Duane’s Retraction Syndrome

- Motility disorder featuring:
  1) Retraction of globe on attempted adduction
  2)
  3) an abnormal eye movement
Duane’s Retraction Syndrome

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

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  1) Retraction of globe on attempted adduction
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- 90% sporadic, 10% AD
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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
Duane’s Retraction Syndrome

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

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It is sporadic

Is there a sex predilection?
Yes, M is twice as likely to be affected as F
Duane’s Retraction 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
What is its noneponymous name?

(Hint: Note that the ‘O’ in the vertical Goldenhar below is a different color than the other letters…)

Very convenient mnemonic
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?
- 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
Duane's Retraction Syndrome

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Goldenhar

OAV syndrome

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

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

Very convenient mnemonic

Goldenhar OAV syndrome

This mnemonic helps remember the findings associated with Goldenhar syndrome:

Gold: Goldenhar
Oav: Oculo-Auriculo-Vertebral
Har: Hemivertebrae
Duane's Retraction Syndrome

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Where specifically are dermoids commonly located in Goldenhar?
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Are they cognitively impaired?
- A minority (5-15%) have mental retardation

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

Very convenient mnemonic: L.O.G.O.S. - L (Lid colobomas) OAV syndrome Goldenhar syndrome
Duane’s Retraction Syndrome

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

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Another syndrome of ophthalmic concern includes butterfly vertebrae as a finding. What is it?
What is its noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

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

Another syndrome of ophthalmic concern includes butterfly vertebrae as a finding. What is it? Alagille syndrome. If you want more info on Alagille syndrome—and if you don’t know it, you should—check out the slide-set on anterior segment dysgenesis.

Goldenhar syndrome
OAV syndrome

Very convenient mnemonic

Goldenhar
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?

Goldenhar syndrome

Lid coloboma
Dermoid

Very convenient mnemonic

Goldenhar OAV syndrome
 Duane's Retraction Syndrome

Motility disorder featuring:

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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.
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What is the ‘full’ name of the dermoid in question? dermoid

Very convenient mnemonic

Goldenhar OAV syndrome
Lid coloboma

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

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

What is the ‘full’ name of the dermoid in question?
Epibulbar dermoid

Note: There is another legit answer, so if you came up with that one, no worries (we’ll identify it shortly)
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Where specifically are epibulbar dermoids commonly located in 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.
Duane's Retraction Syndrome
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Oculo-Auriculo-Vertebral (OAV) syndrome

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

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

Is there a relationship between epibulbar dermoids and lipodermoids (aka dermolipomas)?
Yes. The relationship is that, like dermoids, lipodermoids are associated with Goldenhar

Where are dermolipomas typically located?
The temporal fornix

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

Very convenient mnemonic

Goldenhar syndrome
OAV syndrome
Lid coloboma
Dermoid

Goldenhar

Looking for a mnemonic? Try:

**ENHAR**

E = Epibulbar
dermoid
N = Nothing starts with 'N'
H = Hemifacial microsomia
A = At the limbus
R = Retardation in 5-15%
Duane's Retraction Syndrome

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Goldenhar syndrome

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Very convenient mnemonic
What is its noneponymous name? 
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**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)

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

- Very convenient mnemonic
What is its noneponymous name? 
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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? 
I have no idea
What is its noneponymous name? Oculo-Auriculo-Vertebral (OAV) syndrome

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

Lid coloboma
Dermoid
Ear abnormalities
Nothing starts w/ ‘N’
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Very convenient mnemonic

Very

convenient

mnemonic

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

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Why the right side?

I have no idea

Very convenient mnemonic
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Why the right side? I have no idea
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Oculo-Auriculo-Vertebral (OAV) syndrome
Lid coloboma
Dermoids of the cornea
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Hemifacial microsomia

Very convenient mnemonic:

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

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Limbal dermoids

Goldenhar syndrome
- OAV syndrome
- Lid coloboma
- Dermoid
- Ear abnormalities
- Nothing starts w/ ‘N’
- Hemifacial microsomia
- At the limbus

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

Goldenhar
OAV syndrome
Lid coloboma
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90% sporadic, 10% AD

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Can be associated with
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Where specifically are epibulbar dermoids commonly located in Goldenhar?
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Are Goldenhar individuals cognitively impaired?
A minority (~10%) have mental retardation
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- F > M
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  • **90%** sporadic, **10%** AD
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    • Can be associated with **Goldenhar** syndrome
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Q

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

- 90% sporadic, 10% AD
- Usually isolated
  - Can be associated with Goldenhar syndrome

- F > M
- OS > OD

These are the opposite of what they are in another strab condition syndrome
Duane’s Retraction Syndrome

Motility disorder featuring:
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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
  - These are the opposite of what they are in Brown syndrome

- OS > OD
  - Bilateral in ~15%
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’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’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
  - **Exotropich** Type 2: Limited adduction
  - **Orthotropic** 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: 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
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
  - Cranial nerve 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
Q

Duane’s Retraction Syndrome cont

Three types of Duane’s are recognized:

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

*How does this lead to the hallmark of Duane’s retraction syndrome (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

Wait—I get why the MR should contract during attempted adduction, but why does it say the LR should relax? This makes it sound like the LR is obligated in some sense to relax. What’s this all about?
The LR is obligated to relax. This obligation stems from one of the fundamental laws governing motor control, that being the law of reciprocal innervation, which states that increased innervation to a given EOM is accompanied by a reciprocal decrease in innervation to its antagonist. Thus, in an intact EOM control system, the increased MR innervation associated with attempted adduction would be accompanied by a proportional decrease in innervation to the ipsilateral LR.

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

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

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

**law of reciprocal innervation**

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

Sherrington’s law (of reciprocal innervation)
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?**

**Sherrington’s 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, in an intact EOM control system, the increased MR innervation associated with attempted adduction would be accompanied by a proportional decrease in innervation to the ipsilateral LR.

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

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

Three types of Duane's are recognized:

- **Type 1**: Limited abduction
- **Type 2**: Limited adduction
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Most common type: I in >50% of cases

Etiology of Duane's:

- Absent CN6 nucleus
- **CN3 innervates LR**
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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
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An aside: Duane syndrome is a congenital condition in which CN3 (dys)innervates the LR. What is the general term for such congenital cranial dysinnervation disorders? They are called ‘congenital cranial dysinnervation disorders’

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

Marcus-Gunn jaw-winking syndrome (MGJW)

What is the clinical hallmark of MGJW?

- ...decreases with attempted abduction
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 of the jaw

- ...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: **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

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)**

- Marcus–Gunn jaw-winking syndrome is a congenital cranial dysinnervation disorder involving an ophthalmic movement (lid elevation) that occurs in response to voluntary masticatory movements.

What are the muscles of mastication?

- **Temporalis**

What are the muscles of mastication?

- **Masseter**
- **Medial Pterygoid**
- **Lateral Pterygoid**

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 with attempted adduction
  - ...decreases with attempted abduction

A

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

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)**

- Marcus Gunn jaw-winking syndrome is FQ3

What is the clinical hallmark of MGJW?

A ptotic lid elevates in response to voluntary *masticatory movements*

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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:

- **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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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)**

Marcus-Gunn jaw-winking syndrome occurs in ~5-8% of cases

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

What cranial nerve innervates them?

- Trigeminal (V)

---

...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)**

- Marcus-Gunn syndrome in 5-10% of cases

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

Which cranial nerve innervates them?

The trigeminal (V)
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...
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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’

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

- 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

Which cranial nerve innervates them?
- The trigeminal (V)

Which branch of the trigeminal?
Duane's Retraction Syndrome

Three types of Duane's are recognized:

- **Type 1**: Limited abduction
- **Type 2**: Limited adduction
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Most common type: I in >50% of cases

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- Absent CN6 nucleus
- CN3 innervates LR
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  ...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)**

Marcus Gunn syndrome, occurring in >50%

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

Which cranial nerve innervates them?
**The trigeminal (V)**

Which branch of the trigeminal?
**The mandibular (V₃)**
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
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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)**

- May due to defective CN3 innervation: 5-10%

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?

**What are the muscles of mastication?**

--Medial (or internal) pterygoid
--Lateral (or external) pterygoid
--Masseter
--Temporalis

**Which cranial nerve innervates them?**

The trigeminal (V)

**Which branch of the trigeminal?**

The mandibular (V₃)

...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
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  - Increases with attempted adduction
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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)**

Marcus-Gunn jaw-winking syndrome is very rare—5-6%

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?
- **V₃ (dys)innervates the levator**
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…
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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

Is the ptosis of MGJW unilateral, or bilateral?

- …decreases with attempted abduction
Duane's Retraction Syndrome

Three types of Duane's are recognized:

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

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- Absent CN6 nucleus
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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 of the jaw

Is the ptosis of MGJW unilateral, or bilateral? Unilateral
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: **Type 1** 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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--Protrusion
--Wide opening
--Clenching

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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’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

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  ● Still another: Look for narrowing of the palpebral fissure on attempted adduction
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  - Another useful observation: Assess the patient’s muscle balance in primary gaze
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    - In Duane’s, is usually straight
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Q

- 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
Q

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

  *Is there any surgical procedure that will normalize ocular rotations?*
  
  No

  *If you can’t normalize rotations, why do you operate?*
Duane’s Retraction Syndrome: Management

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

- Operate only if:
  - 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’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**
Q

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

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

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

- Type 1 (ET type): surgery
A

- **Duane’s Retraction Syndrome: Management**
  - Operate only if:
    - Deviated in **primary** OR
    - Abnormal **head position** OR
    - Marked **retraction** OR
    - Large **upshoot/downshoot**
  - Type 1 (ET type): **Ipsilateral MR recession**
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 >20° ET
Duane’s Retraction Syndrome: Management

- Operate only if:
  - Deviated in primary  \textbf{OR}
  - Abnormal head position  \textbf{OR}
  - Marked retraction  \textbf{OR}
  - Large upshoot/downshoot
- Type 1 (ET type): Ipsilateral MR \textit{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 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 >20° 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 >20Δ ET
- Most surgeons refrain from **LR resection**

Type 2 (XT 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
  - Add contralateral MR recession if >20° ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type): Ipsilateral LR 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 contralateral MR recession if >20° ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type): Ipsilateral LR recession
  - Add surgery if >20° ET
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 >20Δ ET
  - Most surgeons refrain from LR resection

- Type 2 (XT type): Ipsilateral LR recession
  - Add contralateral LR 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 \( >20\Delta \) ET
  - Most surgeons refrain from **LR resection**

- Type 2 (XT type): **Ipsilateral LR recession**
  - Add **contralateral LR recession** if \( >20\Delta \) ET
  - **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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- Type 1 (ET type): Ipsilateral MR recession
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Duane’s Retraction Syndrome: Management

- Operate only if:
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- Type 2 (XT type): Ipsilateral LR recession
  - Add contralateral LR recession if >20Δ ET
  - 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:
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  - Add contralateral LR recession if >20° ET
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  - Operate only if:
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    - Recess both LR and MR to reduce
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None of the surgeries discussed thus far address upshoot or downshoot. How should these be managed?
Q/A

**Duane’s Retraction Syndrome: Management**

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  - Deviated in primary  **OR**
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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.
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