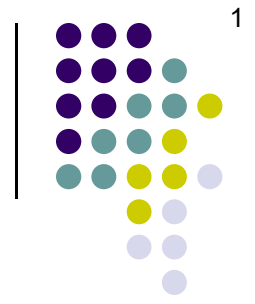


Q

Orbital Rhabdomyosarcoma

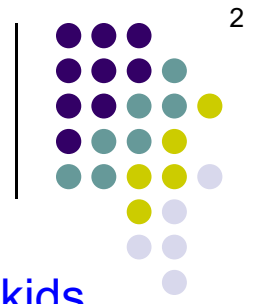
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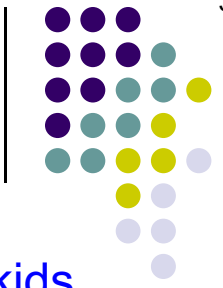


A

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





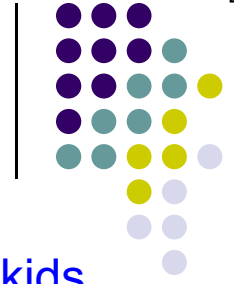
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-
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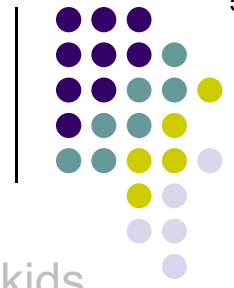
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- Osteosarcoma
- Chondrosarcoma
- Fibrosarcoma



Q

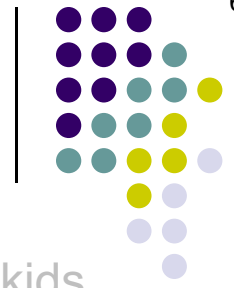
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- Ewing sarcoma?*

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



A

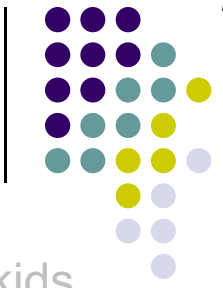
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What about Ewing sarcoma? Doesn't it present in the orbit?
Yes, but as a metastasis



Q

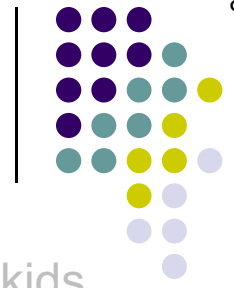
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Of these, which is most common?



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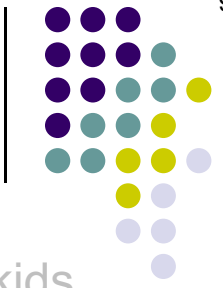
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--Osteosarcoma

--Chondrosarcoma

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Rhabdo, by a mile



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How much more common is rhabdo than the others?



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



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



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Somewhere between 25 and 100



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



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Retinoblastoma (Rb) (But note: To convey an increased risk of sarcoma, the Rb mutation must be in the **germline**)



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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 can clue you in to the fact that a child has rhabdo, not cellulitis?



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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!)



Q

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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



Q

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?
[] to [] years



A

Orbital Rhabdomyosarcoma

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

Orbital Rhabdomyosarcoma



- 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.



Q

Orbital Rhabdomyosarcoma

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



A

Orbital Rhabdomyosarcoma

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



Q

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?



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



Q

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?



A

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?
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Q

Orbital Rhabdomyosarcoma

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



A

Orbital Rhabdomyosarcoma

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5 to 7 years
- What percent are diagnosed prior to age one year? 5
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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



Q

Orbital Rhabdomyosarcoma

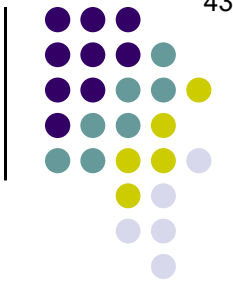
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- What are the 4 histological subtypes?
 - [Redacted]
 - [Redacted]
 - [Redacted]
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A

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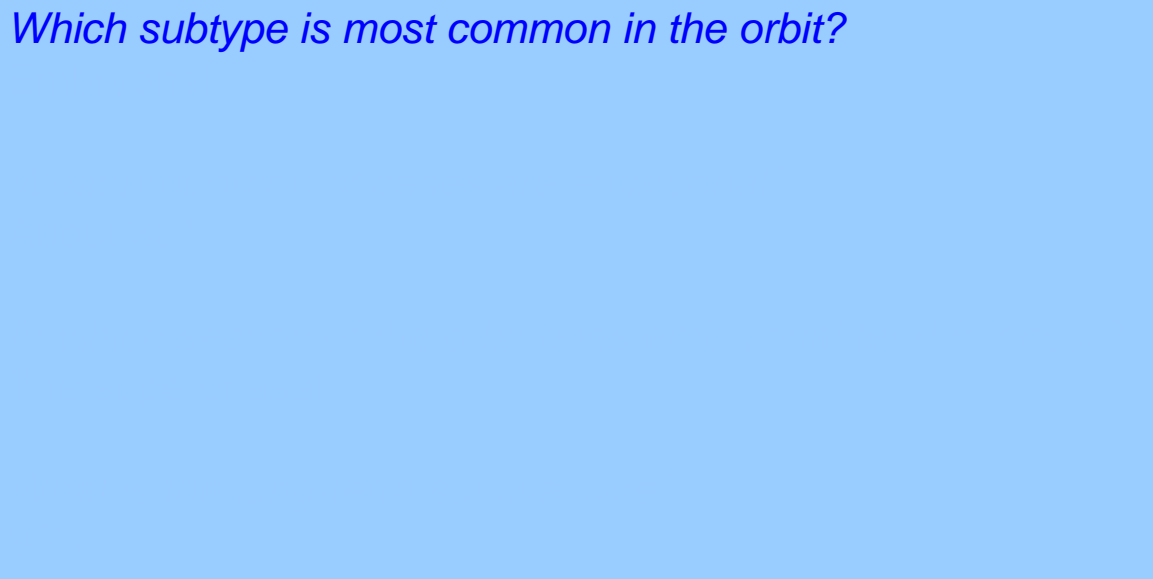


Q

Orbital Rhabdomyosarcoma

● What is the average age at diagnosis?

5 to 7



Which subtype is most common in the orbit?

● What age one year?

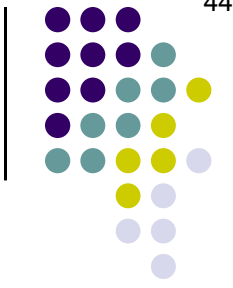
age one

● What years

16

● What

- Embryonal:
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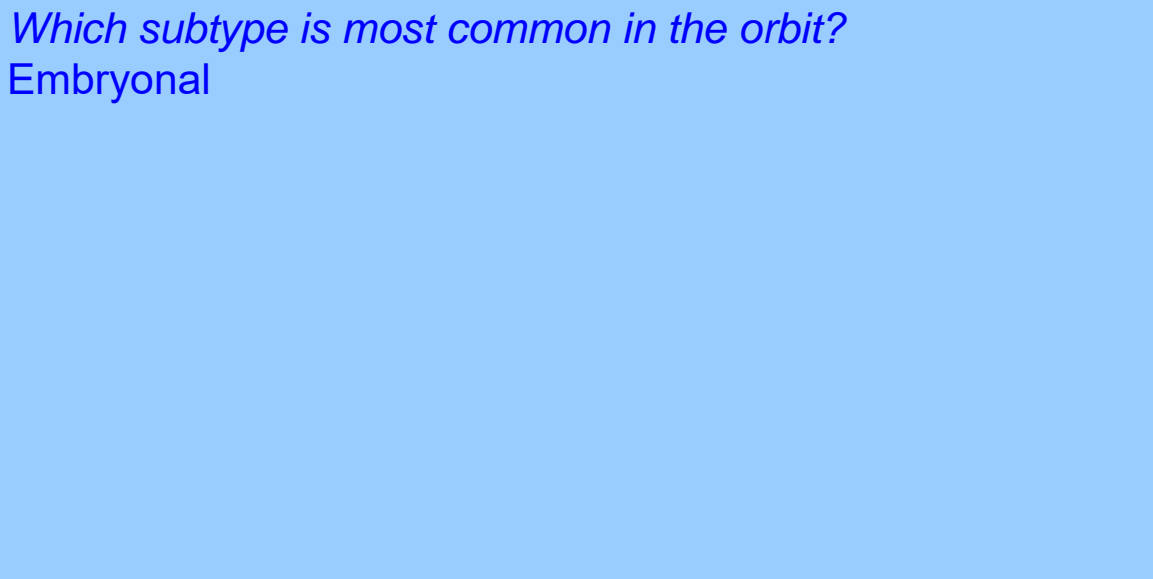


A

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● What is the average age at diagnosis?

5 to 7



Which subtype is most common in the orbit?
Embryonal

● What is the average age one year?

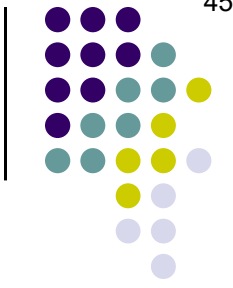
16

● What is the average age 16 years?

16

● What is the average age 16 years?

- Embryonal:
- Alveolar:
- Botryoid:
- Pleomorphic:



Q

Orbital Rhabdomyosarcoma

● What is the average age at diagnosis?

5 to 7

Which subtype is most common in the orbit?
Embryonal

What proportion of cases are embryonal?

16

● What age group?

● What age group?

● What age group?

- **Embryonal:**
- Alveolar:
- Botryoid:
- Pleomorphic:



A

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype is most common in the orbit?

Embryonal

- What age group?

What proportion of cases are embryonal?

About 80%

- What age group?

age one

- What age group?

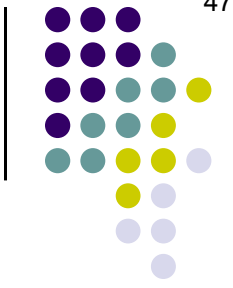
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- Alveolar:

- Botryoid:

- Pleomorphic:



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

● What

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

The superonasal

- What

- **Embryonal**: 80% of cases. Superonasal.

- Alveolar:

- Botryoid:

- Pleomorphic:



Q

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype is most common in the orbit?

Embryonal

- What is the average age at diagnosis in 16-year-olds?

What proportion of cases are embryonal?

About 80%

- What is the average age at diagnosis in 16-year-olds?

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

The superonasal

- What is the 5-year survival rate for embryonal rhabdo?

What is the 5-year survival rate for embryonal rhabdo?

- **Embryonal**: 80% of cases. Superonasal.

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95%

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- Alveolar:

- Botryoid:

- Pleomorphic:



Q

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype has the poorest prognosis?

- What is the average age at diagnosis? age one

- What is the average age at diagnosis? age 16

- What is the average age at diagnosis? ?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- **Alveolar**:

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Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype has the poorest prognosis?

Alveolar

- What is the average age at diagnosis for the embryonal subtype?

age one

- What is the average age at diagnosis for the alveolar subtype?

age 16

- What is the average age at diagnosis for the botryoid subtype?

?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- **Alveolar**:

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Q

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype has the poorest prognosis?

Alveolar

- What is the average age at diagnosis in children under 1 year?

What proportion of cases are alveolar?

age one

- What is the average age at diagnosis in children 1 to 16 years?

e 16

- What is the average age at diagnosis in children over 16 years?

?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

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- **Pleomorphic**:



A

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype has the poorest prognosis?

Alveolar

- What is the average age at diagnosis in children under 1 year of age?

What proportion of cases are alveolar?

About 10%

- What is the average age at diagnosis in children 1 to 16 years of age?

- What is the average age at diagnosis in children over 16 years of age?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- **Alveolar**: 10% of cases.

- **Botryoid**:

- **Pleomorphic**:



Q

Orbital Rhabdomyosarcoma

- What is the average age at diagnosis?

5 to 7

Which subtype has the poorest prognosis?

Alveolar

- What is the average age at diagnosis for alveolar cases?

What proportion of cases are alveolar?

About 10%

- What is the average age at diagnosis for embryonal cases?

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

- What is the average age at diagnosis for pleomorphic cases?

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- **Alveolar**: 10% of cases.

- **Botryoid**:

- **Pleomorphic**:



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5 to 7

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Alveolar

- What age group?

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About 10%

- What age group?

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

The inferior

- What?

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- **Botryoid**:

- **Pleomorphic**:



Q

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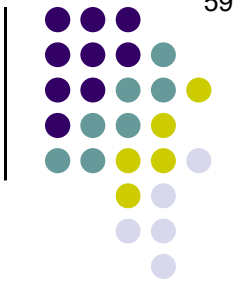
75%

- **Embryonal**: 80% of cases. Superonasal. 95% survival.

- **Alveolar**: 10% of cases. Inferior. 75% survival.

- **Botryoid**:

- **Pleomorphic**:



Orbital Rhabdomyosarcoma

● What is the average age at diagnosis?

5 to 7

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?
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What is the 5-year survival rate for alveolar rhabdo?
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● What age one

● What e 16

● What ?

● **Embryonal**: 80% of cases. Superonasal. 95% survival.

● **Alveolar**: 10% of cases. Inferior. 75% survival,

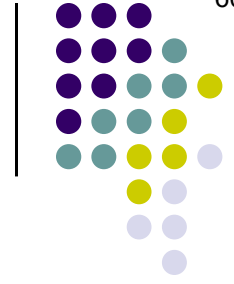
but only 10% at 10 years?

● **Botryoid**:

● **Pleomorphic**:

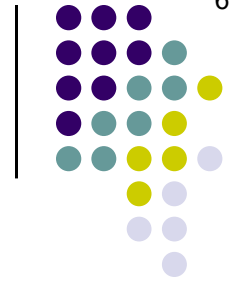
The *Orbit* book mentions a 10-year survival rate of only 10% for alveolar. I'm not sure what to make of this--perhaps it's referring to untreated disease? Caveat emptor.

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**: 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

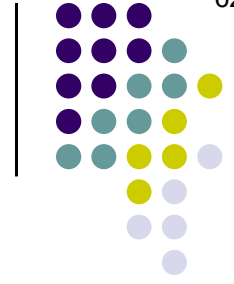


Q

Orbital Rhabdomyosarcoma

- Treatment

- Small/well localized tumor:



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Orbital Rhabdomyosarcoma

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



Q

Orbital Rhabdomyosarcoma

- 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



A

Orbital Rhabdomyosarcoma

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Q

Orbital Rhabdomyosarcoma

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- Larger/more extensive: and abb.



A

Orbital Rhabdomyosarcoma

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 - Larger/more extensive: **Chemo** and **RT**



Q

Orbital Rhabdomyosarcoma

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



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● Prognosis

- **%** long-term survival



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- Prognosis
 - **90%** long-term survival overall



Q

Orbital Rhabdomyosarcoma

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● Prognosis

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



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Orbital Rhabdomyosarcoma

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Q

Orbital Rhabdomyosarcoma

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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 lymphadenopathy.

systemic metastasis via...



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 cervical and preauricular lymphadenopathy.
systemic metastasis via...CXR, lumbar puncture and bone-marrow biopsy