What is the average age at diagnosis for neuroblastoma in general?
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2 years
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What proportion of Nb pts are diagnosed by age 5?
What is the average age at diagnosis for neuroblastoma in general? 2 years

What proportion of Nb pts are diagnosed by age 5? About 90%
What is the average age at diagnosis for neuroblastoma in general?
2 years

What proportion of Nb pts are diagnosed by age 5?
About 90%

Can Nb occur in adults?
What is the average age at diagnosis for neuroblastoma in general?  
2 years

What proportion of Nb pts are diagnosed by age 5?  
About 90%

Can Nb occur in adults?  
Yes, although it is quite uncommon
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
Q

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood? #1
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood? #1

Where does Nb rank as a cause of cancer in childhood? It is the most common cause of extracranial solid cancer (ie, not leukemia) in childhood.
Q

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

Where does Nb rank as a cause of cancer in childhood? It is the most common cause of extracranial solid cancer (ie, not leukemia) in childhood

How about in infants (ie, prior to age 12 months)?
● What is the average age at diagnosis for neuroblastoma in general? 2 years
● Where does neuroblastoma rank as a cause of cancer in childhood? #1

Where does Nb rank as a cause of cancer in childhood? It is the most common cause of extracranial solid cancer (ie, not leukemia) in childhood.

How about in infants (ie, prior to age 12 months)? It is #1
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood? #1

Where does Nb rank as a cause of cancer in childhood? It is the most common cause of extracranial solid cancer (ie, not leukemia) in childhood.

How about in infants (ie, prior to age 12 months)? It is #1

What proportion of peds cancer deaths are due to Nb?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood? #1

Where does Nb rank as a cause of cancer in childhood? It is the most common cause of extracranial solid cancer (ie, not leukemia) in childhood.

How about in infants (ie, prior to age 12 months)? It is #1.

What proportion of peds cancer deaths are due to Nb? 20%
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital cancer? #1

*Orbital Neuroblastoma*

With regard to ophthalmic mets in general (ie, not Nb), a fundamental difference obtains between children and adults. What is it?
Q&A

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital cancer? #1

**Orbital Neuroblastoma**

*With regard to ophthalmic mets in general (ie, not Nb), a fundamental difference obtains between children and adults. What is it?*

As a rule: In adults, mets go to the **eye** (specifically, to the **choroid**); but in children, mets go to the **orbit**.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital cancer? #1

**Orbital Neuroblastoma**

With regard to ophthalmic mets in general (ie, not Nb), a fundamental difference obtains between children and adults. What is it? As a rule: In adults, mets go to the **eye** (specifically, to the **choroid**); but in children, mets go to the **orbit**
Q

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

Of all orbital mets in children, what proportion are due to Nb?
- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

Of all orbital mets in children, what proportion are due to Nb? Almost 90%!
What is the average age at diagnosis for neuroblastoma in general?  
2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer?  #1

What are the two locations for the primary?
● What is the average age at diagnosis for neuroblastoma in general? 2 years
● Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
● What are the two locations for the primary? The adrenals, and the sympathetic chain
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

Which specific portion of the adrenal glands are we talking about here?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain.

Which specific portion of the adrenal glands are we talking about here? The adrenal medulla.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain.

What do the adrenal medulla and sympathetic chain have to do with one another?
Q/A

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
- What are the two locations for the primary? The adrenals, and the sympathetic chain.

What do the adrenal medulla and sympathetic chain have to do with one another?
A lot, actually, and they can be divvied up into two categories:

? → ?
● What is the average age at diagnosis for neuroblastoma in general? 2 years
● Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
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**Orbital Neuroblastoma**

What do the adrenal medulla and sympathetic chain have to do with one another? A lot, actually, and they can be divvied up into two categories:

- Functional
- Embryological
Q

- What is the average age at diagnosis for neuroblastoma in general? 2 years
- Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
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**Orbital Neuroblastoma**

**What do the adrenal medulla and sympathetic chain have to do with one another?**
A lot, actually, and they can be divvied up into two categories:

- **Functional**
  - Functionally, both the adrenal medulla and the sympathetic chain are components of the division of the nervous system.

- **Embryological**
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The **adrenals**, and the **sympathetic chain**

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A lot, actually, and they can be divvied up into two categories:

- **Functional**
- **Embryological**

Functionally, both the adrenal medulla and the sympathetic chain are components of the sympathetic division of the autonomic nervous system.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

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**What do the adrenal medulla and sympathetic chain have to do with one another?**
A lot, actually, and they can be divvied up into two categories:

**Functional**
Functionally, both the adrenal medulla and the sympathetic chain are components of the sympathetic division of the autonomic nervous system. In essence, the adrenal medulla is a modified sympathetic ganglion, one that secretes catecholamines into the bloodstream rather than into a synapse.

"Orbital Neuroblastoma"
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

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**Orbital Neuroblastoma**

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- **Functional**: Functionally, both the adrenal medulla and the sympathetic chain are components of the sympathetic division of the autonomic nervous system. In essence, the adrenal medulla is a modified sympathetic ganglion, one that secretes catecholamines into the bloodstream rather than into a synapse.

- **Embryological**: The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain.

**adrenals**  
**sympathetic chain**

What do the adrenal medulla and sympathetic chain have to do with one another?
A lot, actually, and they can be divvied up into two categories:

- **Functional**  
  Functionally, both the adrenal medulla and the sympathetic chain are components of the sympathetic division of the autonomic nervous system. In essence, the adrenal medulla is a modified sympathetic ganglion, one that secretes catecholamines into the bloodstream rather than into a synapse.

- **Embryological**  
  The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from neuroblasts.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain.

What do the adrenal medulla and sympathetic chain have to do with one another?
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Functionally, both the adrenal medulla and the sympathetic chain are components of the sympathetic division of the autonomic nervous system. In essence, the adrenal medulla is a modified sympathetic ganglion, one that secretes catecholamines into the bloodstream rather than into a synapse.

**Embryological**
The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from neuroblasts, which in turn derive from the neural crest cell line.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What do the adrenal medulla and sympathetic chain have to do with one another? A lot, actually, and they can be divvied up into two categories:

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Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

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- **Embryological**
  - The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from neuroblasts, which in turn derive from the neural crest cell line.

What is the cell of origin in neuroblastoma?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What do the adrenal medulla and sympathetic chain have to do with one another? A lot, actually, and they can be divvied up into two categories:

- **Functional**
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- **Embryological**
  The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from neuroblasts, which in turn derive from the neural crest cell line.

What is the cell of origin in neuroblastoma?

**Neuroblasts**
What is the average age at diagnosis for neuroblastoma in general? 2 years
Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
What are the two locations for the primary? The adrenals, and the sympathetic chain

If the primary involves the superior portion of the cervical sympathetic chain, what clinical presentation might occur?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and **the sympathetic chain**

*If the primary involves the superior portion of the cervical sympathetic chain, what clinical presentation might occur? A Horner syndrome*
Orbital Neuroblastoma

Primary tumor in sympathetic chain

Horner syndrome 2ndry to Nb
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit?
• What is the average age at diagnosis for neuroblastoma in general? 2 years
• Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
• What are the two locations for the primary? The adrenals, and the sympathetic chain
• What percent of neuroblastomas metastasize to the orbit? 20
• What is the average age at diagnosis for neuroblastoma in general? 2 years
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• What are the two locations for the primary? The adrenals, and the sympathetic chain
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• What are the two locations for the primary? The adrenals, and the sympathetic chain
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• What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis? and/or eyelid ecchymosis?

Of the two, which is more common?
What is the average age at diagnosis for neuroblastoma in general? 2 years
Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
What are the two locations for the primary? The adrenals, and the sympathetic chain
What percent of neuroblastomas metastasize to the orbit? 20
What is the classic presentation of orbital mets? Proptosis and/or **eyelid ecchymosis**

**Orbital Neuroblastoma**

*Of the two, which is more common? Eyelid ecchymosis*
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis and/or **eyelid ecchymosis**

Of the two, which is more common? **Eyelid ecchymosis**

Is the ecchymosis unilateral, or bilateral?
• What is the average age at diagnosis for neuroblastoma in general? 2 years
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• What are the two locations for the primary? The adrenals, and the sympathetic chain
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• What is the classic presentation of orbital mets? Proptosis and/or **eyelid ecchymosis**

*Of the two, which is more common? **Eyelid ecchymosis**

*Is the ecchymosis unilateral, or bilateral?* It can be either
Q

- What is the average age at diagnosis for neuroblastoma in general? 2 years
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- What is the classic presentation of orbital mets? Proptosis and/or **eyelid ecchymosis**

Of the two, which is more common? **Eyelid ecchymosis**

*Is the ecchymosis unilateral, or **bilateral**?*
It can be either

*What critter-based description is used for the appearance of bilateral ecchymoses in Nb?*
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

Of the two, which is more common? Eyelid ecchymosis

Is the ecchymosis unilateral, or bilateral? It can be either

What critter-based description is used for the appearance of bilateral ecchymoses in Nb? ‘Raccoon eyes’
Orbital Neuroblastoma

Nb: ‘Raccoon eyes’
What is the average age at diagnosis for neuroblastoma in general? 2 years
Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1
What are the two locations for the primary? The adrenals, and the sympathetic chain
What percent of neuroblastomas metastasize to the orbit? 20
What is the classic presentation of orbital mets?

**Proptosis** and/or eyelid ecchymosis

*Is the proptosis unilateral, or bilateral?*
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

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What is the classic presentation of orbital mets? **Proptosis** and/or eyelid ecchymosis

Is the proptosis unilateral, or bilateral? It can be either
Orbital Neuroblastoma

Nb: Proptosis
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? **Proptosis** and/or eyelid ecchymosis

Is the proptosis **unilateral** or bilateral? It can be either

What is the DDx for acute-onset unilateral proptosis in an infant/young child? -- Neuroblastoma --
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? **Proptosis** and/or eyelid ecchymosis

Is the proptosis unilateral or bilateral? It can be either

**What is the DDx for acute-onset unilateral proptosis in an infant/young child?**

--Neuroblastoma
--Orbital inflammation
--Leukemia
--Orbital cellulitis
--Rhabdomyosarcoma
Orbital Neuroblastoma

- What is the average age at diagnosis for neuroblastoma in general? 2 years
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Is the proptosis unilateral or bilateral? It can be either

What is the DDx for acute-onset unilateral proptosis in an infant/young child?
-- Neuroblastoma
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What is the DDx for acute-onset bilateral proptosis in an infant/young child?
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A

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- What is the classic presentation? Proptosis and/or eyelid ecchymosis

**Orbital Neuroblastoma**

- Is the proptosis **unilateral**, or **bilateral**? It can be either
- **What is the DDx for acute-onset bilateral proptosis in an infant/young child?**
  -- Neuroblastoma
  -- Orbital inflammation
  -- Leukemia
  -- Orbital cellulitis
  -- Rhabdomyosarcoma

- **What is the DDx for acute-onset unilateral proptosis in an infant/young child?**
  -- Neuroblastoma
  -- Orbital inflammation
  -- Leukemia
  -- Orbital cellulitis
  -- Rhabdomyosarcoma

Orbital cellulitis and rhabdo are very, very unlikely to present bilaterally.
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

What if the child is <1 year at diagnosis—is the prognosis better or worse?
● What is the average age at diagnosis for neuroblastoma in general? 2 years
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**What is the difference in 5-year survival rates between cases that present prior to vs after age 1 year?**
● What is the average age at diagnosis for neuroblastoma in general? 2 years
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● What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

What is the difference in 5-year survival rates between cases that present prior to vs after age 1 year?
It is cut roughly in half
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

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What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

What is the classic ocular paraneoplastic association?
• What is the average age at diagnosis for neuroblastoma in general? 2 years
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• What is the classic ocular paraneoplastic association? Opsoclonus
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What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

What is the classic ocular paraneoplastic association? Opsoclonus

What is opsoclonus? A saccadic oscillation
What is the average age at diagnosis for neuroblastoma in general? 2 years

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What is the classic ocular paraneoplastic association? **Opsoclonus**

### Orbital Neuroblastoma

**What is opsoclonus?**
A saccadic oscillation

**Ah, so it's a form of nystagmus, then?**
No, it's a form of saccadic disorder
Q

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**What is opsoclonus?**
A saccadic oscillation

**What’s the difference between a nystagmus and a saccadic disorder?**

Ah, so it’s a form of nystagmus, then? No, it’s a form of saccadic disorder
• What is the average age at diagnosis for neuroblastoma in general? 2 years
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What is opsoclonus? A saccadic oscillation. Ah, so it's a form of nystagmus, then? No, it's a form of saccadic disorder.

What's the difference between a nystagmus and a saccadic disorder? Both are involuntary repetitive eye movements. However, nystagmus initiates with a slow movement off the visual target, whereas a saccadic disorder initiates with a fast movement.
What is the average age at diagnosis for neuroblastoma in general? 2 years

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What is the classic ocular paraneoplastic association? Opsoclonus

What is opsoclonus? A saccadic oscillation

Ah, so it’s a form of nystagmus, then? No, it’s a form of saccadic disorder.

What’s the difference between a nystagmus and a saccadic disorder? Both are involuntary repetitive eye movements. However, nystagmus initiates with a slow movement off the visual target, whereas a saccadic disorder initiates with a fast movement.

By what other name is opsoclonus known?
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

What is the classic ocular paraneoplastic association? Opsoclonus

By what other name is opsoclonus known? Saccadomania

What is opsoclonus? A saccadic oscillation

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In opsoclonus... ...in what direction(s) do the eyes move? Large
--eyelid closure? Yes
--sleep? Yes
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In opsoclonus...
...in what direction(s) do the eyes move? It is multivectorial, so they move in all directions

Opsoclonus
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**In opsoclonus**…
...**in what direction(s) do the eyes move?** It is multivectorial, so they move in all directions
...**are the movements of small, or large amplitude?** Opsoclonus
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**In opsoclonus…**
- **…in what direction(s) do the eyes move?** It is multivectorial, so they move in all directions
- **…are the movements of small, or large amplitude?** Large
- **…do the movements persist during:**
  - **--eyelid closure?**
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So opsoclonus is secondary to Nb metastasizing to the orbit?
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So opsoclonus is secondary to Nb metastasizing to the orbit? No--as indicated by the question, it is a paraneoplastic process, ie, it is related to antigen cross-reactivity, not tumor bulk
Q

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**Orbital Neuroblastoma**

Another Nb-related paraneoplastic process is a syndrome involving myoclonus and ataxia along with opsoclonus. What is it called?
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*Another Nb-related paraneoplastic process is a syndrome involving myoclonus and ataxia along with opsoclonus. What is it called? It is called **opsoclonus-myoclonus-ataxia syndrome** (OMAS)*
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So, neuroblastoma can affect the eyes in a number of ways:

No question—advance when ready
● What is the average age at diagnosis for neuroblastoma in general? 2 years

● Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

● What are the two locations for the primary? The adrenals, and the sympathetic chain—Horner syndrome

● What percent of neuroblastomas metastasize to the orbit? 20%

● What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

● What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

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**Orbital Neuroblastoma**

If the primary is located in the cervical sympathetic chain, it can produce a **Horner syndrome**...

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So, *neuroblastoma can affect the eyes in a number of ways:*
What is the average age at diagnosis for neuroblastoma in general? 2 years

Where does neuroblastoma rank as a cause of orbital mets in childhood cancer? #1

What are the two locations for the primary? The adrenals, and the sympathetic chain (Horners)

What percent of neuroblastomas metastasize to the orbit? 20

What is the classic presentation of orbital mets? Proptosis and/or eyelid ecchymosis

If the primary is located in the cervical sympathetic chain, it can produce a Horner syndrome...

...it can metastasize to the orbit, resulting in proptosis and/or ecchymosis (both either unilateral or bilateral)

What if the child is <1 year at diagnosis—is the prognosis better or worse? The prognosis is better

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Orbital Neuroblastoma

So, neuroblastoma can affect the eyes in a number of ways:

Summary slide—no question