Before you begin: This is a big topic, and big topics beget big slide-sets. There’s a couple of natural breaks (around slide 215, and again around 415); I placed *break time!* slides at those locations.
All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they?

?  ?

(Obviously, there are multiple legit ways to answer this question, but there’s one best way in the context of this slide-set)
All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they?

Pigmented

Nonpigmented

(Obviously, there are multiple legit ways to answer this question, but there’s one best way in the context of this slide-set)
All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they? Which lesions can present pigmented, and which can present nonpigmented?

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(Don’t spend too much time sweating this—it’s kinda rhetorical)
**Melanocytic Eyelid and Epibulbar Lesions**

All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they? Which lesions can present pigmented, and which can present nonpigmented?

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No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they? Which lesions can present pigmented, and which can present nonpigmented?

Point #1:
Regardless of cell of origin, any eyelid or epibulbar lesion can be pigmented.

No question—proceed when ready
All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they? Which lesions can present pigmented, and which can present nonpigmented?

**Pigmented**
- All of them
- All of them
- All of them
- All of them
- All of them
- All of them
- All of them

**Nonpigmented**
- All of them
- All of them

**Point #1:**
Regardless of cell of origin, any eyelid or epibulbar lesion can be pigmented. **This includes melanocytic lesions** (duh).

No question—proceed when ready
All lesions of the eyelid and epibulbar tissue can be said to have one of two appearances—what are they? Which lesions can present pigmented, and which can present nonpigmented?

Point #2:
Lesions of melanocytic origin can be nonpigmented

No question—proceed when ready
All melanocytic lesions of the eyelid and epibulbar tissue arise from one of two cell types—what are they?

(OTOH, this question only has one legit answer)
Melanocytic Eyelid and Epibulbar Lesions

All melanocytic lesions of the eyelid and epibulbar tissue arise from one of two cell types—what are they?

Nevus cells

(OTOH, this question only has one legit answer)
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive?
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Briefly, what’s the backstory on neural crest cells?
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

Briefly, what’s the backstory on neural crest cells? NCCs are a subtype of embryo cell type.
Melanocytes

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Melanocytes

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Melanocytes

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Let's consider the embryology of melanocytes. From which primordial cell do they derive?
Neural crest cells (NCCs)

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Melanocytic Eyelid and Epibulbar Lesions

Neural crest cells…
Melanocytic Eyelid and Epibulbar Lesions

Neural crest cells...and their derivatives
Let’s consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

**neuroectodermal cells**

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Other than giving rise to melanocytes, does neuroectodermal tissue play any role in eye morphogenesis?
Melanocytes

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Other than giving rise to melanocytes, does neuroectodermal tissue play any role in eye morphogenesis? It does indeed—neuroectoderm gives rise to the neurosensory retina and the RPE, for example.
Melanocytes

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So, the NS retina and RPE derive from NCCs?
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So, the NS retina and RPE derive from NCCs? No no no, don’t get it twisted—they derive from neuroectoderm, which is the same primordial tissue that gives rise to NCCs.
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive?
Neural crest cells (NCCs)

Briefly, what’s the backstory on neural crest cells?
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There is a condition—well known to you, dear reader—that consists of a multitude of lesions,
most of which are either melanocytic or neuroglial in origin. What is this condition?
Neurofibromatosis type 1 (NF1)
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

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Neural crest cells (NCCs)

Briefly, what’s the backstory on neural crest cells?

**In one word, what sort of condition is NF1?**

**Neurofibromatosis type 1 (NF1)**

The cohort of NCCs from which melanocytes derive gives rise also to neurons and glial cells.
Melanocytes

Let's consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

Briefly, what's the backstory on neural crest cells?

In one word, what sort of condition is NF1? A phakomatosis

Neurofibromatosis type 1 (NF1)

There is a condition—well known to you, dear reader—that consists of a multitude of lesions, most of which are either melanocytic or neuroglial in origin. What is this condition? Looking at the cohort of NCCs from which melanocytes derive gives rise also to neurons and glial cells.
Let's consider the embryology of melanocytes. From which primordial cell do they derive?
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Briefly, what’s the backstory on neural crest cells?

In one word, what sort of condition is NF1?
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Neurofibromatosis type 1 (NF1)

What are the four classic neuroglial lesions in NF1?
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

Briefly, what’s the backstory on neural crest cells? In one word, what sort of condition is NF1? A phakomatosis

Neurofibromatosis type 1 (NF1)

Neuroglial lesions
--Nodular neurofibromas
--Plexiform neurofibromas
--Optic glioma
--Prominent corneal nerves

What are the four classic neuroglial lesions in NF1?
Melanocytic Eyelid and Epibulbar Lesions

NF1: Neuroglial lesions

- Neuroglial lesions
  - Plexiform neurofibroma
  - Nodular neurofibroma
  - Optic nerve glioma

Images of various melanocytic eyelid and epibulbar lesions.
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

Briefly, what’s the backstory on neural crest cells?

In one word, what sort of condition is NF1?

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Neurofibromatosis type 1 (NF1)

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Melanocytic lesions

--?
--?
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--?

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Let’s consider the embryology of melanocytes. From which primordial cell do they derive? Neural crest cells (NCCs)

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Neurofibromatosis type 1 (NF1)

Melanocytes

Neuroglial lesions
  --Nodular neurofibromas
  --Plexiform neurofibromas
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  --Prominent corneal nerves

Melanocytic lesions
  --Café au lait spots
  --Axillary/inguinal freckles
  --Lisch nodules
  --Choroidal lesions

What are the four classic melanocytic lesions in NF1?
Melanocytic Eyelid and Epibulbar Lesions

Lisch nodules

Café-au-lait spots

Axillary freckling

NF1: Melanocytic lesions
Melanocytes

Let’s consider the embryology of melanocytes. From which primordial cell do they derive?
Neural crest cells (NCCs)

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In one word, what sort of condition is NF1?
Neurofibromatosis type 1 (NF1)
A phakomatosis

In what fundamental way do these lesions differ (other than the cell type of origin, duh)?

**Melanocytic lesions**
-- Café au lait spots
-- Axillary/inguinal freckles
-- Lisch nodules
-- Choroidal lesions

**Neuroglial lesions**
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neurons and glial cells
Melanocytic Eyelid and Epibulbar Lesions

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In what fundamental way do these lesions differ (other than the cell type of origin, duh)? 

The lesions are of no clinical significance beyond establishing the diagnosis, whereas the lesions are associated with significant ocular and/or systemic morbidity
Melanocytes

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Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Let's consider the embryology of melanocytes. From which primordial cell do they derive?
Neural crest cells (NCCs)

Briefly, what's the backstory on neural crest cells?

For more on NF1 and other phakomatoses, see slide-set P10

Neuroglial lesions
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Melanocytic lesions
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Melanocytes

Next let’s consider the function of surface melanocytes. What do they do?
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What is the name of the membrane-bound structure in which melanin is contained?
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What is the name of the membrane-bound structure in which melanin is contained? A melanosome
Melanocytes

*Next let’s consider the function of surface melanocytes. What do they do?*
A surprising number of things, but for our purposes their function is manufacturing *melanin*, the main pigment of the body surface

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*Do melanocytes hang onto their melanosomes?*
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Do melanocytes hang onto their melanosomes? No—once packaged in melanosomes, melanin is transferred to neighboring cells (eg, skin melanocytes transfer their melanin to nearby keratinocytes).

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Melanocytic Eyelid and Epibulbar Lesions

Melanocyte and its keratinocytes
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Some people have darker skin than others. (Thanks, Captain Obvious.) Is it the case that darker-complected individuals have more melanocytes?
Melanocytes

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Some people have darker skin than others. (Thanks, Captain Obvious.) Is it the case that darker-completed individuals have more melanocytes? No, the number of melanocytes does not vary with degree of pigmentation. People with darker complexion have more melanin in their keratinocytes.
Melanocytes

Now let’s consider the histology of surface melanocytes. Where do they reside?
Now let’s consider the histology of surface melanocytes. Where do they reside? Most are found in the basal layer of the epidermis (if we’re talking about skin) or the basal layer of the epithelium (if we’re talking about the conjunctiva). Some skin melanocytes are sub-epidermal, located in the dermis. Likewise, some conj melanocytes are subepithelial, often located in the conjunctival stroma (aka the substantia propria), or in the underlying episcleral tissue.
Melanocytes

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Melanocytic Eyelid and Epibulbar Lesions

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In what key way do epithelial and dermal melanocytes differ?
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*In what key way do epithelial and dermal melanocytes differ?* Under normal conditions, dermal melanocytes do not produce melanin.
Now let’s turn our attention to nevus cells. What are they?
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Nevus cells are a type of melanocyte
Nevus cells

Now let’s turn our attention to nevus cells. *What are they?* Nevus cells are a type of melanocyte. *However, they differ from typical melanocytes in two key ways:*

--With respect to attribute #1

--With respect to attribute #2
Now let’s turn our attention to nevus cells. What are they? Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:

---With respect to **shape**

---With respect to **distribution**
Melanocytic Eyelid and Epibulbar Lesions

Now let’s turn our attention to nevus cells. What are they? Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:
--With respect to shape: Typical melanocytes are described as dendritic, whereas nevus cells are round.
--With respect to distribution:
Melanocytic Eyelid and Epibulbar Lesions

Nevus cells

Now let’s turn our attention to nevus cells. What are they?
Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:
--With respect to shape: Typical melanocytes are described as dendritic, whereas nevus cells are round
--With respect to distribution
Melanocytic Eyelid and Epibulbar Lesions

Don’t be fooled by the round cell body! Note the extensive network of processes snaking out amongst the keratinocytes.

Melanocyte

Melanocytes vs nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Don’t be fooled by the round cell body! Note the extensive network of processes snaking out amongst the keratinocytes. It’s because of all these processes that melanocytes are described as ‘dendritic.’

Melanocytes vs nevus cells
Melanocytic Eyelid and Epibulbar Lesions

If this is here, it means I have yet to find a satisfactory rendering of a nevus cell

Melanocyte  Nevus cell

Melanocytes vs nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Nevus cells

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--With respect to distribution:

There is an exception to this rule, i.e., there is one specific subset of nevus cells that have dendritic processes. With what “colorful” nevus are these cells associated?
Melanocytic Eyelid and Epibulbar Lesions

Nevus cells

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Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:

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--With respect to distribution:

There is an exception to this rule, ie, there is one specific subset of nevus cells that have dendritic processes. With what ‘colorful’ nevus are these cells associated? Blue nevi
Melanocytic Eyelid and Epibulbar Lesions

Nevus cells

Now let’s turn our attention to nevus cells. What are they? Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:

--With respect to **shape**: Typical melanocytes are described as *dendritic*, whereas nevus cells are *round*. dendritic or...?

--With respect to **distribution**

There is an exception to this rule, ie, there is one specific subset of nevus cells that have dendritic processes. With what ‘colorful’ nevus are these cells associated? **Blue nevi** (Note: Sometimes the term *spindly* is used to describe their shape rather than ‘dendritic,’ so don’t be thrown if you see that word used to describe them)
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Nevus cells

Now let’s turn our attention to nevus cells. What are they? Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:
- With respect to shape: Typical melanocytes are described as dendritic, whereas nevus cells are round. ‘Dendritic’ or…’spindly’
- With respect to distribution

There is an exception to this rule, ie, there is one specific subset of nevus cells that have dendritic processes. With what ‘colorful’ nevus are these cells associated? Blue nevi (Note: Sometimes the term spindly is used to describe their shape rather than ‘dendritic,’ so don’t be thrown if you see that word used to describe them)
Nevus cells

*Now let’s turn our attention to nevus cells. What are they?*
Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:

--With respect to **shape**: Typical melanocytes are described as *dendritic*, whereas nevus cells are *round*.

--With respect to **distribution**: Typical melanocytes are found to be evenly dispersed throughout the tissue in which they reside, whereas nevus cells are clustered together.
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--With respect to **shape**: Typical melanocytes are described as *dendritic*, whereas nevus cells are *round*.

--With respect to **distribution**: Typical melanocytes are found to be *evenly dispersed* throughout the tissue in which they reside, whereas nevus cells are *clustered together*. 
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Nevus cells

*Now let’s turn our attention to nevus cells. What are they?*
Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:
--With respect to **shape**: Typical melanocytes are described as dendritic, whereas nevus cells are round
--With respect to **distribution**: Typical melanocytes are found to be evenly dispersed throughout the tissue in which they reside, whereas nevus cells are clustered together

What is the $1$ term for these clusters?
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Nevus cells

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What is the $1 term for these clusters?
‘Nests’
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Nevus cells

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--With respect to **shape**: Typical melanocytes are described as dendritic, whereas nevus cells are round.

--With respect to **distribution**: Typical melanocytes are found to be evenly dispersed throughout the tissue in which they reside, whereas nevus cells are **clustered together**.

*What is the $1 term for these clusters?* ‘Nests’

*What is the $10 term for these nests?*
Melanocytic Eyelid and Epibulbar Lesions

Melanocytes

Nevus cells

Now let’s turn our attention to nevus cells. What are they?
Nevus cells are a type of melanocyte. However, they differ from typical melanocytes in two key ways:
--With respect to shape: Typical melanocytes are described as dendritic, whereas nevus cells are round
--With respect to distribution: Typical melanocytes are found to be evenly dispersed throughout the tissue in which they reside, whereas nevus cells are clustered together

What is the $1 term for these clusters?
‘Nests’

What is the $10 term for these nests?
‘Theques’
At long last we’re ready to start talking about Melanocytic lesions of the eyelid skin and epibulbar tissue. As we will see, equivalent lesions are found in each tissue type, so *if you can remember a lesion in one tissue, you can deduce the equivalent lesion in the other*. In order to facilitate this sort of understanding, we will review the lesions in tandem.
At long last we’re ready to start talking about Melanocytic lesions of the eyelid skin and epibulbar tissue. As we will see, equivalent lesions are found in each tissue type, so if you can remember a lesion in one tissue, you can deduce the equivalent lesion in the other. In order to facilitate this sort of understanding, we will review the lesions in tandem.

Two notes before proceeding: This review is an amalgam of material from the Path, Plastics, External Disease and Peds books. As usual, small inconsistencies exist among the books; I have smoothed these over as best I can. (Regarding skin lesions I leaned into the Plastics book; for epibulbar lesions, External Disease.) Further, for some lesions the BCSC emphasized the cell type of origin, whereas for others it didn’t; I followed suit. As always, caveat emptor.
All melanocytic lesions of the eyelid skin can be traced to one of three cell sources:
All melanocytic lesions of the eyelid skin can be traced to one of three cell sources:
Melanocytes in the epidermis, melanocytes in the dermis, or nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes  | Dermal melanocytes | Nevus cells

*All melanocytic lesions of the eyelid skin can be traced to one of three cell sources: Melanocytes in the epidermis, melanocytes in the dermis, or nevus cells*

**Epibulbar tissue**

?  | ?  | ?  | ?

*The analogous cell-sources of melanocytic lesions of the epibulbar tissue are:*
All melanocytic lesions of the eyelid skin can be traced to one of three cell sources:
Melanocytes in the epidermis, melanocytes in the dermis, or nevus cells

The analogous cell-sources of melanocytic lesions of the epibulbar tissue are:
Melanocytes in the epithelium, melanocytes in the subepi tissues, or nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Melanocytic lesions in both the lid and epibulbar locations can be classified as…

**Epibulbar tissue**

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Melanocytic lesions in both the lid and epibulbar locations can be classified as…
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes

**Benign**

**Pre-malignant**

**Malignant**

*Melanocytic lesions in both the lid and epibulbar locations can be classified as…Benign, pre-malignant or malignant*

**Epibulbar tissue**

- Epithelial melanocytes
- Subepithelial melanocytes

**Benign**

**Pre-malignant**

**Malignant**

*Melanocytic lesions in both the lid and epibulbar locations can be classified as…Benign, pre-malignant or malignant*
Let’s start our review of melanocytic lesions with **benign lesions deriving from epidermal and epithelial melanocytes**.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Benign

Pre-malignant

Malignant

Eyelid Skin

There are two benign eyelid skin lesions attributable to epidermal melanocytes—what are they?

Epibulbar tissue

Benign

Pre-malignant

Malignant

Nevus cells
There are two benign eyelid skin lesions attributable to epidermal melanocytes—what are they?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Benign

What are the equivalent lesions of epibulbar tissue?

Pre-malignant

Malignant

Nevus cells
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines

- Dermal melanocytes

**Epibulbar tissue**

- Epithelial melanocytes
  - Ephelis
  - CAM

- Subepithelial melanocytes

**What are the equivalent lesions of epibulbar tissue?**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Pre-malignant

Malignant

What is the layperson word for ephelis?

'Freckle'

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Benign

Pre-malignant

Malignant

Nevus cells

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells

**Epibulbar tissue**

- Epithelial melanocytes
- Subepithelial melanocytes
- Nevus cells

**Benign**

- Ephelis
  - Lentigines

**Pre-malignant**

- Ephelis

**Malignant**

- CAM

*What is the layperson word for ephelis?*

‘Freckle’
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epithelial melanocytes

Nevus cells

What is the layperson word for ephelis?
‘Freckle’

Hol up…You’re saying freckles of the ocular surface are a thing?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epithelial melanocytes

Nevus cells

What is the layperson word for ephelis?

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Hol up…You’re saying freckles of the ocular surface are a thing?

Apparently, yes
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Ephelis
Lentigines

Dermal melanocytes
Nevus cells

Benign
Pre-malignant
Malignant

What is the layperson word for ephelis?
‘Freckle’

Hol up…You’re saying freckles of the ocular surface are a thing? Apparently, yes
Are they, like, a big deal?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign

- Ephelis
- Lentigines

Pre-malignant

What is the layperson word for ephelis? ‘Freckle’

Malignant

- Ephelis
- CAM

Hol up… You’re saying freckles of the ocular surface are a thing? Apparently, yes

Are they, like, a big deal?
Apparently, no—the External Dz book mentions ‘ocular-surface ephelis’ in a Table, but doesn’t address them at all in text. Because the book doesn’t delve into them, neither will we.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

CAM

Pre-malignant

Malignant

What is the layperson word for ephelis?
‘Freckle’

What is the plural of ephelis?

Next Q

Nevus cells

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Dermal melanocytes

Nevus cells

Benign

Ephelis

Lentigines

Pre-malignant

Epibulbar tissue

What is the layperson word for ephelis?
‘Freckle’

What is the plural of ephelis?
‘Ephelides’

Benign

Ephelis

CAM

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

What is the basic issue (ie, cause) underlying ephelides?

Pre-malignant

Malignant

Nevus cells

Nevus cells

What is the layperson word for ephelis?

'Ephelis'

What is the plural of ephelis?

'Ephelides'

CAM

Simple an increase in the production of melanin by typical melanocytes typically located (ie, in the basal layer of the epidermis)

Are ephelides more likely in fair-skinned, or dark-skinned individuals?

Fair skinned

Do ephelides change in response to sunlight exposure?

Yes—they darken
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Ephelis

Benign

Pre-malignant

Malignant

Epithelial melanocytes

Subepithelial melanocytes

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Ephelis

Lentigines

Epithelial melanocytes

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Benign

Pre-malignant

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Nevus cells

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Freckle

Ephelides
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Are ephelides more likely in fair-skinned, or dark-skinned individuals?
Fair skinned.
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells

Benign
- Ephelis
- Lentigines

Pre-malignant
- Nevus cells

Malignant
- Epithelial melanocytes
- Subepithelial melanocytes

**What is the basic issue (ie, cause) underlying ephelides?**
Simply an increase in the production of melanin by typical melanocytes typically located (ie, in the basal layer of the epidermis)

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Epithelial melanocytes
Subepithelial melanocytes

Benign
Ephelis
Lentigines

Pre-malignant
CAM

Malignant

What is the basic issue (ie, cause) underlying ephelides?
Simply an increase in the production of melanin by typical melanocytes typically located (ie, in the basal layer of the epidermis)

Are ephelides more likely in fair-skinned, or dark-skinned individuals?
Fair skinned

Do ephelides change in response to sunlight exposure?
Yes—they darken (and in the absence of exposure, they fade)
Melanocytic Eyelid and Epibulbar Lesions

Classic ephelides concentration across the sun-exposed malar region
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Ephelis

Lentigines

In what fundamental ways do lentigines differ from ephelides?

--

Pre-malignant

Malignant

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Benign

Ephelis

CAM

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
- **Dermal melanocytes**

**Pre-malignant**
- Benign
- Lentigines

**Malignant**
- Ephelis

**Epibulbar tissue**

- **Epithelial melanocytes**
- **Subepithelial melanocytes**

**Pre-malignant**
- Benign
- Ephelis
- CAM

**Malignant**

- Nevus cells

---

In what fundamental ways do lentigines differ from ephelides?

- Lentigines are a little larger.
- Unlike in ephelides, the number of melanocytes in lentigines is increased.

larger vs smaller
In what fundamental ways do lentigines differ from ephelides?

-- Lentigines are a little larger.

-- Unlike the normal number in ephelides, the number of melanocytes in lentigines is increased.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Pre-malignant

Malignant

Ephelis

Lentigines

In what fundamental ways do lentigines differ from ephelides?
--Lentigines are a little larger
--Unlike the normal number in ephelides, the number of melanocytes in lentigines is increased vs decreased

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Benign

Pre-malignant

Malignant

Ephelis

CAM
In what fundamental ways do lentigines differ from ephelides?

-- Lentigines are a little larger
-- Unlike the normal number in ephelides, the number of melanocytes in lentigines is increased.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

CAM

Pre-malignant

Malignant

There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

Simple lentigines are not related to sun exposure, whereas solar lentigines are.

Ephelis

CAM

Nevus cells
There are two basic types of lentigines—what are they?
Simple lentigo and solar lentigo
There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo are these also known?

' Lentigo simplex' and 'solar lentigo'

Does lentigo simplex and/or solar lentigo have malignant potential?

No
There are two basic types of lentigines—what are they?
Simple lentigo and solar lentigo

By what variants of the terms simple lentigo and solar lentigo are these also known? ‘Lentigo simplex’ and ‘senile lentigo’

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There are two basic types of lentigines—what are they?

**Simple lentigo** and **solar lentigo**

*By what variants of the terms* simple lentigo and solar lentigo *are these also known?*

‘Lentigo simplex’ and ‘senile lentigo’

*Why is it called senile lentigo?*

Because it is more common in the elderly
There are two basic types of lentigines—what are they?

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Epibulbar tissue

Nevus cells

Malignant

Nevus cells

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Why is it called senile lentigo?
Because it is more common in the elderly

Speaking of the elderly…What does Grandma call these lesions?
‘Liver spots’
There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo.

By what variants of the terms simple lentigo and solar lentigo are these also known? ‘Lentigo simplex’ and ‘senile lentigo’.

Why is it called senile lentigo? Because it is more common in the elderly.

Speaking of the elderly…What does Grandma call these lesions? ‘Liver spots’
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Nevus cells

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Nevus cells

Malignant

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Does lentigo simplex and/or solar lentigo have malignant potential?

No

I coulda sworn lentigo simplex had malignant potential.

You sure about this?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Nevus cells

Malignant

Epibulbar tissue

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Does lentigo simplex and/or solar lentigo have malignant potential?

No

I coulda sworn lentigo simplex had malignant potential. You sure about this?

Yes, I’m sure. You’re thinking of lentigo maligna, a pre-malignant melanocytic lesion of the skin.
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes

There are two basic types of lentigines—what are they?

**Simple lentigo** and **solar lentigo**

By what variants of the terms simple lentigo and solar lentigo are these also known?

‘**Lentigo simplex**’ and ‘**senile lentigo**’

**Does lentigo simplex have malignant potential?**

No

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Yes, I’m sure. You’re thinking of **lentigo maligna**, a pre-malignant melanocytic lesion of the skin.
There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo

In what fundamental way do they differ?

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

CAM

Pre-malignant

Malignant

Eyelid Skin

Epibulbar tissue
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

CAM

There are two basic types of lentigines—what are they?
Simple lentigo and solar lentigo

In what fundamental way do they differ?
Simple lentigines are not related to sun exposure, whereas solar lentigines are...
Solar lentigines: Classic location (dorsum of the hand)
Solar lentigo of upper lid
There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?

Peutz-Jeghers syndrome
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Epibulbar tissue

Simple lentigo

There are two basic types of lentigines—what are they?
Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?
Pre-malignant

Malignant

Benign

Pre-malignant

Malignant

Simple lentigo and solar lentigo

Ephelis

Lentigines

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Nevus cells

Epibulbar tissue

Nevus cells

Nevus cells

There are two basic types of lentigines—what are they?
Simple lentigines and solar lentigo

Ephelis

Lentigines

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?
Peutz-Jeghers syndrome
Melanocytic Eyelid and Epibulbar Lesions

Peutz-Jeghers syndrome: Eyelid simple lentigines
There are two basic types of lentigines—what are they?

- Simple lentigo
- Solar lentigo

In what fundamental way do they differ?

- Simple lentigines are not related to sun exposure, whereas solar lentigines are.

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?

Peutz-Jeghers syndrome

How is Peutz-Jeghers pronounced?

Pyoots jeh-grz
There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?

Peutz-Jeghers syndrome

How is Peutz-Jeghers pronounced?

Pyoots jeh-grrz
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
- **Dermal melanocytes**

**Benign**

- **Ephelis**
- **Lentigines**

**Pre-malignant**

- **Simple lentigo**

**Malignant**

- **Peutz-Jeghers syndrome**

There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome? **Peutz-Jeghers syndrome**

Are lentigo simplex eyelid lesions the classic harbinger of Peutz-Jeghers syndrome?
There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

In what fundamental way do they differ?

Simple lentigines are not related to sun exposure, whereas solar lentigines are.

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?

Peutz-Jeghers syndrome

Are lentigo simplex eyelid lesions the classic harbinger of Peutz-Jeghers syndrome?

No, pigmented lesions of the perioral region are the classic/most common finding.
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes

**Benign**

- Lentigines

**Pre-malignant**

- Simple lentigo

**Malignant**

- Ephelis
- CAM

There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo.

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome? Peutz-Jeghers syndrome.

Are lentigo simplex eyelid lesions the classic harbinger of Peutz-Jeghers syndrome? No, pigmented lesions of the perioral region are the classic/most common finding.
Characteristic circumoral pigmentation in a patient with Peutz-Jeghers syndrome
Melanocytic Eyelid and Epibulbar Lesions

Speaking of: Did you notice the pigmented lip lesions in this pic?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo.

Simple lentigines have a *syndromic association with colon cancer*. What is the name of this syndrome?

When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Pre-malignant

Ephelis

CAM

Malignant

There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

Simple lentigines have a syndromic association with colon cancer. What is the name of this syndrome?

Peutz-Jeghers syndrome

When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?

Gardner syndrome and Muir-Torre syndrome
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Gardner syndrome is an important subtype of what class of condition? **Familial adenomatous polyposis**.

What transpires in a familial adenomatous polyposis that is concerning? Pts develop hundreds of colonic polyps, a significant number of which are malignant.

What proportion of untreated Gardner syndrome pts will develop colon cancer? **All** of them.

By what age will this occur? **40, maybe a little later**.

What is the treatment of choice? **Prophylactic colectomy**.
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Prophylactic colectomy is the treatment of choice.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

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Benign

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

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Benign

Pre-malignant

Malignant

Nevus cells
Intraocular Tumors of Childhood

Gardner syndrome: Colonic polyps
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By what age will this occur?

40, maybe a little later

What is the treatment of choice?

**Prophylactic colectomy**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

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What is the treatment of choice?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Benign

Nevus cells

Pre-malignant

Malignant

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Prophylactic colectomy
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells

**Benign**
- Lentigines

**Pre-malignant**
- Simple lentigo

**Malignant**
- Lentigines
- Ephelis

There are two basic types of lentigines—what are they?
- Simple lentigo
- Solar lentigo

In what fundamental way do they differ?
- Simple lentigines are not related to sun exposure, whereas solar lentigines are.

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What ocular finding is associated with Gardner syndrome?

- CHRPE-like lesions

What does **CHRPE** stand for in this context?

- Congenital hypertrophy of the RPE

How does CHRPE present?

- As solitary or grouped hyperpigmented lesions
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- As solitary or grouped hyperpigmented lesions

Malignant

Epibulbar tissue

Nevus cells

Benign

Epidermal melanocytes

Dermal melanocytes

Pre-malignant

Lentigines

Ephelis

Epithelial melanocytes

Subepithelial melanocytes

Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells
- Nevis cells
- Ephelis
- Lentigines
- Epithelial melanocytes
- Subepithelial melanocytes

There are two basic types of lentigines—what are they?
- Simple lentigo
- Solar lentigo

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When ‘colon cancer’ and ‘ophthalmic lesion’ are mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?
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How does CHRPE present?
- As solitary or grouped hyperpigmented lesions
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Nevus cells

- Ephelis
- Lentigines

Benign
Pre-malignant
Malignant

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CHRPE-like lesions

What does CHRPE stand for in this context?
Congenital hypertrophy of the RPE
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

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How does CHRPE present?

As solitary or grouped hyperpigmented lesions
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign

Epibulbar tissue

Ephelis
Lentigines

Pre-malignant

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What ocular finding is associated with Gardner syndrome?

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What does CHRPE stand for in this context?

Congenital hypertrophy of the RPE

How does CHRPE present?

As solitary or grouped hyperpigmented lesions

Nevus cells

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Gardner syndrome and Muir-Torre syndrome

Gardner syndrome is an important subtype of what class of condition?

Familial adenomatous polyposis
Intraocular Tumors of Childhood

Solitary

Grouped

CHRPE
There are two basic types of lentigines—what are they? 
They are simple lentigo and solar lentigo.

Simple lentigines have a **syndromic association with colon cancer**. What is the name of this syndrome?

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40, maybe a little later.

What is the treatment of choice?

Prophylactic colectomy.

What ocular finding is associated with Gardner syndrome?

CHRPE-like lesions.

What does CHRPE stand for in this context?

Congenital hypertrophy of the RPE.

How do CHRPE present?

As solitary or grouped hyperpigmented lesions.

What characteristics of a CHRPE-like presentation increase the likelihood that it is actually a manifestation of Gardner syndrome?

--?

--?

--?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes | Dermal melanocytes
---|---

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

Malignant

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What ocular finding is associated with Gardner syndrome?

CHRPE-like lesions

What does **CHRPE** stand for in this context?

Congenital hypertrophy of the RPE

How do CHRPE present?

As solitary or grouped hyperpigmented lesions

What characteristics of a CHRPE-like presentation increase the likelihood that it is actually a manifestation of Gardner syndrome?

- If it is bi-lat/unilateral (regular CHRPE is almost always uni-lat/unilateral)
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

Solar lentigo

Malignant

Nevus cells

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Simple lentigo and solar lentigo

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By what age will this occur?

40, maybe a little later

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Prophylactic colectomy

What ocular finding is associated with Gardner syndrome?

CHRPE-like lesions

What does **CHRPE** stand for in this context?

Congenital hypertrophy of the RPE

How do CHRPE present?

As solitary or grouped hyperpigmented lesions

What characteristics of a CHRPE-like presentation increase the likelihood that it is actually a manifestation of Gardner syndrome?

--If it is bilateral (regular CHRPE is almost always unilateral )

--
CHRPE-like lesions of Gardner syndrome: Bilateral presentation
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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- Simple lentigo
- Solar lentigo

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- Familial adenomatous polyposis

What transpires in a familial adenomatous polyposis that is concerning?
- Patients develop hundreds of colonic polyps, a significant number of which are malignant

What proportion of untreated Gardner syndrome patients will develop colon cancer?
- All of them

By what age will this occur?
- 40, maybe a little later

What is the treatment of choice?
- Prophylactic colectomy

What ocular finding is associated with Gardner syndrome?
- CHRPE-like lesions

What does **CHRPE** stand for in this context?
- Congenital hypertrophy of the RPE

How does CHRPE present?
- As solitary or grouped hyperpigmented lesions

What characteristics of a CHRPE-like presentation increase the likelihood that it is actually a manifestation of Gardner syndrome?
- Bilateral (regular CHRPE is almost always unilateral)
- Scattered throughout multiple sectors of the eyes (not grouped)
- Distribution pattern
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign
Pre-malignant
Malignant

Ephelis
Lentigines
Nevus cells

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- Simple lentigo
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What characteristics of a CHRPE-like presentation increase the likelihood that it is actually a manifestation of Gardner syndrome?
-- If it is bilateral (regular CHRPE is almost always unilateral)
-- If the lesions are scattered throughout multiple sectors of the eyes (ie, not ‘grouped’)
CHRPE-like lesions of Gardner syndrome: Scattered distribution
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

Malignant

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By what age will this occur?

40, maybe a little later

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Prophylactic colectomy

What ocular finding is associated with Gardner syndrome?

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--If the lesions are scattered throughout multiple sectors of the eyes (ie, not ‘grouped’)

--If the shape of the lesions is pisciform
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign
Pre-malignant
Malignant

Ephelis
Lentigines

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By what age will this occur? 40, maybe a little later

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What ocular finding is associated with Gardner syndrome? CHRPE-like lesions

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign

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Ephelis
Lentigines

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--If the shape of the lesions is pisciform

What does pisciform mean?
It means ‘fish-shaped’

What proportion of untreated Gardner syndrome pts will develop colon cancer?
All of them

By what age will this occur?
40, maybe a little later

What is the treatment of choice?
Prophylactic colectomy
Melanocytic Eyelid and Epibulbar Lesions

Epidermal melanocytes

Dermal melanocytes

Benign

Epithelial melanocytes

Subepithelial melanocytes

Ephelis

Lentigines

Pre-malignant

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--If the lesions are scattered throughout multiple sectors of the eyes (i.e., not ‘grouped’)
--If the shape of the lesions is **pisciform**

What does pisciform mean?

It means ‘fish-shaped’

What proportion of untreated Gardner syndrome pts will develop colon cancer?

All of them

By what age will this occur?

40, maybe a little later

What is the treatment of choice?

Prophylactic colectomy
Melanocytic Eyelid and Epibulbar Lesions

CHRPE-like lesions of Gardner syndrome: Pisciform shape
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

There are two basic types of lentigines—what are they?
Simple lentigo and solar lentigo

Simple lentigines have a **syndromic association with colon cancer**.
What is the name of this syndrome?
Peutz-Jeghers syndrome

When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind.
One is Peutz-Jeghers syndrome. What are the other two?
Gardner syndrome and Muir-Torre syndrome

How is Muir-Torre pronounced?
mure (rhymes with ‘pure’) tore-

Benign

Pre-malignant

Malignant

Ephelis

Lentigines

Nevus cells

Nevus cells

Epibulbar tissue

Camelid

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

Malignant

Nevus cells

There are two basic types of lentigines—what are they?

- Simple lentigo
- Solar lentigo

Simple lentigines have a **syndromic association with colon cancer**. What is the name of this syndrome?

When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?

- Gardner syndrome
- Muir-Torre syndrome

How is Muir-Torre pronounced?

Mure (rhymes with pure) tore-ay
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Benign

Epheilis

Lentigines

Pre-malignant

Simple lentigo and solar lentigo

Malignant

Simple lentigines have a **syndromic association with colon cancer**. What is the name of this syndrome?

What is the main ophthalmic manifestation of Muir-Torre syndrome?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

There are two basic types of lentigines—what are they?
-Simple lentigo
-Solar lentigo

Simple lentigos have a syndromic association with colon cancer. What is the name of this syndrome?

When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?
-Gardner syndrome and Muir-Torre syndrome

What is the main ophthalmic manifestation of Muir-Torre syndrome?
-Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

Pre-malignant

Benign

Malignant

Ephelis

Lentigines

Epibulbar tissue

Nevus cells

Nevus cells
There are two basic types of lentigines—what are they?

Simple lentigo and solar lentigo

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What is the name of this syndrome?

**Peutz-Jeghers syndrome**

When 'colon cancer + ophthalmic issue' is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?

**Gardner syndrome and Muir-Torre syndrome**

What is the main ophthalmic manifestation of Muir-Torre syndrome?

Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

What sort of sebaceous lesions?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign

Ephelis
Lentigines

Pre-malignant

Simple lentigo

Malignant

Nevus cells

There are two basic types of lentigines—what are they?
Simple lentigo and solar lentigo

Simple lentigines have a **syndromic association with colon cancer**.
What is the name of this syndrome?
When ‘colon cancer + ophthalmic issue’ is mentioned, three syndromes should come to mind.
One is Peutz-Jeghers syndrome. What are the other two?
Gardner syndrome and Muir-Torre syndrome

What is the main ophthalmic manifestation of Muir-Torre syndrome?
Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

What sort of sebaceous lesions?
Mainly sebaceous-cell adenomas and carcinomas
Multiple skin-colored to yellow–pink papules (arrows) on the face of a 64-year-old woman with a history of colon and cervical cancer. A skin biopsy confirmed a diagnosis of sebaceous adenoma resulting from Muir–Torre syndrome.
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes  Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Simple lentigo

Malignant

There are two basic types of lentigines—what are they? Simple lentigo and solar lentigo

Simple lentigines have a **syndromic association with colon cancer**. What is the name of this syndrome?

What is the main opthalmic manifestation of Muir-Torre syndrome? Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

What sort of sebaceous lesions? Mainly sebaceous-cell adenomas and carcinomas

Does Muir-Torre present with multiple adenomatous polyps of the colon as in Peutz-Jeghers and Gardner syndrome?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells

**Benign**
- Ephelis
- Lentigines

**Pre-malignant**
- Simple lentigo

**Malignant**
- Epithelial melanocytes
- Subepithelial melanocytes

*There are two basic types of lentigines—what are they?*
- Simple lentigo
- Solar lentigo

Simple lentigines have a **syndromic association with colon cancer**.

What is the name of this syndrome?
- Peutz-Jeghers syndrome

When 'colon cancer + ophthalmic issue' is mentioned, three syndromes should come to mind. One is Peutz-Jeghers syndrome. What are the other two?
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- Muir-Torre syndrome

What is the main ophthalmic manifestation of Muir-Torre syndrome?
- Multiple sebaceous lesions of (but not necessarily limited to) the eyelids

What sort of sebaceous lesions?
- Mainly sebaceous-cell adenomas and carcinomas

Does Muir-Torre present with multiple adenomatous polyps of the colon as in Peutz-Jeghers and Gardner syndrome?
- No; Muir-Torre is an example of a disease spectrum called Hereditary Non-Polyposis Colorectal Cancer
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Epibulbar tissue

- Epithelial melanocytes
- Subepithelial melanocytes

Benign
- Ephelis

Pre-malignant

Malignant

What does CAM stand for in this context?

- Racial melanosis
- Benign acquired melanosis
- Benign epithelial melanosis
- Primary conjunctival melanosis
- Acquired hypermelanosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Ephelis
Lentigines

Benign
Pre-malignant
Malignant

Epibulbar tissue

Epithelial melanocytes
Subepithelial melanocytes

Ephelis

Benign
Pre-malignant
Malignant

What does CAM stand for in this context?
Complexion-associated melanosis

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes

**Epibulbar tissue**

- Epithelial melanocytes
  - Ephelis
- Subepithelial melanocytes

---

**What does CAM stand for in this context?**
Complexion-associated melanosis

**CAM is known by several other names—what are they?**
- ?
- ?
- ?
- ?
- ?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Ephelis
- Lentigines

**Epibulbar tissue**

- Epithelial melanocytes
- Subepithelial melanocytes

What does CAM stand for in this context?

*Complexion-associated melanosis*

CAM is known by several other names—what are they?

--Racial melanosis
--Benign acquired melanosis
--Benign epithelial melanosis
--Primary conjunctival melanosis
--Acquired hypermelanosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign

Epithelial melanocytes
Subepithelial melanocytes

Pre-malignant

Who is prone to developing CAM?

Malignant

Eyeball skin
Nevus cells

Benign

CAM

Pre-malignant

Malignant

Ephelis
Lentigines

Who is prone to developing CAM?

It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?
The perilimbal region.

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea?
Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none
Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.
Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

At what life-stage does CAM typically first appear?

Essentially none
Young adulthood
No, it typically progresses with advancing age
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

At what life-stage does CAM typically first appear?
- Young adulthood

Benign

Pre-malignant

Malignant

Epithelial melanocytes
- Ephelis

Nevus cells

Eyelid Skin

Cam

Pre-malignant

Malignant

Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
- Essentially none

Can the palpebral conj be involved? The caruncle? The cornea?

Yes.

Does it tend to be static?
- No, it typically progresses with advancing age
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Benign

Pre-malignant

Malignant

Epithelial melanocytes

Who is prone to developing CAM?

Nevus cells

At what life-stage does CAM typically first appear?
Young adulthood

Does it tend to be static?

Yes. Yes. Yes—it's called striate melanokeratosis.

Essentially none

At what life-stage does CAM typically first appear?
Young adulthood

Does it tend to be static?

Bilateral (and fairly symmetrically so)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes

Benign

Pre-malignant

Malignant

Nevus cells

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

At what life-stage does CAM typically first appear?
Young adulthood

Does it tend to be static?
No, it typically progresses with advancing age
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**

**Benign**

**Pre-malignant**

**Malignant**

**Who is prone to developing CAM?**
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

**Which portion of the conj is most likely to be involved?**

**Epithelial melanocytes**

- Ephelis

**Benign**

**CAM**

**Pre-malignant**

**Malignant**

Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential? Essentially none.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Nevus cells

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?
The perilimbal region

CAM

Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none
Melanocytic Eyelid and Epibulbar Lesions

CAM: Perilimbal involvement
Melanocytic Eyelid and Epibulbar Lesions

Epelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?
The perilimbal region

Upon close inspection, CAM lesions often exhibit a subtle but distinctive pattern—what is it?

Essentially none
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Benign
Pre-malignant
Malignant

Epithelial melanocytes
Subepithelial melanocytes

Who is prone to developing CAM?
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Can the palpebral conj be involved? The caruncle? The cornea?
Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none

Upon close inspection, CAM lesions often exhibit a subtle but distinctive pattern—what is it?
‘Microfolds’ (see the next slide)
Complexion-associated melanosis. Slit-lamp photograph of a 73-year-old Black man that demonstrates conjunctival pigmentation with limbal “microfolds” [inset].
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?
The perilimbal region.

Does it present in unilateral, or bilateral fashion?

Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Malignant

Ephelis

Who is prone to developing CAM?

It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?

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Does it present in unilateral, or bilateral fashion?

Bilateral (and fairly symmetrically so)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

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Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so).

Can the palpebral conj be involved?
Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?
The perilimbal region

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved?
Yes

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Epilid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?
The perilimbal region

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle?
Yes

Essentially none
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Epithelial melanocytes
Dermal melanocytes
Subepithelial melanocytes

Benign
Pre-malignant
Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?
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Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle?
Yes. Yes.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

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The perilimbal region

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea??!!
Yes. Yes.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Epithelial melanocytes

Ephelis

Lentigines

Pre-malignant

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Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea??!!
Yes. Yes. Yes—it’s called

two words
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes
- Nevus cells

Benign
- Ephelis
- Lentigines

Pre-malignant
- Ephelis

Malignant
- CAM

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?
The perilimbal region

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea??!!
Yes. Yes. Yes—it’s called striate melanokeratosis.
Melanocytic Eyelid and Epibulbar Lesions

CAM: Striate melanokeratosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples

Which portion of the conj is most likely to be involved?
The perilimbal region

Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea??!!
Yes. Yes. Yes—it’s called striate melanokeratosis.

What is its malignant potential?
Essentially none
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Epilysis
Lentigines

Benign
Pre-malignant
Malignant

Nevus cells

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Does it present in unilateral, or bilateral fashion?
Bilateral (and fairly symmetrically so)

Can the palpebral conj be involved? The caruncle? The cornea??!!
Yes. Yes. Yes—it's called striate melanokeratosis.

What is its malignant potential?
Essentially none
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis

Lentigines

Pre-malignant

Nevus cells

Malignant

Nevus cells

What is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?

There’s a simple, commonsense reason why these highly pigmented lesions have essentially no malignant potential. What is it?

Yes. Yes. Yes—it’s called 'striped melanokeratosis'.

What is its malignant potential?
Essentially none

There’s a simple, commonsense reason why these highly pigmented lesions have essentially no malignant potential. What is it?

Yes. Yes. Yes—it’s called 'striped melanokeratosis'.

What is its malignant potential?
Essentially none
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Epithelial melanocytes
Subepithelial melanocytes

Dermal melanocytes
Nevus cells

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?
There’s a simple, commonsense reason why these highly pigmented lesions have essentially no malignant potential. What is it?
It’s because the increased pigment in CAM doesn’t stem from the proliferation of melanocytes (with its attendant risk of malignant transformation).

Yes. Yes. Yes—it’s called striate melanokeratosis.

What is its malignant potential?
Essentially none
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes

**Benign**
- Ephelis
- Lentigines

**Pre-malignant**

**Malignant**

Who is prone to developing CAM?
It can arise in any racial group, but is commonly associated with more darkly pigmented peoples.

Which portion of the conj is most likely to be involved?

There’s a simple, commonsense reason why these highly pigmented lesions have essentially no malignant potential. What is it?

It’s because the increased pigment in CAM doesn’t stem from the proliferation of melanocytes (with its attendant risk of malignant transformation), but rather (as mentioned earlier) from an increase in the rate of melanin synthesis and transfer to adjacent basal epithelial cells.

What is its malignant potential?
Essentially none.
CAM is a *nonproliferative* process: The number of melanocytes is normal, they just produce an increased amount of melanin that gets transferred to the surrounding keratinocytes.
Complexion-associated melanosis. A, Clinical appearance. B, Histologic examination shows a normal density of small, morphologically unremarkable melanocytes confined mainly to the basal layer of the epithelium (arrows) with variable extension of pigment into more superficial epithelial layers.
(This is a good point in the set to take a break)
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines
- **Dermal melanocytes**

Benign

Pre-malignant

Malignant

Next, let’s look at **benign lesions deriving from dermal and subepithelial melanocytes**

**Epibulbar tissue**

- **Epithelial melanocytes**
  - Ephelis
  - CAM
- **Subepithelial melanocytes**

Benign

Pre-malignant

Malignant

No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - ?
  - ?

There are three benign eyelid skin lesions attributable to dermal melanocytes—what are they?

**Epibulbar tissue**

- **Epithelial melanocytes**
  - Ephelis
  - CAM

- **Subepithelial melanocytes**

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Ephelis
Lentigines

Dermal melanocytes
Blue nevus
(Oculo)dermal melanocytosis

Benign

Pre-malignant

Malignant

There are three benign eyelid skin lesions attributable to dermal melanocytes—what are they?
(Note: Oculodermal melanocytosis and dermal melanocytosis are separate conditions)

Epibulbar tissue

Epithelial melanocytes
Ephelis
CAM

Subepithelial melanocytes

Benign

Pre-malignant

Malignant

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines
- **Dermal melanocytes**
  - Blue nevus
  - (Oculo)dermal melanocytosis

Epibulbar tissue

- **Epithelial melanocytes**
  - Ephelis
  - CAM
- **Subepithelial melanocytes**
  - ?
  - ?

What are the equivalent lesions of epibulbar tissue?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines
- **Dermal melanocytes**
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Epibulbar tissue**

- **Epithelial melanocytes**
  - Ephelis
  - CAM
- **Subepithelial melanocytes**
  - Blue nevus?
  - Ocular(dermal) melanocytosis

What are the equivalent lesions of epibulbar tissue?

*(Ditto for oculodermal melanocytosis and ocular melanocytosis)*
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

<table>
<thead>
<tr>
<th>Benign</th>
<th>Pre-malignant</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidermal melanocytes</td>
<td>Dermal melanocytes</td>
<td></td>
</tr>
<tr>
<td>Ephelis</td>
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<td>(Oculo)dermal melanocytosis</td>
</tr>
<tr>
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<td></td>
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</table>

Epibulbar tissue

<table>
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<td>Ephelis</td>
<td>Blue nevus?*</td>
<td>Ocular(dermal) melanocytosis</td>
</tr>
<tr>
<td>CAM</td>
<td></td>
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</tr>
</tbody>
</table>

What are the equivalent lesions of epibulbar tissue?

(Ditto for oculodermal melanocytosis and ocular melanocytosis)

*We’ll unpack the question mark shortly
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - (Oculo) **dermal melanocytosis**

**Note:** The terms **dermal** melanocytosis and **ocular** melanocytosis refer to related but separate conditions at each location

Epibulbar tissue

- **Epithelial melanocytes**
  - Ephelis
  - CAM

- **Subepithelial melanocytes**
  - Blue nevus?
  - **Ocular** (dermal) melanocytosis

<table>
<thead>
<tr>
<th>Benign</th>
<th>Pre-malignant</th>
<th>Malignant</th>
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</table>

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - **Oculo dermal** melanocytosis

Note: But **oculodermal** melanocytosis refers to a single condition involving both locations

**Epibulbar tissue**

- Epithelial melanocytes
  - Ephelis
  - CAM
- Subepithelial melanocytes
  - Blue nevus?
  - **Oculo dermal** melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Ephelis
Lentigines

Dermal melanocytes

Blue nevus

(Oculo)dermal melanocytosis

First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

Nevus cells

Benign

Pre-malignant

Malignant

First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, they probably are. But the latest edition (at the time of this writing) of the Plastics book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.
First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, they probably are. But the latest edition (at the time of this writing) of the Plastics book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.

Diameter-wise, are blue nevi large, or small?
First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, they probably are. But the latest edition (at the time of this writing) of the Plastics book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.

Diameter-wise, are blue nevi large, or small?
Small—less than # and units

Benign
- Ephelis
- Lentigines

Pre-malignant
- Blue nevus?

Malignant
- Ocular(odermal) melanocytosis
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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<tbody>
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<td>Epidermal melanocytes</td>
<td>Nevus cells</td>
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<td>(Oculo)dermal melanocytosis</td>
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<td>Lentigines</td>
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**First things first.** Surely something called a **blue nevus** is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, **they probably are**. But the latest edition (at the time of this writing) of the *Plastics* book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.

**Diameter-wise, are blue nevi large, or small?**
Small—less than 10 mm
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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<td>Dermal melanocytes</td>
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First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those 'blue nevus cells' mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, they probably are. But the latest edition (at the time of this writing) of the Plastics book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.

Diameter-wise, are blue nevi large, or small?
Small—less than 10 mm

Are they flat, or elevated?

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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</tr>
<tr>
<td>melanocytes</td>
<td>Dermal</td>
<td>(Oculo)dermal</td>
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<tr>
<td>Ephelis</td>
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<td>Blue nevus</td>
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First things first. Surely something called a blue nevus is made up of nevus cells—specifically, those ‘blue nevus cells’ mentioned on an earlier slide? You’d think so, wouldn’t you? And in truth, they probably are. But the latest edition (at the time of this writing) of the Plastics book refers to the responsible cells as “dermal melanocytes.” Caveat emptor.

Diameter-wise, are blue nevi large, or small?
Small—less than 10 mm

Are they flat, or elevated?
Elevated—looks like a small dark dome
Melanocytic Eyelid and Epibulbar Lesions

Blue nevus (not eyelid, obvs)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - (Oculo)dermal melanocytosis

Epibulbar tissue

- Epithelial melanocytes
  - Ephelis
  - CAM
- Subepithelial melanocytes
  - Blue nevus?
  - Ocular(dermal) melanocytosis

About that question mark: The latest (again, as I write this) edition of the *External Dz* book doesn’t mention blue nevi of the conj. The *Path book* might (it’s not entirely clear in context).
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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**Epibulbar tissue**

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<tr>
<td>Ephelis</td>
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<tr>
<td>CAM</td>
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About that question mark: The latest (again, as I write this) edition of the *External Dz* book doesn’t mention blue nevi of the conj. The *Path* book might (it’s not entirely clear in context). My take: The likelihood of encountering an OKAP question about blue nevi of the conj is too low to warrant fretting about (like I’m doing right now).
Melanocytic Eyelid and Epibulbar Lesions

Just because: Blue nevus of the conj
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Epithelial melanocytes
Subepithelial melanocytes
Epibulbar tissue

Dermal melanocytes
Blue nevus
Ocular (odermal) melanocytosis
(Nevus cells

Benign

Pre-malignant

Malignant

How does dermal melanocytosis present?

With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?

It is unilateral in about 95% of cases

Is there a gender predilection?

Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?

Yes, those of Hispanic, African or Asian descent are at increased risk
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo-)dermal melanocytosis

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Benign
- CAM

Pre-malignant

Malignant
- Ocular (odermal) melanocytosis

Does it present as unilateral or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Benign

Epheles

Lentigines

Pre-malignant

Malignant

Blue nevus

(Oculo)dermal melanocytosis

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

The pigment tends to be distributed in a particular pattern—what is it?

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

The pigment tends to be distributed in a particular pattern—what is it?

- V1 (aka the ophthalmic nerve)
- V2 (aka the maxillary nerve)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
  - Ephelis
  - Lentigines

Pre-malignant
- Nevus cells

Malignant
- Nevus cells
  - Ocular (odermal) melanocytosis

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation.

The pigment tends to be distributed in a particular pattern—what is it?
It tends to be limited to the dermatomes of
- V1 (aka the ophthalmic nerve)
- V2 (aka the maxillary nerve)
  - another nerve

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases.

Is there a gender predilection?
Yes, ♀ are more likely to be affected.

Does it manifest an ethnicity predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk.
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - *dermal melanocytosis*

**Benign**

- Ephelis
- Lentigines

**Pre-malignant**

**Malignant**

- CAM
- Ocular (odermal) melanocytosis

---

**How does dermal melanocytosis present?**

With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation.

**The pigment tends to be distributed in a particular pattern—what is it?**

It tends to be limited to the dermatomes of \( V_1 \) (aka the **ophthalmic nerve**) and \( V_2 \) (aka the **maxillary nerve**).
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
  - *(Oculo)* dermal melanocytosis

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

The pigment tends to be distributed in a particular pattern—what is it?
It tends to be limited to the dermatomes of $V_1$ (aka the *ophthalmic nerve*) and $V_2$ (aka the *maxillary nerve*)
Melanocytic Eyelid and Epibulbar Lesions

Dermal melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Benign**

**Pre-malignant**

**Malignant**

**How does dermal melanocytosis present?**
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

**Does it tend to be unilateral, or bilateral?**

- Yes, females are more likely to be affected
- Yes, those of Hispanic, African or Asian descent are at increased risk
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Benign**

**Pre-malignant**

**Malignant**

- Nevus cells
- Epibulbar tissue

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

- CAM
- Ocular (odermal) melanocytosis

- Nevus cells
- dermal melanocytosis

- Benign
- Pre-malignant
- Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Benign

Pre-malignant

Malignant

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Nevus cells

Benign

Pre-malignant

Malignant

CAM

Ocular (dermal) melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, ♀ are more likely to be affected

Does it manifest an ethnic predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Benign

Ephelis
Lentigines

Pre-malignant

Nevus cells

Malignant

Blue nevus
(Oculo)dermal melanocytosis

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, females are more likely to be affected
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Benign**
- Ephelis
- Lentigines

**Pre-malignant**
- Nevus cells

**Malignant**
- Nevus cells
  - CAM
  - Ocular (dermal) melanocytosis

---

**How does dermal melanocytosis present?**
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

**Does it tend to be unilateral, or bilateral?**
It is unilateral in about 95% of cases

**Is there a gender predilection?**
Yes, ♀ are more likely to be affected
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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<tr>
<td>Epidermal melanocytes</td>
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<td>Blue nevus (Oculo)</td>
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<tr>
<td>Lentigines</td>
<td></td>
<td>(Oculo)dermal melanocytosis</td>
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**How does dermal melanocytosis present?**
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

**Does it tend to be unilateral, or bilateral?**
It is unilateral in about 95% of cases

**Is there a gender predilection?**
Yes, ♀ are more likely to be affected

**Does it manifest an ethnicity predilection?**
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

<table>
<thead>
<tr>
<th>Epidermal melanocytes</th>
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<td>(Oculo) dermal melanocytosis</td>
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**Benign**

**Pre-malignant**

**Malignant**

---

*How does dermal melanocytosis present?*

With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

*Does it tend to be unilateral, or bilateral?*

It is unilateral in about 95% of cases

*Is there a gender predilection?*

Yes, ♀ are more likely to be affected

*Does it manifest an ethnicity predilection?*

Yes, those of Hispanic, African, or Asian descent are at increased risk

---

*Benign*  

*Pre-malignant*  

*Malignant*
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Nevus cells

Benign
- Ephelis
- Lentigines

Pre-malignant

Malignant

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Benign
- Nevus cells

Pre-malignant

Malignant
- Nevus cells

Both the Path and Plastics books state that dermal melanocytosis is a form of another melanocytic lesion—which one? (It's already been mentioned.)

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - **dermal melanocytosis**

**Benign**
- Eyelid Skin
  - Nevus cells

**Pre-malignant**
- How does dermal melanocytosis present?
  - With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

**Malignant**
- Does it tend to be unilateral, or bilateral?
  - It is unilateral in about 95% of cases

- **Is there a gender predilection?**
  - Yes, ♀ are more likely to be affected

- **Does it manifest an ethnicity predilection?**
  - Yes, those of Hispanic, African or Asian descent are at increased risk

Both the Path and Plastics books state that dermal melanocytosis is a form of another melanocytic lesion—**which one?** (It’s already been mentioned.) Both books refer to it as a form of ‘blue nevus.’
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines
- **Dermal melanocytes**
  - Blue nevus
  - Ocular (odermal) melanocytosis

**Benign**

- Nevus cells

**Pre-malignant**

**Malignant**

- Nevus cells

---

How does dermal melanocytosis present?

- With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?

- It is unilateral in about 95% of cases

Is there a gender predilection?

- Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?

- Yes, those of Hispanic, African or Asian descent are at increased risk

---

*Both the Path and Plastics books state that dermal melanocytosis is a form of another melanocytic lesion—which one? (It’s already been mentioned.) Both books refer to it as a form of ‘blue nevus.’ Note that, with respect to cell of origin, this puts us in the same dilemma we were with blue nevi.*
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- dermal melanocytosis

Benign
- Nevus cells

Pre-malignant
- Nevus cells

Malignant
- Nevus cells

How does dermal melanocytosis present?
With eyelid and periocular skin containing patches of diffusely brown, gray or blue pigmentation

Does it tend to be unilateral, or bilateral?
It is unilateral in about 95% of cases

Is there a gender predilection?
Yes, ♀ are more likely to be affected

Does it manifest an ethnicity predilection?
Yes, those of Hispanic, African or Asian descent are at increased risk

Both the Path and Plastics book state that dermal melanocytosis is a form of another melanocytic lesion— which one? (It's already been mentioned.)

Speaking of the Plastics book, an important observation: The current (2020-21) version states that dermal melanocytosis is aka Nevus of Ota. Per every other BCSC book that addresses the topic, as well as per other Academy sources, this assertion is erroneous—it is oculo
dermal melanocytosis (to be discussed very shortly) that is aka nevus of Ota, not dermal melanocytosis.
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus

**Benign**

**Pre-malignant**

**Malignant**

**Epithelial melanocytes**

- Ephelis
- CAM

- **Ocular (odermal) melanocytosis**

---

**How does ocular melanocytosis present?**

- With slate-gray patches of episcleral pigmentation

- It is usually unilateral

- An eye with ocular melanocytosis is at increased risk of potentially blinding ocular condition — about 10% of these eyes develop it

- Does it manifest an ethnicity predilection?
  - Like the dermal version, it is more common among individuals of Hispanic, African, and/or Asian descent
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

How does ocular melanocytosis present?
With slate-gray patches of tissue pigmentation

Epithelial melanocytes
- Ephelis
- CAM

Ocular (odermal) melanocytosis

Benign
- Nevus cells

Pre-malignant

Malignant

Benign

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
  - Ephelis
  - Lentigines

- Dermal melanocytes
  - Blue nevus

- Epithelial melanocytes
  - Ephelis
  - CAM

Nevus cells

How does ocular melanocytosis present?
With slate-gray patches of episcleral pigmentation

Ocular (odermal) melanocytosis

How does ocular melanocytosis present?
With slate-gray patches of episcleral pigmentation

An eye with ocular melanocytosis is at increased risk of what potentially blinding ocular condition?
Glaucoma—about 10% of these eyes develop it

Does it manifest an ethnicity predilection?
Like the dermal version, it is more common among individuals of Hispanic, African, and/or Asian descent
Melanocytic Eyelid and Epibulbar Lesions

Ocular melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Ocular melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

A, Clinical photograph illustrating slate-gray patches of pigmentation of the scleral surface.
B, Histologic examination shows an increased population of intensely pigmented spindle and dendritic melanocytes in the deep episclera (E), sclera (S), and uveal tract (U).

Ocular melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

Ephelis

Lentigines

How does ocular melanocytosis present?
With slate-gray patches of episcleral pigmentation

Does it tend to be unilateral, or bilateral?

Benign

Pre-malignant

Malignant

Epithelial melanocytes
- Ephelis
- CAM

Nevus cells

Ocular (odermal) melanocytosis

Benign

Pre-malignant

Malignant

Ocular melanocytosis

With slate-gray patches of episcleral pigmentation

Does it tend to be unilateral, or bilateral?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines

- Dermal melanocytes
  - Blue nevus

**Nevus cells**

- Benign
- Pre-malignant
- Malignant

**Epithelial melanocytes**

- Ephelis
- CAM

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**Ocular (odermal) melanocytosis**

How does ocular melanocytosis present?
With slate-gray patches of episcleral pigmentation

Does it tend to be unilateral, or bilateral?
It is usually unilateral

An eye with ocular melanocytosis is at increased risk of what potentially blinding ocular condition?
Glaucoma—about 10% of these eyes develop it

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Ocular (odermal) melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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Is the glaucoma open angle, or closed?
Open

Is the glaucoma primary or secondary?
Secondary

Ah, so they're at risk for POAG?
No. Remember, the P in POAG stands for primary; the glaucoma in ocular melanocytosis is secondary.
Melanocytic Eyelid and Epibulbar Lesions

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Nevus of Ota

Oculo dermal melanocytosis

Remind me: Oculodermal melanocytosis is aka…

Yes

Yes

Yes

Yes

Yes
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Remind me: Oculodermal melanocytosis is aka... Nevus of Ota

Oculo dermal melanocytosis

Oculo dermal melanocytosis

Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis?
Yes

Does it convey a risk of melanoma in white folk like ocular melanocytosis?
Yes

Does it have a female preponderance like dermal melanocytosis?
Yes

Does it convey a risk of glaucoma like ocular melanocytosis?
Yes

Does it have an Asian/African/Hispanic preponderance like both ocular and dermal melanocytosis?
Yes
Melanocytic Eyelid and Epibulbar Lesions

Oculodermal melanocytosis (nevus of Ota)
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*Does it have an Asian/African/Hispanic preponderance like both ocular and dermal melanocytosis?* Yes
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

Ephelis

Lentigines

Pre-malignant

Benign

Malignant

Remind me: Oculodermal melanocytosis is aka... Nevus of Ota

Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis? Yes

Does it convey a risk of melanoma in white folk like ocular melanocytosis? Yes

Nevus cells

Oculo dermal melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eye lid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

Ephelis

Lentigines

Benign

Pre-malignant

Malignant

Nevus of Ota

Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis? Yes

Does it convey a risk of melanoma in white folk like ocular melanocytosis? Yes

Does it have a female preponderance like dermal melanocytosis?

Remind me: Oculodermal melanocytosis is aka… Nevus of Ota

Oculo dermal melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus

Oculo dermal melanocytosis

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- Oculo dermal melanocytosis

Benign

Pre-malignant

Malignant

Remind me: Oculodermal melanocytosis is aka... Nevus of Ota

Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis? Yes

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Does it have a female preponderance like dermal melanocytosis? Yes

Does it convey a risk of glaucoma like ocular melanocytosis?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - **Oculo dermal melanocytosis**

**Remind me:** Oculodermal melanocytosis is aka... **Nevus of Ota**

* Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis? Yes
* Does it convey a risk of melanoma in white folk like ocular melanocytosis? Yes
* Does it have a female preponderance like dermal melanocytosis? Yes
* Does it convey a risk of glaucoma like ocular melanocytosis? Yes

**Oculo dermal melanocytosis**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Epithelial melanocytes

Dermal melanocytes
Subepithelial melanocytes

Ephelis
Nevus cells
Lentigines
Nevus cells

Benign

Pre-malignant

Malignant

Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis? Yes

Does it convey a risk of melanoma in white folk like ocular melanocytosis? Yes

Does it have a female preponderance like dermal melanocytosis? Yes

Does it convey a risk of glaucoma like ocular melanocytosis? Yes

Does it have an Asian/African/Hispanic preponderance like both ocular and dermal melanocytosis?

Remind me: Oculodermal melanocytosis is aka... Nevus of Ota

Oculo dermal melanocytosis

Oculo dermal melanocytosis
<table>
<thead>
<tr>
<th>Benign</th>
<th>Epidermal melanocytes</th>
<th>Dermal melanocytes</th>
<th>Nevus cells</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Ephelis</td>
<td>Blue nevus</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lentigines</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Eyelid Skin**

**Remind me:** Oculodermal melanocytosis is aka… **Nevus of Ota**

*Does nevus of Ota tend to occur in a V1/V2 distribution like dermal melanocytosis?* Yes

*Does it convey a risk of melanoma in white folk like ocular melanocytosis?* Yes

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*Does it convey a risk of glaucoma like ocular melanocytosis?* Yes

*Does it have an Asian/African/Hispanic preponderance like both ocular and dermal melanocytosis?* Yes

**Oculo dermal melanocytosis**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - (Oculo)dermal melanocytosis

Pre-malignant

Malignant

Epibulbar tissue

Benign
- Epithelial melanocytes
  - Ephelis
  - CAM
- Subepithelial melanocytes
  - Blue nevus
  - Ocular(oculodermal) melanocytosis

Pre-malignant

Malignant

Next, let’s look at **benign lesions deriving from nevus cells**

No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Nevus cells

What benign eyelid skin lesion is attributable to nevus cells?

Epibulbar tissue

Epithelial melanocytes
- Ephelis
- CAM

Subepithelial melanocytes
- Blue nevus
- Ocular(odermal) melanocytosis

Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
  - Ephelis
  - Lentigines

- **Dermal melanocytes**
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Epibulbar tissue**

- **Epithelial melanocytes**
  - Ephelis
  - CAM

- **Subepithelial melanocytes**
  - Blue nevus
  - Ocular(dermal) melanocytosis

What benign eyelid skin lesion is attributable to nevus cells?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
  - Ephelis
  - Lentigines
- Dermal melanocytes
  - Blue nevus
  - (Oculo)dermal melanocytosis

**Epibulbar tissue**

- Epithelial melanocytes
  - Ephelis
  - CAM
- Subepithelial melanocytes
  - Blue nevus
  - Ocular(odermal) melanocytosis

---

**What is its epibulbar equivalent?**

- Nevus cells

---

**Benign**

- Pre-malignant

**Malignant**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Epibulbar tissue

Epithelial melanocytes
- Ephelis
- CAM

Subepithelial melanocytes
- Blue nevus
- Ocular(dermal) melanocytosis

Nevus cells
- Nevus

What is its epibulbar equivalent?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign

Epidermal melanocytes
Ephelis

Pre-malignant

Epithelial melanocytes
Ephelis
CAM

Malignant

Dermal melanocytes
Blue nevus

Nevus cells

Are congenital eyelid nevi visible at birth?

Nevus

Benign

Subepithelial melanocytes
Blue nevus
Ocular(dermal) melanocytosis

Pre-malignant

Malignant

Nevus cells

Nevus
Are congenital eyelid nevi visible at birth?
You’d think so, but no
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
- Ephelis

Pre-malignant

Malignant
- Nevus cells
- Nevus
- Subepithelial melanocytes
- Blue nevus
- Ocular(odermal) melanocytosis

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages?

To where (ie, which skin layer) the nevus cells are found at that stage:
- The dermis-epidermis junction. Small, pigmented, flat lesion
- Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
- Dermis only (after involution of the epidermal component).
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

<table>
<thead>
<tr>
<th>Nevi</th>
<th>Benign</th>
<th>Pre-malignant</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Epidermal melanocytes</td>
<td>Dermal melanocytes</td>
<td>Nevus cells</td>
</tr>
<tr>
<td></td>
<td>Ephelis</td>
<td>Blue nevus</td>
<td>Nevus</td>
</tr>
</tbody>
</table>

- **Are congenital eyelid nevi visible at birth?**
  - You’d think so, but no

- **Nevi have a three-stage ‘life cycle.’ What are the stages?**
  - Junctional
  - Compound
  - Dermal

- **Epithelial melanocytes**
  - Ephelis
  - CAM

- **Subepithelial melanocytes**
  - Blue nevus
  - Ocular(odermal) melanocytosis

### Notes

- Nevus cells
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
- Ephelis

Pre-malignant
- Junctional
- Compound

Malignant
- Dermal melanocytes
- Blue nevus
- Nevus cells
- Nevus

Are congenital eyelid nevi visible at birth? You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?

Benign
- Epithelial melanocytes
- CAM

Pre-malignant
- Subepithelial melanocytes
- Blue nevus
- Ocular (odermal) melanocytosis

Malignant
- Nevus cells
- Nevus
Are congenital eyelid nevi visible at birth? You’d think so, but no.

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?

To where (ie, which skin layer) the nevus cells are found at that stage
---Junctional
---Compound
---Dermal

Are congenital eyelid nevi visible at birth? You’d think so, but no.

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?

To where (ie, which skin layer) the nevus cells are found at that stage
---Junctional
---Compound
---Dermal
Are congenital eyelid nevi visible at birth? You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer? Where are the nevus cells at each stage?

To where (ie, which skin layer) the nevus cells are found at that stage

--Junctional: ?
--Compound
--Dermal
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
- Ephelis

Pre-malignant
- Subepithelial melanocytes
- Blue nevus

Malignant
- Ocular (odermal) melanocytosis

Nevoid melanocytosis

---

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?

To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound
--Dermal
Junctional nevus. Nests of nevus cells (pigmented in this case) are apparent at the dermal–epidermal junction.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Nevus cells

- Ephelis
- Blue nevus

Benign
- Are congenital eyelid nevi visible at birth?
  You’d think so, but no

Pre-malignant
- Nevus have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
- Where are the nevus cells at each stage?
  - Junctional: The dermis-epidermis junction
  - Compound: ?

Malignant
- To where (ie, which skin layer) the nevus cells are found at that stage
  - Junctional: The dermis-epidermis junction
  - Compound: ?
- Nevus cells
  - Epithelial melanocytes
  - Subepithelial melanocytes
  - Nevus cells
  - Ephelis
  - CAM
  - Blue nevus
  - Ocular(odermal) melanocytosis

Next Q
**Melanocytic Eyelid and Epibulbar Lesions**

## Eyelid Skin

<table>
<thead>
<tr>
<th>Stage</th>
<th>Nevus Cells</th>
<th>Benign</th>
<th>Pre-malignant</th>
<th>Malignant</th>
</tr>
</thead>
<tbody>
<tr>
<td>Junctional</td>
<td>Nevus cells at the dermis-epidermis junction.</td>
<td>Epidermal melanocytes</td>
<td>Blue nevus</td>
<td>Ocular (dermal) melanocytosis</td>
</tr>
<tr>
<td>Compound</td>
<td>Nevus cells at the dermis-epidermis junction.</td>
<td>Epidermal melanocytes</td>
<td>Dermal melanocytes</td>
<td>Nevus cells</td>
</tr>
<tr>
<td>Dermal</td>
<td>Nevus cells in the dermis only.</td>
<td>Ephelis</td>
<td>Nevus</td>
<td>Nevus</td>
</tr>
</tbody>
</table>

### Are congenital eyelid nevi visible at birth?
You’d think so, but no

### Nevils have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?

- **Junctional:** The dermis-epidermis junction
- **Compound:** Extending down into the dermis, up through the epidermis
- **Dermal**
**Melanocytic Eyelid and Epibulbar Lesions**

**Compound** nevus. Nests of nevus cells are present in the dermis (*arrows*) as well as at the dermal–epidermal junction (*arrowheads*).
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Ephelis

Blue nevus

Benign

Pre-malignant

Malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?

To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound: Extending down into the dermis, up through the epidermis
--Dermal: ?

Epithelial melanocytes

Subepithelial melanocytes

Ephelis

Blue nevus

CAM

Ocular(dermal) melanocytosis

Nevus cells

Nevus

Oculo(dermal) melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes
Ephelis
Blue nevus

Benign
Pre-malignant
Malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound: Extending down into the dermis, up through the epidermis
--Dermal: Dermis only

Epithelial melanocytes
Subepithelial melanocytes
Nevus cells
Ephelis
Blue nevus
CAM
Ocular(odermal) melanocytosis

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound: Extending down into the dermis, up through the epidermis
--Dermal: Dermis only
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes
- Ephelis
- Lentigines
- Blue nevus
- Nevus cells

Benign

Pre-malignant

Malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound: Extending down into the dermis, up through the epidermis

What happens to the portion of the nevus that was up in the epidermis?
It involutes
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Benign

Ephelis

Blue nevus

Pre-malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction
--Compound: Extending down into the dermis, up through the epidermis

Malignant

Dermal: Dermis only

What happens to the portion of the nevus that was up in the epidermis?
It involutes

Pre-malignant

Malignant
Dermal nevus. The nests of nevus cells are confined to the dermis, and there is no junctional component. The superficial extent of the nevus cell nests is indicated with *arrowheads*. 
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Ephelis

Dermal melanocytes
Blue nevus

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage? What does each stage look like?

To where (ie, which skin layer) the nevus cells are found at that stage:

---Junctional: The dermis-epidermis junction.
---Compound: Extending down into the dermis, up through the epidermis
---Dermal: Dermis only

Ephelis
CAM

Subepithelial melanocytes
Blue nevus
Ocular(odermal) melanocytosis

Benign
Pre-malignant
Malignant

Oculo)dermal melanocytosis

Nevus cells

Next Q

(this is part of it too)
Are congenital eyelid nevi visible at birth? You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer? Where are the nevus cells at each stage? What does each stage look like?

To where (ie, which skin layer) the nevus cells are found at that stage

--- Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion
--- Compound: Extending down into the dermis, up through the epidermis
--- Dermal: Dermis only
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Ephelis

Dermal melanocytes
Blue nevus

Benign

Pre-malignant

Malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage? What does each stage look like?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion
--Compound: Extending down into the dermis, up through the epidermis. ?
--Dermal: Dermis only

Epithelial melanocytes
Ephelis
CAM

Subepithelial melanocytes
Blue nevus
Ocular(odermal) melanocytosis

Nevus cells

Nevus
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
- Dermal melanocytes

Benign

- Ephelis
- Blue nevus

Pre-malignant

- Nevus cells

Malignant

- Epithelial melanocytes
- Subepithelial melanocytes

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer? Where are the nevus cells at each stage? What does each stage look like?

- Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion
- Compound: Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
- Dermal: Dermis only

To where (ie, which skin layer) the nevus cells are found at that stage
Compound nevus
Melanocytic Eyelid and Epibulbar Lesions

Eyelid margin nevus
Are congenital eyelid nevi visible at birth? You'd think so, but no.

Nevi have a three-stage 'life cycle.' What are the stages? To what do the stage-names refer? Where are the nevus cells at each stage? What does each stage look like?

To where (ie, which skin layer) the nevus cells are found at that stage:
--Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion
--Compound: Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
--Dermal: Dermis only.?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
- **Dermal melanocytes**
- **Ephelis**
- **Blue nevus**

Are congenital eyelid nevi visible at birth? You’d think so, but no.

Nevi have a three-stage ‘life cycle.’ What are the stages? **To what do the stage-names refer?**

Where are the nevus cells at each stage? **What does each stage look like?**

To where (ie, which skin layer) the nevus cells are found at that stage:

--Junctional: The dermis-epidermis junction. **Small, pigmented, flat lesion**
--Compound: Extending down into the dermis, up through the epidermis. **Somewhat larger, somewhat more pigmented, somewhat elevated lesion**
--Dermal: Dermis only. **Varies, but usually unpigmented and flat**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign

Epidermal melanocytes

Dermal melanocytes

Pre-malignant

Ephelis

Lentigines

Blue nevus

Malignant

Nevus cells

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?

Where are the nevus cells at each stage? What does each stage look like?

To where (ie, which skin layer) the nevus cells are found at that stage

--Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion

When do junctional nevi appear?
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
- **Dermal melanocytes**

**Ephelis**

**Lentigines**

**Blue nevus**

**Nevus cells**

- **Benign**
  - Ephelis
  - CAM
  - CAM
- **Pre-malignant**
- **Malignant**
  - Ocular(odermal) melanocytosis

---

**Are congenital eyelid nevi visible at birth?**

You’d think so, but no

**Nevi have a three-stage ‘life cycle.’** What are the stages? To what do the stage-names refer?

Where are the nevus cells at each stage? What does each stage look like?

To where (i.e., which skin layer) the nevus cells are found at that stage:

- **Junctional**: The dermis-epidermis junction. Small, pigmented, flat lesion
- **Compound**: Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
- **Dermal**: Dermis only. Varies, but usually unpigmented and flat

When do junctional nevi appear?

In childhood
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
- **Dermal melanocytes**

**Nevus cells**

- **Benign**
  - Ephelis
  - CAM

- **Pre-malignant**
  - Ocular (or dermal) melanocytosis

- **Malignant**
  - Blue nevus

---

**Are congenital eyelid nevi visible at birth?**
You’d think so, but no.

**Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?**
Where are the nevus cells at each stage? What does each stage look like?
To where (ie, which skin layer) the nevus cells are found at that stage:

- **Junctional**: The dermis-epidermis junction. **Small, pigmented, flat lesion**
- **Compound**: Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
- **Dermal**: Dermis only. Varies, but usually unpigmented and flat

**When do junctional nevi appear?**
- In childhood

A small flat pigmented lesion—sounds like an ephelis to me. How are junctional nevi and ephelides differentiated clinically?
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Ephelis

Blue nevus

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage? What does each stage look like?
To where (ie, which skin layer) the nevus cells are found at that stage

--Junctional: The dermis-epidermis junction. Smaller, pigmented, flat lesion
--Compound: Extending down into the dermis, up through the epidermis. Somewhat larger,
somewhat more pigmented, somewhat elevated lesion

When do junctional nevi appear?
--In childhood

A small flat pigmented lesion—sounds like an ephelis to me. How are junctional nevi and ephelides differentiated clinically?
They’re not (by which I mean they can’t be—they are indistinguishable)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Ephelis

Blue nevus

Nevus cells

Benign

Pre-malignant

Malignant

Are congenital eyelid nevi visible at birth?
You’d think so, but no

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
Where are the nevus cells at each stage? What does each stage look like?
To where (ie, which skin layer) the nevus cells are found at that stage
--Junctional: The dermis-epidermis junction. Small, pigmented, flat lesion
--Compound: Extending down into the dermis, up through the epidermis. Somewhat larger, somewhat more pigmented, somewhat elevated lesion
--Dermal: Dermis only. Varies, but usually unpigmented and flat

At what age do junctional nevi start evolving into compound nevi?
Are congenital eyelid nevi visible at birth? You’d think so, but no.

Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer? Where are the nevus cells at each stage? What does each stage look like?

- **Junctional:** The dermis-epidermis junction. Small, pigmented, flat lesion.
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At what age do junctional nevi start evolving into compound nevi? Adolescence.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Ephelis

Blue nevus

Pre-malignant

Nevus cells

Malignant

At what age do compound nevi evolve into dermal nevi?

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To where (ie, which skin layer) the nevus cells are found at that stage

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At what age do compound nevi evolve into dermal nevi?

The BCSC doesn’t specify, but the Plastics book does say that virtually all nevi have reached the dermal stage by age 70

A small nonpigmented lesion—sounds like unaffected skin to me. How are dermal nevi distinguished from the surrounding skin?

If they’re no longer elevated, they might not be distinguishable

This is why the elderly—like young children—seem to have few (if any) nevi.

As the dermatologists say: ‘We come into the world without moles, and we leave without moles.’
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# Melanocytic Eyelid and Epibulbar Lesions

## Eyelid Skin

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## Benign

- **Ephelis**
- **Blue nevus**

## Pre-malignant

- Nevi have a three-stage ‘life cycle.’ What are the stages? To what do the stage-names refer?
- Where are the nevus cells at each stage? What does each stage look like?

### Stages
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## Malignant

### Questions
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Benign

Pre-malignant

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Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

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Eyelid Skin

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During what period of life do conj nevi typically appear?
During what period of life do conj nevi typically appear? The first or second decade—similar to lid nevi
**Melanocytic Eyelid and Epibulbar Lesions**

### Eyelid Skin

**Benign**
- Epidermal melanocytes
  - Ephelis
  - Lentigines

**Dermal melanocytes**
- Blue nevus
- (Oculo)dermal melanocytosis

**Pre-malignant**

**Malignant**

---

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?

---

Yes—about 1/3 are nearly devoid of pigment

---

Benign

Pre-malignant

Malignant
During what period of life do conj nevi typically appear? The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
--Juxtalimbal
--Plica
--Caruncle

Eyelid Skin

Benign
Epithelial melanocytes
Ephelis
CAM

Pre-malignant

Malignant
Nevus cells
Nevus
(Oculo)dermal melanocytosis

Benign
Epidermal melanocytes
Ephelis
Lentigines

Dermal melanocytes
Blue nevus

Nevus

Pre-malignant

Malignant
(Neo)epithelial melanocytes
Subepithelial melanocytes

Nevus cells

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Nevus

Nevus cells
Conjunctival nevus: Typical locations

- Juxtalimbal
- Plica
- Caruncle
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Benign
- Nevus cells

Pre-malignant

Malignant

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
---?
---?

On which portions of the conj are nevi typically not found?
---?
---?

Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

What should you do if you find a nevus-like lesion in one of these areas?
Biopsy it

Nevus cells

Nevus

Melanocytosis

Benign

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Benign
- Epidermal melanocytes
  - Ephelis
- Lentigines

Pre-malignant

Malignant
- Dermal melanocytes
  - Blue nevus
  - (Oculo)dermal melanocytosis

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
- Juxtalimbal
- Plica
- Caruncle

Are they usually unilateral, or bilateral?
Unilateral

Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

On which portions of the conj are nevi typically not found?
- Forniceal
- Palpebral

What should you do if you find a nevus-like lesion in one of these areas?
Biopsy it
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

- Epidermal melanocytes
  - Ephelis
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- Dermal melanocytes
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Benign
- Pre-malignant
- Malignant

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
- Juxtalimbal
- Plica
- Caruncle

Are they usually unilateral, or bilateral?
Unilateral

Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

On which portions of the conj are nevi typically not found?
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What should you do if you find a nevus-like lesion in one of these areas?
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Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

**Benign**

**Pre-malignant**

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During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what two locations are they most commonly found?
- Forniceal
- Palpebral

On which portions of the conj are nevi typically not found?
- Forniceal
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What should you do if you find a nevus-like lesion in one of these areas?
Biopsy it

Nevus cells

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The first or second decade—similar to lid nevi

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- Plica?
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Which location is the most common?
Juxtalimbal
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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The first or second decade—similar to lid nevi

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Nevus cells

Nevus
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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In what three locations are they most commonly found?
- Juxtalimbal
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- Caruncle

Regarding juxtalimbal nevi:
Do they tend to be elevated, or flat?

Benign
- CAM

Pre-malignant

Malignant

Benign

Pre-malignant

Malignant

Nevus cells

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Ocular(dermal) melanocytosis
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
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Benign

Pre-malignant

Malignant

Which location is the most common?
- Juxtalimbal

Regarding juxtalimbal nevi:
Do they tend to be elevated, or flat?
They are essentially always flat

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
- Juxtalimbal
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Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

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**Melanocytic Eyelid and Epibulbar Lesions**

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**Benign**
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Epilid Skin

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Are epithelial inclusion cysts a common finding?

Benign
- Nevus cells

Pre-malignant

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
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Yes—about \% of all nevi at this location contain them

Nevus cells

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Ocular(odermal) melanocytosis

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Regarding juxtalimbal nevi:
- Do they tend to be elevated, or flat?
  They are essentially always flat

Are epithelial inclusion cysts a common finding?
Yes—about half of all nevi at this location contain them
A, Clinical appearance with characteristic cystic areas (arrows). B, Histologically, the nevus cells have round, oval, or pear-shaped nuclei with a moderate amount of cytoplasm, mostly arranged in nests (arrowheads). Note the epithelial inclusion cysts (asterisks) within the lesion, correlating with the clinical appearance.

Conj nevus
Conj nevus: Epithelial inclusion cysts on AS-OCT
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
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- (Oculo)dermal melanocytosis

Benign

Pre-malignant

Malignant

Which location is the most common?

Juxtalimbal

Regarding juxtalimbal nevi:
- Do they tend to be elevated, or flat?
  They are essentially always flat
- Are epithelial inclusion cysts a common finding?
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Epithelial inclusion cysts

Epithelial cysts can cause nevus enlargement. What is the mechanism for this?

During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
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Are they usually unilateral, or bilateral?
Unilateral

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Yes—about 1/3 are nearly devoid of pigment

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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Nevus cells

During what period of life do conj nevi typically appear? The first or second decade—similar to lid nevi

In what three locations are they most commonly found? Juxtalimbal, Plica, Caruncle

Which location is the most common? Juxtalimbal

Regarding juxtalimbal nevi:
Do they tend to be elevated, or flat? They are essentially always flat

Are epithelial inclusion cysts a common finding? Yes—about half of all nevi at this location contain them

Epithelial cysts can cause nevus enlargement. What is the mechanism for this? Cells in the cyst wall secrete mucin, causing the cyst (and lesion) to enlarge
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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During what period of life do conj nevi typically appear?
The first or second decade—similar to lid nevi

In what three locations are they most commonly found?
--Juxtalimbal
--Plica
--Caruncle

Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

Ocular (odermal) melanocytosis

Nevus cells

Nevus
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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Eyelid Skin

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Benign

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Pre-malignant

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**Benign**
- Ephelis
- Lentigines

**Pre-malignant**

**Malignant**
- Juxtalimbal

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Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

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Benign

Ephelis

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Melanocytic Eyelid and Epibulbar Lesions

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**Eyelid Skin**

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**There's a simple, commonsense reason why these pigmented lesions have a nonzero malignancy risk. What is it?**
- It's that the evolution of a nevus does involve replication of melanocytes, which introduces the opportunity for malignant transformation.
Melanocytic Eyelid and Epibulbar Lesions

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Is the risk of malignant transformation large, or small?

Quite small (<1%)

Is the concern justified—does malignant transformation occur at this age?

Essentially never

Speaking of enlargement…If a limbal nevus has been present since the first decade, it will often be observed to enlarge at a later date. During what time period does this tend to occur?

The second decade, specifically during puberty

So you’re saying conj nevi carry essentially no risk of malignant transformation?

No, I’m saying the risk during puberty is essentially zero. It can occur later in life.

There’s a simple, commonsense reason why these pigmented lesions have a nonzero malignancy risk. What is it?

It’s that the evolution of a nevus does involve replication of melanocytes, which introduces the opportunity for malignant transformation.
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

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**During what period of life do conj nevi typically appear?**
- The first or second decade—similar to lid nevi

**In what three locations are they most commonly found?**
- Juxtalimbal
- Plica
- Caruncle

Speaking of enlargement...If a limbal nevus has been present since the first decade, it will often be observed to enlarge at a later date. During what time period does this tend to occur?
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**So you're saying conj nevi carry essentially no risk of malignant transformation?**
- No, I’m saying the risk during puberty is essentially zero. It can occur later in life.

**Is the risk of malignant transformation quite small (<1%)?**

**Is the concern justified—does malignant transformation occur at this age?**
- Essentially never. Reassure the parent that such enlargement is common, expected, and of no clinical concern.

Epithelial cysts can cause nevus enlargement. Why? Goblet cells in the cyst wall secrete mucin, causing...

**For this reason, most conj nevi should be followed with serial photography every 6-12 months or so**
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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In what three locations are they most commonly found?
Juxtalimbal
Plica
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Are they usually unilateral, or bilateral?
Unilateral

Can they be nonpigmented?
Yes—about 1/3 are nearly devoid of pigment

Regarding juxtalimbal nevi:
Do they tend to be elevated, or flat?
They are essentially always flat

Are epithelial inclusion cysts a common finding?
Yes—about half of all nevi at this location contain them

Epithelial cysts can cause nevus enlargement. What is the mechanism for this?
Goblet cells in the cyst wall secrete mucin, causing the cyst (and lesion) to enlarge

Speaking of enlargement...If a limbal nevus has been present since the first decade, it will often be observed to enlarge at a later date. During what time period does this tend to occur? The second decade, specifically during puberty

So you're saying conj nevi carry essentially no risk of malignant transformation? No, I'm saying the risk during puberty is essentially zero. It can occur later in life.

Is the risk of malignant transformation quite small (≤1%)? Quite small (≤1%)

Is the concern justified—does malignant transformation occur at this age? Essentially never. Reassure the parent that such enlargement is common, expected, and of no clinical concern.

Wait, most? Which nevi shouldn't be followed like this?
Palpebral or forniceal conj

There's a simple, commonsense reason why these pigmented lesions have a nonzero malignancy risk. What is it? The evolution of a nevus does involve replication of melanocytes, introducing the opportunity for malignant transformation

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Benign
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The second decade, specifically during puberty

Does this enlargement tend to occur slowly, or rapidly?
Rapidly

A parent notes rapid enlargement of a pigmented spot on her child's eye. Not surprisingly, what is the concern?
Malignant transformation

Is the concern justified—does malignant transformation occur at this age?
Essentially never. Reassure the parent that such enlargement is common, expected, and of no clinical concern.

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No, I'm saying the risk during puberty is essentially zero. It can occur later in life.

Is the risk of malignant transformation during puberty...If a limbal nevus has been present since the first decade, it will often be observed to enlarge at a later date. During what time period does this tend to occur?

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There's a simple, commonsense reason why these pigmented lesions have a nonzero malignancy risk. What is it?
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What nevi should be followed like this?
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Melanocytic Eyelid and Epibulbar Lesions

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Melanocytic Eyelid and Epibulbar Lesions

PAM of the palpebral conjunctiva
# Melanocytic Eyelid and Epibulbar Lesions

## Eyelid Skin

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## Epithelial cysts can cause nevus enlargement. What is the mechanism for this?
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### Melanocytic Eyelid and Epibulbar Lesions

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Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- **Epidermal melanocytes**
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**Nevus cells**

**Benign**

**Pre-malignant**

**Malignant**

*During what period of life do conj nevi typically appear?*

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Malignant transformation

*Is the concern justified—does malignant transformation occur at this age?*

Essentially never. Reassure the parent that such enlargement is common, expected, and of no clinical cern.

Finally: Enlargement during a related life-event is also common and not a harbinger of malignant transformation. What is this related life-event?
Melanocytic Eyelid and Epibulbar Lesions

## Eyelid Skin

- **Epidermal melanocytes**
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### Nevus cells

- **Benign**
  - Ephelis
  - Lentigines

- **Pre-malignant**

- **Malignant**
  - Juxtalimbal

---

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- Pregnancy

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### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

- **Epidermal melanocytes**
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---

**Benign**

- Ephelis
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**Pre-malignant**

**Malignant**

**Ocular(dermal) melanocytosis**

---

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Melanocytic Eyelid and Epibulbar Lesions

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Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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**Nevus**

- Benign
  - Ephelis
  - Lentigines
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Conjunctival nevus: Nonpigmented
Conjunctival nevus: Nonpigmented

But...But I can still see it—it clearly has a pinkish color. How can it be called ‘nonpigmented’?
But...But I can still see it—it clearly has a pinkish color. How can it be called ‘nonpigmented’? The term *nonpigmented* here doesn’t mean ‘lacking in color’; it means lacking in *pigment*, specifically the pigment known as melanin. (The more term *amelanotic* is more accurate, probably preferable.)

Conjunctival nevus: Nonpigmented
Do conj nevi go through the same three-stage life cycle as lid nevi?

- Junctional: The epithelial-stromal/subepi junction. Small, pigmented, flat lesion
- Compound: Extending down into the stroma/subepi, and up through the epithelium. When the epithelial component involutes late in life, the nevus enter the stromal/subepi stage.
- Dermal – Stromal/subepithelial?
**Melanocytic Eyelid and Epibulbar Lesions**

*Eyelid Skin*

- Epidermal melanocytes
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**Do conj nevi go through the same three-stage life cycle as lid nevi?**

Essentially yeah, but of course there’s no dermis in the conj, so there can’t be a dermal stage.

---

**Benign**

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Do conj nevi go through the same three-stage life cycle as lid nevi?
Essentially yeah, but of course there’s no dermis in the conj, so there can’t be a dermal stage.

As noted earlier in the set, the conj ‘dermis’ is the subepithelium or stroma.

Where are the nevus cells at each stage?
Note that the conj has an epithelium, not an epidermis, so the ‘junction’ of the junctional stage is that of the epithelium and stroma/subepi. Similarly, the ‘compound stage’ involves nevus cells extending down into the stroma/subepi and up through the epithelium. When the epithelial component involutes late in life, the nevus enter the stromal/subepi stage.

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Melanocytic Eyelid and Epibulbar Lesions

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Conjunctival nevus: Compound
**Melanocytic Eyelid and Epibulbar Lesions**

**Eyelid Skin**

Epidermal melanocytes  Dermal melanocytes  Nevus cells

---

Do conj nevi go through the same three-stage life cycle as lid nevi? Essentially yeah, but of course there’s no dermis in the conj, so there can’t be a dermal stage. As noted earlier in the set, the conj ‘dermis’ is the subepithelium or stroma.

Where are the nevus cells at each stage? Note that the conj has an epithelium, not an epidermis, so the ‘junction’ of the junctional stage is that of the epithelium and stroma/subepi. Similarly, the ‘compound stage’ involves nevus cells extending down into the stroma/subepi and up through the epithelium.

--Junctional: The epithelial-stromal/subepi junction
--Compound: Extending down into the stroma/subepi, and up through the epithelium
--Stromal/subepithelial: ?

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Benign

Epithelial melanocytes

Ephelis

CAM

Pre-malignant

Subepithelial melanocytes

Blue nevus

Ocular(odermal) melanocytosis

Malignant

Nevus cells

---

Next Q
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

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--Junctional: The epithelial-stromal/subepi junction
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Pre-malignant

Malignant

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Ephelis
CAM

Blue nevus
Ocular(odermal) melanocytosis
Conjunctival nevus: Stromal

Melanocytic Eyelid and Epibulbar Lesions
(This is a good point in the set to take a break)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Dermal melanocytes

Nevus cells

Benign

Ephelis

Lentigines

Blue nevus

Nevus

(Oculo)dermal melanocytosis

Pre-malignant

Malignant

Epibulbar tissue

Epithelial melanocytes

Subepithelial melanocytes

Nevus cells

Benign

Ephelis

CAM

Blue nevus

Nevus

Ocular(odermal) melanocytosis

Pre-malignant

Malignant

Next, let’s look at pre-malignant lesions deriving from dermal and subepithelial melanocytes

No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Nevus cells

Benign
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Pre-malignant?

Malignant

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- CAM

Subepithelial melanocytes
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Nevus cells

Benign
- Ephelis
- Nevus

Pre-malignant

Malignant

What pre-malignant eyelid skin lesion is attributable to epidermal melanocytes?
(We mentioned it previously)
**Melanocytic Eyelid and Epibulbar Lesions**

### Eyelid Skin

**Epidermal melanocytes**
- Ephelis
- Lentigines

**Dermal melanocytes**
- Blue nevus
- (Oculo)dermal melanocytosis

**Nevus cells**

### Epibulbar tissue

**Epithelial melanocytes**
- Ephelis
- CAM

**Subepithelial melanocytes**
- Blue nevus
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**Nevus cells**

---

**What pre-malignant eyelid skin lesion is attributable to epidermal melanocytes?**

(We mentioned it previously)
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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| Malignant      | Nevus cells           |                      |             |

#### Epibulbar tissue

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| Malignant      | Nevus cells            |                      |             |

? What is its epibulbar equivalent?
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

**Epidermal melanocytes**
- Ephelis
- Lentigines

**Dermal melanocytes**
- Blue nevus
- (Oculo)dermal melanocytosis

**Epibulbar tissue**

**Epithelial melanocytes**
- Ephelis
- CAM

**Subepithelial melanocytes**
- Blue nevus
- Ocular(odermal) melanocytosis

**What is its epibulbar equivalent?**

Nevus cells
- Nevus
- Nevus
Melanocytic Eyelid and Epibulbar Lesions

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- PAM

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What does PAM stand for in this context?
Melanocytic Eyelid and Epibulbar Lesions

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- Pre-malignant
  - Lentigo maligna

- Malignant

---

**Pre-malignant**

- **PAM**

**Malignant**

---

*What does PAM stand for in this context?*

*Primary acquired melanosis*
Melanocytic Eyelid and Epibulbar Lesions

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Who is at risk for lentigo maligna?
Old white people—the older and whiter they are, the greater the risk

When lentigo maligna of the lids is present, it often has spread from what location?
That sun-exposed part of the face, specifically the malar region

How can lentigo maligna be distinguished from simple and solar lentigines?
Lentigo maligna lesions tend to be larger, to have borders that are more irregular, and to have more color variation within them than do the lentigines

Are lentigo maligna lesions flat, or elevated?
Flat

Pre-malignant
- PAM

Malignant

Melanocytic Eyelid and Epibulbar Lesions

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“The whiter”—what does this mean exactly?

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Simply that individuals with a fairer complexion are at greater risk
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Pre-malignant
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Lentigo maligna that has spread from the malar region to the lid
Melanocytic Eyelid and Epibulbar Lesions

Callback to a pic from earlier in the set that referred to the malar region as “sun-exposed”

Classic ephelides concentration across the sun-exposed malar region
Melanocytic Eyelid and Epibulbar Lesions

## Eyelid Skin

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Melanocytic Eyelid and Epibulbar Lesions

Lentigo maligna. Note the color variation, irregular borders
Solar lentigines for comparison. Within each lesion note the regular borders, uniform coloring.
Melanocytic Eyelid and Epibulbar Lesions

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Melanocytic Eyelid and Epibulbar Lesions

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**Benign**

- Nevus cells

**Pre-malignant**

- Lentigo maligna

**Malignant**

- PAM

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**Who is at risk for lentigo maligna?**
Old white people—the older and whiter they are, the greater the risk

**When lentigo maligna of the lids is present, it often has spread from what location?**
That sun-exposed part of the face, specifically the malar region

**How can lentigo maligna be distinguished from simple and solar lentigines?**
Lentigo maligna lesions tend to be **larger and larger and larger** and to have **more irregular borders** and **more color variation within them** than do the lentigines.

**The size of a lentigo maligna lesion differ from that of the lentigines in another important way—what is it?**
The size of the maligna lesion expands—rapid radial growth is the rule.
Melanocytic Eyelid and Epibulbar Lesions

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The size of the maligna lesion expands—**rapid radial growth** is the rule

**What is the underlying cause of this radial growth?**
Unchecked intradermal proliferation of melanocytes and larger and larger and...
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Old white people—the older and whiter they are, the greater the risk

When lentigo maligna of the lids is present, it often has spread from what location?
That sun-exposed part of the face, specifically the malar region

How can lentigo maligna be distinguished from simple and solar lentigines?
Lentigo maligna lesions tend to be larger and larger and larger and more irregular, and to have more color variation within them than do the lentigines.

The size of a lentigo maligna lesion differs from that of the lentigines in another important way—what is it?
The size of the maligna lesion expands—rapid radial growth is the rule

What is the underlying cause of this radial growth?
Unchecked intradermal proliferation of melanocytes
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

Ephelis

Lentigines

Dermal melanocytes

Blue nevus

(Oculo)dermal melanocytosis

Benign

Pre-malignant

Lentigo maligna

Malignant

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rapid radial growth is the rule

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Epithelial melanocytes

Subepithelial melanocytes

Pre-malignant

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Can an eyelid lentigo maligna lesion spread across the gray line to the palpebral conj?
Indeed it can

rapid radial growth is the rule

What is the underlying cause of this radial growth?
Unchecked intradermal proliferation of melanocytes

Pre-malignant

PAM

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Lentigo maligna crossing onto the palpebral conjunctiva.
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
- Ephelis
- Lentigines

Dermal melanocytes
- Blue nevus
- (Oculo)dermal melanocytosis

Benign

Pre-malignant
- Lentigo maligna

Malignant

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Are lentigo maligna lesions flat, or elevated?
Flat
**Melanocytic Eyelid and Epibulbar Lesions**

### Eyelid Skin

<table>
<thead>
<tr>
<th>Type</th>
<th>Condition</th>
<th>Cells Description</th>
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<tbody>
<tr>
<td>Epidermal</td>
<td>Melanocytes</td>
<td></td>
</tr>
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<td><strong>Lentigo maligna</strong></td>
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- (Oculo)dermal melanocytosis

Benign
- Nevus cells

Pre-malignant
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Malignant
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What is suggested by the presence of an elevated component to an otherwise flat lentigo maligna lesion?
That this portion of the lesion is now undergoing vertical growth

Vertical growth is an ominous sign—why?
It indicates this portion has transformed from lentigo maligna into full-blown melanoma

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- PAM

Malignant
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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Melanocytic Eyelid and Epibulbar Lesions

Lentigo maligna lesion containing several areas of vertical growth
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**Eyelid Skin**

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**Benign**

Nevus cells

**Pre-malignant**

Lentigo maligna

**Malignant**

PAM

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Is this a common occurrence—for lentigo maligna to transform into melanoma?
Not really—only 2-3% will do so
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

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Subepithelial melanocytes

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Pre-malignant
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Subepithelial melanocytes

Who is at risk for developing PAM?

Same people at risk for lentigo maligna—older (it presents in middle age) white folk

How do you differentiate between PAM and a plain ol' nevus at the slit lamp?

You don’t—this can only be done at the pathologist's bench

Are PAM lesions flat, or elevated?

Flat (like lentigo maligna)

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?

Yes

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence?

Yes, but…(we will unpack this shortly)
Melanocytic Eyelid and Epibulbar Lesions

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PAM

Nevus

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life-stage

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PAM

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Pre-malignant  PAM  Malignant
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Ephelis
- Blue nevus

**Benign**
- Nevus cells

**Pre-malignant**
- PAM

**Malignant**

Who is at risk for developing PAM?
Same people at risk for lentigo maligna—older (it presents in middle age) white folk

How do you differentiate between PAM and a plain ol' nevus at the slit lamp?
You don't—this can only be done at the pathologist's bench

To be clear: **What I'm saying is, if you encounter a lesion suspicious for PAM, you must perform an excisional biopsy!**

No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes

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Pre-malignant

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Malignant
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--Whereas \( \% \) of conj nevi are cystic, the proportion of PAM lesions that are cystic is \( \% \)
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Are there any exam findings suggestive that a lesion is PAM rather than a nevus?
Yes; these include:
--Whereas half of conj nevi are cystic, the proportion of PAM lesions that are cystic is none
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Melanocytic Eyelid and Epibulbar Lesions

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Epidermal melanocytes

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Blue nevus

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--Whereas half of conj nevi are cystic, the proportion of PAM lesions that are cystic is none
--PAM pigment is often distributed in a pattern described as spicy

Pre-malignant

PAM

Malignant
Melanocytic Eyelid and Epibulbar Lesions

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Epidermal melanocytes

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Subepithelial melanocytes

Pre-malignant

PAM

Malignant

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*Are there any exam findings suggestive that a lesion is PAM rather than a nevus?*
Yes; these include:

--Whereas half of conj nevi are cystic, the proportion of PAM lesions that are cystic is none

--PAM pigment is often distributed in a pattern described as ‘peppery’
Primary acquired melanosis (PAM). Slit-lamp photograph of a 72-year-old white man that shows “peppery” pigmentation of the perilimbal conjunctiva [rectangle].
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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#### Benign Nevi

<table>
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<tr>
<th>Nevus cells</th>
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#### Oculo(dermal) melanocytosis

<table>
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### Questions

**Who is at risk for developing PAM?**
Same people at risk for lentigo maligna—older (it presents in middle age) white folk

**How do you differentiate between PAM and a plain ol’ nevus at the slit lamp?**
You don’t—this can only be done at the pathologist’s bench

**Are there any exam findings suggestive that a lesion is PAM rather than a nevus?**
Yes; these include:

- Whereas half of conj nevi are cystic, the proportion of PAM lesions that are cystic is none
- PAM pigment is often distributed in a pattern described as ‘peppery’
- If the lesion continues to enlarge after puberty

**Are PAM lesions flat, or elevated?**
Flat (like lentigo maligna)

**As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?**
Yes

**Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence?**
Yes, but…(we will unpack this shortly)

**Pre-malignant**

**PAM**

**Malignant**
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At what lesion-size should you start being concerned?

- continues to enlarge
- If the lesion continues to enlarge after puberty

Pre-malignant PAM Malignant

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- PAM pigment may be distributed in a pattern described as ‘peppery’
- If the lesion continues to enlarge

At what lesion-size should you start being concerned?
The BCSC books differ (modestly) on this: Per the External Disease book, start worrying when a lesion is larger than 2 clock hrs; per the Path book, when it’s 3+
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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--PAM pigment is often distributed in a pattern described as ‘peppery’
--If the lesion continues to enlarge after puberty
--If nodules develop within it
--
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

- Epidermal melanocytes
- Dermal melanocytes
- Ephelis
- Blue nevus

**Benign**

- Nevus cells
- (Oculo)dermal melanocytosis

**Pre-malignant**

- PAM

Who is at risk for developing PAM?
Same people at risk for lentigo maligna—older (it presents in middle age) white folk

How do you **differentiate between PAM and a plain ol’ nevus at the slit lamp?**
You don’t—this can only be done at the pathologist’s bench

**Are there any exam findings suggestive that a lesion is PAM rather than a nevus?**
Yes; these include:

- Whereas half of conj nevi are cystic, the proportion of PAM lesions that are cystic is none
- PAM pigment is often distributed in a pattern described as ‘peppy’
- If the lesion continues to enlarge after puberty
- If nodules develop within it

Malignant
Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

Epidermal melanocytes

Dermal melanocytes

Ephelis

Lentigines

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-- If it has...two words
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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- If nodules develop within it
- If it has feeder vessels
Note the nodularity, and feeder vessels (disclosure: this is a melanoma, not PAM)
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Epidermal melanocytes
Dermal melanocytes

Ephelis
Lentigines
Blue nevus

Epithelial melanocytes
Subepithelial melanocytes

Benign
Pre-malignant
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Pre-malignant
PAM
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**Melanocytic Eyelid and Epibulbar Lesions**

*Eyelid Skin*

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Flat (like lentigo maligna)

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**Pre-malignant**  
**PAM**

**Malignant**
Melanocytic Eyelid and Epibulbar Lesions

PAM, bulbar conj: Large, flat, no cysts; pt white, elderly (I’m inferring they’re aged by the fact they’re s/p CE)
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Malignant
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The usual suspects, including:
--Mitotic figures
--Cells that are:
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----‘Epithelioid’ in appearance
--Nuclei that are:
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----Pleomorphic
----Hyperchromatic

Pre-malignant

Malignant
**Melanocytic Eyelid and Epibulbar Lesions**

*Eyelid Skin*

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--- abn big vs abn small
--- something-'oid'
--- in appearance

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Melanocytic Eyelid and Epibulbar Lesions

**Eyelid Skin**

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Pre-malignant

Malignant
### Melanocytic Eyelid and Epibulbar Lesions

#### Eyelid Skin

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Abnormal vs normal

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Pre-malignant
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PAM with atypia. Atypical, melanin-laden cells are present approximately midway through the epithelium.
Melanocytic Eyelid and Epibulbar Lesions

Eye lid Skin

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Pre-malignant  PAM  Malignant
Melanocytic Eyelid and Epibulbar Lesions

A, CAM. Recall that CAM is a nonproliferative process: The number of melanocytes is normal, they just produce an increased amount of melanin that gets transferred to the surrounding keratinocytes.
Melanocytic Eyelid and Epibulbar Lesions

A, CAM. Recall that CAM is a *nonproliferative* process: The number of melanocytes is normal, they just produce an increased amount of melanin that gets transferred to the surrounding keratinocytes.

B, PAM without atypia, or with mild atypia. There is both increased pigment production and an increased number of melanocytes, but no or very mild change in melanocyte morphology.
A. **CAM.** Recall that CAM is a *nonproliferative* process: The number of melanocytes is normal, they just produce an increased amount of melanin that gets transferred to the surrounding keratinocytes.

B. **PAM without atypia, or with mild atypia.** There is both increased pigment production and an increased number of melanocytes, but no or very mild change in melanocyte morphology.

C. **PAM with moderate to severe atypia.** There is increased pigment production and number of melanocytes, and 1) migration of melanocytes into the more superficial epithelial layers, as well as 2) atypical melanocyte morphology.

**Note:**
- **PAM w/ moderate atypia**
  - Cells less dendritic-y
- **PAM w/ severe atypia**
  - Cells epithelioid-ish
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Speaking of the pathologist…She divvies PAM into two types. What are they?
She further divides PAM with atypia into three subtypes. What are they?
--PAM without atypia: Melanocytes confined to the basal epithelial layer; lack atypical features
--PAM with atypia: Melanocytes found in the superficial epithelial layers; display atypical features
----PAM with mild atypia
----PAM with moderate atypia
----PAM with severe atypia

How do you differentiate between PAM and a plain ol' nevus at the slit lamp?
You don’t—this can only be done at the pathologist's bench.

Are PAM lesions flat, or elevated?
Flat (like lentigo maligna)

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?
Yes

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence?
Yes, but…(we will unpack this shortly)

Pre-malignant (PAM)
Malignant

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…
No question—proceed when ready
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

Speaking of the pathologist…She divvies PAM into two types. What are they?
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It’s time to unpack it!

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

How do you differentiate between PAM and a plain old nevus at the slit lamp? You don’t—this can only be done at the pathologist’s bench.

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

- PAM without atypia
- PAM with mild atypia

PAM without atypia and PAM with mild atypia behave alike

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You don’t—this can only be done at the pathologist’s bench.

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- **PAM without atypia**
- **PAM with mild atypia**
- **PAM with moderate atypia**
- **PAM with severe atypia**

**PAM without atypia** and **PAM with mild atypia** behave alike

**PAM with moderate** and **PAM with severe atypia** behave alike

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

PAM without atypia

PAM with mild atypia

PAM with moderate atypia

PAM with severe atypia

PAM without atypia and PAM with mild atypia behave alike. So group them in your head like this! PAM with moderate and PAM with severe atypia behave alike.

How do you differentiate between PAM and a plain-old nevus at the slit lamp? You don’t—this can only be done at the pathologist’s bench.

Are PAM lesions flat, or elevated?
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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

- **PAM without atypia**
  - Both PAM without atypia and PAM with mild atypia have a near-zero probability of malignant transformation.

- **PAM with mild atypia**
  - PAM with moderate atypia
  - PAM with severe atypia

It's time to unpack it!

Are PAM lesions flat, or elevated?
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- **PAM with mild atypia**
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  - PAM with severe atypia

Both *PAM without atypia* and *PAM with mild atypia* have a **near-zero chance** of malignant transformation.

How do you differentiate between PAM and a plain old nevus at the slit lamp?
You don’t—this can only be done at the **pathologist’s bench**.

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Melanocytic Eyelid and Epibulbar Lesions

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If the biopsy returns as one of these, the appropriate management is...

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

PAM without atypia

PAM with mild atypia

PAM with moderate atypia

PAM with severe atypia

Both PAM without atypia and PAM with mild atypia have a near-zero chance of malignant transformation.

If the biopsy returns as one of these, the appropriate management is observation.

It's time to unpack it!

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence? Yes, but…(we will unpack this shortly)

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development? Yes

How do you differentiate between PAM and a plain ol' nevus at the slit lamp? You don’t—this can only be done at the pathologist’s bench.

Who is at risk for developing PAM? Same people at risk for lentigo maligna—older white folk

How do you differentiate between PAM and a plain ol' nevus at the slit lamp? You don’t—this can only be done at the pathologist’s bench.

Are PAM lesions flat, or elevated? Flat (like lentigo maligna)

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**Melanocytic Eyelid and Epibulbar Lesions**

When it comes to prognosis, this is how PAM shakes out:

- **PAM without atypia**
- **PAM with mild atypia**
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  - PAM with severe atypia

Both **PAM without atypia** and **PAM with mild atypia** have a **near-zero chance** of malignant transformation.

If the biopsy returns as one of these, the appropriate management is **observation**—every **amount of time** or so.

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?

Yes

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence?

Yes, but…(we will unpack this shortly)

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

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- **PAM with mild atypia**
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If the biopsy returns as one of these, the appropriate management is **observation** — every 6-12 months or so (~12 if *without*; closer to 6 if *with*).

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development? **Yes**

*Also like lentigo maligna:* Is malignant transformation of PAM an unlikely occurrence? **Yes,** but…(we will unpack this shortly)

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…

It's time to unpack it!
Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

- **PAM without atypia**
  - Both PAM without atypia and PAM with mild atypia have a near-zero chance of malignant transformation.

- **PAM with mild atypia**
  - In contrast, PAM with moderate or severe atypia carry a significant risk (10%–15%) of transformation.

If the biopsy returns as one of these, the appropriate management is observation—every 6-12 months or so (~12 if without; closer to 6 if with).

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Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

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If the biopsy returns as one of these, the appropriate management is observation—every 6-12 months or so (~12 if without; closer to 6 if with).

If the biopsy returns as one of these, the appropriate management is complete excision.

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence? Yes, but…(we will unpack this shortly)

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…
Melanocytic Eyelid and Epibulbar Lesions

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As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?

Yes.

Also like lentigo maligna: Is malignant transformation of PAM an unlikely occurrence?

Yes, but...(we will unpack this shortly).

Remember when we hedged regarding how likely PAM is to undergo malignant transformation...

What if complete excision is not feasible?

Adjuvant chemotherapy can be employed with topical MMC.
Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

- **PAM without atypia**
  - Both PAM without atypia and PAM with mild atypia have a **near-zero chance** of malignant transformation.

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- **PAM with moderate atypia**
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- **As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development?** Yes

- **What if complete excision is not feasible?**
  - Adjuvant chemotherapy can be employed with topical MMC route & med

Also like lentigo maligna: Is malignant transformation an inevitable inevitability? Yes, but…(we will unpack this shortly)

Pre-malignant (PAM)

Malignant

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…
Melanocytic Eyelid and Epibulbar Lesions

When it comes to prognosis, this is how PAM shakes out:

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- **PAM with severe atypia**: If the biopsy returns as one of these, the appropriate management is complete excision.
- **PAM without mild atypia**: If the biopsy returns as one of these, the appropriate management is observation—every 6-12 months or so (~12 if without; closer to 6 if with).

As is the case with their lentigo maligna cousin, is the presence of a vertical growth component an alarming development? Yes

Also like lentigo maligna: Is malignant transformation an unlikely occurrence? Yes, but…(we will unpack this shortly)

What if complete excision is not feasible? Adjuvant chemotherapy can be employed with topical MMC.

Remember when we hedged regarding how likely PAM is to undergo malignant transformation…
## Melanocytic Eyelid and Epibulbar Lesions

### Eyelid Skin

<table>
<thead>
<tr>
<th>Category</th>
<th>Epidermal Melanocytes</th>
<th>Dermal Melanocytes</th>
<th>Nevus Cells</th>
</tr>
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<tbody>
<tr>
<td>Benign</td>
<td>Ephelis</td>
<td>Blue nevus</td>
<td>Nevus</td>
</tr>
<tr>
<td>Pre-malignant</td>
<td>Lentigines</td>
<td>(Oculo)dermal melanocytosis</td>
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<tr>
<td>Malignant</td>
<td>Lentigo maligna</td>
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</table>

Next, let’s look at **malignant lesions, specifically melanoma**

### Epibulbar tissue

<table>
<thead>
<tr>
<th>Category</th>
<th>Epithelial Melanocytes</th>
<th>Subepithelial Melanocytes</th>
<th>Nevus Cells</th>
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<td>Pre-malignant</td>
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<td>Ocular(odermal) melanocytosis</td>
<td>Nevus</td>
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<td>Malignant</td>
<td>Melanoma</td>
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*No question—proceed when ready*
## Melanocytic Eyelid and Epibulbar Lesions

### Eyelid Skin

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<tr>
<th>Cell Type</th>
<th>Lesion Type</th>
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<tbody>
<tr>
<td>Epidermal melanocytes</td>
<td>Ephelis, Lentigines</td>
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<th>Status</th>
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<tr>
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There are four types of cutaneous melanoma, one of which accounts for almost all melanomas of the eyelid. Which one is that?

- Lentigo maligna

- Punch biopsy is preferred in establishing a diagnosis of melanoma because it allows for the determination of tumor thickness, which is important for its prognostic value.
There are four types of cutaneous melanoma, one of which accounts for almost all melanomas of the eyelid. Which one is that? Lentigo maligna melanoma.
There are four types of cutaneous melanoma, one of which accounts for almost all melanomas of the eyelid. Which one is that? Lentigo maligna melanoma

Which biopsy technique is preferred in establishing a diagnosis of melanoma? Punch biopsy

Because it allows for the determination of tumor thickness

Why is it important to determine tumor thickness? Because of its prognostic value
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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Why is punch the preferred technique?

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## Melanocytic Eyelid and Epibulbar Lesions

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Which biopsy technique is preferred in establishing a diagnosis of melanoma? **Punch biopsy**

Does punch biopsy increase the risk of metastasis? **No**

Why is punch the preferred technique? Because it allows for the determination of tumor thickness

Why is it important to determine tumor thickness? Because of its prognostic value
## Melanocytic Eyelid and Epibulbar Lesions

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#### Eyelid Skin

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What is the preferred tx for eyelid melanoma?
- Complete tumor removal via wide surgical excision, with intra-op confirmation of clean margins by Pathology using permanent sections

The Plastics book mentions another option described as a "first-line treatment." What is it?
- Immunotherapy drugs
Melanocytic Eyelid and Epibulbar Lesions

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Benign
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three words
Melanocytic Eyelid and Epibulbar Lesions

Eyelid Skin

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<tr>
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<tbody>
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**Further Information:**

- **Immunotherapy drugs** as a first-line treatment.
# Melanocytic Eyelid and Epibulbar Lesions

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### Benign
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- **Benign**
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  - Dermal melanocytes: Blue nevus, (Oculo)dermal melanocytosis

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All three are indicated only for the really bad cases: Metastatic, advanced, unresectable or recurrent
In terms of incidence, how do these stack up: Choroidal, skin, and conj melanoma?

Skin is an order of magnitude more common than choroidal, which is in turn an order more common than conj.

Just how (un)common is conj melanoma?

In white folk the incidence is only 1 per 2 million; in black and Asian pops it's probably an order of magnitude less common.

Is there a gender predilection?

No.

Is there an age predilection?

Yes. Conj melanoma is a disease of the middle-aged and elderly.

Are all conj melanomas pigmented?

No. A surprisingly high proportion (25%) are amelanotic.

What are the three origins for a conj melanoma, ie, what can they arise from?

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A certain subpopulation of conj melanomas are especially likely to be amelanotic— which population is that?

Is there an age predilection? Yes—conj melanoma is a disease of the middle-aged and elderly.

Is there a gender predilection? No.

Does it matter whether the original melanoma was pigmented? Nope—pigmented melanomas also give rise to amelanotic recurrences.
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**Pre-malignant**

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**Is recurrence a significant issue in conj melanoma?**

Indeed it is—a whopping 50% of cases will recur.

What implication does this appalling rate carry regarding managing these pts?

That they require close follow-up for the rest of their lives.
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### Benign

- Melanocytic nevus
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### Pre-malignant

- Lentigo maligna
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### Malignant

- Lentigo maligna melanoma
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(Don’t @me, bro—I know they don’t sum to 100. They’re my attempt at a compromise among the varying numbers across the BCSC.)
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**Is mortality generally secondary to local spread, or metastatic dz?**

Metastatic dz.

---

**By what route does choroidal melanoma usually metastasize?**

Hematogenous.

---

**By what route does eyelid (ie, skin) melanoma usually metastasize?**

Lymphatics.

---

**Does conj melanoma metastasize hematogenously like choroidal does, or lymphatically like skin?**

Lymphatically, like skin.

---

**To what distant location does choroidal melanoma tend to spread?**

The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement).

---

**To what distant location does skin melanoma tend to spread?**

Widely to many tissues—no one in particular.

---

**Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?**

Widely like skin.
## Melanocytic Eyelid and Epibulbar Lesions

<table>
<thead>
<tr>
<th>Lesion Type</th>
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<th>Malignant</th>
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<td>Lentigines</td>
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<tr>
<td>Blue nevus</td>
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### By what route does choroidal melanoma usually metastasize?
- Hematogenous

### By what route does eyelid (skin) melanoma usually metastasize?
- Lymphatics

### Is conj melanoma carry a significant mortality rate?
- Indeed it does

### What is the overall mortality rate for conj melanoma?
There's some disagreement among the BCSC books, but 25% is a reasonable compromise.

### Is mortality generally secondary to local spread, or metastatic dz?
- Metastatic dz

### Is conj melanoma metastasize hematogenously like choroidal, or lymphatically like skin?
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### To what distant location does choroidal melanoma tend to spread?
- The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement).

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### Does conj melanoma metastasize to the liver like choroidal melanoma, or widely like skin?
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Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate? Indeed it does.

What is the overall mortality rate for conj melanoma? There’s some disagreement among the BCSC books, but 25% is a reasonable compromise.

Is mortality gender dependent? No.

Is there an age predilection? Yes—conj melanoma is a disease of the middle-aged and elderly.

Are all conj melanomas pigmented? No. A surprisingly high proportion (25%) are amelanotic.

What are the three origins for a conj melanoma, ie, what can they arise from?

- De novo: ~25%
- From a nevus: ~2%
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Is mortality generally seen as metastatic dz? Yes

By what route does choroidal melanoma usually metastasize? Hematogenous

By what route does eyelid (ie, skin) melanoma usually metastasize? Lymphatics

Is there a gender predilection? No

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What does this indicate about the first place metastatic conj melanoma will appear?
Local lymph nodes

What are the two locations for nodes to which the conj drains?
Preauricular nodes, and submandibular nodes

When you’re presented on the boards with a pic and history strongly suggestive of conj melanoma, what are you going to tell the examiner you’ll do as part of your initial exam?
Check these locations for signs of metastasis
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**Melanocytic Eyelid and Epibulbar Lesions**

| Epidermal melanocytes | Dermal melanocytes | Ephelis | Lentigines | Blue nevus
|----------------------|--------------------|---------|------------|-------------
| Benjamin             | Pre-malignant      |        |            | Malignant   |

| Eyelid Skin          | Epibulbar tissue   | Nevus cells | Nevus cells | Melanoma
|----------------------|--------------------|-------------|-------------|-----------
| Benjamin             | Pre-malignant      | Malignant   |             |           |

In terms of incidence, how do these stack up: Choroidal, skin, and conj melanoma?

- Skin is an order of magnitude more common than choroidal, which is in turn an order of magnitude more common than conj.

Just how (un)common is conj melanoma?

- In white folk the incidence is only 1 per 2 million.
- In black and Asian populations it's probably an order of magnitude less common.

Is there a gender predilection?

- No.

Is there an age predilection?

- Yes—conj melanoma is a disease of the middle-aged and elderly.

Are all conj melanomas pigmented?

- No. A surprisingly high proportion (25%) are amelanotic.

What are the three origins for a conj melanoma, i.e., what can they arise from?

- De novo: ~25%
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Is conj melanoma carry a significant mortality rate?

- Indeed it does.

What is the overall mortality rate for conj melanoma?

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Is mortality generally secondary to local spread, or metastatic disease?

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By what route does choroidal melanoma usually metastasize?

- Hematogenous.

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- Lymphatics.

Does conj melanoma metastasize hematogenously like choroidal melanoma, or lymphatically like skin?

- Lymphatically, like skin.

What does this indicate about the first place metastatic conj melanoma will appear?

- That it will be in local lymph nodes.

What are the two locations for nodes to which the conj drains?

- Preauricular nodes, and submandibular nodes.

When you're presented on the Boards with a picture and history strongly suggestive of conj melanoma, what are you going to tell the examiner you'll do as part of your initial exam?

- Check these locations for signs of metastasis.
Melanocytic Eyelid and Epibulbar Lesions

<table>
<thead>
<tr>
<th>Melanocytes</th>
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<th>Lentigines</th>
<th>Blue nevus</th>
<th>Nevus</th>
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By what route does choroidal melanoma usually metastasize? Hematogenously.

By what route does eyelid (ie, skin) melanoma usually metastasize? Lymphatically.

Does conj melanoma metastasize hematogenously like choroidal melanoma, or lymphatically like skin? Lymphatically, like skin.

What does this indicate about the first place metastatic conj melanoma will appear? That it will be in local lymph nodes.

What are the two locations for nodes to which the conj drains? The preauricular nodes, and the submandibular nodes.
Melanocytic Eyelid and Epibulbar Lesions

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**Is conj melanoma carry a significant mortality rate?**
Indeed it does

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There’s some disagreement among the BCSC books, but 25% is a reasonable compromise.

**Is mortality generally secondary to local spread, or metastatic dz?**
Metastatic dz

### By what route does choroidal melanoma usually metastasize?
Hematogenous

**By what route does eyelid (ie, skin) melanoma usually metastasize?**
Lymphatics

**Does conj melanoma metastasize hematogenously like choroidal melanoma, or lymphatically like skin?**
Lymphatically, like skin

**To what distant location does choroidal melanoma tend to spread?**
The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

**To what distant location does skin melanoma tend to spread?**
Widely to many tissues—no one in particular

**Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?**
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Is conj melanoma more common than choroidal melanoma?
No

Is conj melanoma more common than skin melanoma?
Yes

In terms of incidence, how do these stack up: Choroidal, skin, and conj melanoma?
Skin is an order of magnitude more common than choroidal, which is in turn an order of magnitude more common than conj melanoma.

Just how (un)common is conj melanoma?
In white folk the incidence is only 1 per 2 million; in black and Asian populations it’s probably an order of magnitude less common.

Is there a gender predilection?
No.

Is there an age predilection?
Yes—conj melanoma is a disease of the middle-aged and elderly.

Are all conj melanomas pigmented?
No. A surprisingly high proportion (25%) are amelanotic.

What are the three origins for a conj melanoma, ie, what can they arise from?

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Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate? Indeed it does.

What is the overall mortality rate for conj melanoma? There’s some disagreement among the BCSC books, but 25% is a reasonable compromise.

By what route does choroidal melanoma usually metastasize? Hematogenous

By what route does eyelid (ie, skin) melanoma usually metastasize? Lymphatics

Does conj melanoma metastasize hematogenously like choroidal melanoma, or lymphatically like skin? Lymphatically, like skin

To what distant location does choroidal melanoma tend to spread? The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

To what distant location does skin melanoma tend to spread? Widely to many tissues—no one in particular

Does conj melanoma metastasize to the liver like choroidal does, or widely like skin? Widely like skin

By what route does PAM usually metastasize? Lymphatics

Pre-malignant

Benign

Malignant
Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate? Indeed it does

What is the overall mortality rate for conj melanoma? There's some disagreement among the BCSC books, but 25% is a reasonable compromise.

Is mortality generally

Ben

Pre-malignant

Malignant

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**Melanocytic Eyelid and Epibulbar Lesions**

**Is conj melanoma carry a significant mortality rate?**
Indeed it does

**What is the overall mortality rate for conj melanoma?**
There is a real mortality risk. The overall mortality rate is 25%

**Is metastatic conj melanoma more common than choroidal?**
No

**Are some conj melanomas pigmented?**
No. A surprisingly high proportion (25%) are amelanotic

**What are the three origins for a conj melanoma, and what proportion of conj melanomas arise from each?**
--- De novo: ~25%
--- From a nevus: ~2%
--- From PAM: ~70%

**Is conj melanoma carry a significant mortality rate?**
Indeed it does

**What is the overall mortality rate for conj melanoma?**
There's some disagreement among the BCSC books, but 25% is a reasonable compromise

**Is mortality generally from local spread, or metastatic disease?**
Metastatic disease

**By what route does choroidal melanoma usually metastasize?**
Hematogenous

**By what route does eyelid (ie, skin) melanoma usually metastasize?**
Lymphatics

**Does conj melanoma metastasize hematogenously like choroidal, or lymphatically like skin?**
Lymphatically, like skin

**To what distant location does choroidal melanoma tend to spread?**
The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

**To what distant location does skin melanoma tend to spread?**
Widely to many tissues—no one in particular

**Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?**
Widely like skin
Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate?
Indeed it does

What is the overall mortality rate for conj melanoma?
There is no mortality rate for conj melanoma, which is in turn an order of magnitude less common than choroidal, which is in turn an order of magnitude less common than skin melanoma.

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

Choroidal melanoma
- To what distant location does choroidal melanoma tend to spread?
The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement).

Skin melanoma
- To what distant location does skin melanoma tend to spread?
Widely to many tissues—no one in particular

 Conj melanoma
- Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?
Widely like skin

Benign
- Nevi
- Lentigines
- Ephelis

Pre-malignant
- Lentigo maligna
- Premalignant nevi

Malignant
- Melanoma
- Melanoma
- Melanoma

Eyelid Skin
- Epibulbar tissue

Nevus cells
- Nevus cells

Melanoma Melanoma Melanoma

In terms of incidence, how do these stack up: Choroidal, skin, and conj melanoma?
Skin is an order of magnitude more common than choroidal, which is in turn an order of magnitude less common than skin melanoma.

Just how (un)common is conj melanoma?
In white folk the incidence is only 1 per 2 million; in black and Asian pops it's probably an order of magnitude less common.

Is there a gender predilection?
No

Is there an age predilection?
Yes—conj melanoma is a disease of the middle-aged and elderly.

Are all conj melanomas pigmented?
No. A surprisingly high proportion (25%) are amelanotic.
Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate?
Indeed it does

What is the overall mortality rate for conj melanoma?
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What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How wide should the margins be around the lesion?
Quite wide—2 mm at least

How should the lesion be handled intraoperatively?
It shouldn't—ie, a 'no touch' technique should be employed. That is, at no point during the case should the surgeon make contact with the lesion itself.

To what distant location does choroidal melanoma tend to spread?
The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

To what distant location does skin melanoma tend to spread?
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Melanocytic Eyelid and Epibulbar Lesions

**Is conj melanoma carry a significant mortality rate?**
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Complete tumor removal via wide surgical excision.

**How wide should the margins be around the lesion?**
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**Is conj melanoma metastasize to the liver like choroidal does, or widely like skin?**
Widely like skin.

**Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?**

**To what distant location does choroidal melanoma tend to spread?**
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**To what distant location does skin melanoma tend to spread?**
Widely to many tissues—no one in particular.

**Is there an age predilection?**
Yes—conj melanoma is a disease of the middle-aged and elderly.

**Is there a gender predilection?**
No.

**In terms of incidence, how do these stack up: Choroidal, skin, and conj melanoma?**
Skin is an order of magnitude more common than choroidal, which is in turn an order of magnitude more common than conj.

**Just how (un)common is conj melanoma?**
In white folk the incidence is only 1 per 2 million; in black and Asian pops it’s probably an order of magnitude less common.

**Is mortality generally secondary to local spread, or metastatic dz?**
Metastatic dz.

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Hematogenous.

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Lymphatics.

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Lymphatically, like skin.

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Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate? Indeed it does

What is the overall mortality rate for conj melanoma?
There is a real mortality associated with conj melanoma, which is in turn an order of magnitude more common than choroidal, which is an order of magnitude more common than skin.

What is the preferred treatment for conj melanoma?
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Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate?
Indeed it does

What is the overall mortality rate for conj melanoma?
There is significant variation in mortality, but a reasonable estimate is 25% over 25 years
of follow-up. The mortality rate is lower for PAM.

Is melanoma, skin, and conj melanoma?
Choroidal melanoma is the most common, followed by skin melanoma, and then conj melanoma.

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How wide should the margins be around the lesion?
Quite wide—2 mm at least

How should the lesion be handled intraoperatively?
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To what distant location does skin melanoma tend to spread?
Widely to many tissues—no one in particular

Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?
Widely like skin

Is there a gender predilection?
No

Is there an age predilection?
Yes—conj melanoma is a disease of the middle-aged and elderly

Are all conj melanomas pigmented?
No. A surprisingly high proportion (25%) are amelanotic

What are the three origins for a conj melanoma, ie, what can they arise from?

--De novo: ~25%
--From a nevus: ~2%
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To what distant location does choroidal melanoma tend to spread? The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

Does conj melanoma metastasize to the liver like choroidal does, or widely like skin? Widely like skin

To what distant location does skin melanoma tend to spread? Widely to many tissues—no one in particular

Is there a gender predilection? No

Is there an age predilection? Yes—conj melanoma is a disease of the middle-aged and elderly

Are melanomas malignant? Yes—> Malignant

Are melanomas benign? No—> Benign

Are melanomas pre-malignant? Yes—> Pre-malignant

Are melanoma, choroidal, skin, and conj melanoma? Yes—> Malignant
Melanocytic Eyelid and Epibulbar Lesions

**Is conj melanoma carry a significant mortality rate?**
Indeed it does.

**What is the overall mortality rate for conj melanoma?**
The mortality of conj melanoma is low, which is in turn an order of magnitude lower than skin.

**What is the preferred treatment for conj melanoma?**
Complete tumor removal via wide surgical excision.

**How wide should the margins be around the lesion?**
Quite wide—2 mm at least.

**How should the lesion be handled intraoperatively?**
It shouldn’t—i.e., a ‘no touch’ technique should be employed. That is, at no point during the case should the surgeon make contact with the lesion itself.

What is the concern motivating the ‘no touch’ technique?

The concern motivating the ‘no touch’ technique is that intraop lesion manipulation might seed the ocular surface with tumor cells.

In light of this concern, what alternative to excisional biopsy is obviously unacceptable?
Incisional biopsy.

Is conj melanoma metastasize to the liver like choroidal does, or widely like skin?
Widely like skin.

To what distant location does skin melanoma tend to spread?
Widely to many tissues—no one in particular.

What is the concern motivating the ‘no touch’ technique?

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What is the concern motivating the ‘no touch’ technique?

That intraop lesion manipulation might seed the ocular surface with tumor cells.
Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate?
Indeed it does

What is the overall mortality rate for conj melanoma?
There is quite a bit of a real

Is melanoma malignant in nature?
Just how (un)common is conj melanoma?
In white folk the incidence is only 1 per 2 million; in black and Asian populations it's probably an order of magnitude less common

Is there a gender predilection?
No

Is there an age predilection?
Yes—conj melanoma is a disease of the middle-aged and elderly

Are all conj melanomas pigmented?
No. A surprisingly high proportion (25%) are amelanotic

What are the three origins for a conj melanoma, i.e., what can they arise from?

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What is the concern motivating the ‘no touch’ technique?
That intraop lesion manipulation might seed the ocular surface with tumor cells

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How wide should the margins be around the lesion?
Quite wide—2 mm at least

How should the lesion be handled intraoperatively?
It shouldn’t—i.e., a ‘no touch’ technique should be employed. That is, at no point during the case should the surgeon make contact with the lesion itself. Instead, the lesion should be removed using a technique that does not disrupt the epidermis, and the surgical area should be kept dry by avoiding BSS irrigation.

What is the concern motivating the ‘no touch’ technique?
That intraop lesion manipulation might seed the ocular surface with tumor cells

In light of this concern, what alternative to excisional biopsy is obviously unacceptable?
Incisional biopsy

The liver is the site to which choroidal melanoma most commonly metastasizes, and 33% of such cases have only liver involvement

To what distant location does skin melanoma tend to spread?
Widely to many tissues—no one in particular

Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?
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Widely to many tissues—no one in particular

Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?
Widely like skin
Is conj melanoma carry a significant mortality rate? Indeed it does.

What is the overall mortality rate for conj melanoma? There is no real consensus, but a reasonable estimate is 25%.

What is the preferred treatment for conj melanoma? Complete tumor removal via wide surgical excision.

How wide should the margins be around the lesion? Quite wide—2 mm at least.

How should the lesion be handled intraoperatively? It shouldn’t—i.e., a ‘no touch’ technique should be employed. That is, at no point during the case should the surgeon make contact with the lesion itself. Further, some surgeons will insist on a ‘dry’ field (i.e., no periodic re-wetting with BSS), as the act of wetting the surface may disseminate cancer cells.

What is the concern motivating the ‘no touch’ technique? That intraop lesion manipulation might seed the ocular surface with tumor cells.

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To what distant location does skin melanoma tend to spread? Widely to many tissues—no one in particular.

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Melanocytic Eyelid and Epibulbar Lesions

Is conj melanoma carry a significant mortality rate?
Indeed it does

What is the overall mortality rate for conj melanoma?
There is a real variation, with some authors suggesting a rate of 5% to 15%

Is mortality generally secondary to local spread, or metastatic dz?
Metastatic dz

How wide should the margins be around the lesion?
Quite wide—2 mm at least

How should the lesion be handled intraoperatively?
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To what distant location does skin melanoma tend to spread?
Widely to many tissues—no one in particular

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How is incidence of choroidal, skin, and conj melanoma?
Skin is an order of magnitude more common than choroidal, which is in turn an order of magnitude more common than conj melanoma

Just how (un)common is conj melanoma?
In white folk the incidence is only 1 per 2 million; in black and Asian pops it’s probably an order of magnitude less common

Is there a gender predilection?
No

Is there an age predilection?
Yes—conj melanoma is a disease of the middle-aged and elderly

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How is mortality of conj melanoma?
There's some disagreement among the BCSC books, but 25% is a reasonable compromise

Is mortality generally secondary to local spread, or metastatic dz?
Metastatic dz

By what route does choroidal melanoma usually metastasize?
Hematogenous

By what route does eyelid (ie, skin) melanoma usually metastasize?
Lymphatics

Does conj melanoma metastasize hematogenously like choroidal does, or lymphatically like skin?
Lymphatically, like skin

To what distant location does choroidal melanoma tend to spread?
The liver. Of pts who succumb to choroidal melanoma, 95% have liver involvement (and 33% of such cases have only liver involvement)

To what distant location does skin melanoma tend to spread?
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Does conj melanoma metastasize to the liver like choroidal does, or widely like skin?
Widely like skin

What is the preferred treatment for conj melanoma?
Complete tumor removal via wide surgical excision

How wide should the margins be around the lesion?
Quite wide—2 mm at least

How should the lesion be handled intraoperatively?
It shouldn’t—ie, a ‘no touch’ technique should be employed. That is, at no point during the case should the surgeon make contact with the lesion itself.

Further, some surgeons will insist on a ‘dry’ field (ie, no periodic re-wetting with BSS), as the act of wetting the surface may disseminate cancer cells.

What is the concern motivating the ‘no touch’ technique?
That intraop lesion manipulation might seed the ocular surface with tumor cells

In light of this concern, what alternative to excisional biopsy is obviously unacceptable?
Incisional biopsy

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Melanocytic Eyelid and Epibulbar Lesions

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A melanoma of the ciliary body extending through the sclera. Don’t be fooled by this lesion!
Melanoma of the ciliary body with extrascleral extension, presenting as an ocular surface mass. Note that there is no PAM surrounding the nodule, a clue that the lesion might have an intraocular origin. Also note that the lesion is associated with deep episcleral/scleral vessels (sentinel vessels, *arrow*) and does not obscure the overlying conjunctival vessels. This indicates that the lesion is deep to the conjunctiva.

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