

Q

Orbital Neuroblastoma



- What is the average age at diagnosis for neuroblastoma in general?

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What proportion of Nb pts are diagnosed by age 5?

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



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Can Nb occur in adults?

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

Can Nb occur in adults?
Yes, although it is quite uncommon

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

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Where does Nb rank as a cause of cancer in childhood?

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*Where does Nb rank as a cause of cancer in childhood?
It is the most common cause of extracranial solid
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How about in infants (ie, prior to age 12 months)?



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What proportion of peds cancer deaths are due to Nb?



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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%

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mets in childhood

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

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As a rule: In adults, mets go to the [] (specifically, to the []); but in children, mets go to the []

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mets in childhood

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**



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Of all orbital mets in children, what proportion are due to Nb?



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Of all orbital mets in children, what proportion are due to Nb?
Almost 90%!

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The adrenals, and the sympathetic chain



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Which specific portion of the adrenal glands are we talking about here?



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The adrenal medulla



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

Q/A

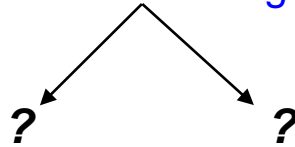
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A lot, actually, and they can be divvied up into two categories:



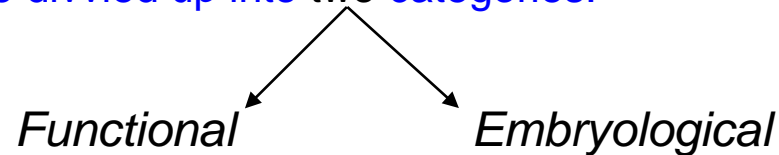


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Functionally, both the adrenal medulla and the sympathetic chain are components of the division of the nervous system.



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The secretory cells of the adrenal medulla and the neurons of the sympathetic chain both derive from neuroblasts, which in turn derive from the [redacted] cell line



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If the primary involves the superior portion of the cervical sympathetic chain, what clinical presentation might occur?



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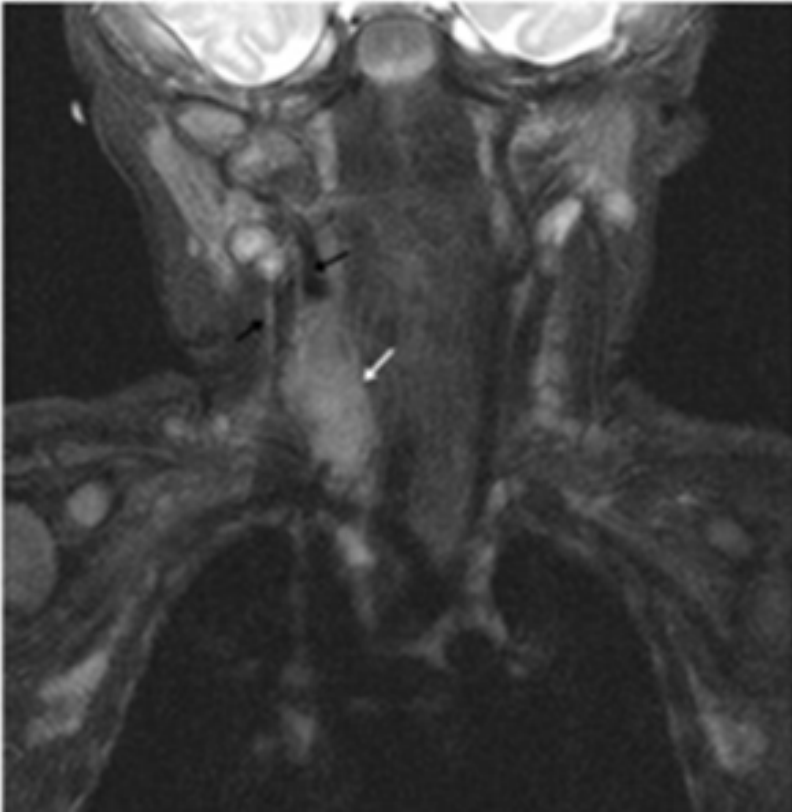
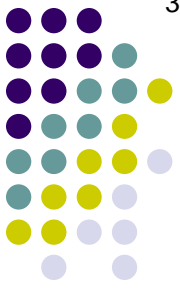
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A Horner syndrome

Orbital Neuroblastoma



Primary tumor in sympathetic chain



Horner syndrome 2ndry to Nb



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Is the ecchymosis unilateral, or bilateral?



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What critter-based description is used for the appearance of bilateral ecchymoses in Nb?



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'Raccoon eyes'*

Orbital Neuroblastoma



Nb: 'Raccoon eyes'



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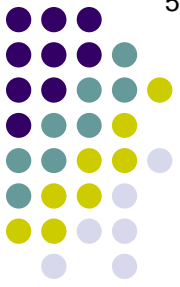
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Nb: Proptosis



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What is the DDX for acute-onset unilateral proptosis
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- Neuroblastoma
- Orbital inflammation
- Leukemia
- Orbital cellulitis
- Rhabdomyosarcoma



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What is the DDX for acute-onset bilateral proptosis in an infant/young child?

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- Neuroblastoma
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- ~~Rhabdomyosarcoma~~

Orbital cellulitis and rhabdo are very, very unlikely to present bilaterally

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



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What is the difference in 5-year survival rates between cases that present prior to vs after age 1 year?



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It is cut roughly in half



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- What is the classic ocular paraneoplastic association?

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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*



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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



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The adrenals, and the **sympathetic chain** **Horners**
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Proptosis and/or **eyelid ecchymosis**

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

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...and finally, it can produce a paraneoplastic syndrome resulting in **opsoclonus**

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