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
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90% sporadic, 10% AD
Duane’s Retraction Syndrome

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● **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**
Duane’s Retraction Syndrome

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1) Retraction of globe on attempted adduction
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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?

A craniofacial malformation
Duane’s Retraction Syndrome

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Q/A

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Goldenhar syndrome

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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Not craniosynostoses

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

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Not craniosynostoses

-?
-?
-?

Which non-craniosynostosis conditions are addressed in the Peds book?
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Craniosynostoses
  --Crouzon
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  --Saethre-Chotzen

Not craniosynostoses
  --Goldenhar
  --Treacher Collins
  --Pierre Robin sequence
  --Fetal alcohol

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As we will see, the word Goldenhar provides a very convenient mnemonic for remembering the important features of Goldenhar syndrome!
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- 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 epibulbar dermoids commonly located in Goldenhar?

- At the limbus

Are they cognitively impaired?

- A minority (5-15%) have mental retardation

Goldenhar

- OAV syndrome
- Lid colobomas
- Dermoids
- Ear abnormalities
- Hemifacial microsomia

Nothing starts w/ 'N'

- Retardation in 5-15%

Very convenient mnemonic

Goldenhar → GOLDENHAR

Goldenhar syndrome
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Very convenient mnemonic: GLDENHAR
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What is the classic vertebral finding?
Hemivertebrae, aka "butterfly vertebrae"

Very convenient mnemonic

Goldenhar OAV syndrome

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

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

Very convenient mnemonic

Goldenhar OAV syndrome
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Goldenhar syndrome: Butterfly vertebrae
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Goldenhar syndrome
OAV syndrome

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

- Motility disorder featuring:
  1. Retraction of globe on attempted adduction
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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)
- Hemifacial microsomia (maxillary/mandibular hypoplasia)

Where specifically are dermoids commonly located in Goldenhar?
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What other ocular/periocular abnormalities are common in Goldenhar?

-- Lid coloboma
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What nonocular findings are usually present?
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Very convenient mnemonic

Goldenhar
OAV syndrome

LDENHAR

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

Very convenient mnemonic

Goldenhar
OAV syndrome
Lid coloboma
Dermoid

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?

Goldenhar syndrome

Very convenient mnemonic

Goldenhar
OAV syndrome
Lid coloboma
Dermoid
Duane’s Retraction Syndrome

- Retraction of globe on attempted adduction
- 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

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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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 ‘full’ name of the dermoid in question?
dermoid

Very convenient mnemonic

Goldenhar
OAV syndrome
Lid coloboma
Dermoid
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What other ocular/periocular abnormalities are common in Goldenhar?
--Lid coloboma
--Dermoids of the cornea

What are 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)

Very convenient mnemonic:
Goldenhar syndrome
OAV syndrome
Lid coloboma
Dermoid

Goldenhar syndrome
Goldenhar syndrome: Epibulbar dermoid
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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

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

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

Very convenient mnemonic...
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Lid coloboma
Dermoid

ENHAR
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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...

**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?**
- At the limbus

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- A minority (5-15%) have mental retardation

**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)
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Goldenhar OAV syndrome
Lid coloboma
Dermoid; Duane’s

Very convenient mnemonic
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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

Very convenient mnemonic

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

Goldenhar syndrome
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--Hemifacial microsomia (maxillary/mandibular hypoplasia)

Goldenhar syndrome

Very convenient mnemonic

G - Goldenhar  
OAV - Oculo-Auriculo-Vertebral
L - Lid coloboma  
D - Dermoid; Duane’s  
E - Ear abnormalities  
N - Nothing start w/ ‘N’  
H - Hemifacial microsomia  
A -  
R -
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)

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

Very convenient mnemonic

Goldenhar syndrome
- Lid coloboma
- Dermoid; Duane’s
- Ear abnormalities
- Nothing starts w/ ‘N’
- Hemifacial microsomia

Goldenhar syndrome
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Goldenhar
OAV syndrome
Lid coloboma
Dermoid; Duane’s
Ear abnormalities
Nothing starts w/ ‘N’
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The right

Why the right side?

Very convenient mnemonic:

Goldenhar
OAV syndrome
Lid coloboma
Dermoid; Duane’s
Ear abnormalities
Nothing starts w/ ‘N’
Hemifacial microsomia
A
R

Goldenhar 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

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

Why the right side?
I have no idea
**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

**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)
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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

---

Goldenhar syndrome

- Lid coloboma
- Dermoid; Duane’s
- Ear abnormalities
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- Hemifacial microsomia

**Very convenient mnemonic**

- G: Goldenhar
- OAV: Oculo-Auriculo-Vertebral
- L: Lid coloboma
- D: Dermoid; Duane’s
- E: Ear abnormalities
- N: Nothing starts w/ ‘N’
- H: Hemifacial microsomia
- A: A
- R: R
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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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Very convenient mnemonic
Goldenhar
OAV syndrome
Lid coloboma
Dermoid; Duane’s
Ear abnormalities
Nothing starts w/ ‘N’
Hemifacial microsomia
At the limbus
R
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

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

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

Several slides ago I acknowledged that epibulbar dermoids had another legit name. At long last--what is it?
One word: dermoids
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)
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

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

Goldenhar syndrome

Goldenhar
OAV syndrome
Lid coloboma
Dermoid; Duane’s
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Nothing starts w/ ‘N’
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
At the limbus

Are Goldenhar individuals cognitively impaired?
R

Goldenhar
OAV syndrome
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Goldenhar syndrome
What is its noneponymous name?
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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

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

- 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
Duane’s Retraction Syndrome

- Motility disorder featuring:
  1) Retraction of globe on attempted adduction
  2) At least some limitation of horizontal movement
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- Usually isolated
  - Can be associated with Goldenhar syndrome

- F > M

- OS > OD
Duane’s Retraction Syndrome

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- 90% sporadic, 10% AD

- Usually isolated
  - Can be associated with Goldenhar syndrome

- F > M
  - These are the opposite of what they are in another strab condition

- 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

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%
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 *Orthotropic* 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: \[\frac{1}{3}>\%\] 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
- **Type 3**: Both abduction and adduction limited

Most common type: 1 in >50% of cases

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
- **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
- Innervates LR
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
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
- **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 **adduction**
      - …**decreases** with attempted **abduction**
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 (i.e., globe retraction)?

- CN3 innervates LR
  - Paradoxical innervation to LR...
    - ...increases with attempted adduction
    - ...decreases with attempted abduction
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
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 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
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
Duane’s Retraction Syndrome cont

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

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 adduction
- ...decreases with attempted abduction

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.

**What is the eponymous name of this law?**

**Law of reciprocal innervation**
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 would be accompanied by a 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
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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 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 to result in a proportional decrease in 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 (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

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

Most common type: **I** in >50% of cases

**Etiology of Duane's syndrome**:
- Absent CN6 nucleus
- CN3 innervates LR
- Paradoxical innervation to LR:
  - Increases with attempted adduction
  - Decreases with attempted abduction

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

What is the clinical hallmark of MGJW?

- ...decreases with attempted abduction
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: 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

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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
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? A ptotic lid elevates in response to voluntary masticatory movements.

- ...decreases with attempted abduction
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… decreases…

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? A ptotic lid elevates in response to voluntary masticatory movements

What are the muscles of mastication?
- Medial (or internal) pterygoid
- Lateral (or external) pterygoid
- Masseter
- Temporalis

...decreases with attempted abduction
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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Which cranial nerve innervates them?

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Which cranial nerve innervates them?

The trigeminal (V)

---

…decreases with attempted abduction
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- Masseter
- Temporalis

Which cranial nerve innervates them?

The trigeminal (V)

Which branch of the trigeminal?

...decreases with attempted abduction
Duane's Rejection 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
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  - Which branch of the trigeminal?
    - The mandibular (V3)
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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.

So, putting it all together: In MGJW, which cranial nerve (dys)innervates what muscle?

- Which cranial nerve innervates them?
  - The trigeminal (V)
- Which branch of the trigeminal?
  - The mandibular (V₃)
- What are the muscles of mastication?
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  - Temporalis

...decreases with attempted abduction
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So, putting it all together: In MGJW, which cranial nerve (dys)innervates what muscle? V₃ (dys)innervates the levator

What are the muscles of mastication?
- Medial (or internal) pterygoid
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Which cranial nerve innervates them? The trigeminal (V)

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

- **...decreases** with attempted abduction
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**Which jaw movements can be involved?**
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- Lateral displacement
- Protrusion
- Wide opening
- Clenching

- decreases with attempted abduction
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  - Mom says the infant's lid 'twitches' while nursing
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- …decreases with attempted abduction
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 muscle balance in clinical exam component (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
- In CN6 palsy, is usually 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**
  - In CN6 palsy, is usually **esotropic**
  - In Duane’s, 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
  - In CN6 palsy, is usually esotropic
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Duane’s Retraction Syndrome cont

- Key observation to differentiate Duane’s syndrome from CN6 palsy: Retraction on attempted adduction
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- 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’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
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
Is there any surgical procedure that will normalize ocular rotations?
No

If you can’t normalize rotations, why do you operate?
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 OR
- 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 OR
  - Abnormal head position OR
  - Marked
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
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 >20Δ 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 >20Δ 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
  - Most surgeons refrain from 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
  - 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):...
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° 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
    - 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**
  - **Type 2 (XT type):** **Ipsilateral LR recession**
    - Add **LR resection surgery** if >20Δ XT
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): Ipsilateral LR recession
- Add contralateral LR recession if >20° XT
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

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

- Type 3 (Ortho type)
  - No surgery will improve
**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):** 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

- 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): 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
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in **primary** OR
  - Abnormal **head position** OR
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  - 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 **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**
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 $\Delta ET > 20$°
- Most surgeons refrain from LR resection

Type 2 (XT type): Ipsilateral LR recession
- Add contralateral LR recession if $\Delta XT > 20$°
- 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

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

- Type 2 (XT type): Ipsilateral LR recession
  - Add contralateral LR recession if >20Δ 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

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.
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
  - 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

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.