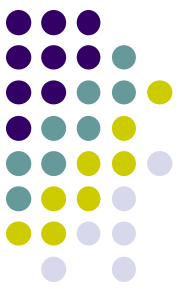


# Q



- **Duane's Retraction Syndrome**

- Motility disorder featuring:
  - 1) 

an abnormal  
eye movement

 of globe on attempted adduction
  - 2)
  - 3)

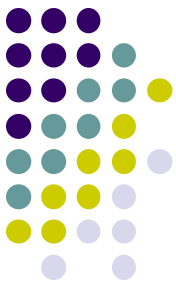


# A

- **Duane's Retraction Syndrome**
  - Motility disorder featuring:
    - 1) **Retraction** of globe on attempted adduction
    - 2)
    - 3)



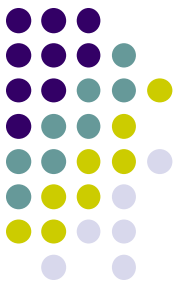
Duane syndrome: Globe retraction



# Q

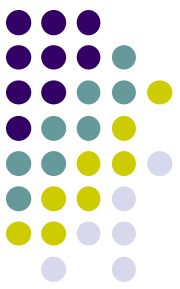
## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of a normal eye movement
  - 3)

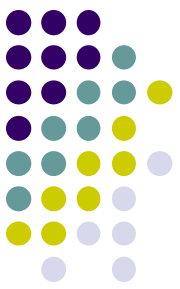


# A

- **Duane's Retraction Syndrome**
  - Motility disorder featuring:
    - 1) **Retraction** of globe on attempted adduction
    - 2) At least some limitation of **horizontal movement**
    - 3)



Duane syndrome: Horizontal movement limitation

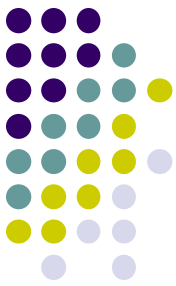


# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) **Retraction** of globe on attempted adduction
  - 2) At least some limitation of **horizontal movement**
  - 3) Up- or downshoot in 

eye position

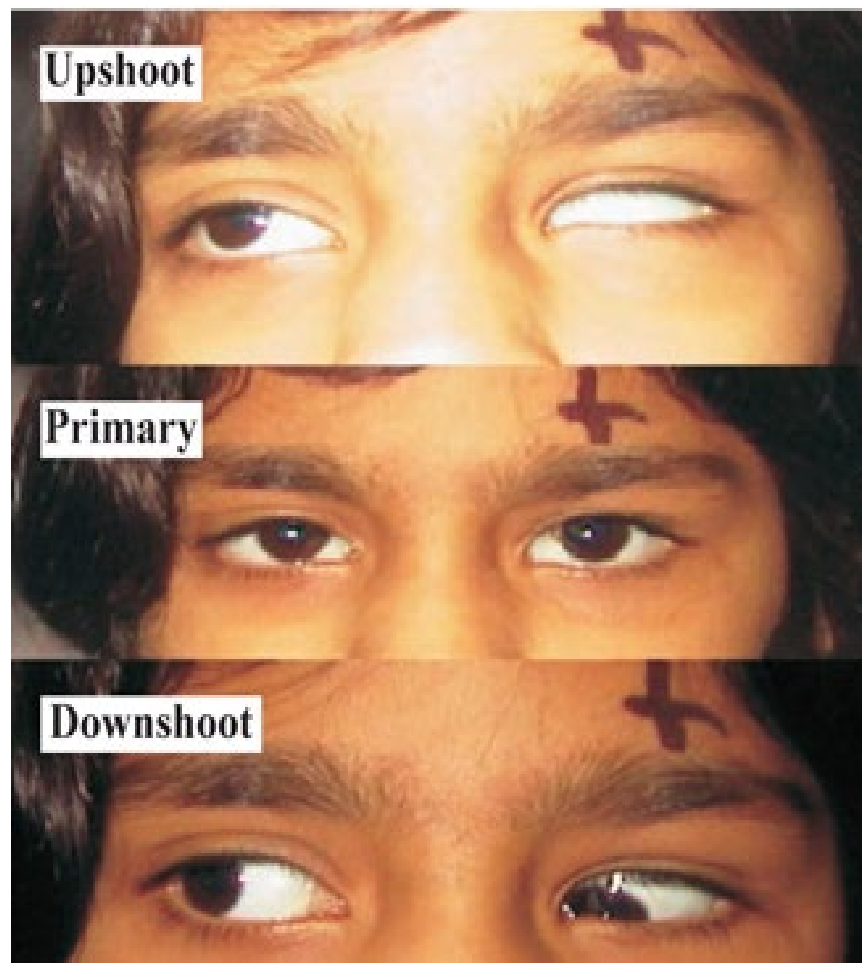


# A

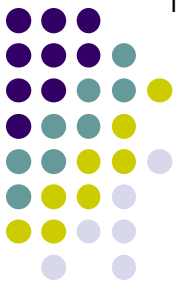
- **Duane's Retraction Syndrome**

- Motility disorder featuring:
  - 1) **Retraction** of globe on attempted adduction
  - 2) At least some limitation of **horizontal movement**
  - 3) Up- or downshoot in **adduction**





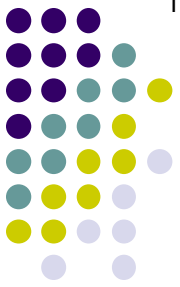
Duane syndrome: Upshoot/downshoot



# Q

## ● Duane's Retraction Syndrome

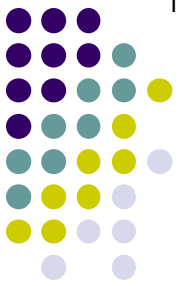
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- % sporadic, % AD



# A

## ● Duane's Retraction Syndrome

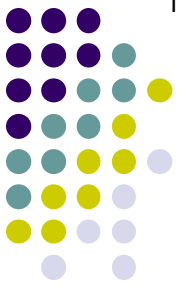
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD



# Q

## ● Duane's Retraction Syndrome

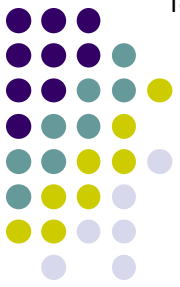
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with [redacted] syndrome



# A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome



# Q

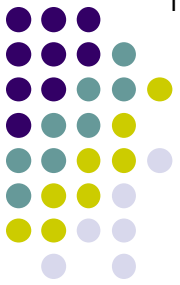
- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**



# A

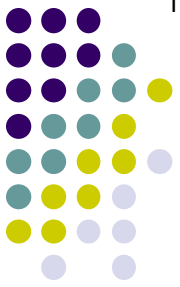
- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*  
About 1/4000 live births

attempted adduction  
of horizontal movement  
duction

**Goldenhar** syndrome



# Q

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

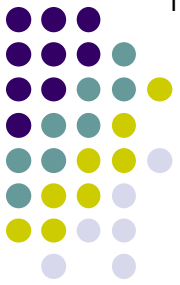
About 1/4000 live births

*What is its inheritance pattern?*

attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**





# A

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

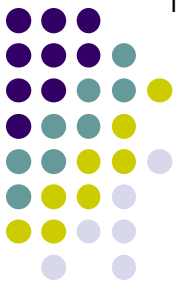
It is sporadic

attempted adduction

of horizontal movement

duction

**Goldenhar** syndrome



# Q

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

*Is there a sex predilection?*

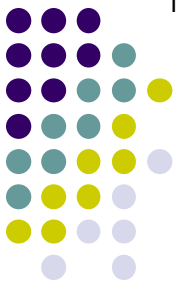
attempted adduction

of horizontal movement

duction

)

**Goldenhar syndrome**



# Q/A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

*Is there a sex predilection?*

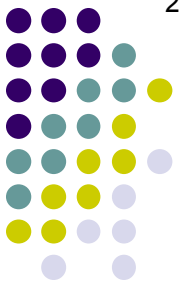
Yes, **M v F** are **# x** as likely to be affected

attempted adduction

of horizontal movement

duction

**Goldenhar** syndrome



# A

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

*Is there a sex predilection?*

Yes, males are twice as likely to be affected

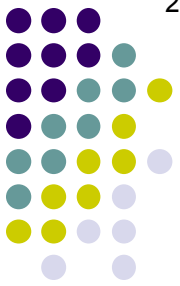
attempted adduction

of horizontal movement

duction

)

**Goldenhar syndrome**



# Q

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

*Is there a sex predilection?*

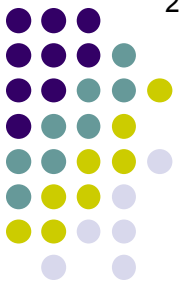
Yes, males are twice as likely to be affected

*In two words, what sort of condition is Goldenhar?*

A

attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**



# A

- Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

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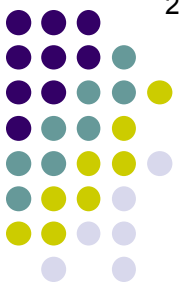
Yes, males are twice as likely to be affected

*In two words, what sort of condition is Goldenhar?*

A craniofacial malformation

attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**



# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

*What is its inheritance pattern?*

It is sporadic

*Is there a sex predilection?*

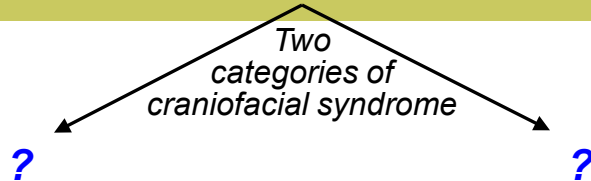
Yes, males are twice as likely to be affected

*In two words, what sort of condition is Goldenhar?*

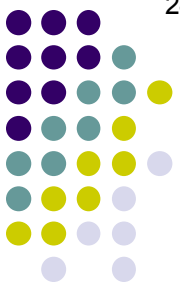
A **craniofacial malformation**

attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**



*What are the two categories of craniofacial syndrome?*



# A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

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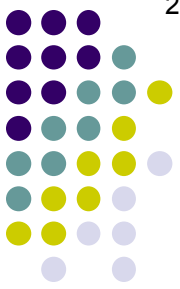
Two  
categories of  
craniofacial syndrome

**Craniosynostoses**

**Not craniosynostoses**

*What are the two categories of craniofacial syndrome?*





# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

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duction

**Goldenhar syndrome**

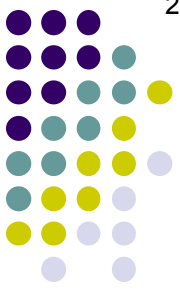
Two  
categories of  
craniofacial syndrome

**Craniosynostoses**

**Not craniosynostoses**

--?  
--?  
--?  
--?

*Which craniosynostosis syndromes  
are addressed in the Peds book?*



# A

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- Motility disorder featuring:

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duction

**Goldenhar syndrome**

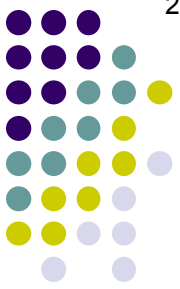
Two  
categories of  
craniofacial syndrome

**Craniosynostoses**

- Crouzon
- Apert
- Pfeiffer
- Saethre-Chotzen

**Not craniosynostoses**

*Which craniosynostosis syndromes  
are addressed in the Peds book?*



# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

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attempted adduction  
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**Goldenhar syndrome**

Two  
categories of  
craniofacial syndrome

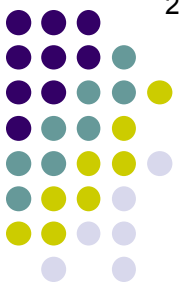
**Craniosynostoses**

- Crouzon
- Apert
- Pfeiffer
- Saethre-Chotzen

**Not craniosynostoses**

- ?
- ?
- ?
- ?

*Which non-craniosynostosis conditions  
are addressed in the Peds book?*



# A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:

*What is the incidence of Goldenhar?*

About 1/4000 live births

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*In two words, what sort of condition is Goldenhar?*

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attempted adduction  
of horizontal movement  
duction

**Goldenhar syndrome**

Two  
categories of  
craniofacial syndrome

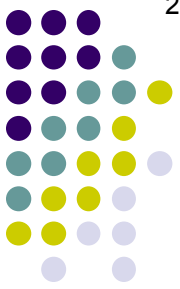
**Craniosynostoses**

- Crouzon
- Apert
- Pfeiffer
- Saethre-Chotzen

**Not craniosynostoses**

- Goldenhar
- Treacher Collins
- Pierre Robin sequence
- Fetal alcohol

*Which non-craniosynostosis conditions  
are addressed in the Peds book?*



## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) [redacted] of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in [redacted]

- [redacted] sporadic, [redacted] AD

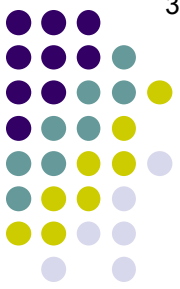
- Use "Goldenhar"

● C

G  
O  
L  
D  
E  
N  
H  
A  
R

th **Goldenhar syndrome**

As we will see, the word *Goldenhar* provides a very convenient mnemonic for remembering the important features of Goldenhar syndrome!



What is its noneponymous name?

O



ent

Goldenhar

O



L

D

E

N

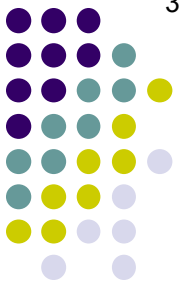
H

A

R

Very  
convenient  
mnemonic

Goldenhar syndrome



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

ent

- Us
- O

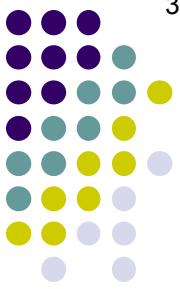
**Goldenhar**

**OAV syndrome**

th **Goldenhar syndrome**

*Very  
convenient  
mnemonic*

**L  
D  
E  
N  
H  
A  
R**



What is its noneponymous name?

Oculo-Auriculo-**Vertebral** (OAV) syndrome

What is the classic vertebral finding?

- Us
- O

**Goldenhar**

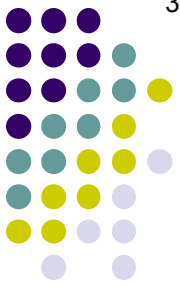
**OAV** syndrome

Very  
convenient  
mnemonic

**L  
D  
E  
N  
H  
A  
R**

**Goldenhar** syndrome





What is its noneponymous name?

Oculo-Auriculo-**Vertebral** (OAV) syndrome

What is the classic vertebral finding?

Hemivertebrae, aka perty...

ent

- Us
- OAV syndrome

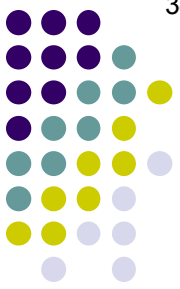
Very  
convenient  
mnemonic

**Goldenhar**

**OAV** syndrome

**L  
D  
E  
N  
H  
A  
R**

**Goldenhar syndrome**



What is its noneponymous name?

Oculo-Auriculo-**Vertebral** (OAV) syndrome

What is the classic vertebral finding?

Hemivertebrae, aka **butterfly vertebrae**

ent

- Us
- OAV syndrome

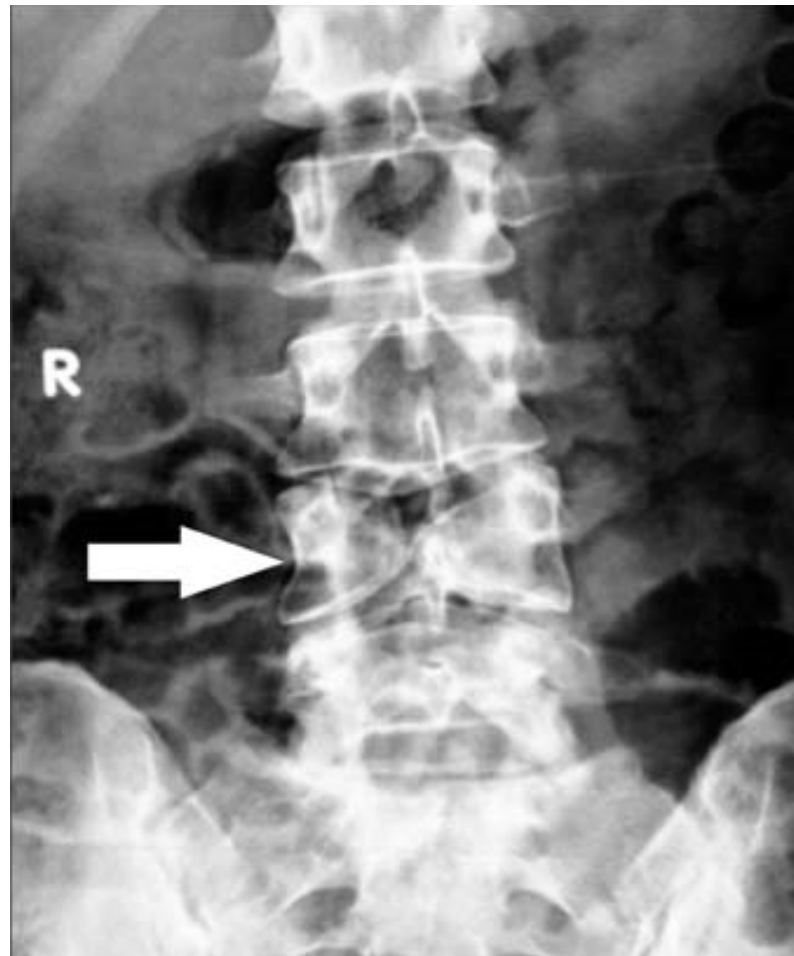
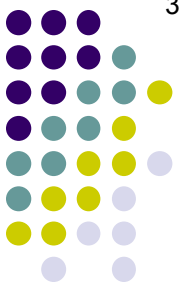
Very  
convenient  
mnemonic

**Goldenhar**

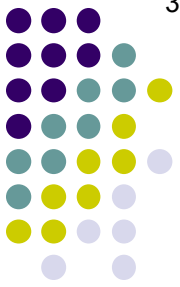
**OAV** syndrome

**L  
D  
E  
N  
H  
A  
R**

th **Goldenhar** syndrome



Goldenhar syndrome: Butterfly vertebrae



What is its noneponymous name?

Oculo-Auriculo-**Vertebral** (OAV) syndrome

What is the classic vertebral finding?

Hemivertebrae, aka **butterfly vertebrae**

Another syndrome of ophthalmic concern includes butterfly vertebrae as a finding. What is it?

ent

- Us
- OAV syndrome

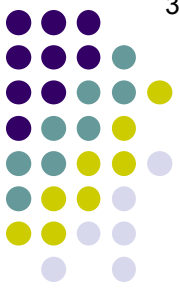
Very  
convenient  
mnemonic

**Goldenhar**

**OAV syndrome**

**L  
D  
E  
N  
H  
A  
R**

th **Goldenhar syndrome**



What is its noneponymous name?

Oculo-Auriculo-**Vertebral** (OAV) syndrome

What is the classic vertebral finding?

Hemivertebrae, aka **butterfly vertebrae**

Another syndrome of ophthalmic concern includes butterfly vertebrae as a finding. What is it?

**Alagille syndrome.** If you want more info on Alagille syndrome--and if you don't know it, you should--check out the slide-set on *anterior segment dysgenesis*.

ent

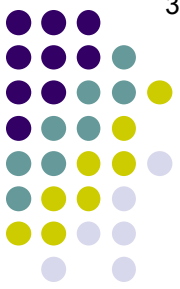
Goldenhar

OAV syndrome

L  
D  
E  
N  
H  
A  
R

Very  
convenient  
mnemonic

Goldenhar syndrome



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--L ←  
--D ←

ent

- Us
- O

**Goldenhar**

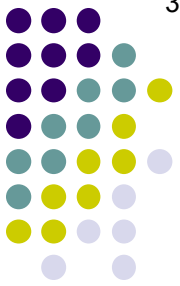
**OAV syndrome**

L ←  
D ←

Very  
convenient  
mnemonic

**E  
N  
H  
A  
R**

th **Goldenhar syndrome**



*What is its noneponymous name?*

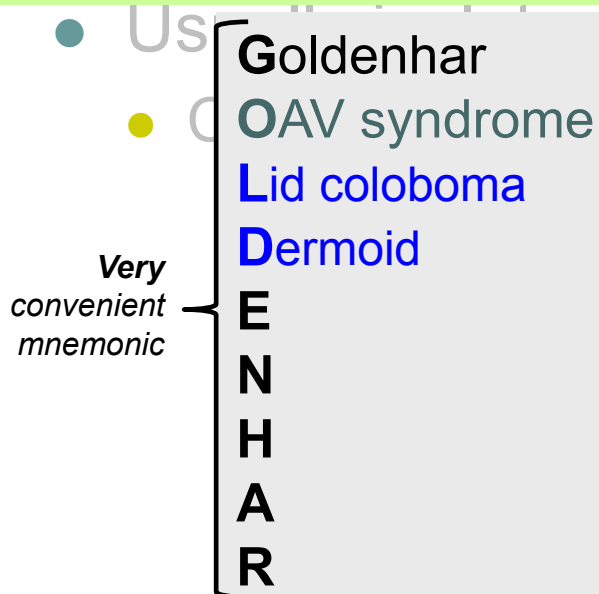
**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

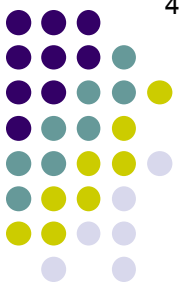
--**Lid coloboma**

--**Dermoids of the cornea**

ent



**Goldenhar syndrome**



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--**Lid coloboma**

--Dermoids of the cornea

*Does the coloboma tend to be in the upper lid, or the lower?*

ent

- Us
  - C
- Very convenient mnemonic
- E**  
**N**  
**H**  
**A**  
**R**

**Goldenhar**

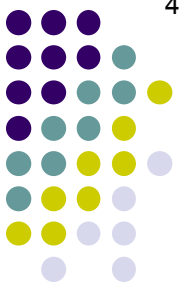
**OAV syndrome**

**Lid coloboma**

**Dermoid**

th **Goldenhar syndrome**





What is its noneponymous name?

Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

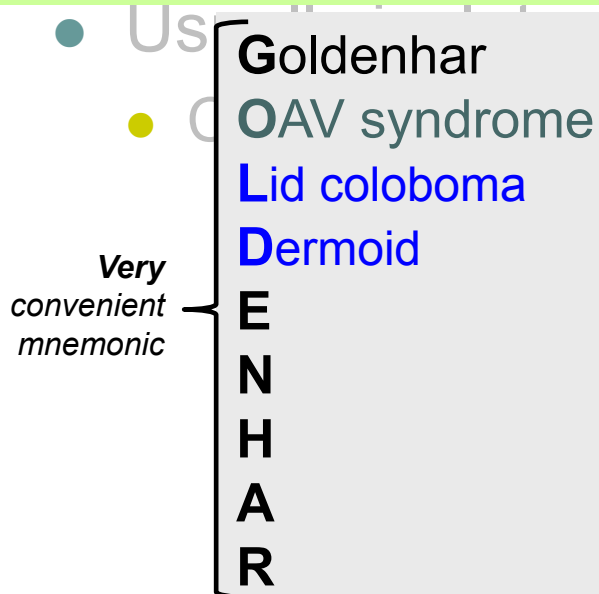
--Lid coloboma

--Dermoids of the cornea

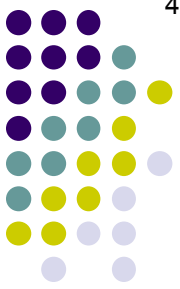
Does the coloboma tend to be in the upper lid, or the lower?

Depends on who you ask. The BCSC Cornea book says the upper, whereas the *Plastics* book indicates the lower. (The *Peds* book doesn't address this issue.) Caveat emptor.

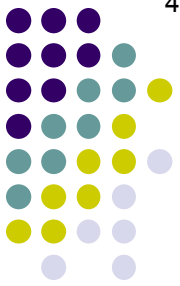
ent



Goldenhar syndrome



Goldenhar syndrome: Lid coloboma



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

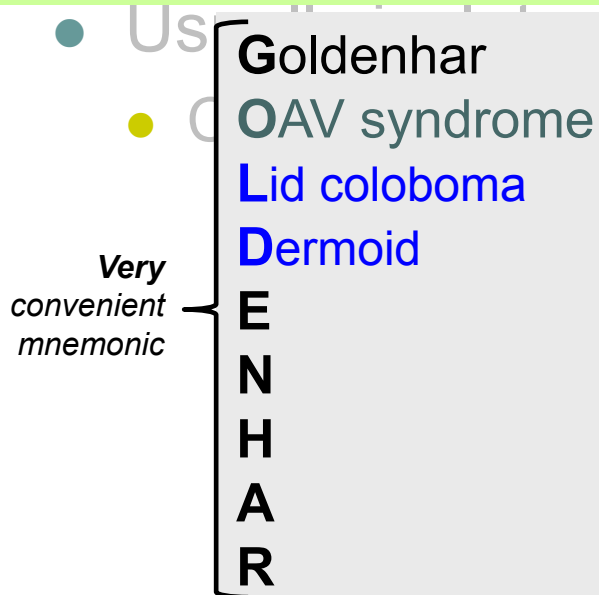
--Lid coloboma

--**Dermoids of the cornea**

*What is the 'full' name of the dermoid in question?*

one word dermoid

ent



th **Goldenhar syndrome**



What is its noneponymous name?

**Oculo-Auriculo-Vertebral (OAV) syndrome**

What other ocular/periocular abnormalities are common in Goldenhar?

--Lid coloboma

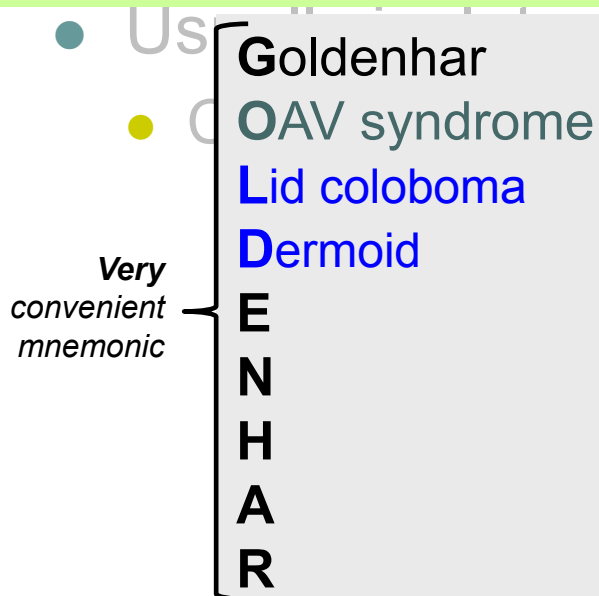
--**Dermoids of the cornea**

What is the 'full' name of the dermoid in question?

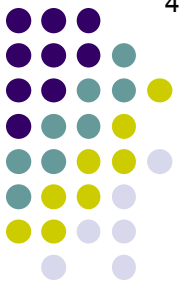
**Epibulbar** dermoid

*Note: There is another legit answer, so if you came up with **that** one, no worries (we'll identify it shortly)*

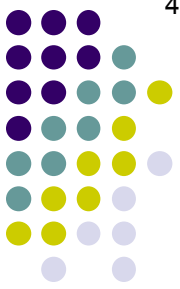
ent



Goldenhar syndrome



Goldenhar syndrome: Epibulbar dermoid



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

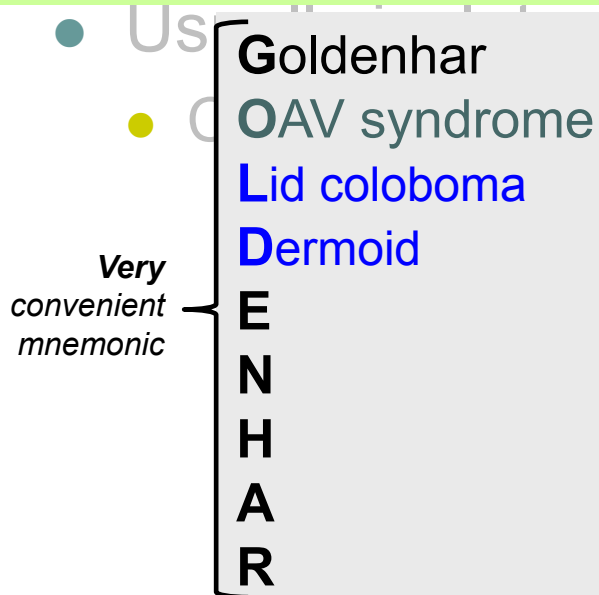
--Lid coloboma

--**Dermoids of the cornea**

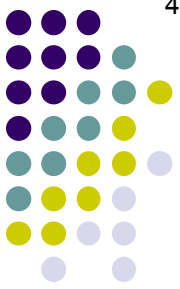
*What is the 'full' name of the dermoid in question?*

**Epibulbar** dermoid

*Is there a relationship between epibulbar dermoids and lipodermoids (aka dermolipomas)?*



th **Goldenhar syndrome**



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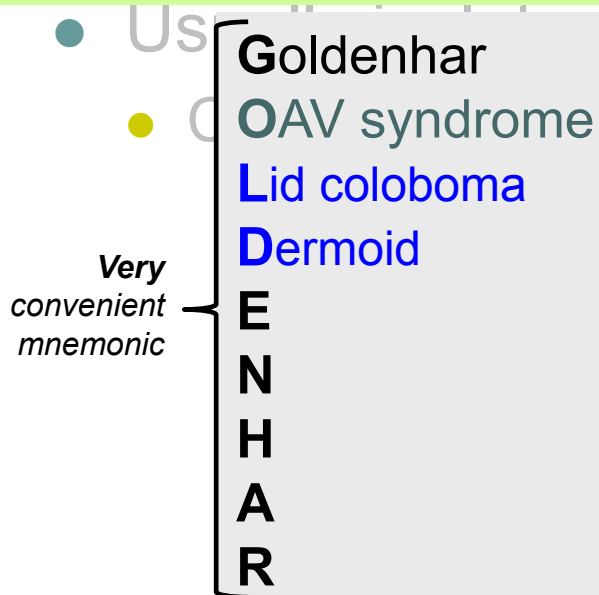
--**Dermoids of the cornea**

*What is the 'full' name of the dermoid in question?*

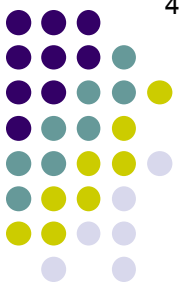
**Epibulbar** dermoid

*Is there a relationship between epibulbar dermoids and lipodermoids (aka dermolipomas)?*

Yes. The relationship is that, like dermoids, lipodermoids are associated with Goldenhar



Goldenhar syndrome



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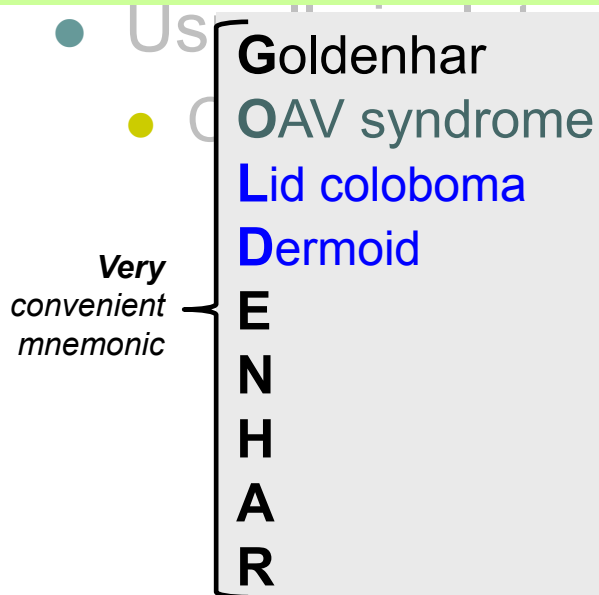
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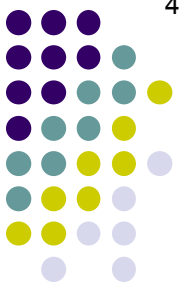
Yes. The relationship is that, like dermoids, lipodermoids are associated with Goldenhar

*Where are dermolipomas typically located?*



Goldenhar syndrome





*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--Lid coloboma

--**Dermoids of the cornea**

*What is the 'full' name of the dermoid in question?*

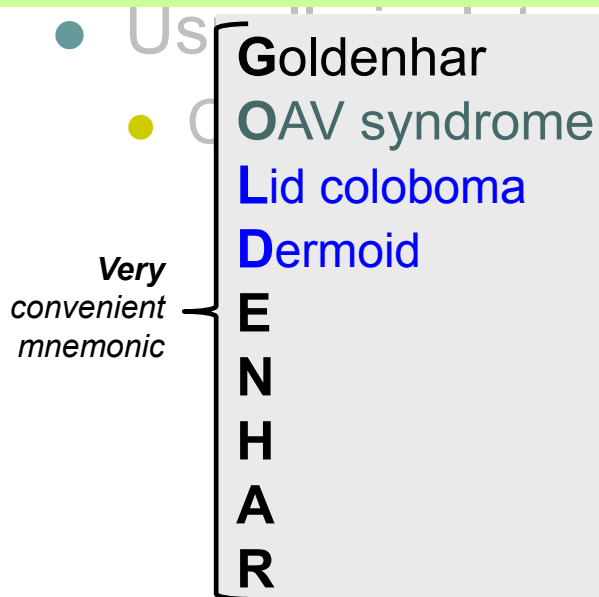
**Epibulbar** dermoid

*Is there a relationship between epibulbar dermoids and lipodermoids (aka dermolipomas)?*

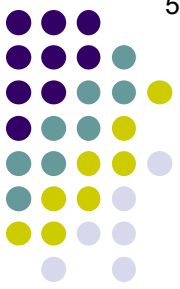
Yes. The relationship is that, like dermoids, lipodermoids are associated with Goldenhar

*Where are dermolipomas typically located?*

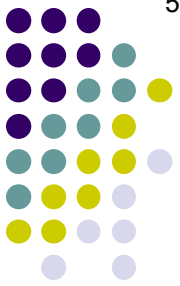
The temporal fornix



th **Goldenhar syndrome**



Goldenhar syndrome: Dermalipoma



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

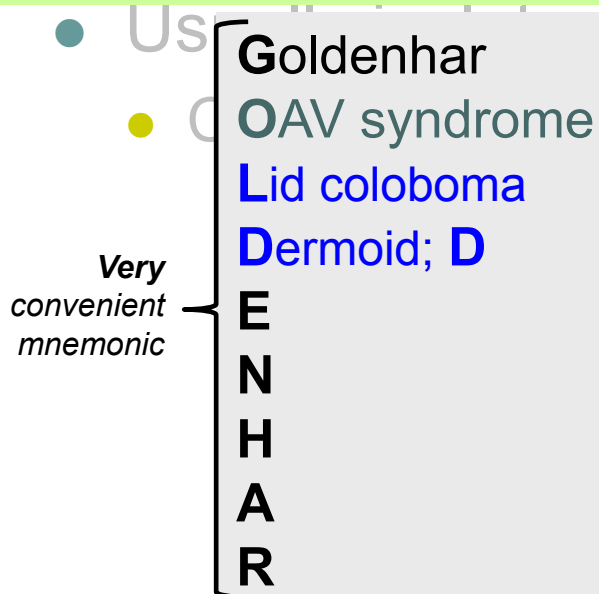
*What other ocular/periocular abnormalities are common in Goldenhar?*

--Lid coloboma

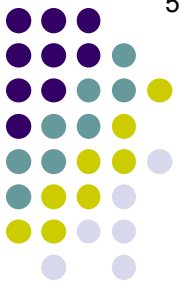
--**Dermoids of the cornea; D...**

*There is another 'D' association with Goldenhar that I am absolutely positive you know. What is it?*

ent



Goldenhar syndrome



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--Lid coloboma

--**Dermoids of the cornea; Duane syndrome**

*There is another 'D' association with Goldenhar that I am absolutely positive you know. What is it?*

**Duane syndrome** (aka the subject of the slide-set you're currently reading)

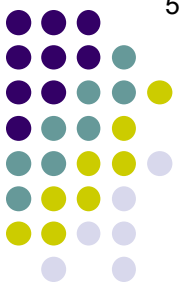
ent

● Us  
 ● O

Very  
 convenient  
 mnemonic

Goldenhar  
 OAV syndrome  
 Lid coloboma  
 Dermoid; Duane's  
**E  
N  
H  
A  
R**

th **Goldenhar syndrome**



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--**Lid coloboma**

--**Dermoids of the cornea; Duane syndrome**

*What **nonocular** findings are usually present?*

--**E** ←

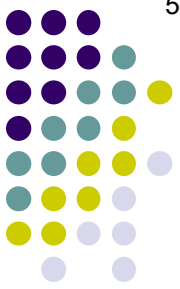
--**H** ←

ent

Very convenient mnemonic

- Goldenhar
- OAV syndrome
- Lid coloboma
- Dermoid; Duane's
- E** ←
- Nothing starts w/ 'N'
- H** ←
- A**
- R**

**Goldenhar syndrome**



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--**Lid coloboma**

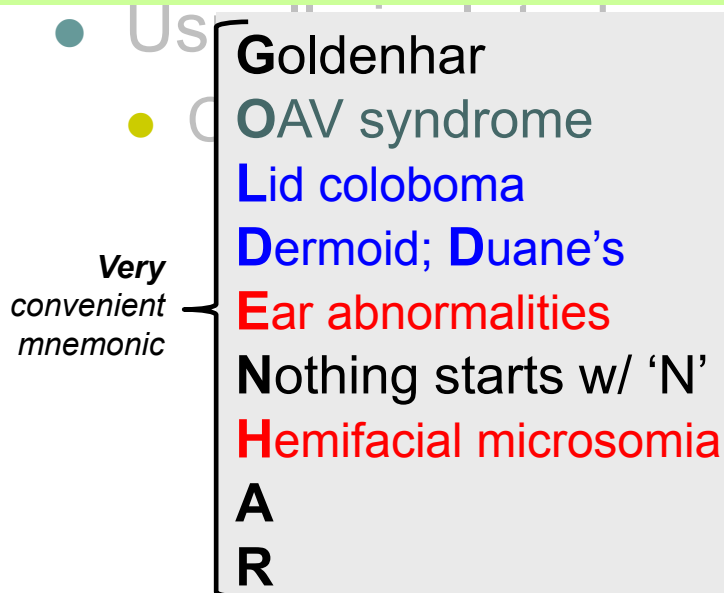
--**Dermoids of the cornea; Duane syndrome**

*What **nonocular** findings are usually present?*

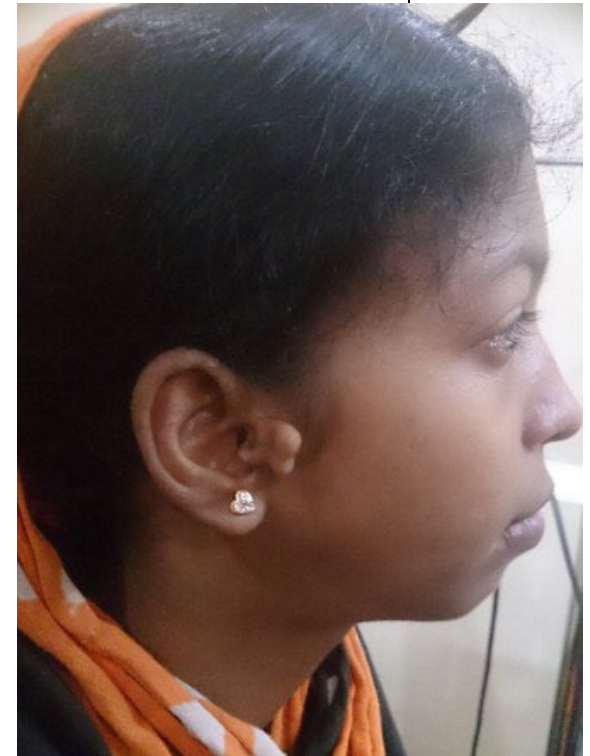
--**Ear abnormalities** (pre-auricular appendages; aural fistulae)

--**Hemifacial microsomia** (maxillary/mandibular hypoplasia)

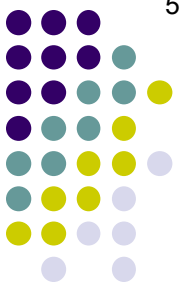
ent



th **Goldenhar syndrome**



Goldenhar syndrome: Ear abnormalities



What is its noneponymous name?

**Oculo-Auriculo-Vertebral (OAV) syndrome**

What other ocular/periocular abnormalities are common in Goldenhar?

--Lid coloboma

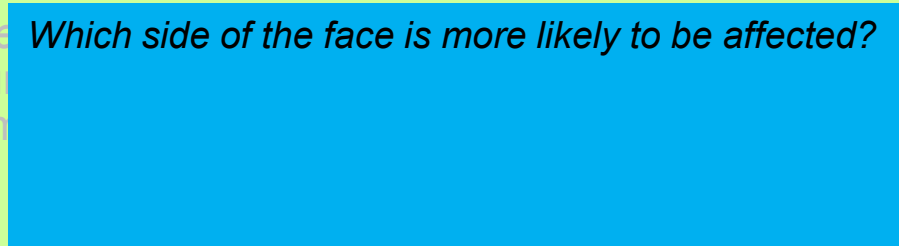
--Dermoids of the cornea; Duane syndrome

What *nonocular* findings are

--Ear abnormalities (pre-auditory)

--**Hemifacial microsomia** (not

Which side of the face is more likely to be affected?

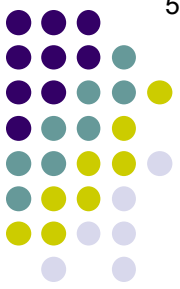


ent

- Us
  - O
- Very convenient mnemonic
- G**oldenhar
  - O**AV syndrome
  - L**id coloboma
  - D**ermoid; **D**uane's
  - E**ar abnormalities
  - N**othing starts w/ 'N'
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  - A**
  - R**

th **Goldenhar syndrome**





What is its noneponymous name?

**Oculo-Auriculo-Vertebral (OAV) syndrome**

What other ocular/periocular abnormalities are common in Goldenhar?

--Lid coloboma

--Dermoids of the cornea; Duane syndrome

What *nonocular* findings are

--Ear abnormalities (pre-auditory)

--**Hemifacial microsomia** (mandibular hypoplasia)

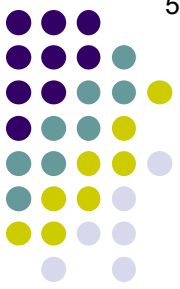
Which side of the face is more likely to be affected?

The right

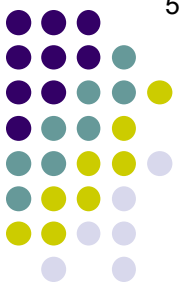
ent

- Us
  - O
- Very convenient mnemonic
- G**oldenhar
  - O**AV syndrome
  - L**id coloboma
  - D**ermoid; **D**uane's
  - E**ar abnormalities
  - N**othing starts w/ 'N'
  - H**emifacial microsomia
  - A**
  - R**

th **Goldenhar syndrome**



Goldenhar syndrome: Hemifacial microsomia



What is its noneponymous name?

**Oculo-Auriculo-Vertebral (OAV) syndrome**

What other ocular/periocular abnormalities are common in Goldenhar?

--Lid coloboma

--Dermoids of the cornea; Duane syndrome

What *nonocular* findings are common in Goldenhar?

--Ear abnormalities (pre-auricular pits)

--**Hemifacial microsomia** (more common)

Which side of the face is more likely to be affected?

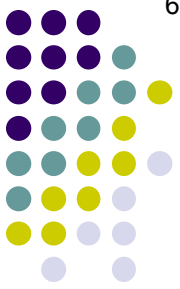
The right

Why the right side?

ent

- Us
  - O
- Very convenient mnemonic
- G**oldenhar
  - O**AV syndrome
  - L**id coloboma
  - D**ermoid; **D**uane's
  - E**ar abnormalities
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th **Goldenhar syndrome**



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**Oculo-Auriculo-Vertebral (OAV) syndrome**

What other ocular/periocular abnormalities are common in Goldenhar?

--Lid coloboma

--Dermoids of the cornea; Duane syndrome

What *nonocular* findings are common in Goldenhar?

--Ear abnormalities (pre-auditory)

--**Hemifacial microsomia** (mandibulofacial dysostosis)

Which side of the face is more likely to be affected?

The right

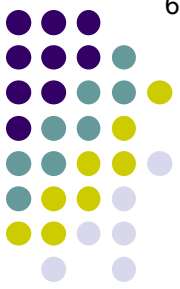
Why the right side?

I have no idea

ent

- Us
  - O
- Very convenient mnemonic
- G**oldenhar
  - O**AV syndrome
  - L**id coloboma
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th **Goldenhar syndrome**



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--**Lid coloboma**

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--**Ear abnormalities** (pre-auricular appendages; aural fistulae)

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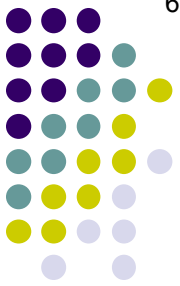
*Where specifically are epibulbar dermoids commonly located in Goldenhar?*

**A** ←

ent

- Us
  - Goldenhar
  - OAV syndrome
  - Lid coloboma
  - Dermoid; Duane's
  - Ear abnormalities
  - Nothing starts w/ 'N'
  - Hemifacial microsomia
  - **A** ←
  - R
- Very convenient mnemonic

th **Goldenhar syndrome**



*What is its noneponymous name?*

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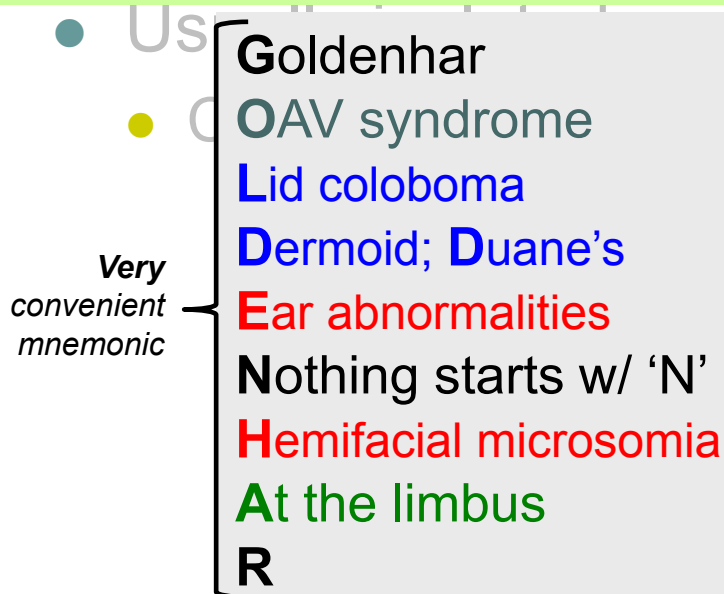
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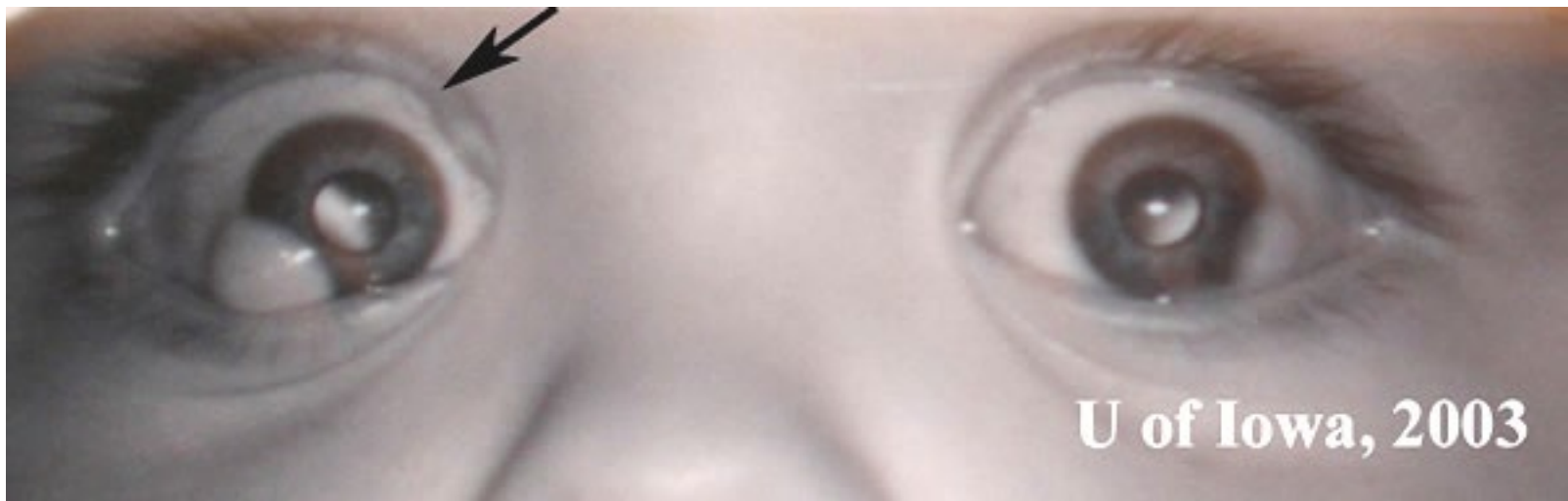
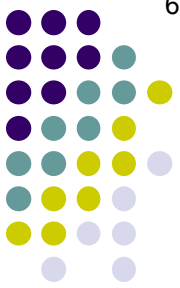
*Where specifically are epibulbar dermoids commonly located in Goldenhar?*

**At the limbus**

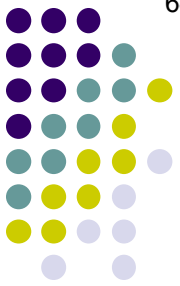
ent



th **Goldenhar syndrome**



Goldenhar syndrome: Limbal (epibulbar) dermoids.  
Note also the lid coloboma (arrow)



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

--**Lid coloboma**

--**Dermoids of the cornea; Duane syndrome**

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--**Ear abnormalities** (pre-auricular appendages; aural fistulae)

--**Hemifacial microsomia** (maxillary/mandibular hypoplasia)

*Where specific?*

**At the limbus**

*Several slides ago I acknowledged that epibulbar dermoids had another legit name. At long last--what is it?*

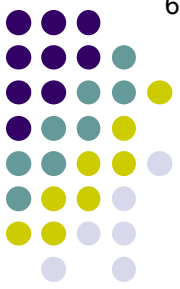
one word

**dermoids**

- Us
- O
- Very convenient mnemonic {
  - Goldenhar**
  - OAV syndrome**
  - Lid coloboma**
  - Dermoid; Duane's**
  - Ear abnormalities**
  - Nothing starts w/ 'N'**
  - Hemifacial microsomia**
  - At the limbus**
  - R**

th **Goldenhar syndrome**





*What is its noneponymous name?*

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*Where specific?*

**At the limbus**

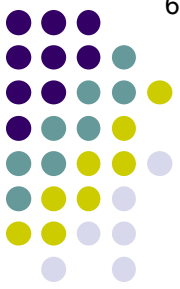
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**Limbal** dermoids

ent

- Us
  - O
- Very convenient mnemonic
- G**oldenhar
  - O**AV syndrome
  - L**id coloboma
  - D**ermoid; **D**uane's
  - E**ar abnormalities
  - N**othing starts w/ 'N'
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  - A**t the limbus
  - R**

th **Goldenhar syndrome**



*What is its noneponymous name?*

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--**Dermoids of the cornea; Duane syndrome**

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--**Hemifacial microsomia** (maxillary/mandibular hypoplasia)

*Where specifically are epibulbar dermoids commonly located in Goldenhar?*

**At the limbus**

*Are Goldenhar individuals cognitively impaired?*

**R** ←

**Goldenhar**

**OAV syndrome**

**Lid coloboma**

**Dermoid; Duane's**

**Ear abnormalities**

**Nothing starts w/ 'N'**

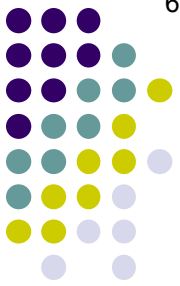
**Hemifacial microsomia**

**At the limbus**

**R** ←

Very  
convenient  
mnemonic

**Goldenhar syndrome**



*What is its noneponymous name?*

**Oculo-Auriculo-Vertebral (OAV) syndrome**

*What other ocular/periocular abnormalities are common in Goldenhar?*

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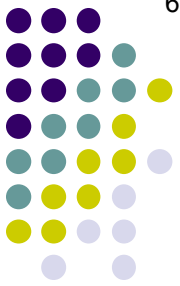
**At the limbus**

*Are Goldenhar individuals cognitively impaired?*

**Retardation** is present in a minority (~10%)

- Us
  - Goldenhar
  - OAV syndrome
  - Lid coloboma
  - Dermoid; Duane's
  - Ear abnormalities
  - Nothing starts w/ 'N'
  - Hemifacial microsomia
  - At the limbus
  - Retardation in ~10%
- Very convenient mnemonic

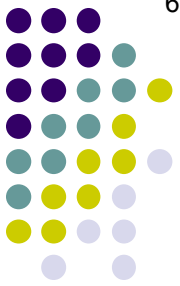
th **Goldenhar syndrome**



# Q

## ● Duane's Retraction Syndrome

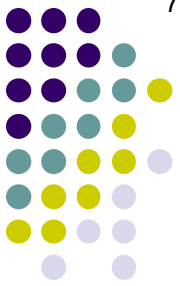
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- F  $\begin{matrix} > \\ < \\ = \end{matrix}$  M



# A

## ● Duane's Retraction Syndrome

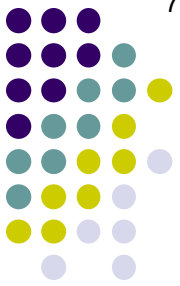
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  - Can be associated with Goldenhar syndrome
- F > M



# Q

## ● Duane's Retraction Syndrome

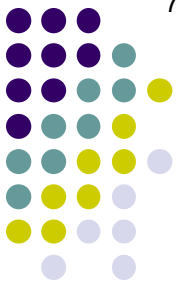
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
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- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- F > M
- OS  $\begin{matrix} > \\ < \\ = \end{matrix}$  OD



# A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
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- F > M
- OS > OD



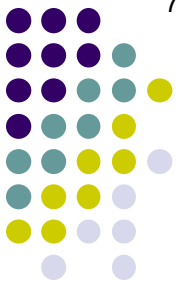
# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- $F > M$
- $OS > OD$

*These are the opposite of what they are in another strab condition syndrome*





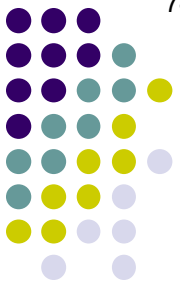
# A

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- $F > M$
- $OS > OD$

*These are the opposite of what they are in Brown syndrome*

*Brown syndrome:*  
 $F < M$   
 $OS < OD$

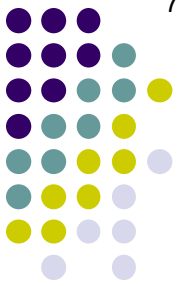


# Q

## ● Duane's Retraction Syndrome

- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- $F > M$
- $OS > OD$
- Bilateral in %

*These are the opposite of what they are in Brown syndrome*

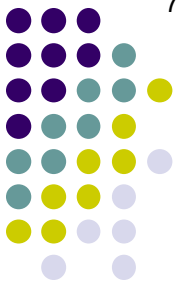


# A

## ● Duane's Retraction Syndrome

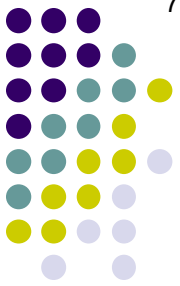
- Motility disorder featuring:
  - 1) Retraction of globe on attempted adduction
  - 2) At least some limitation of horizontal movement
  - 3) Up- or downshoot in adduction
- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome
- F > M
- OS > OD
- Bilateral in ~15%

*These are the opposite of what they are in Brown syndrome*



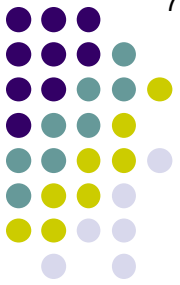
# Q

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type ?*
    - *Type ?*
    - *Type ?*



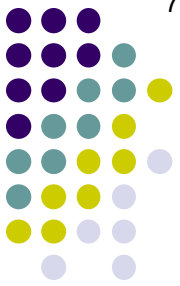
# A

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*
    - *Type 2*
    - *Type 3*



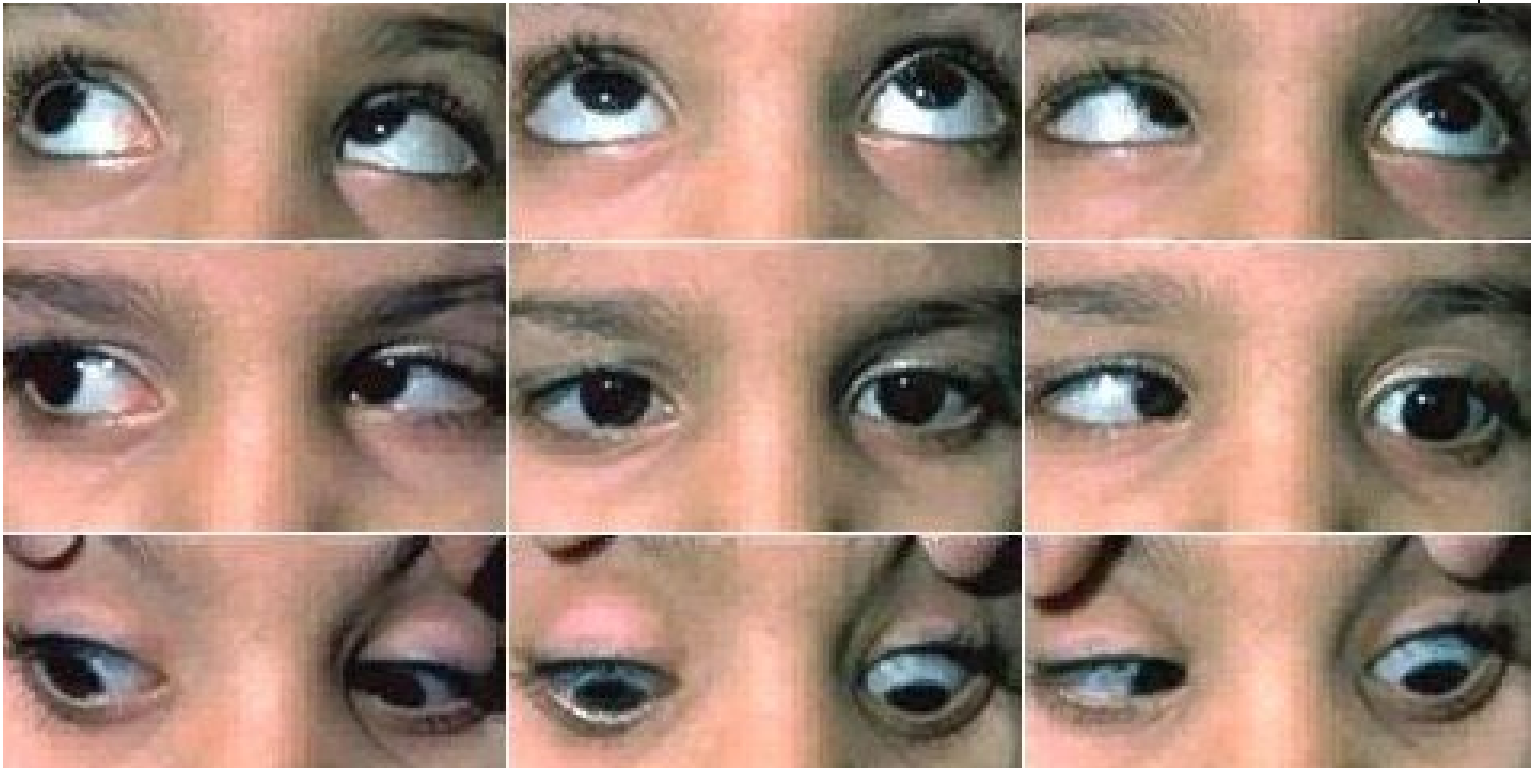
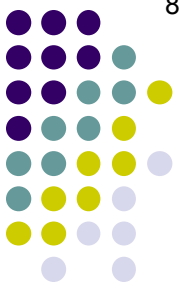
# Q

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited movement
    - *Type 2*
    - *Type 3*



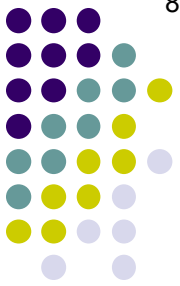
# A

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited abduction
    - *Type 2*
    - *Type 3*



Duane syndrome Type 1

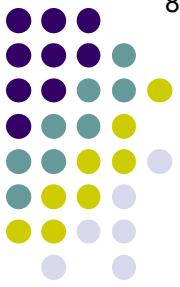




# Q

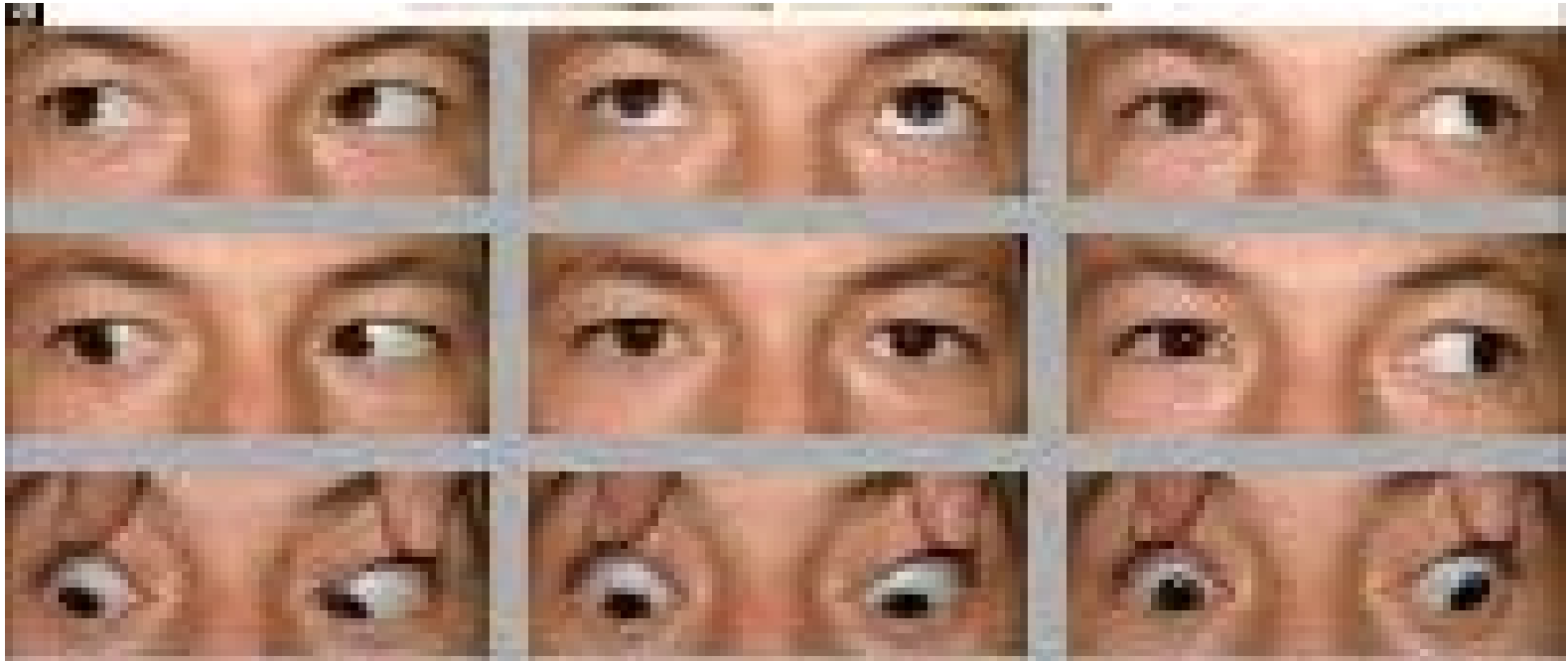
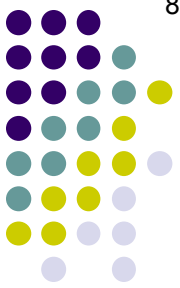
## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - *Type 1*: Limited abduction
  - *Type 2*: Limited movement
  - *Type 3*

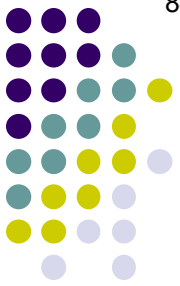


# A

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited abduction
    - *Type 2*: Limited adduction
    - *Type 3*

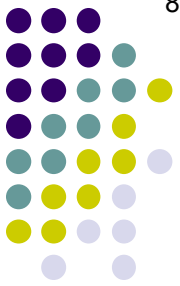


Duane syndrome Type 2



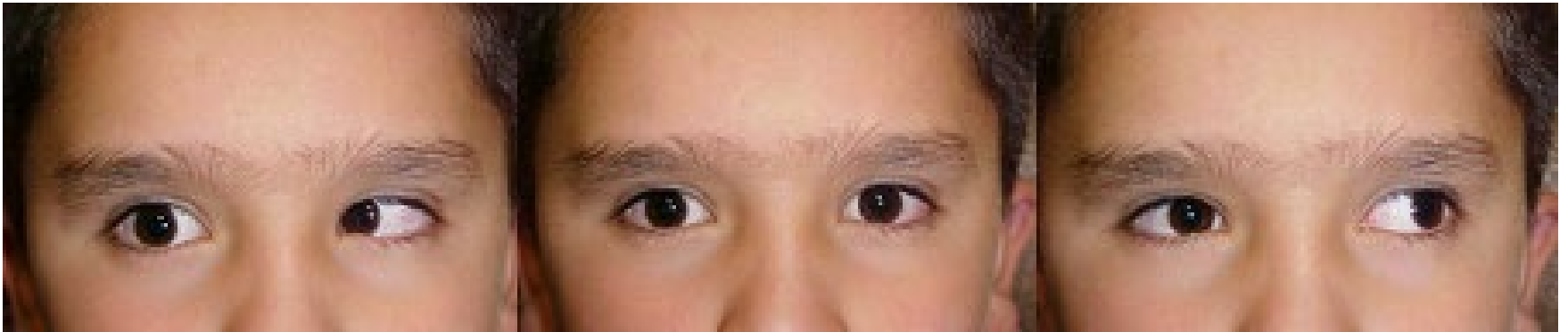
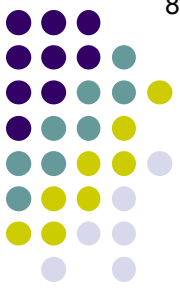
# Q

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited abduction
    - *Type 2*: Limited adduction
    - *Type 3*: Both movement and movement limited



# A

- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited abduction
    - *Type 2*: Limited adduction
    - *Type 3*: Both abduction and adduction limited



Duane syndrome Type 3

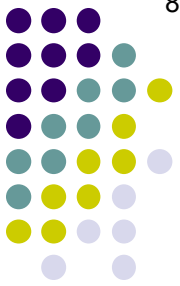


## ● Duane's Retraction Syndrome cont

**Mnemonic:**  
The number of 'Ds'  
= type of Duane's

● Three types of Duane's are recognized:

- *Type 1:* Limited **abduction** (1)
- *Type 2:* Limited **adduction** (2)
- *Type 3:* Both **abduction** and **adduction** limited (3)



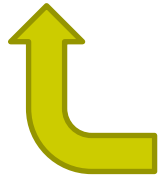
- **Duane's Retraction Syndrome cont**

- Three types of Duane's are recognized:

*Esotropic Type 1*: Limited abduction

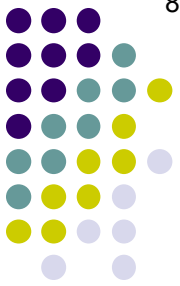
*Exotropic Type 2*: Limited adduction

*OrthoType 3*: Both abduction and adduction limited



The three are known also as the *Esotropic*,  
*Exotropic* and *Orthotropic* types respectively

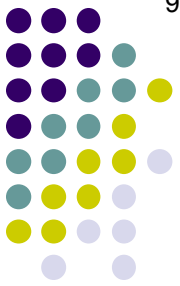




# Q

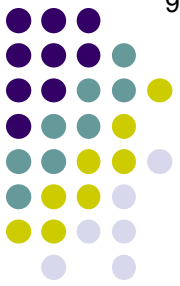
## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - *Type 1*: Limited abduction
  - *Type 2*: Limited adduction
  - *Type 3*: Both abduction and adduction limited
- Most common type:  $\frac{1}{2/3}$  in  $>\%$  of cases



# A

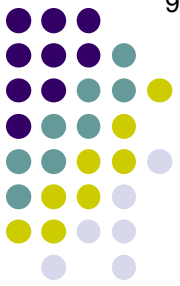
- **Duane's Retraction Syndrome cont**
  - Three types of Duane's are recognized:
    - *Type 1*: Limited abduction
    - *Type 2*: Limited adduction
    - *Type 3*: Both abduction and adduction limited
  - Most common type: 1 in >50% of cases



# Q

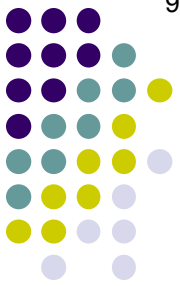
## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - *Type 1*: Limited abduction
  - *Type 2*: Limited adduction
  - *Type 3*: Both abduction and adduction limited
- Most common type: 1 in >50% of cases
- Etiology of Duane's:
  - Absent cranial nerve nucleus



# A

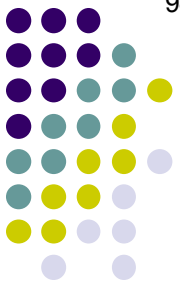
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  - Three types of Duane's are recognized:
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    - *Type 2*: Limited adduction
    - *Type 3*: Both abduction and adduction limited
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# Q

## ● Duane's Retraction Syndrome cont

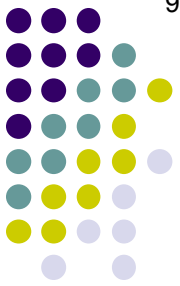
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  - *Type 2*: Limited adduction
  - *Type 3*: Both abduction and adduction limited
- Most common type: 1 in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - cranial nerve innervates LR



# A

## ● Duane's Retraction Syndrome cont

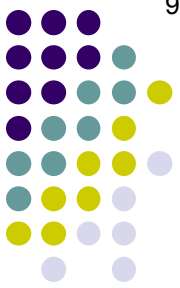
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  - *Type 2*: Limited adduction
  - *Type 3*: Both abduction and adduction limited
- Most common type: 1 in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - CN3 innervates LR



# Q

## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - *Type 1*: Limited abduction
  - *Type 2*: Limited adduction
  - *Type 3*: Both abduction and adduction limited
- Most common type: 1 in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - CN3 innervates LR
  - Paradoxical innervation to LR...
    - ...**increases** with attempted movement
    - ...**decreases** with attempted movement

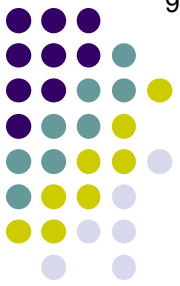


# A

## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
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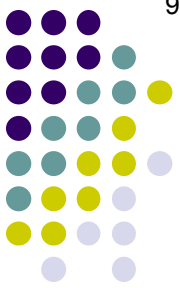
# Q

## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - Type 1: Limited abduction
  - Type 2: Limited adduction
  - Type 3: Both abduction and adduction limited

*How does this lead to the hallmark of Duane's **retraction** syndrome (ie, globe retraction)?*

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted adduction
  - ...**decreases** with attempted abduction



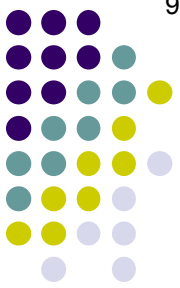
# A

## ● Duane's Retraction Syndrome cont

- Three types of Duane's are recognized:
  - Type 1: Limited abduction
  - Type 2: Limited adduction
  - Type 3: Both abduction and adduction limited

*How does this lead to the hallmark of Duane's **retraction** syndrome (ie, globe retraction)?*  
 During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted adduction
  - ...**decreases** with attempted abduction



# Q

## ● Duane's Retraction Syndrome cont

*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)? During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.*

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# Q/A

## ● Duane's Retraction Syndrome cont

*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the *law of reciprocal innervation*, which states that **innervation** to a given EOM is accompanied by a reciprocal **in** innervation to its antagonist.

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- **CN3** innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# A

## ● Duane's Retraction Syndrome cont

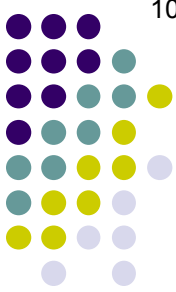
*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the *law of reciprocal innervation*, which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist.

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# A

## ● Duane's Retraction Syndrome cont

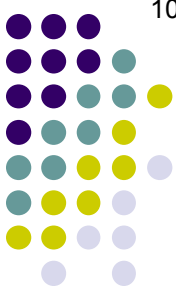
*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the *law of reciprocal innervation*, which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist. Thus, in an intact EOM control system, the increased MR innervation associated with attempted adduction would be accompanied by a proportional decrease in innervation to the ipsilateral LR.

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# Q

## • Duane's Retraction Syndrome cont

*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

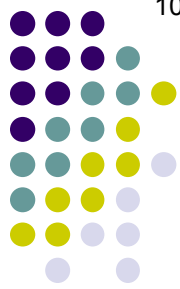
The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the **law of reciprocal innervation**, which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist. Thus, the increased MR innervation associated with attempted adduction is proportional decrease in innervation to the ipsilateral LR.

*What is the eponymous name of this law?*

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and **the LR should relax**. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# A

## • Duane's Retraction Syndrome cont

*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the **law of reciprocal innervation**, which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist. The **Sherrington's law** (of reciprocal innervation) proportional decrease in innervation to the ipsilateral LR.

*What is the eponymous name of this law?*

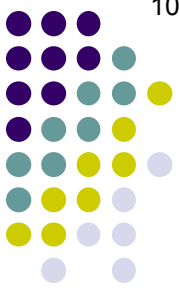
**Sherrington's law** (of reciprocal innervation)

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and **the LR should relax**. But instead of relaxing, in Duane's the LR contracts as well. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**





## ● Duane's Retraction Syndrome cont

*Wait—I get why the MR should contract during attempted adduction, but why does it say the LR **should** relax? This makes it sound like the LR is obligated in some sense to relax. What's this all about?*

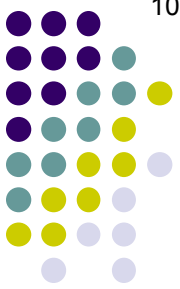
The LR **is** obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the **law of reciprocal innervation** which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist. Thus, in an intact EOM control system, the increased MR

*Thus, we can see that Duane's is a condition that violates Sherrington's law!*

*How does this lead to the hallmark of Duane's Retraction Syndrome (ie, globe retraction)?*

During attempted adduction, the MR should contract and the LR should relax. But instead of relaxing, **in Duane's the LR contracts as well**. What's the globe going to do if a muscle on either side of it contracts at the same time? It's going to move backwards (ie, retract) into the orbit.

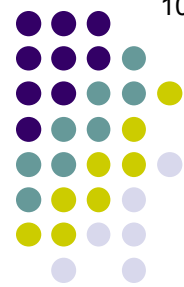
- CN3 innervates LR
- Paradoxical innervation to LR...
  - ...**increases** with attempted **adduction**
  - ...**decreases** with attempted **abduction**



# Q

*An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders?*

- Most common type: I in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - **CN3 innervates LR**
  - Paradoxical innervation to LR...
    - ...increases with attempted adduction
    - ...decreases with attempted abduction

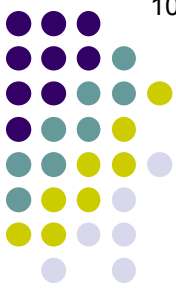


# A

*An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders?*

They are called '**congenital cranial dysinnervation disorders**'

- Most common type: **I** in **>50%** of cases
- Etiology of Duane's:
  - Absent **CN6** nucleus
  - **CN3 innervates LR**
  - Paradoxical innervation to LR...
    - ...**increases** with attempted adduction
    - ...**decreases** with attempted abduction



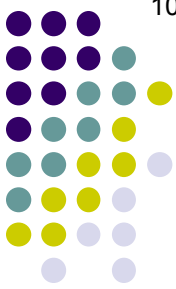
# Q

*An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders?*

They are called '**congenital cranial dysinnervation disorders**'

*Another congenital cranial dysinnervation disorder involving an ophthalmic movement (lid elevation) should readily come to mind-- what is it?*

- Most common type: I in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - **CN3 innervates LR**
  - Paradoxical innervation to LR...
    - ...increases with attempted adduction
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# A

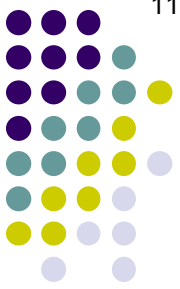
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They are called '**congenital cranial dysinnervation disorders**'

*Another congenital cranial dysinnervation disorder involving an ophthalmic movement (lid elevation) should readily come to mind-- what is it?*

Marcus-Gunn jaw-winking syndrome (MGJW)

- Most common type: I in >50% of cases
- Etiology of Duane's:
  - Absent CN6 nucleus
  - **CN3 innervates LR**
  - Paradoxical innervation to LR...
    - ...increases with attempted adduction
    - ...decreases with attempted abduction



# Q

*An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders?*

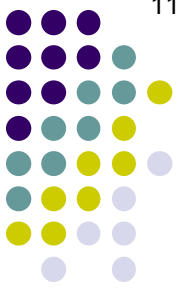
They are called '**congenital cranial dysinnervation disorders**'

*Another congenital cranial dysinnervation disorder involving an ophthalmic movement (lid elevation) should readily come to mind--what is it?*

**Marcus-Gunn jaw-winking syndrome (MGJW)**

*What is the clinical hallmark of MGJW?*

- ...**decreases** with attempted abduction



# A

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They are called '**congenital cranial dysinnervation disorders**'

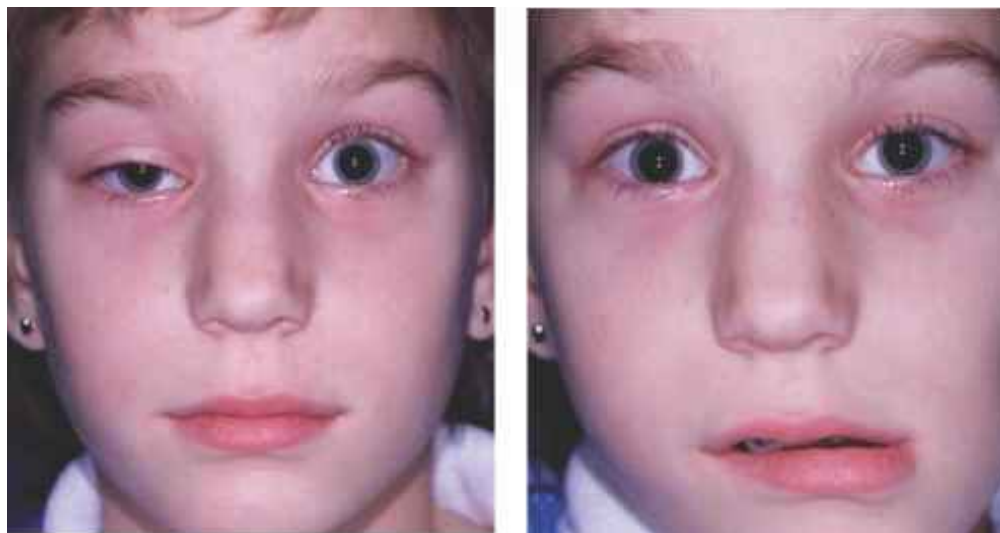
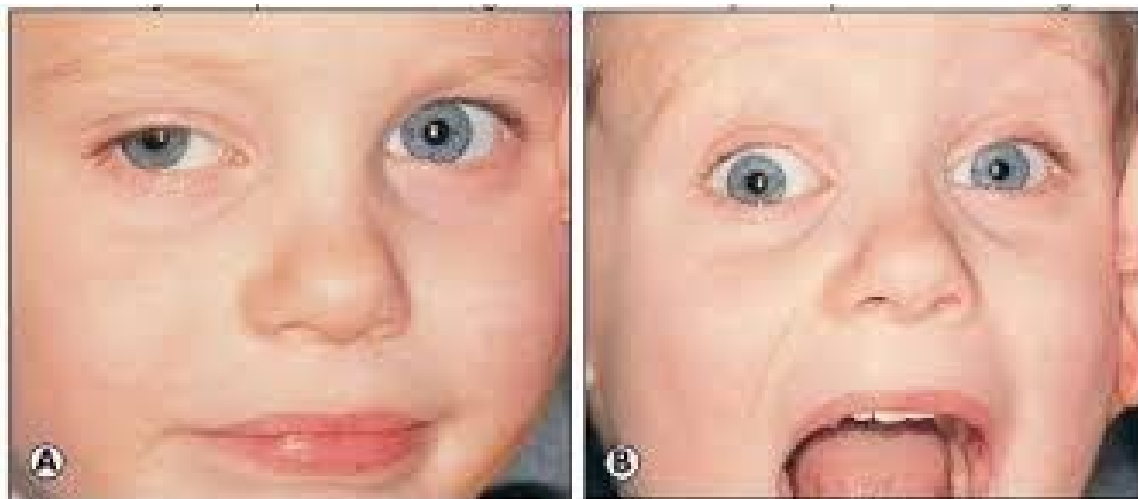
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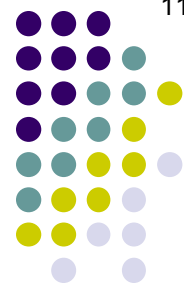
A ptotic lid elevates in response to voluntary masticatory movements of the jaw

- ...**decreases** with attempted abduction



MGJW. Note the resolution of ptosis (second pictures) with a jaw movement





# Q

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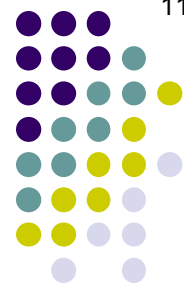
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--  
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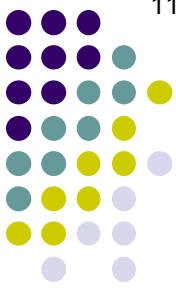
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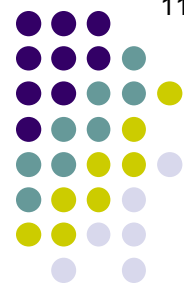
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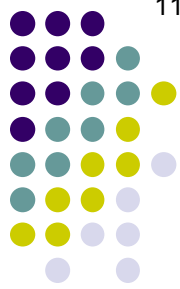
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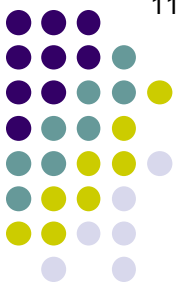
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*Which branch of the trigeminal?*

- ...decreases with attempted abduction

# A



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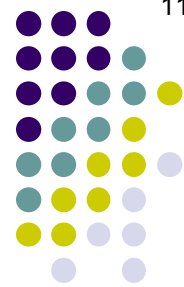
*Which cranial nerve innervates them?*

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**The mandibular (V<sub>3</sub>)**

- ...decreases with attempted abduction



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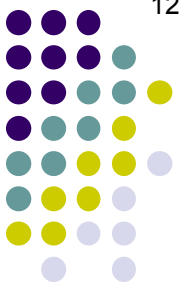
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**V<sub>3</sub> (dys)innervates the levator**

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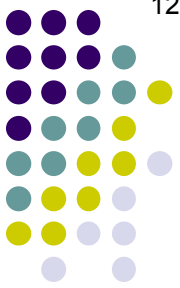
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*Is the ptosis of MGJW unilateral, or bilateral?*

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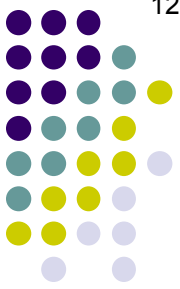
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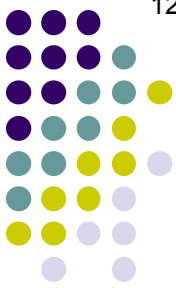
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*Which jaw movements can be involved?*

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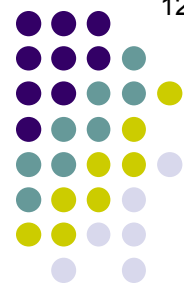
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Unilateral

*Which jaw movements can be involved?*

- Lateral displacement
- Protrusion
- Wide opening
- Clenching

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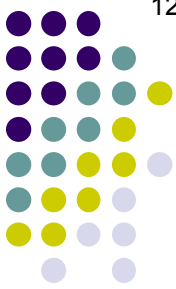
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*Unilateral*

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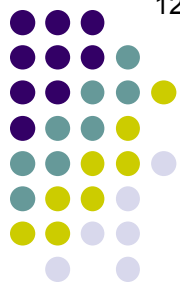
*What is the classic story regarding when parents first note their infant has MGJW?*

Unilateral Mom says the infant's lid 'twitches' while nursing

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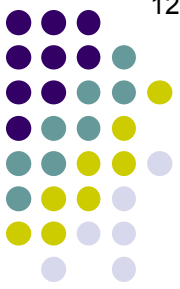
*Another one involving an ophthalmic movement—lateral gaze **this time**—what should come to mind as well. What is it?*

**Marcus**

What is the  
Aptosis

Who  
Is the first  
Unilateral

Which jaw  
--Lateral d  
--Protrusio  
--Wide ope  
--Clenchin



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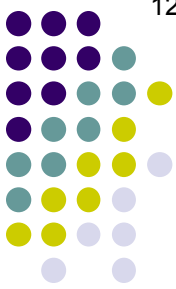
**Marcus** Möbius syndrome

*What is the first?*  
Aptosis

*What is the first?*  
Unilateral Möbius

*Which jaw?*  
--Lateral deviation  
--Protrusion  
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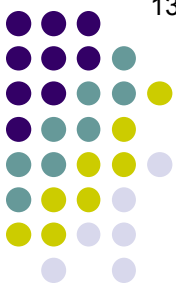
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*In a nutshell, what findings are typical in Möbius syndrome?*

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# Q/A



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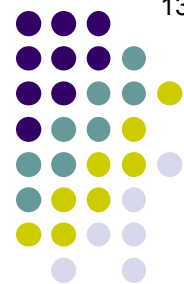
*Which jaw*

—Lateral d

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—Wide ope

—Clenchin



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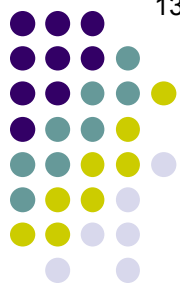
*Which jaw*

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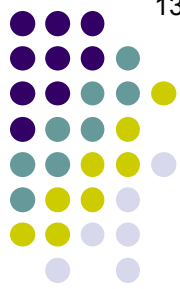
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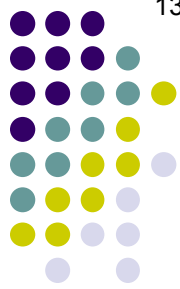
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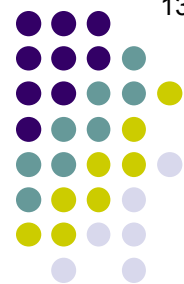
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*What is the classic descriptor of the expressionless facies resulting from the bilateral CN7 palsies?*

Which jaw  
--Lateral d  
--Protrusio  
--Wide op  
--Clenchin



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'Mask-like'

What is the  
Aptosis

What is the first  
Unilateral

Which jaw  
--Lateral d  
--Protrusio  
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# Q

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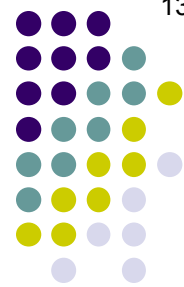
*Are vertical eye movements affected as well?*

What is the  
Aptosis

What is the first  
Unilateral

Which jaw  
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No, they are intact

*Which jaw*

--Lateral d

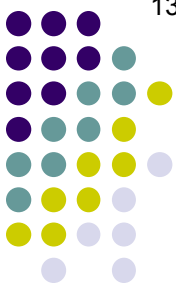
--Protrusio

--Wide op

--Clenchin



Möbius syndrome. **A**, orthotropia in primary. **B**, inability to smile. **C**, bilaterally absent adduction and severely limited abduction. **D**, Vertical movements are not affected.



# Q

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Another  
ophthal  
what is  
Marcus

Speaking of congenital cranial dysinnervation disorders:

Another on  
should con

**Mobius syndrome**

What tops the DDx for a Mobius-like presentation in a newborn?

What is the  
Aptosis

What  
Is the first  
Unilateral

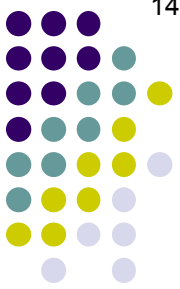
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# A

*An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders?*

They are called '**congenital cranial dysinnervation disorders**'

*Another  
ophtha  
what is  
Marcus*

Speaking of congenital cranial dysinnervation disorders:

*Another on  
should con*

*What tops the DDx for a Möbius-like presentation in a newborn?*  
Congenital myasthenia

**Möbius syndrome**

*What is the  
Aptosis like*

*What  
Is the first  
Unilateral*

*Which jaw  
--Lateral d  
--Protrusio  
--Wide open  
--Clenching*

*In a nutshell  
Bilateral C  
defects often*

*What is the  
Orthophoria*

*What is the  
the bilateral CN7 palsies?*  
'Mask-like'

*Are vertical eye movements affected as well?*  
No, they are intact



# Q

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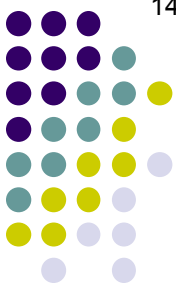
What is the  
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# Q/A



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**Möbius syndrome**

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Because moms. That is, if mom has circulating antibodies (recall those are the culprit in MG), they can cross the placenta and enter the neonate's bloodstream, thereby producing clinical MG.

two words

What is the  
Aptosis  
What  
Is the first  
Unilateral

In a nutshell  
Bilateral C  
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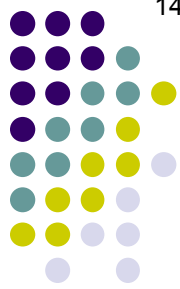
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# Q

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*Another ophthalmic what is Marcus*

Speaking of congenital cranial dysinnervation disorders:

*Another one should consider*

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*Is such an infant doomed to a lifetime of MG?*

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Because moms. That is, if mom has circulating ACh receptor antibodies (recall those are the culprit in MG), they can cross the placenta and enter the neonate's bloodstream, thereby producing clinical MG.

Is such an infant doomed to a lifetime of MG?

Nah—the condition resolves spontaneously and rapidly

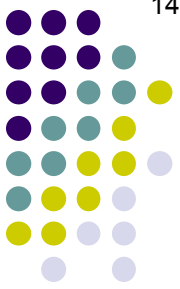
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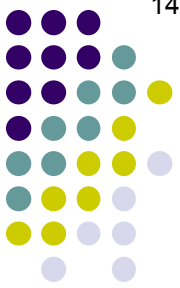
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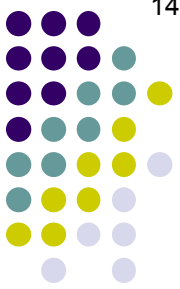
# Q

- **Duane's Retraction Syndrome cont**
  - Key observation to differentiate Duane's syndrome from CN6 palsy:



# A

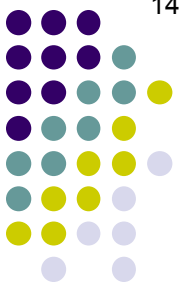
- **Duane's Retraction Syndrome cont**
  - Key observation to differentiate Duane's syndrome from CN6 palsy: **Retraction on attempted adduction**



# Q

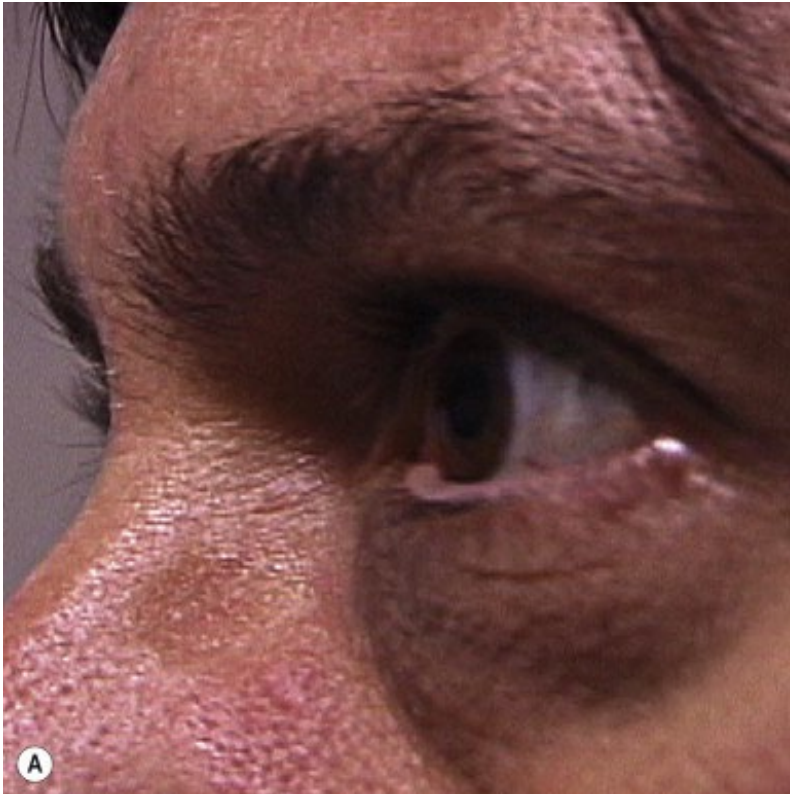
- **Duane's Retraction Syndrome cont**

- Key observation to differentiate Duane's syndrome from CN6 palsy: **Retraction on attempted adduction**
  - Observe patient in this position to assess

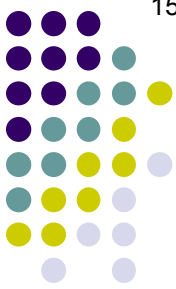


# A

- **Duane's Retraction Syndrome cont**
  - Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
    - Observe patient from the side to assess



Duane syndrome: Retraction



# Q

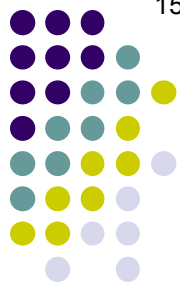
- **Duane's Retraction Syndrome cont**

- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's

clinical exam component (two words)

in

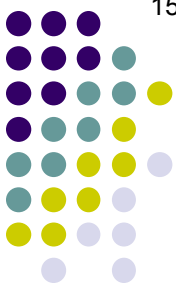
two words



# A

- **Duane's Retraction Syndrome cont**
  - Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
    - Observe patient from the side to assess
  - Another useful observation: Assess the patient's muscle balance in primary gaze



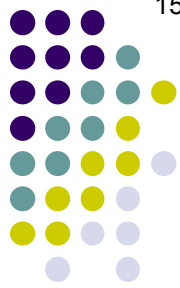


# Q

- **Duane's Retraction Syndrome cont**

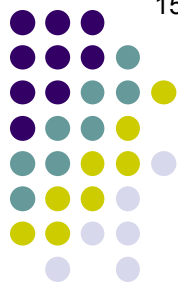
- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's muscle balance in primary gaze
  - In CN6 palsy, is usually 

straight vs esotropic
--------------------------



# A

- **Duane's Retraction Syndrome cont**
  - Key observation to differentiate Duane's syndrome from CN6 palsy: **Retraction on attempted adduction**
    - Observe patient **from the side** to assess
  - Another useful observation: Assess the patient's **muscle balance** in **primary gaze**
    - In CN6 palsy, is usually **esotropic**

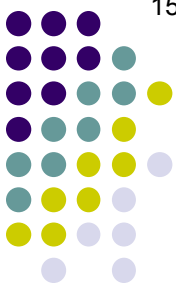


# Q

## ● Duane's Retraction Syndrome cont

- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's muscle balance in primary gaze
  - In CN6 palsy, is usually esotropic
  - In Duane's, is usually

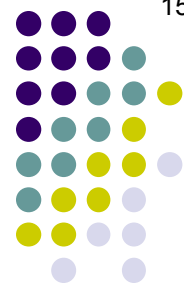
straight vs  
esotropic



# A

- **Duane's Retraction Syndrome cont**

- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's muscle balance in primary gaze
  - In CN6 palsy, is usually esotropic
  - In Duane's, is usually straight



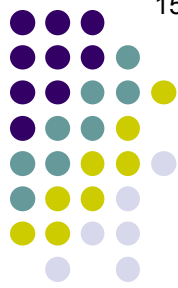
# Q

## ● Duane's Retraction Syndrome cont

- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's muscle balance in primary gaze
  - In CN6 palsy, is usually esotropic
  - In Duane's, is usually straight
- Still another: Look for  of the   on attempted adduction
 

second word

first word of two word phrase



# A

## ● Duane's Retraction Syndrome cont

- Key observation to differentiate Duane's syndrome from CN6 palsy: Retraction on attempted adduction
  - Observe patient from the side to assess
- Another useful observation: Assess the patient's muscle balance in primary gaze
  - In CN6 palsy, is usually esotropic
  - In Duane's, is usually straight
- Still another: Look for narrowing of the palpebral fissure on attempted adduction

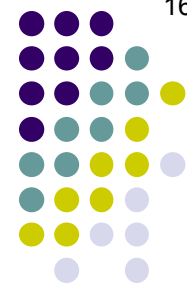


Duane syndrome: Fissure narrowing on adduction

# Q

- **Duane's Retraction Syndrome: Management**

*Is there any surgical procedure that will normalize ocular rotations?*



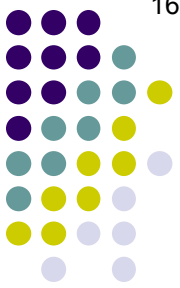


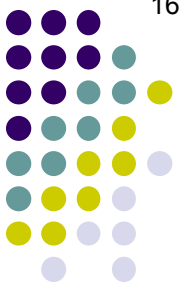
# A

- **Duane's Retraction Syndrome: Management**

*Is there any surgical procedure that will normalize ocular rotations?*

No





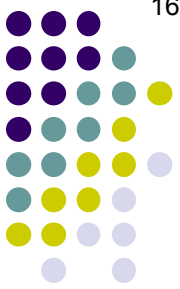
# Q

- **Duane's Retraction Syndrome: Management**

*Is there any surgical procedure that will normalize ocular rotations?*

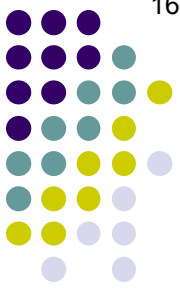
No

*If you can't normalize rotations, why do you operate?*



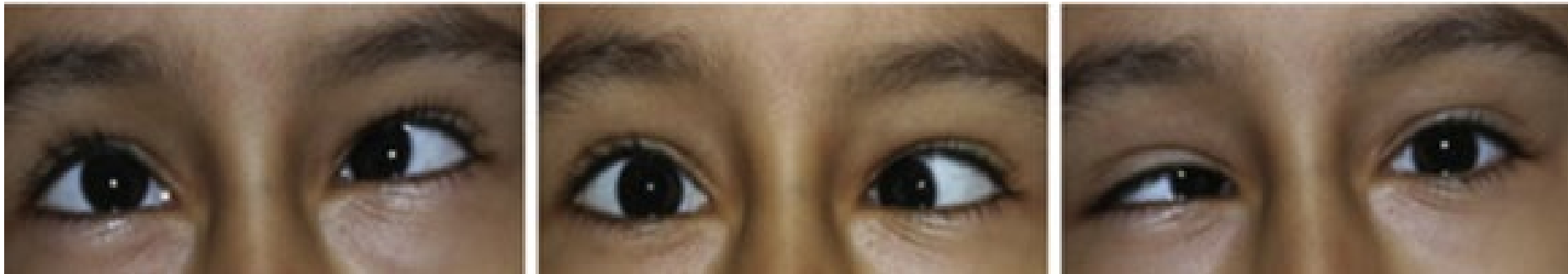
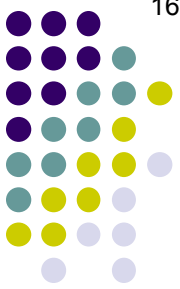
# Q

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in

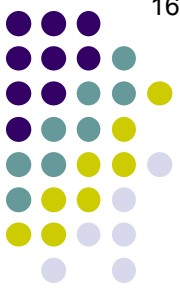


# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary**

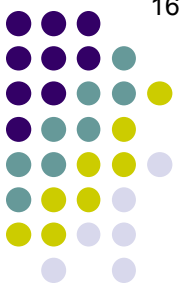


Duane syndrome: Deviated in primary



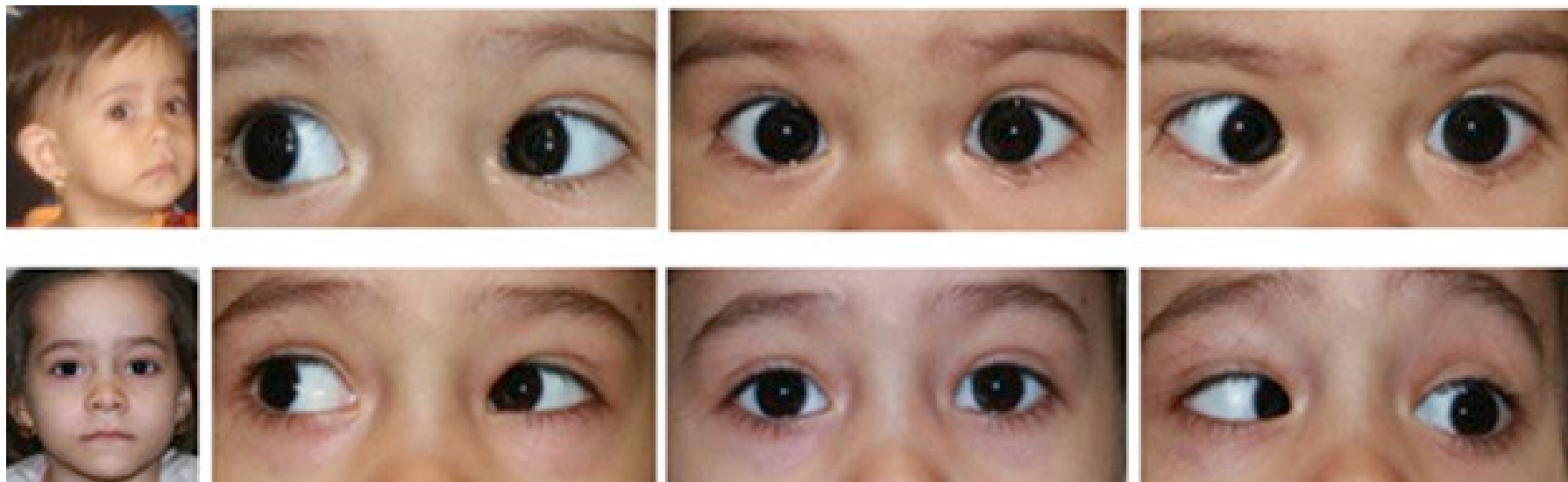
# Q

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal



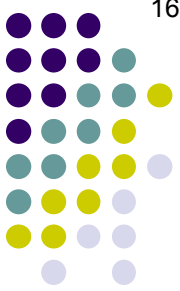
# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal **head position**



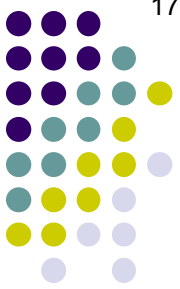
Duane syndrome with severe face turn (top row).  
Bottom row, s/p left MRM recession.





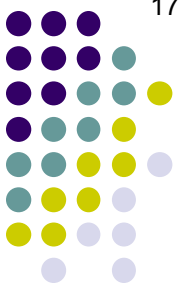
# Q

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal **head position** *OR*
    - Marked

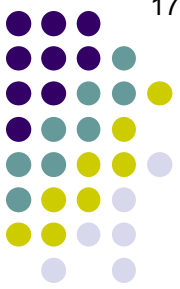


# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal **head position** *OR*
    - Marked **retraction**



Duane syndrome: Marked retraction



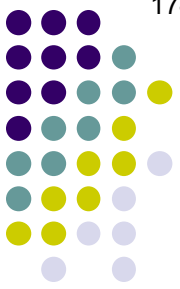
# Q

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal **head position** *OR*
    - Marked **retraction** *OR*
    - Large

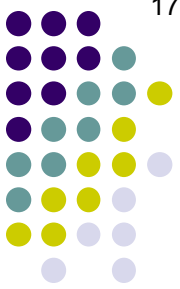


# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** *OR*
    - Abnormal **head position** *OR*
    - Marked **retraction** *OR*
    - Large **upshoot/downshoot**



Duane syndrome: Marked retraction and upshoot

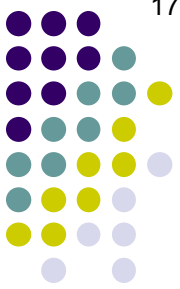


# Q

## ● Duane's Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary** *OR*
  - Abnormal **head position** *OR*
  - Marked **retraction** *OR*
  - Large **upshoot/downshoot**
- Type 1 (ET type):

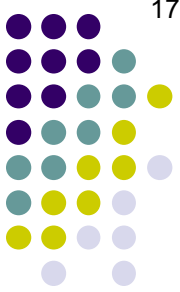
surgery



# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** **OR**
    - Abnormal **head position** **OR**
    - Marked **retraction** **OR**
    - Large **upshoot/downshoot**
  - Type 1 (ET type): **Ipsilateral MR *recession***





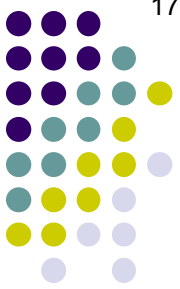
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- Operate only if:
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  - Marked **retraction** *OR*
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add 

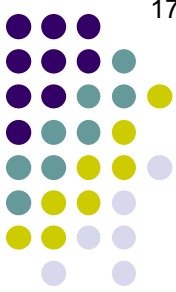
surgery

 if  $>20\Delta$  ET



# A

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
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    - Abnormal **head position** **OR**
    - Marked **retraction** **OR**
    - Large **upshoot/downshoot**
  - Type 1 (ET type): **Ipsilateral MR recession**
    - Add **contralateral MR recession** if  $>20\Delta$  ET

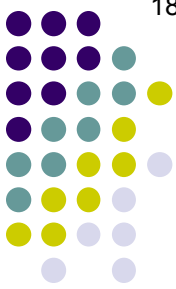


# Q

## ● Duane's Retraction Syndrome: Management

- Operate only if:
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  - Marked **retraction** **OR**
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from 

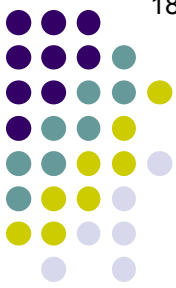
surgery



# A

## ● Duane's Retraction Syndrome: Management

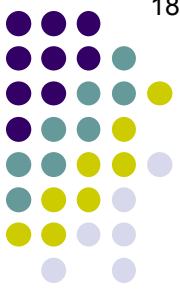
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  - Marked **retraction** **OR**
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from **LR resection**



## ● Duane's Retraction Syndrome: Management

- Operate only if:
  - Deviated in primary **OR**
  - Abnormal head position **OR**
  - Marked retraction **OR**
  - Large upshoot/downshoot
- Type 1 (ET type): Ipsilateral MR *recession*
  - Add contralateral MR recession if  $>20\Delta$  ET
  - **Most surgeons refrain from LR resection**

At one time, it was an ironclad rule that one must avoid resection procedures in Duane's. (The thinking was, resections would only worsen the retraction.) And per the latest edition of the *Peds* book, most surgeons still don't favor performing LR resection in Type 1/ET type Duane's. That said, the book also mentions that, in cases where LR co-contraction is minimal, some surgeons have found that *small* LR resections can improve abduction significantly.

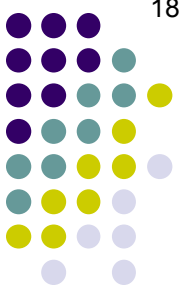


# Q

## ● Duane's Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary** **OR**
  - Abnormal **head position** **OR**
  - Marked **retraction** **OR**
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from **LR resection**
- Type 2 (XT type): 

surgery



# A

## ● Duane's Retraction Syndrome: Management

- Operate only if:
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  - Abnormal **head position** **OR**
  - Marked **retraction** **OR**
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from **LR resection**
- Type 2 (XT type): **Ipsilateral LR recession**



# Q

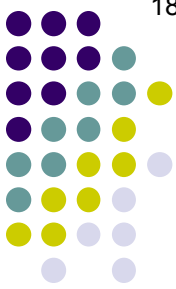
## • Duane's Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary** **OR**
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  - Marked **retraction** **OR**
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- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from **LR resection**
- Type 2 (XT type): **Ipsilateral LR recession**
  - Add 

surgery

 if  $>20\Delta$  XT

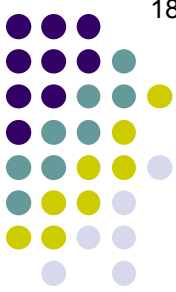




# A

## ● Duane's Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary** **OR**
  - Abnormal **head position** **OR**
  - Marked **retraction** **OR**
  - Large **upshoot/downshoot**
- Type 1 (ET type): **Ipsilateral MR recession**
  - Add **contralateral MR recession** if  $>20\Delta$  ET
  - Most surgeons refrain from **LR resection**
- Type 2 (XT type): **Ipsilateral LR recession**
  - Add **contralateral LR recession** if  $>20\Delta$  XT



# Q

- **Duane's Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** **OR**
    - Abnormal **head position** **OR**
    - Marked **retraction** **OR**
    - Large **upshoot/downshoot**
  - Type 1 (ET type): **Ipsilateral MR recession**
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  - Type 2 (XT type): **Ipsilateral LR recession**
    - Add **contralateral LR recession** if  $>20\Delta$  XT
    - All surgeons refrain from 

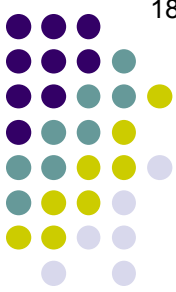
surgery

 in Type 2/XT type



# A

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    - Add **contralateral LR recession** if  $>20\Delta$  XT
    - All surgeons refrain from **MR resection** in Type 2/XT type



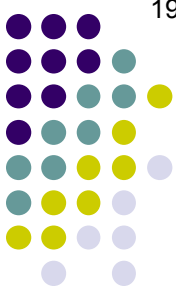
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    - Most surgeons refrain from **LR resection**
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    - All surgeons refrain from **MR resection** in Type 2/XT type
  - Type 3 (Ortho type)
    - No surgery will improve



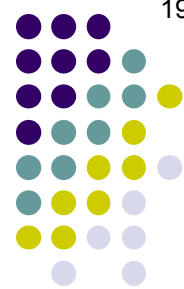
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    - Add **contralateral LR recession** if  $>20\Delta$  XT
    - All surgeons refrain from **MR resection** in Type 2/XT type
  - Type 3 (Ortho type)
    - No surgery will improve **excursion**



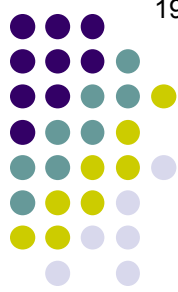
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    - All surgeons refrain from **MR resection** in Type 2/XT type
  - Type 3 (Ortho type)
    - No surgery will improve **excursion**
    - Recess both LR and MR to reduce



# A

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    - All surgeons refrain from **MR resection** in Type 2/XT type
  - Type 3 (Ortho type)
    - No surgery will improve **excursion**
    - Recess both LR and MR to reduce **retraction**



# Q

## • Duane's Retraction Syndrome: Management

### • Operate only if:

- Deviated in primary OR
- Abnormal head position OR
- Marked retraction OR
- **Large upshoot/downshoot**

### • Type 1 (EI type): Ipsilateral MR recession

*None of the surgeries discussed thus far address upshoot or downshoot. How should these be managed?*

### • Type 2 (XT type): Ipsilateral LR recession

- Add contralateral LR recession if  $>20\Delta$  XT
- All surgeons refrain from MR resection in Type 2/XT type

### • Type 3 (Ortho type)

- No surgery will improve excursion
- Recess both LR and MR to reduce retraction





# Q/A

## • Duane's Retraction Syndrome: Management

### • Operate only if:

- Deviated in primary OR
- Abnormal head position OR
- Marked retraction OR
- **Large upshoot/downshoot**

### • Type 1 (EI type): Ipsilateral MR recession

*None of the surgeries discussed thus far address upshoot or downshoot. How should these be managed?*

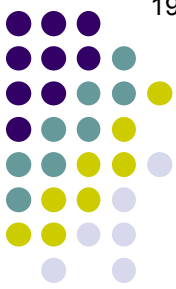
Several procedures are employed; the most popular involves two terms the LR

### • Type 2 (XT type): Ipsilateral LR recession

- Add contralateral LR recession if  $>20\Delta$  XT
- All surgeons refrain from MR resection in Type 2/XT type

### • Type 3 (Ortho type)

- No surgery will improve excursion
- Recess both LR and MR to reduce retraction



# A

## • Duane's Retraction Syndrome: Management

### • Operate only if:

- Deviated in primary OR
- Abnormal head position OR
- Marked retraction OR
- **Large upshoot/downshoot**

### • Type 1 (EI type): Ipsilateral MR recession

*None of the surgeries discussed thus far address upshoot or downshoot. How should these be managed?*

Several procedures are employed; the most popular involves Y-splitting the LR

### • Type 2 (XT type): Ipsilateral LR recession

- Add contralateral LR recession if  $>20\Delta$  XT
- All surgeons refrain from MR resection in Type 2/XT type

### • Type 3 (Ortho type)

- No surgery will improve excursion
- Recess both LR and MR to reduce retraction