Motility Disorders: Overview

Which cranial nerves innervate the extraocular muscles (EOMs)?
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Motility Disorders: Overview

What is the name for the collections of neurons that give rise to each of these cranial nerves? (This is not a trick question--the answer is as obvious as it seems.)
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Motility Disorders: Overview

Um, Dr Flynn, 4 comes before 6. Why are these nuclei listed out of order?

CN3 Nucleus
CN6 Nucleus
CN4 Nucleus
Motility Disorders: Overview

Um, Dr Flynn, 4 comes before 6. Why are these nuclei listed out of order? This will be explained shortly.

CN3 Nucleus

CN6 Nucleus

CN4 Nucleus
(As we shall soon see, this ‘nuclear level’ serves as a useful point around which to organize the EOM-control pathway.)
With respect to pathology of the EOM control pathways, there are four major ‘locations.’ One of these (the nuclear) has been identified already. What are the other three? (Hint: Their names reflect the relationship each has to the nuclear level.)
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...you may not be with this one, although you’ll agree it makes sense in context. (Further, and importantly, it is used in the BCSC Neuro book.)
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...you may not be with this one, although you’ll agree it makes sense in context. (Further, and importantly, it is used in the BCSC Neuro book.)
What sorts of inputs comprise the supranuclear pathways?

**Supranuclear**

**Nuclear**

- CN3 Nucleus
- CN6 Nucleus
- CN4 Nucleus

**Infranuclear**

An important rule-of-thumb can be stated regarding supranuclear motility disorders and diplopia—what is it? It is this: With four important exceptions, supranuclear pts do not complain of diplopia.
What sorts of inputs comprise the supranuclear pathways? Those from the cortex, cerebellum, vestibular system, and a number of brainstem nuclei.

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Relationship between level of pathology and diplopia: 

*The rule of thumb*
Motility Disorders: Overview

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Why don’t pts with supranuclear disorders have diplopia?

Because supranuclear disorders affect both eyes in a symmetric fashion.

Relationship between level of pathology and diplopia: The rule of thumb.
Motility Disorders: Overview

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What are the four supranuclear disorders in which pts c/o diplopia?
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Relationship between level of pathology and diplopia: The rule of thumb.
### Supranuclear

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What are the four supranuclear disorders in which pts c/o diplopia?

- Skew deviation
- Divergence insufficiency
- Convergence insufficiency
- Convergence spasm

### Nuclear

### Infranuclear

Relationship between level of pathology and diplopia: The rule of thumb.
What sorts of inputs comprise the supranuclear pathways? Those from the cortex, cerebellum, vestibular system, and a number of brainstem nuclei.

An important rule-of-thumb concerning supranuclear motility disorders is: if you don’t see it, you don’t get it.

It is this: With four important exceptions, supranuclear pts do not complain of diplopia.

Why don’t pts with supranuclear disorders have diplopia? Because supranuclear disorders affect both eyes in a symmetric fashion.

Relationship between level of pathology and diplopia: The rule of thumb

What are the four supranuclear disorders in which pts c/o diplopia?

--Skew deviation
--Divergence insufficiency
--Convergence insufficiency
--Convergence spasm

What are some of the supranuclear disorders that present typically, ie, without diplopia?

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**Supranuclear**

- What are the four supranuclear disorders in which pts c/o diplopia?
  - Skew deviation
  - Divergence insufficiency
  - Convergence insufficiency
  - Convergence spasm

- What are some of the supranuclear disorders that present typically, ie, without diplopia?
  - Gaze palsies, eg, Parinaud syndrome
  - Congenital ocular motor apraxia (COMA)
  - Progressive supranuclear palsy (PSP)
  - Saccadic disorders

- Relationship between level of pathology and diplopia:
  *The rule of thumb*

**Nuclear**

**Infranuclear**

- What sorts of inputs comprise the supranuclear pathways?
  - Those from the cortices, cerebellum, vestibular system, and a number of brainstem nuclei.

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Supranuclear

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  - Those from the cortex, cerebellum, vestibular system, and a number of brainstem nuclei.

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  - Congenital ocular motor apraxia
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  - Skew deviation
  - Divergence insufficiency
  - Convergence insufficiency
  - Convergence spasm

- Why don’t pts with supranuclear disorders have diplopia?
  - Because supranuclear disorders affect both eyes in a symmetric fashion.

Nuclear

- Each of these is addressed in detail in other slide-sets--check the ToC

Infranuclear

Relationship between level of pathology and diplopia:
The rule of thumb
Are lesions of the CN3 nucleus commonly encountered in clinical practice?
Motility Disorders: Overview

Are lesions of the CN3 nucleus commonly encountered in clinical practice?
No, they are rare
Are lesions of the CN4 nucleus commonly encountered in clinical practice?
Motility Disorders: Overview

Supranuclear

Nuclear

Supranuclear

Internuclear

CN3 Nucleus

CN4 Nucleus

CN6 Nucleus

Infranuclear

Are lesions of the CN4 nucleus commonly encountered in clinical practice?
No, these are even rarer
Are lesions of the CN6 nucleus commonly encountered in clinical practice?
Motility Disorders: Overview

**Nuclear**

- CN3 Nucleus
- CN6 Nucleus
- CN4 Nucleus

**Supranuclear**

**Infranuclear**

Are lesions of the CN6 nucleus commonly encountered in clinical practice? While not common, they are a well-known clinical entity.
Which two nuclei share an internuclear connection that is of well-established clinical importance?
Motility Disorders: Overview

Which two nuclei share an internuclear connection that is of well-established clinical importance?
3 and 6
(Apropos a previous question: This is why the nuclei are not in numeric order!)
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?
The medial longitudinal fasciculus (MLF)
Motility Disorders: Overview

What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei? The **medial longitudinal fasciculus (MLF)**

In which direction do impulses travel along the MLF?

**Internuclear**

**CN3**

**MLF**

**CN6**

**Supranuclear**

**Nuclear**

**Infranuclear**
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei? The **medial longitudinal fasciculus** (MLF).

*In which direction do impulses travel along the MLF?* From the CN6 to the CN3 nucleus.
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei? The **medial longitudinal fasciculus** (MLF)

In which direction do impulses travel along the MLF? From the CN6 to the CN3 nucleus

What condition is caused by disruption of the MLF? **Internuclear ophthalmoplegia** (INO)
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?
The medial longitudinal fasciculus (MLF)

In which direction do impulses travel along the MLF?
From the CN6 to the CN3 nucleus

What condition is caused by disruption of the MLF?
Internuclear ophthalmoplegia (INO)
What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?
The **medial longitudinal fasciculus** (MLF)

In which direction do impulses travel along the MLF?
From the CN6 to the CN3 nucleus

What condition is caused by disruption of the MLF?
**Internuclear ophthalmoplegia (INO)**

INO is addressed in detail in its own slide-set (N20)
Next we will turn our attention to the *infranuclear pathway*, which proceeds in an ordered fashion from the nuclei to the extraocular muscles themselves.
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

Infranuclear

The first portion of the nerve as it leaves the nucleus, but before leaving the substance of the brainstem.
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

Fascicular

Infranuclear

The first portion of the nerve as it leaves the nucleus, but before leaving the substance of the brainstem
The cranial-nerve nuclei and their fascicles are located within the brainstem. Given this, it shouldn’t come as a surprise that, generally speaking, lesions of the nuclei and/or fascicles do not present with *isolated* EOM abnormalities; ie, the ophthalmoparesis is almost always accompanied by *nonocular* signs and symptoms of CNS damage.
The cranial-nerve nuclei and their fascicles are located within the brainstem. Given this, it shouldn’t come as a surprise that, generally speaking, lesions of the nuclei and/or fascicles do not present with isolated EOM abnormalities; ie, the ophthalmoparesis is almost always accompanied by nonocular signs and symptoms of CNS damage.

What general term is used to describe conditions presenting with motility dysfunction secondary to fascicle damage + non-ocular CNS findings?
The cranial-nerve nuclei and their fascicles are located within the brainstem. Given this, it shouldn’t come as a surprise that, generally speaking, lesions of the nuclei and/or fascicles do not present with isolated EOM abnormalities; ie, the ophthalmoparesis is almost always accompanied by nonocular signs and symptoms of CNS damage.

What general term is used to describe conditions presenting with motility dysfunction 2ndry to fascicle damage + non-ocular CNS findings?

Fascicular syndrome
Speaking of fascicular syndromes…the Neuro book describes four involving the CN3 fascicle, and two for the CN6. Name them.

CN3 fascicular syndromes:
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Speaking of fascicular syndromes…the Neuro book describes four involving the CN3 fascicle, and two for the CN6. Name them.

**CN3 fascicular syndromes:**
--Weber syndrome
--Benedikt syndrome
--Claude syndrome
--Nothnagel syndrome
Speaking of fascicular syndromes… the Neuro book describes four involving the CN3 fascicle, and two for the CN6. Name them.

**CN3 fascicular syndromes:**
-- Weber syndrome
-- Benedikt syndrome
-- Claude syndrome
-- Nothnagel syndrome

**CN6 fascicular syndromes:**
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--
Speaking of fascicular syndromes…the Neuro book describes four involving the CN3 fascicle, and two for the CN6. Name them.

**CN3 fascicular syndromes:**
--Weber syndrome
--Benedikt syndrome
--Claude syndrome
--Nothnagel syndrome

**CN6 fascicular syndromes:**
--Foville syndrome
--Millard-Gubler syndrome
Speaking of fascicular syndromes…the Neuro book describes four involving the CN3 fascicle, and two for the CN6. Name them.

**CN3 fascicular syndromes:**
--Weber syndrome
--Benedikt syndrome
--Claude syndrome
--Nothnagel syndrome

**CN6 fascicular syndromes:**
--Foville syndrome
--Millard-Gubler syndrome

The fascicular syndromes are addressed in detail in their own slide-set (N14)
The next portion commences once the fascicles exit the brainstem--now they’re a **nerve**. Named for the space in which the nerves travel.
Motility Disorders: Overview

Fascicular

Subarachnoid

The next portion commences once the fascicles exit the brainstem--now they’re a nerve. Named for the space in which the nerves travel.
Which cause of ophthalmoparesis—common among vasculopathies—is attributed to damage occurring to the subarachnoid segments?
Motility Disorders: Overview

 WHICH CAUSE OF OPHTHALMOPARESIS—COMMON AMONG VASCOLOGY—IS ATTRIBUTED TO DAMAGE OCCURRING TO THE SUBARACHNOID SEGMENTS?

Ischemic palsies (ie, a so-called ‘diabetic third’ or ‘diabetic sixth’)
The nerves then leave the subarachnoid space by diving into a space of a very different sort. This portion is named for the space entered into.
The nerves then leave the subarachnoid space by diving into a space of a very different sort. This portion is named for the space entered into.
What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously.

Which nerves may be involved?

- CN3
- CN4
- CN6
- V1
- V2
- Sympathetics
What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process? The involvement of two or more cranial nerves simultaneously.
What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process?
The involvement of two or more cranial nerves simultaneously

Which nerves may be involved?
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What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process? The involvement of two or more cranial nerves simultaneously.

Which nerves may be involved?

- Involvement manifests as...
- Involvement manifests as...
Motility Disorders: Overview

**Supranuclear**

**Nuclear**

**Internuclear**

**Infranuclear**

What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process?
The involvement of two or more cranial nerves simultaneously.

Which nerves may be involved?

- Involvement manifests as ophthalmoplegia
- Involvement manifests as facial hypoesthesia
- Involvement manifests as Horner's

CN3 Nucleus $\rightarrow$ MLF $\rightarrow$ CN6 Nucleus

CN4 Nucleus
What is the hallmark of ophthalmoplegia secondary to a cavernous sinus process? The involvement of two or more cranial nerves simultaneously.

Which nerves may be involved?
- CN3
- CN4
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- V1
- V2
- Sympathetics

Involvement manifests as ophthalmoplegia
Involvement manifests as facial hypoesthesia
Involvement manifests as Horner's
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN6 Nucleus

CN4 Nucleus

Fascicular

Subarachnoid

Cavernous sinus

Getting pretty close now. Post-cavernous sinus, another well-defined space.

Infranuclear

?
Getting pretty close now. Post-cavernous sinus, another well-defined space.
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN6 Nucleus

CN4 Nucleus

Fascicular

Subarachnoid

Cavernous sinus

Superior orbital fissure

Orbital

Superior orbital fissure

BTW, the answer superior orbital fissure is just as good here as ‘orbit’ (if not better, as the Neuro book breaks out the fissure as a separate structure in the pathway)
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN6 Nucleus

CN4 Nucleus

Fascicular

Subarachnoid

Cavernous sinus

Superior orbital fissure

Orbital apex

Orbital

Likewise, the answer orbital apex would also be reasonable at this junction

??
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

Fascicular

Subarachnoid

Cavernous sinus
Superior orbital fissure
Orbital apex
Orbital

Motility disorders 2ndry to pathology in these areas are addressed in detail in their own slide-set (N19)
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

Fascicular
Subarachnoid
Cavernous sinus
Orbital
?
?

Where the journey ends for the nerves.
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN4 Nucleus

CN6 Nucleus

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Where the journey ends for the nerves.

Infranuclear

Where the journey ends for the nerves.
Per the Neuro book, what is the “prototypical” disease of the neuromuscular junction?
Per the Neuro book, what is the “prototypical” disease of the neuromuscular junction?
Myasthenia gravis
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus \(\rightarrow\) MLF \(\rightarrow\) CN6 Nucleus

CN4 Nucleus

Infranuclear

Fascicular
Subarachnoid
Cavernous sinus
Orbital
Neuromuscular junction

? 

And finally... Don’t forget pathology here when evaluating motility disorders!
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

Fascicular
Subarachnoid
Cavernous sinus
Orbital
Neuromuscular junction
Extraocular muscle

And finally...Don't forget pathology here when evaluating motility disorders!
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN6 Nucleus

CN4 Nucleus

Fascicular

Subarachnoid

Cavernous sinus

Infranuclear

Extraocular muscle

What sorts of conditions are included here?

Restrictive (e.g., thyroid eye dz); inflammatory (e.g., orbital myositis); myopathies (e.g., chronic progressive external ophthalmoplegia)
Motility Disorders: Overview

What sorts of conditions are included here?
Restrictive (eg, thyroid eye dz); inflammatory (eg, orbital myositis); myopathies (eg, chronic progressive external ophthalmoplegia)

Extraocular muscle
Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3 Nucleus

MLF

CN6 Nucleus

CN4 Nucleus

Supranuclear:

- Fascicular
- Subarachnoid

Infranuclear:

- Cavernous sinus
- Orbital
- Neuromuscular junction
- Extraocular muscle

Commit this slide to memory, and use it to organize your thought process when dealing with an ophthalmoparesis pt!