Orbital Rhabdomyosarcoma

Which is more common in kids--orbital carcinomas, or sarcomas?
Orbital Rhabdomyosarcoma

Which is more common in kids--orbital carcinomas, or sarcomas?
Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)
Orbital Rhabdomyosarcoma

Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children?
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Orbital Rhabdomyosarcoma

*Which is more common in kids--orbital carcinomas, or sarcomas?*  
Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

*What are the four most common orbital sarcoma primaries in children?*  
--Rhabdomyosarcoma  
--Osteosarcoma  
--Chondrosarcoma  
--Fibrosarcoma
Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children?
--Rhabdomyosarcoma
--Osteosarcoma
--Chondrosarcoma
--Fibrosarcoma
--Ewing sarcoma?

What about Ewing sarcoma? Doesn’t it present in the orbit?
Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children?
--Rhabdomyosarcoma
--Osteosarcoma
--Chondrosarcoma
--Fibrosarcoma
--Ewing sarcoma? Not as primary

What about Ewing sarcoma? Doesn't it present in the orbit? Yes, but as a metastasis
Which is more common in kids--orbital carcinomas, or sarcomas?
Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children?
--Rhabdomyosarcoma?
--Osteosarcoma?
--Chondrosarcoma?
--Fibrosarcoma?

Of these, which is most common?

Rhabdo, by a mile

The incidence of rhabdo is greater than that of the rest combined
Orbital Rhabdomyosarcoma

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How much more common is rhabdo than the others?
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*Rhabdo may be the most common orbital sarcoma, but is it a common condition in an absolute sense?*
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Of these, which is most common? Rhabdo, by a mile

How much more common is rhabdo than the others? The incidence of rhabdo is greater than that of the rest combined

Rhabdo may be the most common orbital sarcoma, but is it a common condition in an absolute sense? No, it is quite rare
Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children? 

--**Rhabdomyosarcoma!**  
--Osteosarcoma  
--Chondrosarcoma  
--Fibrosarcoma

Of these, which is most common?  
Rhabdo, by a mile

How much more common is rhabdo than the others?  
The incidence of rhabdo is greater than that of the rest combined

*Rhabdo may be the most common orbital sarcoma, but is it a common condition in an absolute sense?*  
No, it is quite rare

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

Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

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How much more common is rhabdo than the others? The incidence of rhabdo is greater than that of the rest combined

Rhabdo may be the most common orbital sarcoma, but is it a common condition in an absolute sense? No, it is quite rare

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

Which is more common in kids--orbital carcinomas, or sarcomas? Sarcoma, by a mile (carcinomas are vanishingly rare orbital tumors in kids)

What are the four most common orbital sarcoma primaries in children? --Rhabdomyosarcoma! --Osteosarcoma! --Chondrosarcoma! --Fibrosarcoma!

All that said, the presence of another, different malignancy greatly increases the risk of developing a childhood sarcoma. What is this other malignancy?
Q/A

**Orbital Rhabdomyosarcoma**

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How can a mutation be heritable without being inherited?
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How can a mutation be heritable without being inherited? By representing a sporadic mutation that occurred early enough during embryogenesis to be passed to (and thus present in) germline cells
Q

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This can happen in Rb?
Q/A

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*This can happen in Rb?*  
Not only can it happen, but it does, with rather alarming frequency—about % of Rb cases represent new germline mutations.
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For more on Rb and its genetics, see slide-set R2

*How can a mutation be heritable without being inherited?*

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*This can happen in Rb?*

Not only *can* it happen, but it *does*, with rather alarming frequency—about 1/3 of Rb cases represent new germline mutations. In fact, a heritable (read: germline) Rb mutation is *much* more likely to be new/sporadic than it is to be inherited.
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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?

All that said, the presence of another, different malignancy greatly increases the risk of developing a childhood sarcoma.
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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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What is the cell of origin for rhabdo?
**Orbital Rhabdomyosarcoma**

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Undifferentiated/pluripotent mesenchymal cells (note: **not** the EOMs!)
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What is the classic presentation of orbital rhabdo?
Rapid unilateral proptosis associated with lid edema and discoloration
Orbital Rhabdomyosarcoma

Orbital rhabdo
**Orbital Rhabdomyosarcoma**

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*With what process is it often confused?*
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Orbital cellulitis
Orbital Rhabdomyosarcoma

Orbital rhabdo mimicking orbital cellulitis
**Orbital Rhabdomyosarcoma**

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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 are in no pain. (Beware the child who presents with 'orbital cellulitis' and is playing happily in your waiting room!)
**Orbital Rhabdomyosarcoma**

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Orbital cellulitis?

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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With what process is it often confused?
Orbital cellulitis

Is there a racial and/or gender predilection regarding who gets rhabdo?
Which is more common in kids--orbital carcinomas, or sarcomas?
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With what process is it often confused?
Orbital cellulitis

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
What is the average age at diagnosis?

5 to 7 years

Note: The ‘5-7’ range is from the most recent edition of the Peds book. OTOH, the Orbit book gives a range of 8-10. (FWIW, at the time of this writing the Peds book is the more recent edition.) The EyeWiki entry gives a range of 7-8. Caveat emptor.
What is the average age at diagnosis?
5 to 7 years

What percent are diagnosed prior to age one year?
What is the average age at diagnosis? 5 to 7 years
What percent are diagnosed prior to age one year? 5
Q

- What is the average age at diagnosis? 5 to 7 years
- What percent are diagnosed prior to age one year? 5

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

Orbital Rhabdomyosarcoma
A

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis? 5 to 7 years
- What percent are diagnosed prior to age one year? 5

Does rhabdo in infancy (ie, before age 1) carry a better, or worse prognosis? Worse—it tends to be significantly more aggressive
What is the average age at diagnosis?  
5 to 7 years

What percent are diagnosed prior to age one year? 5

What percent are diagnosed after age 16 years?
What is the average age at diagnosis? 5 to 7 years

What percent are diagnosed prior to age one year? 5

What percent are diagnosed after age 16 years? 10
What is the average age at diagnosis? 5 to 7 years
What percent are diagnosed prior to age one year? 5
What percent are diagnosed after age 16 years? 10

How does rhabdo presentation in a teenager differ from that in a younger child?
What is the average age at diagnosis? 5 to 7 years

What percent are diagnosed prior to age one year? 5

What percent are diagnosed after age 16 years? 10

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
Q

- What is the average age at diagnosis? 5 to 7 years
- What percent are diagnosed prior to age one year? 5
- What percent are diagnosed after age 16 years? 10
- What are the 4 histological subtypes?
  - Embryonal
  - Alveolar
  - Botryoid
  - Pleomorphic
A

Orbital Rhabdomyosarcoma

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- What are the 4 histological subtypes?
  - Embryonal:
  - Alveolar:
  - Botryoid:
  - Pleomorphic: Rare in orbit

Which subtype is most common in the orbit? Embryonal

What proportion of cases are embryonal? About 80%

In which section of the orbit does embryonal tend to occur? The superonasal

What is the long-term survival rate? 95%
What is the average age at diagnosis? 5 to 7 years
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What proportion of cases are embryonal? About 80%
In which section of the orbit does embryonal tend to occur?

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

What percent are diagnosed prior to age one year? 5%

What percent are diagnosed after age 16 years? 10%

What are the 4 histological subtypes?
- Embryonal: 80% of cases. Superonasal.
- Alveolar:
- Botryoid:
- Pleomorphic: Rare in orbit

Which subtype is most common in the orbit? Embryonal

What proportion of cases are embryonal? About 80%

In which section of the orbit does embryonal tend to occur? The superonasal

Embryonal: 80% of cases. Superonasal.
What is the average age at diagnosis? 5 to 7 years
What percent are diagnosed prior to age one year? 5
What percent are diagnosed after age 16 years? 10
What are the 4 histological subtypes?
- Embryonal: 80% of cases. Superonasal.
- Alveolar:
- Botryoid:
- Pleomorphic: Rare in orbit

Which subtype is most common in the orbit? Embryonal
What proportion of cases are embryonal? About 80%
In which section of the orbit does embryonal tend to occur? The superonasal
What is the 5-year survival rate for embryonal rhabdo? 95%

Orbital Rhabdomyosarcoma
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Orbital Rhabdomyosarcoma
What is the average age at diagnosis?
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What are the 4 histological subtypes?
- Embryonal: 80% of cases. Superonasal. 95% survival.
- Alveolar:
- Botryoid:
- Pleomorphic: Rare in orbit

Which subtype has the poorest prognosis?
Alveolar

What proportion of cases are alveolar?
About 10%

In which section of the orbit does alveolar tend to occur?
The inferior

What is the long-term survival rate for alveolar rhabdo?
95%
Orbital Rhabdomyosarcoma

- What is the average age at diagnosis? 5 to 7 years
- What percent are diagnosed prior to age one year? 5
- What percent are diagnosed after age 16 years? 10
- What are the 4 histological subtypes?
  - Embryonal: 80% of cases. Superonasal. 95% survival.
  - Alveolar
  - Botryoid
  - Pleomorphic: Rare in orbit
- Which subtype has the poorest prognosis? Alveolar
- What proportion of cases are alveolar? About 10%
- In which section of the orbit does alveolar tend to occur? The inferior
- What is the long-term survival rate for alveolar rhabdo? 75%
Q

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

What percent are diagnosed prior to age one year? 5

What percent are diagnosed after age 16 years? 10

What are the 4 histological subtypes?

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- **Alveolar**
- **Botryoid**
- **Pleomorphic**: Rare in orbit

Which subtype has the poorest prognosis? Alveolar

What proportion of cases are alveolar? About 10%

In which section of the orbit does alveolar tend to occur? The inferior

What is the long-term survival rate for alveolar rhabdo? 75%
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What proportion of cases are alveolar? About 10%

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**Orbital Rhabdomyosarcoma**
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Long-term survival rate for alveolar rhabdomyosarcoma?
75%

Orbital Rhabdomyosarcoma
**Q**

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

- **What percent are diagnosed prior to age one year?**
  - 5

- **What percent are diagnosed after age 16 years?**
  - 10

- **What are the 4 histological subtypes?**
  - **Embryonal:** 80% of cases. Superonasal. 95% survival.
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  - **Pleomorphic:** Rare in orbit

- **Which subtype has the poorest prognosis?**
  - Alveolar

- **What proportion of cases are alveolar?**
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- **In which section of the orbit does alveolar tend to occur?**
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- **What is the 5-year survival rate for alveolar rhabdo?**
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- **Alveolar**: 10% of cases. Inferior. 75% survival.
- **Botryoid**:
- **Pleomorphic**: Rare in orbit.

Which subtype has the poorest prognosis? **Alveolar**.

What proportion of cases are alveolar? About 10%.

In which section of the orbit does alveolar tend to occur? The inferior.

What is the 5-year survival rate for alveolar rhabdo? 75%.

**Orbital Rhabdomyosarcoma**
Orbital Rhabdomyosarcoma

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

- What percent are diagnosed prior to age one year?
  - About 10%

- What percent are diagnosed after age 16 years?
  - About 10%

- What are the 4 histological subtypes?
  - **Embryonal**: 80% of cases. Superonasal. 95% survival.
  - **Alveolar**: 10% of cases. Inferior. 75% survival, but only 10% at 10 years.
  - **Botryoid**: Rare in orbit
  - **Pleomorphic**

- Which subtype has the poorest prognosis?
  - Alveolar

- What proportion of cases are alveolar?
  - About 10%

- In which section of the orbit does alveolar tend to occur?
  - The inferior

- What is the 5-year survival rate for alveolar rhabdo?
  - 75%

The *Orbit* book mentions a 10-year survival rate of only 10% for alveolar. I’m not sure what to make of this—perhaps it’s referring to untreated disease? Caveat emptor.
What is the average age at diagnosis? 5 to 7 years

What percent are diagnosed prior to age one year? 5

What percent are diagnosed after age 16 years? 10

What are the 4 histological subtypes?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.
- **Alveolar**: 10% of cases. Inferior. 75% survival.
- **Botryoid**: Rare in orbit
- **Pleomorphic**: Rare in orbit

The botryoid and pleomorphic subtypes are rarely found in the orbit.
Q

Orbital Rhabdomyosarcoma

Treatment

- Small/well localized tumor:
A

**Orbital Rhabdomyosarcoma**

- **Treatment**
  - Small/well localized tumor: **Excision**
Treatment

- 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
Orbital Rhabdomyosarcoma

- Treatment
  - Small/well localized tumor: **Excision**
    - Don’t be aggressive in excising, because of risk of involvement of adjacent structures by the surgery
Treatment

- 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: [Blank] and **Abb.**
A

Orbital Rhabdomyosarcoma

- **Treatment**
  - 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**
Orbital Rhabdomyosarcoma

Treatment

- 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**
- Orbital exenteration is **rarely** indicated vs **commonly**
Orbital Rhabdomyosarcoma

- **Treatment**
  - 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**
  - Orbital exenteration is **rarely** indicated
Treatment

- 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**
- Orbital exenteration is **rarely** indicated

Prognosis

- **%** long-term survival overall

*Orbital Rhabdomyosarcoma*
## Treatment

- **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*
- Orbital exenteration is *rarely* indicated

## Prognosis

- *90%* long-term survival overall
Q

Orbital Rhabdomyosarcoma

- **Treatment**
  - 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**
  - Orbital exenteration is **rarely** indicated

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

Treatment

- 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**
- Orbital exenteration is rarely indicated

Prognosis

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

- **Treatment**
  - 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**
  - Orbital exenteration is **rarely** indicated

- **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…

Q/A

- **Orbital Rhabdomyosarcoma**

- **Treatment**
  - 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**
  - Orbital exenteration is rarely indicated

- **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 two areas lymphadenopathy
Orbital Rhabdomyosarcoma

Treatment

- 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**
- Orbital exenteration is rarely indicated

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
Orbital Rhabdomyosarcoma

Treatment
- 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**
- Orbital exenteration is **rarely** indicated

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
Orbital Rhabdomyosarcoma

Treatment

- 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**
- Orbital exenteration is *rarely* indicated

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**