthus

What is the key anatomic difference between CNLDO and dacryoceles?

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Clinically apparent toddlerhood

Epidermoid cysts present above the medial canthus (i.e., above the orbital septum) as post-septal masses. Dacryoceles present below the medial canthus (i.e., below the orbital septum) as pre-septal masses.

**Anatomic issue:** Is the mass...

- **Above canthus:** Meningocele
- **Below canthus:** Dacryoceles

**CNLDO**

**Dacryocele**

**Dacryocele**

**Meningocele**

**CNLDO**
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

#1

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The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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No. In fact, they generally don’t have much epiphora at all.

When and how does dacryocoele present?

At birth, with a bluish swelling below the nasal canthus

What is the key anatomic difference between CNLDO and dacryoceles?

CNLDO involves obstruction at only one site (‘below’ the lacrimal sac), whereas a dacryocoele involves obstruction at two—one below the lacrimal sac (as in CNLDO), but also one ‘above’ it (usually at the common canaliculus)
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A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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When and how does dacryocle present?

At birth, with a bluish swelling below the nasal canthus

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Epi dermoid cysts

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The lesion’s location

In a nutshell, what is involved with the various -oceles?

A herniation of CNS contents through a bony defect

What’s up with the various names?

They reflect the contents of the herniation. If it's meninges only, it's called a meningocele. If it's brain tissue, it's an encephalocele. If both are present, it's a meningoencephalocele.

In addition to its relation to the medial canthus, what other exam finding should alert you to the possibility that one of these 'oceles' is present?

If the mass is pulsatile.
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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Anatomic issue:

Is the mass…

Above canthus

Meningocele

Below canthus

CNLDO

Dacryocele

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If the mass is pulsatile...
Q

Orbital Dermoid Cysts

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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Mass in the nasal orbit of an infant... An anatomic factor—what is it?

If the mass is... Slowly... progressive proptosis

Painful? No

The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it?

It’s whether the mass is above or below the medial canthus

Above canthus

Below canthus

Meningocele

CNLDO

Dacryocele

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In addition to its relation to the medial canthus, what other exam finding should alert you to the possibility that one of these 'oceles' is present?

If the mass is... Pulsatile
In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst? A choristoma.


What embryologic event results in the eventual development of a dermoid cyst? The sequestration of dermoid elements within bony sutures.

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In a nutshell, what is involved with the various -oceles? A herniation of CNS contents through a bony defect.

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In general terms, pre-septal mass in the nasal orbit of an infant should alert you to another anatomic factor—what is it? The mass is below the medial canthus.

Anatomic issue: Is the mass above or below the medial canthus?

<table>
<thead>
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<th>Anatomic issue: Is the mass...</th>
<th>No</th>
<th>Yes</th>
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The differential for a pre-septal mass in the nasal orbit of an infant is influenced by another anatomic factor—what is it? It’s whether the mass is above or below the medial canthus.

What’s the exam finding that should alert you to the possibility that one of these ‘oceles’ is present? If the mass is pulsatile.

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In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

#1

What embryologic event results in the eventual development of a dermoid cyst?

The sequestration of dermoid elements within bony sutures

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

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In a nutshell, what is involved with the various -oceles? A herniation of CNS contents through a bony defect

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In addition to its relation to the medial canthus, what other exam finding should alert you to the possibility that one of these ‘oceles’ is present?

If the mass is pulsatile
Q/A

Orbital Dermoid Cysts

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

A choristoma

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

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What embryologic event results in the eventual development of a dermoid cyst?

The sequestration of dermoid elements within bony sutures

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The lesion’s location

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<th>Proptosis</th>
<th>Painful?</th>
</tr>
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<tbody>
<tr>
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<td>No</td>
<td>Slowly…</td>
<td>No</td>
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<td>Meningocele</td>
<td>Progressive proptosis</td>
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<td>Location</td>
<td>Pre-septal</td>
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<td>---------------</td>
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<tr>
<td>Nasal vs Temporal</td>
<td>Nasal</td>
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<tr>
<td>Adult</td>
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<tr>
<td>Progressive Proptosis</td>
<td>Slowly...</td>
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<td>No</td>
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**Orbital Dermoid Cysts**

In very general terms (think in terms of the Path and/or Fundamentals book), what sort of lesion is a dermoid cyst?

**A choristoma**

Where does dermoid cyst rank as a cause of space-occupying orbital lesions in kids?

**#1**

What embryologic event results in the eventual development of a dermoid cyst?

**The sequestration of dermoid elements within bony sutures**

A closely-related condition presents in a manner identical to that of dermoid cysts, differing only with regard to histology. What is this condition?

**Epidermoid Cyst**

In general terms, dermoid/epidermoid cysts presents in one of two manners. We'll get to what the two presentations are shortly, but first: What factor determines the manner in which it presents?

**The lesion's location**

If the mass is pre-septal?

If it is above the medial canthus, it's a **meningoencephalocele**.

If the mass is below the medial canthus?

If both are present, it's a **meningocele**. If it's brain tissue, it's an **encephalocele**.

In addition to its relation to the medial canthus, what other exam finding should alert you to the possibility that one of these 'oceles' is present?

If the mass is pulsatile?

In a nutshell, what is involved with the various 'oceles'?

A herniation of CNS contents through a bony defect...
Dermoid cysts: Management

- Depends upon whether the cyst in question is pre-septal vs post-septal
Dermoid cysts: Management

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Dermoid cysts: Management

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  - *Pre-septal*: Surgical excision…
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Dermoid cysts: Management

- Depends upon whether the cyst in question is **pre-septal** vs **post-septal**
  - *Pre-septal*: Surgical excision... **around age 1 year**
  - *Post-septal*: Surgical excision...
Dermoid cysts: Management

- Depends upon whether the cyst in question is pre-septal vs post-septal
  
  **Pre-septal**: Surgical excision... around age 1 year

  **Post-septal**: Surgical excision...

  Why this age?
Dermoid cysts: Management

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  - Pre-septal: Surgical excision... around age 1 year
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Why this age?
Because it is around this age when the risk of accidental cyst rupture (due to incidental trauma commonly experienced by unsteady toddlers) outweighs risk of surgical anesthesia.
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*Why is cyst rupture something to be avoided?*
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Good question! We’ll address it shortly.
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- For both locations, every effort must be made to avoid rupturing the cyst during surgical removal
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*Why is it important not to rupture the cyst during excision?*
A

**Orbital Dermoid Cysts**

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  - For both locations, every effort must be made to **avoid rupturing the cyst** during surgical removal

*Why is it important not to rupture the cyst during excision?*
Because the cyst’s contents are highly inflammogenic-- if spilled, they can produce significant local scarring. (And, circling back: This is also why traumatic rupture is something to be avoided.)