Duane’s Retraction Syndrome

- Motility disorder featuring:
  1) Retraction of globe on attempted adduction
  2) An abnormal eye movement
  3)
• Duane’s Retraction Syndrome
  • Motility disorder featuring:
    1) **Retraction** of globe on attempted adduction
    2)
    3)
Duane syndrome: Globe retraction
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)
• 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
- **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*
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
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
Duane’s Retraction Syndrome

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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
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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?

Craniofacial malformation
Duane’s Retraction Syndrome

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Goldenhar syndrome

What are the two categories of craniofacial syndrome?
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Two categories of craniofacial syndrome
Craniosynostoses  Not craniosynostoses

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Which craniosynostosis syndromes are addressed in the Peds book?
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Goldenhar syndrome

Craniosynostoses
--Crouzon
--Apert
--Pfeiffer
--Saethre-Chotzen

Not craniosynostoses

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Goldenhar syndrome

Craniosynostoses

--Crouzon
--Apert
--Pfeiffer
--Saethre-Chotzen

Not craniosynostoses

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

Which non-craniosynostosis conditions are addressed in the Peds book?
Duane’s Retraction Syndrome
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Craniosynostoses
--Crouzon
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Not craniosynostoses
--Goldenhar
--Treacher Collins
--Pierre Robin sequence
--Fetal alcohol

Which non-craniosynostosis conditions are addressed in the Peds book?
Duane’s Retraction Syndrome

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- 90% sporadic, 10% AD

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- Occasionally associated with Goldenhar syndrome

As we will see, the word Goldenhar provides a very convenient mnemonic for remembering the important features of Goldenhar syndrome!
Duane's Retraction Syndrome

Motility disorder featuring:
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3) Up- or downshoot in adduction

90% sporadic, 10% AD

Usually isolated

Can be associated with Goldenhar syndrome

What is its noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

- Upper lid colobomas
- Dermoids of the cornea

What nonocular findings are usually present?

- Ear abnormalities (pre-auricular appendages; aural fistulae)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

Where specifically are dermoids commonly located in Goldenhar?

At the limbus

Are they cognitively impaired?

A minority (5-15%) have mental retardation

Goldenhar syndrome

Very convenient mnemonic
Duane’s Retraction Syndrome

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Goldenhar OAV syndrome

Goldenhar syndrome
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Where specifically are dermoids commonly located in Goldenhar? At the limbus

Are they cognitively impaired? A minority (5-15%) have mental retardation

What is the classic vertebral finding? Hemivertebrae, aka pteryg...

Very convenient mnemonic

Goldenhar
OAV syndrome

Goldenhar syndrome
Duane's Retraction Syndrome
Motility disorder featuring:
1) Retraction of globe on attempted adduction
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Where specifically are dermoids commonly located in Goldenhar?
At the limbus

Are they cognitively impaired?
A minority (5-15%) have mental retardation

What is the classic vertebral finding?
Hemivertebrae, aka butterfly vertebrae

Very convenient mnemonic

Goldenhar syndrome
Goldenhar syndrome: Butterfly vertebrae
Duane’s Retraction Syndrome

Motility disorder featuring:
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Goldenhar OAV syndrome

Lid colobomas
Dermoids
Ear abnormalities
Hemifacial microsomia

A minority (5-15%) have mental retardation

What is the classic vertebral finding? Hemivertebrae, aka butterfly vertebrae

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

Goldenhar syndrome

Very convenient mnemonic

Goldenhar
OAV

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.
Duane's Retraction Syndrome

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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 nonocular findings are usually present?
- Ear abnormalities (pre-auricular appendages; aural fistulae)
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
- At the limbus

Are they cognitively impaired?
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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.
What is its noneponymous name?
Oculo-Auriculo-Vertebral (OAV) syndrome

What other ocular/periocular abnormalities are common in Goldenhar?

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Very convenient mnemonic

Goldenhar OAV syndrome
Duane's Retraction Syndrome

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Are they cognitively impaired?
A minority (5-15%) have mental retardation
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?

Goldenhar
OAV syndrome
Lid coloboma
Dermoid

Very convenient mnemonic

Goldenhar syndrome
Duane's Retraction Syndrome

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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.
Goldenhar syndrome: Lid coloboma
Duane's Retraction Syndrome

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**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?**
DERMID

**What nonocular findings are usually present?**
- Ear abnormalities (pre-auricular appendages; aural fistulae)
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**Where specifically are dermoids commonly located in Goldenhar?**
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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)
Goldenhar syndrome: Epibulbar dermoid
Duane's Retraction Syndrome

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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Are they cognitively impaired?
A minority (5-15%) have mental retardation

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

What is the ‘full’ name of the dermoid in question?
Epibulbar dermoid
Duane's Retraction Syndrome

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Very convenient mnemonic
GOLDENHAR

Goldenhar syndrome
Lid coloboma
Dermoid

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Epibulbar dermoid
What is its noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

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Very convenient mnemonic:

Goldenhar syndrome
OAV syndrome
Lid coloboma
Dermoid

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Very convenient mnemonic

Goldenhar syndrome

OAV syndrome
Lid coloboma
Dermoid

ENHAR
Goldenhar syndrome: Dermolipoma
Duane's Retraction Syndrome

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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?

Very convenient mnemonic

Goldenhar syndrome

OAV syndrome

Lid coloboma

Dermoid; D
Duane's Retraction Syndrome

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2) At least some limitation of horizontal movement
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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)

Goldenhar OAV syndrome
Lid coloboma
Dermoid; Duane’s

Very convenient mnemonic

Gold, En, Ha, R
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

Goldenhar
OAV syndrome
Lid coloboma
Dermoid; Duane’s
E
Nothing starts w/ ‘N’
H

Very convenient mnemonic

Goldenhar syndrome
What is its noneponymous name?
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--Dermoids of the cornea; Duane syndrome

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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 usually present?**
--Ear abnormalities (pre-auricular appendages; aural fistulae)
--Hemifacial microsomia (maxillary/mandibular hypoplasia)

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

I have no idea

Very convenient mnemonic

**Goldenhar syndrome**

- Lid coloboma
- Dermoid; Duane’s
- Ear abnormalities
- Nothing starts w/ ‘N’
- Hemifacial microsomia
- A
- R
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Goldenhar syndrome
Goldenhar syndrome: Hemifacial microsomia
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Are they cognitively impaired?
A minority (5-15%) have mental retardation

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

Why the right side?

Very convenient mnemonic:

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Goldenhar OAV syndrome
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Dermoid; Duane’s
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At the limbus
Goldenhar syndrome: Limbal (epibulbar) dermoids. Note also the lid coloboma (arrow)
What is its noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

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

Goldenhar syndrome

Very convenient mnemonic:

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OAV syndrome
Lid coloboma
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Where specifically are epibulbar dermoids commonly located in Goldenhar? At the limbus

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**Where specifically are epibulbar dermoids commonly located in Goldenhar?**
At the limbus

Are Goldenhar individuals cognitively impaired? **R**

**Very convenient mnemonic**
- Goldenhar
- OAV syndrome
- **Lid coloboma**
- **Dermoid; Duane’s**
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Are Goldenhar individuals cognitively impaired?
Retardation is present in a minority (~10%)
• 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
Duane’s Retraction Syndrome

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- F > M
- OS > OD
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These are the opposite of what they are in another strab syndrome
Duane’s Retraction Syndrome

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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
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
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
Duane’s Retraction Syndrome cont

Three types of Duane’s are recognized:

- Type ?
- Type ?
- Type ?
Duane’s Retraction Syndrome cont

Three types of Duane’s are recognized:

- Type 1
- Type 2
- Type 3
Duane’s Retraction Syndrome cont

Three types of Duane’s are recognized:

- **Type 1**: Limited movement
- **Type 2**
- **Type 3**
Duane’s Retraction Syndrome cont

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

Three types of Duane’s are recognized:

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

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

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)

*Mnemonic:* The number of ‘Ds’ = type of Duane’s
Duane’s Retraction Syndrome cont

- Three types of Duane’s are recognized:

  * **Esotropic Type 1**: Limited abduction
  * **Exotropic Type 2**: Limited adduction
  * **Ortho Type 3**: Both abduction and adduction limited

The three are known also as the *Esotropic, Exotropic* and *Orthotrop*ic types respectively.
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: >50% of cases
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
Duane’s Retraction Syndrome cont

- Three types of Duane’s are recognized:
  - Type 1: Limited abduction
  - Type 2: Limited adduction
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- Etiology of Duane’s:
  - Absent cranial nerve nucleus
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
Duane’s Retraction Syndrome cont

Three types of Duane’s are recognized:
- *Type 1*: Limited abduction
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Most common type: 1 in >50% of cases

Etiology of Duane’s:
- Absent **CN6** nucleus
- cranial nerve innervates LR
Duane’s Retraction Syndrome cont
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Etiology of Duane’s:

- Absent **CN6** nucleus
- **CN3** innervates LR
- Paradoxical innervation to LR…
  - …*increases* with attempted movement
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Q

- Duane’s Retraction Syndrome cont
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How does this lead to the hallmark of Duane’s retraction syndrome (ie, globe retraction)?

- CN3 innervates LR
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  - ...increases with attempted adduction
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Duane’s Retraction Syndrome cont

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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.
Duane’s Retraction Syndrome cont

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- CN3 innervates LR
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  - …*increases* with attempted *adduction*
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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 decrease in innervation to its antagonist.

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How does this lead to the hallmark of Duane’s retraction syndrome (i.e., 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 (i.e., retract) into the orbit.

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- Paradoxical innervation to LR...
  - …*increases* with attempted *adduction*
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Duane’s Retraction Syndrome cont

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- **CN3 innervates LR**
- **Paradoxical innervation to LR**
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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 arises 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 have been expected to decrease LR innervation.

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 (i.e., 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 (i.e., retract) into the orbit.

- CN3 innervates LR
- Paradoxical innervation to LR…
  - …increases with attempted adduction
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Duane’s Retraction Syndrome

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: 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

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)
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**Q**

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- Most common type: 1 in >50% of cases
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What is the clinical hallmark of MGJW?

- ...decreases with attempted abduction
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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?*

A ptotic lid elevates in response to voluntary masticatory movements of the jaw...
MGJW. Note the resolution of ptosis (second pictures) with a jaw movement.
Duane's Retraction Syndrome

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: I in >50% of cases

Etiology of Duane's:

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

**What is the clinical hallmark of MGJW?**

A ptotic lid elevates in response to voluntary masticatory movements

- ...decreases with attempted abduction

**What are the muscles of mastication?**

- Temporalis

Which cranial nerve innervates them?

The trigeminal (V)
Duane's Retraction Syndrome

There are three types of Duane's:

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

The most common type is Type 1, occurring in over 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

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- What is the clinical hallmark of MGJW?
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What are the muscles of mastication?

- Medial (or internal) pterygoid
- Lateral (or external) pterygoid
- Masseter
- Temporalis
Duane's Retraction Syndrome

Three types of Duane's are recognized:

1. **Type 1**: Limited abduction
2. **Type 2**: Limited adduction
3. **Type 3**: Both abduction and adduction limited

Most common type: I in >50% of cases

Etiology of Duane's:

- Absent CN6 nucleus
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Which branch of the trigeminal?

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- Most common in children, arising in 50%

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Speaking of congenital cranial dysinnervation disorders:

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

In a nutshell, what findings are typical in Mobiüs syndrome?

- Bilateral CN6 palsies + bilateral CN7 palsies (tongue, chest, and limb defects often occur as well)

What is the most common gaze status in primary position?

**Orthophoria** (but eso- and exotropic presentations occur as well)

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

'Mask-like'

Are vertical eye movements affected, or intact?

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- Absent CN6 nucleus
- CN3 innervates LR
- Paradoxical innervation to LR…
  - Increases with attempted adduction
  - Decreases with attempted abduction

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**

Speaking of congenital cranial dysinnervation disorders:

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

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

What is the clinical hallmark of MGJW?

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

Is the ptosis of MGJW unilateral, or bilateral?

- Unilateral

Which jaw movements can be involved?

- Lateral displacement
- Protrusion
- Wide opening
- Clenching

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

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**Mobiüs syndrome**

In a nutshell, what findings are typical in Mobiüs syndrome?

- Bilateral CN6 palsies + bilateral CN7 palsies (tongue, chest, and limb defects often occur as well)

What is the most common gaze status in primary position?

- Orthophoria (but eso- and exotropic presentations occur as well)

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

- 'Mask-like'

Are vertical eye movements affected as well?

- Intact
Duane's Retraction Syndrome

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: I in >50% of cases

Etiology of Duane's:

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- Paradoxical innervation to LR…
  - Increases with attempted adduction
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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?
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Speaking of congenital cranial dysinnervation disorders:

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

In a nutshell, what findings are typical in Möbius syndrome?
- Bilateral CN6 palsies + bilateral CN7 palsies (tongue, chest, and limb defects often occur as well)
- Orthophoria (but eso- and exotropic presentations occur as well)
- What is the classic descriptor of the expressionless facies resulting from the bilateral CN7 palsies?
  - 'Mask-like'
  - Are vertical eye movements affected as well?
  - No, they are intact

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

How on earth can an infant be born with myasthenia?
- 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?
- Thankfully no—the condition resolves spontaneously and rapidly.
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Another ophthalmologic condition involving CN3...what is it?

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In a nutshell:

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Duane’s Retraction Syndrome cont

Key observation to differentiate Duane’s syndrome from CN6 palsy:
Duane’s Retraction Syndrome cont
- Key observation to differentiate Duane’s syndrome from CN6 palsy: Retraction on attempted adduction
• 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
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
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
- **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**
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**
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
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- Another useful observation: Assess the patient’s muscle balance in primary gaze
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- Key observation to differentiate Duane’s syndrome from CN6 palsy: **Retraction on attempted adduction**
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- Still another: Look for **narrowing of the palpebral fissure on attempted adduction**
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- Still another: Look for **narrowing** of the **palpebral fissure** on attempted adduction
Duane syndrome: Fissure narrowing on adduction
Duane’s Retraction Syndrome: Management

Is there any surgical procedure that will normalize ocular rotations?
Duane’s Retraction Syndrome: Management

*Is there any surgical procedure that will normalize ocular rotations?* 
No
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
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in primary
Duane syndrome: Deviated in primary
Duane’s Retraction Syndrome: Management

Operate only if:

- Deviated in primary
- Abnormal
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.
Duane’s Retraction Syndrome: Management

Operate only if:

- Deviated in primary
- Abnormal head position
- Marked
Duane’s Retraction Syndrome: Management

Operate only if:

- Deviated in primary OR
- Abnormal head position OR
- Marked retraction
Duane syndrome: Marked retraction
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary**  **OR**
  - Abnormal **head position**  **OR**
  - Marked **retraction**  **OR**
  - Large **or**

172
Duane’s Retraction Syndrome: Management

Operate only if:

- Deviated in primary
- Abnormal head position
- Marked retraction
- Large upshoot/downshoot
Duane syndrome: Marked retraction and upshoot
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
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**
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 surgery if >20° ET
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∆ ET
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in primary \textit{OR} 
  - Abnormal head position \textit{OR}
  - Marked retraction \textit{OR}
  - Large upshoot/downshoot

- Type 1 (ET type): Ipsilateral MR recession
  - Add contralateral MR recession if $>20\Delta$ ET

- Most surgeons refrain from surgery
Duane’s Retraction Syndrome: Management

Operate only if:

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

Type 1 (ET type): Ipsilateral MR recession
- Add contralateral MR recession if >20Δ 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 >20Δ 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.
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° ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type):
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 >20Δ ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type): Ipsilateral LR recession
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Operate only if:
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- Abnormal head position
- Marked retraction
- Large upshoot/downshoot

Type 1 (ET type): Ipsilateral MR recession
- Add contralateral MR recession if >20° ET
- Most surgeons refrain from LR resection

Type 2 (XT type): Ipsilateral LR recession
- Add surgery if >20° XT
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in primary OR
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- Type 1 (ET type): Ipsilateral MR recession
  - Add contralateral MR recession if >20\(\Delta\) ET
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- Deviated in primary
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- Type 2 (XT type): Ipsilateral LR recession
- Add contralateral LR recession if >20° XT
- All surgeons refrain from surgery in Type 2/XT type
Duane’s Retraction Syndrome: Management

Operate only if:
- Deviated in primary OR
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- Add contralateral LR recession if >20° XT
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Type 3 (Ortho type)
- No surgery will improve
Duane’s Retraction Syndrome: Management

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  - Add contralateral MR recession if >20˚ ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type): Ipsilateral LR recession
  - Add contralateral LR recession if >20˚ XT
  - All surgeons refrain from MR resection in Type 2/XT type

- Type 3 (Ortho type)
  - No surgery will improve excursion
Duane’s Retraction Syndrome: Management

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  - No surgery will improve excursion
  - Recess both LR and MR to reduce
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Duane’s Retraction Syndrome: Management

Operate only if:

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

Type 1 (ET type):
- Ipsilateral MR recession
- Add contralateral MR recession if >20° ET

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° 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**
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    - Deviated in primary
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  - No surgery will improve excursion
  - Recess both LR and MR to reduce retraction

*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.*
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° ET

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° 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