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- ____





kids)

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- --Osteosarcoma
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- --Fibrosarcoma



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- --Fibrosarcoma
- --Ewing sarcoma?

What about Ewing sarcoma? Doesn't it present in the orbit?



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- --Fibrosarcoma
- -- Ewing sarcoma? Not as primary

What about Ewing sarcoma? Doesn't it present in the orbit? Yes, but as a metastasis



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How much more common is rhabdo than the others?



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How many cases of orbital rhabdo occur in the US each year?





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How many cases of orbital rhabdo occur in the US each year? Somewhere between 25 and 100

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For more on Rb and its genetics, see slide-set R2

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As if a hx of germline Rb isn't bad enough: One modality used to tx Rb increases the risk of a subsequent sarcoma even more. What is this modality?



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As if a hx of germline Rb isn't bad enough: One modality used to tx Rb increases the risk of a subsequent sarcoma even more. What is this modality? **External-beam radiation treatment (XBRT)**. The increased risk of sarcoma is one reason clinicians use XBRT only reluctantly in the management of Rb.





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Orbital rhabdo



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Orbital rhabdo mimicking orbital cellulitis


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What is the classic presentation of **orbital rhabdo?** Rapid unilateral proptosis associated with lid edema and discoloration



What can clue you in to the fact that a child has rhabdo, not cellulitis? Children with orbital cellulitis are usually **systemically ill**, and the affected eye **hurts**. In contrast, children with rhabdo seem well, and 90% have no pain. (Beware the child who presents with 'orbital cellulitis' and is playing happily in your waiting room!)



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Is there a racial and/or gender predilection regarding who gets rhabdo?





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Is there a racial and/or gender predilection regarding who gets rhabdo? No to racial; there may be a slight male preponderance





What is the average age at diagnosis?
 to years







What is the average age at diagnosis? 5 to 7 years

(Per the Peds and Orbit books; 7-8 per the Path)





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Does rhabdo in infancy (ie, before age 1) carry a better, or worse prognosis?





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Does rhabdo in infancy (ie, before age 1) carry a better, or worse prognosis? Worse—it tends to be significantly more aggressive





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How does rhabdo presentation in a teenager differ from that in a younger child?





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How does rhabdo presentation in a teenager differ from that in a younger child? The course tend to be slower; eg, in a teen, the proptosis develops over a period of weeks rather than days





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 - Embryonal
 - Alveolar
 - Botryoid
 - Pleomorphic





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What is the average age at diagnosis? Which subtype is most common in the orbit? 5 to Embryonal • What year What 16 years • What **Embryonal**:

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57













Pleomorphic

60



61



- Alveolar
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- Embryonal: 80% of cases. Superonasal. 95% survival.
- Alveolar:
- Botryoid:
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 - **Botryoid**: Rare in orbit
 - **Pleomorphic**: Rare in orbit

The *botryoid* and *pleomorphic* subtypes are rarely found in the orbit





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Small/well localized tumor: Excision

don't be vs be very aggressive in excising, because of risk of involvement of adjacent structures by the surgery vs tumor





Small/well localized tumor: Excision

 Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery





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- Larger/more extensive:







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Treatment

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 - Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
- Larger/more extensive Chemo and RT

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Small/well localized tumor: Excision

 Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery

indicated

- Larger/more extensive: Chemo and RT
- Orbital exenteration is rarely vs
 commonly





- Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
- Larger/more extensive: Chemo and RT
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Small/well localized tumor: Excision

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Prognosis

[%] long-term survival overall





- Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
- Larger/more extensive: Chemo and RT
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- Prognosis
 - 90% long-term survival overall





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Prognosis

90% long-term survival overall

An important 'if' must be appended to make this figure true what is it? 90% survival **if**...





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 - Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
- Larger/more extensive: Chemo and RT
- Orbital exenteration is rarely indicated
- Prognosis
 - 90% long-term survival overall

An important 'if' must be appended to make this figure true what is it? 90% survival *if*...the tumor is confined to the orbit, with no bony invasion (survival drops markedly if this is not the case)





Small/well localized tumor: Excision

- Don't be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
- Larger/more extensive: Chemo and RT
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Prognosis

- 90% long-term survival overall
- Orbital rhabdo has best vs worst
 prognosis of any location





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In evaluating a rhabdo pt, make sure you rule out: **Local** metastasis by...





• Treatment

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In evaluating a rhabdo pt, make sure you rule out: **Local** metastasis by...palpating for cervical and preauricular lymphadenopathy



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• Prognosis

- 90% long-term survival overall
- Orbital rhabdo has best prognosis of any location

In evaluating a rhabdo pt, make sure you rule out: Local metastasis by...palpating for cervical and preauricular lymphadenopathy Systemic metastasis via three studies/procedures





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- Larger/more extensive: Chemo and I
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• Prognosis

- 90% long-term survival overall
- Orbital rhabdo has best prognosis of any location

In evaluating a rhabdo pt, make sure you rule out: Local metastasis by...palpating for cervical and preauricular lymphadenopathy Systemic metastasis via CXR, lumbar puncture and bone-marrow biopsy