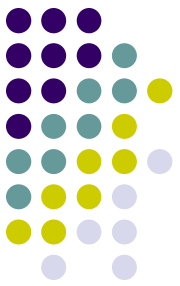


Motility Disorders: Overview



1

CN...

CN...

CN...

Which cranial nerves innervate the extraocular muscles (EOMs)?

Motility Disorders: Overview



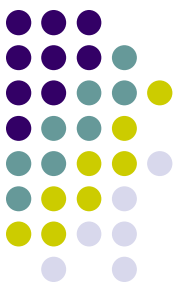
CN3

CN6

CN4

Which cranial nerves innervate the extraocular muscles (EOMs)?

Motility Disorders: Overview



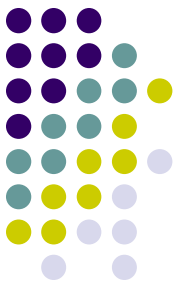
CN3
?

CN6
?

CN4
?

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.)

Motility Disorders: Overview



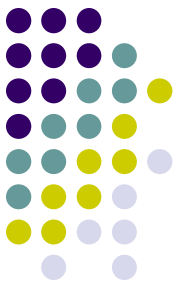
**CN3
Nucleus**

**CN6
Nucleus**

**CN4
Nucleus**

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.)

Motility Disorders: Overview



Nuclear

CN3
Nucleus

CN6
Nucleus

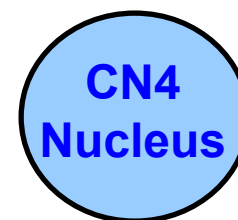
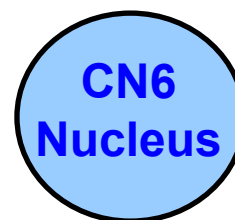
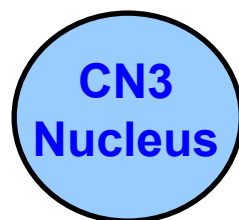
CN4
Nucleus

(As we shall soon see, this 'nuclear level' serves as a useful point around which to organize the EOM-control pathway.)

Motility Disorders: Overview

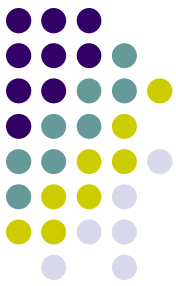


Um, Dr Flynn, 4 comes before 6. Why are these nuclei listed out of order?



Motility Disorders: Overview

7



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

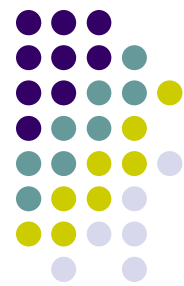
This will be explained shortly

**CN3
Nucleus**

**CN6
Nucleus**

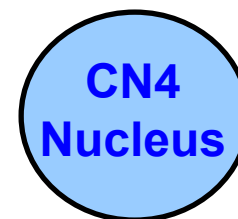
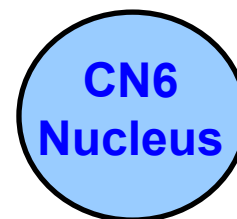
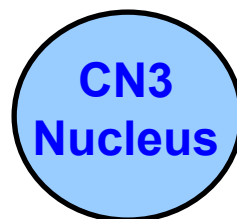
**CN4
Nucleus**

Motility Disorders: Overview



?

Nuclear

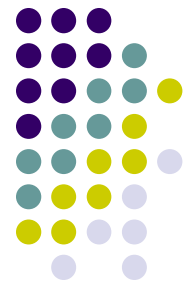


?

?

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.)

Motility Disorders: Overview



?

Note: While you're familiar with these terms...

?

Nuclear

CN3
Nucleus

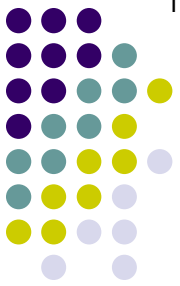
CN6
Nucleus

CN4
Nucleus

?

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.)

Motility Disorders: Overview



?

Note: While you're familiar with these terms...

?

Nuclear

CN3
Nucleus

CN6
Nucleus

CN4
Nucleus

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.)

?

...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.)

Motility Disorders: Overview

Supranuclear

Note: While you're familiar with these terms...

Internuclear

Nuclear

CN3
Nucleus

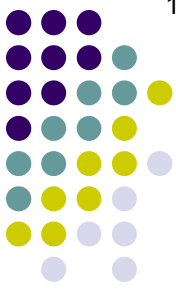
CN6
Nucleus

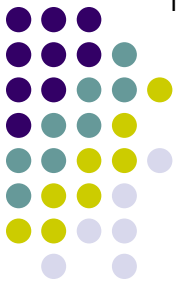
CN4
Nucleus

Infranuclear

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.)

...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.)





Motility Disorders: Overview

Supranuclear

The ***supranuclear pathways*** consists of inputs to the nuclei from centers in the cortex, cerebellum, vestibular system, etc.

Internuclear

Nuclear

CN3
Nucleus

CN6
Nucleus

CN4
Nucleus

Infranuclear



Motility Disorders: Overview

Supranuclear

The ***supranuclear pathways*** consists of inputs to the nuclei from centers in the cortex, cerebellum, vestibular system, etc. These locations are 'supra' in that they carry signals *to* the nuclei.

Internuclear

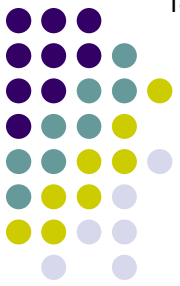
Nuclear

CN3
Nucleus

CN6
Nucleus

CN4
Nucleus

Infranuclear



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

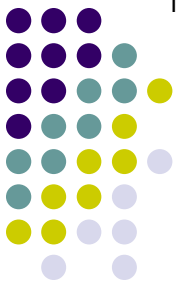
**CN3
Nucleus**

**CN6
Nucleus**

**CN4
Nucleus**

*Are lesions of the CN3 nucleus
commonly encountered in
clinical practice?*

Infranuclear



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

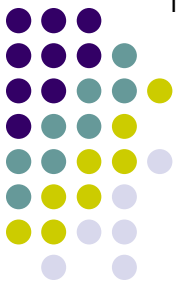
**CN3
Nucleus**

**CN6
Nucleus**

**CN4
Nucleus**

*Are lesions of the CN3 nucleus
commonly encountered in
clinical practice?*
No, they are rare

Infranuclear

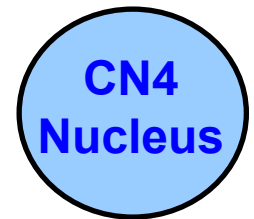
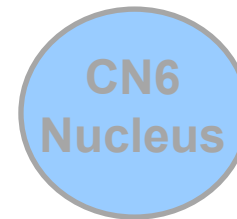


Motility Disorders: Overview

Supranuclear

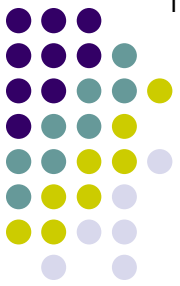
Internuclear

Nuclear



*Are lesions of the CN4 nucleus
commonly encountered in
clinical practice?*

Infranuclear

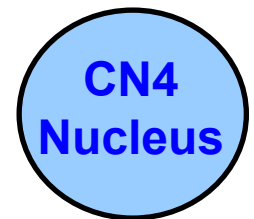
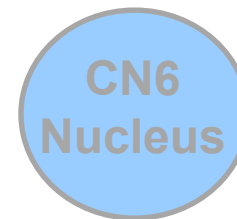


Motility Disorders: Overview

Supranuclear

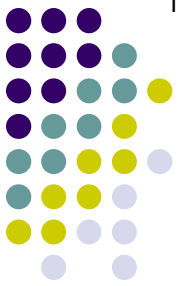
Internuclear

Nuclear



*Are lesions of the CN4 nucleus
commonly encountered in
clinical practice?*
No, these are even rarer

Infranuclear



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

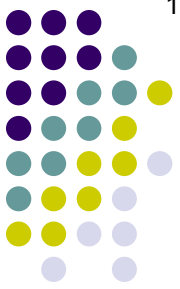
CN3
Nucleus

CN6
Nucleus

CN4
Nucleus

*Are lesions of the CN6 nucleus
commonly encountered in
clinical practice?*

Infranuclear

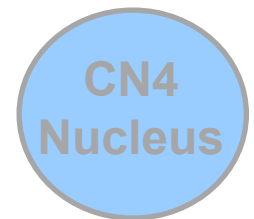
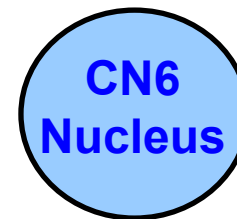
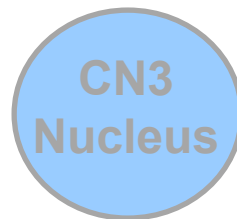


Motility Disorders: Overview

Supranuclear

Internuclear

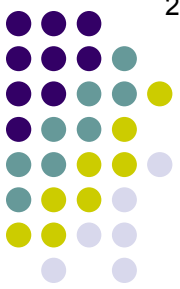
Nuclear



*Are lesions of the CN6 nucleus
commonly encountered in
clinical practice?*

**While not common, they are
a well-known clinical entity**

Infranuclear

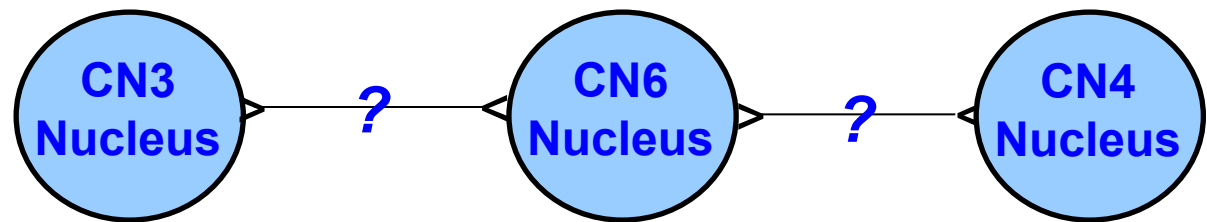


Motility Disorders: Overview

Supranuclear

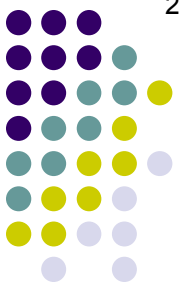
Internuclear

Nuclear



Which two nuclei share an internuclear connection that is of well-established clinical importance?

Infranuclear

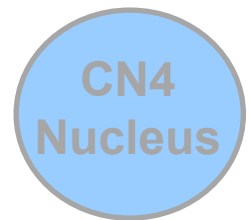
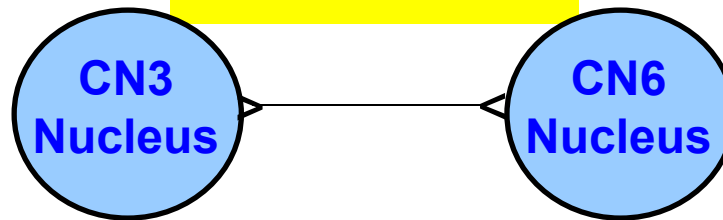


Motility Disorders: Overview

Supranuclear

Nuclear

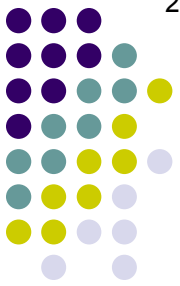
Internuclear



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!)

Infranuclear

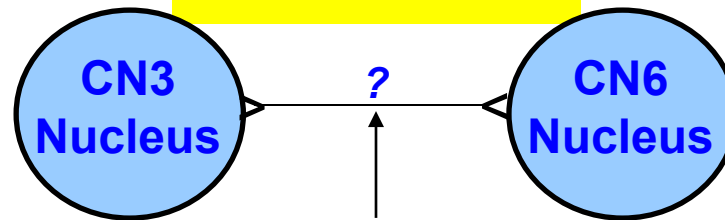


Motility Disorders: Overview

Supranuclear

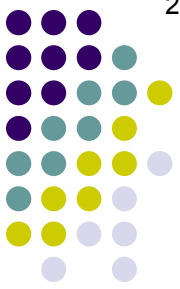
Nuclear

Internuclear



What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?

Infranuclear

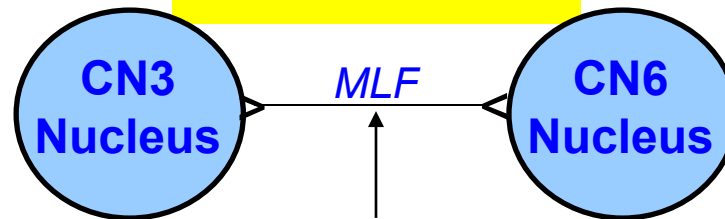


Motility Disorders: Overview

Supranuclear

Nuclear

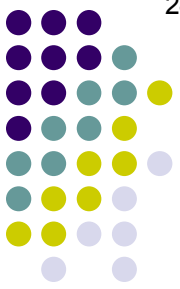
Internuclear



What is the name of the internuclear pathway connecting the CN3 and CN6 nuclei?

The **medial longitudinal fasciculus (MLF)**

Infranuclear

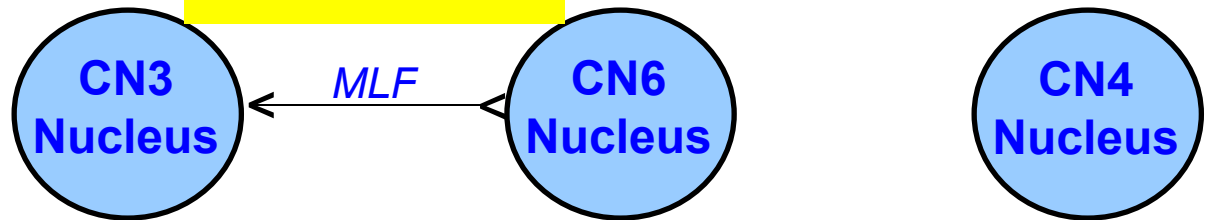


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Infranuclear

The ***infranuclear pathway*** consists of everything below the nuclei: the axons as they run from the nuclei to the neuromuscular junction; the junction itself; and finally the EOMs themselves. (There are many subsections in this pathway; we will identify them shortly.)



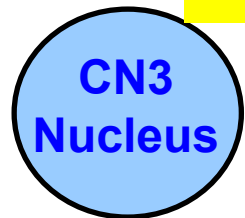
Extraocular muscle

This slide summarizes the basic organization of EOM control.

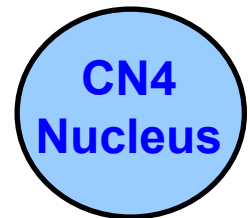
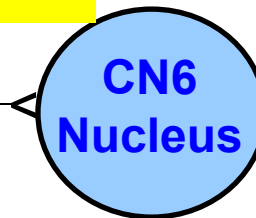
Supranuclear

Nuclear

Internuclear

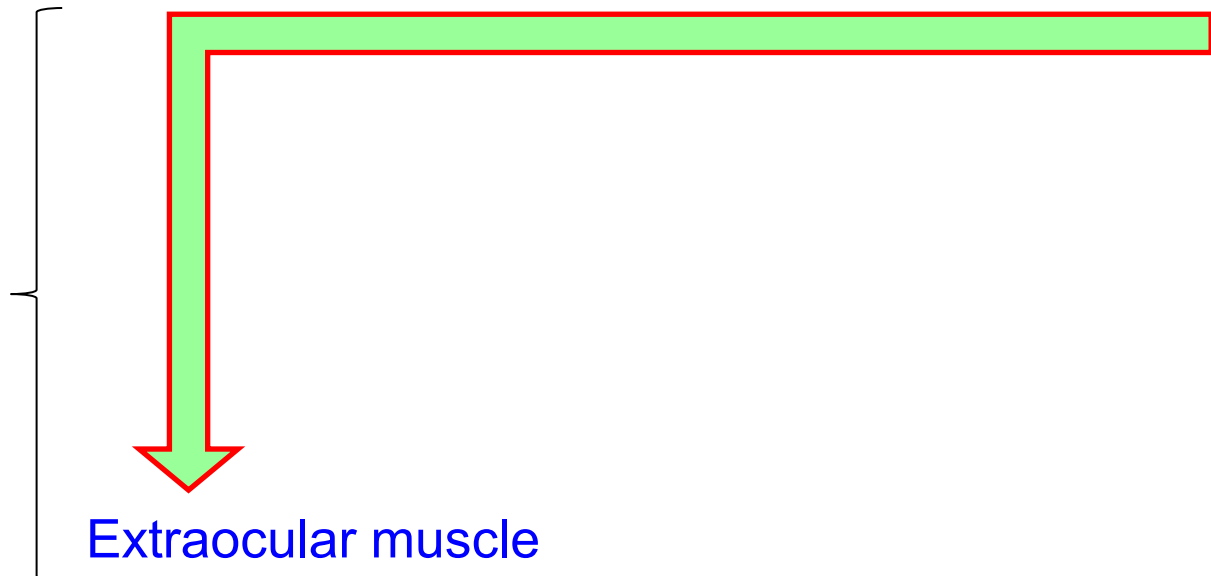


MLF



Infranuclear

Extraocular muscle

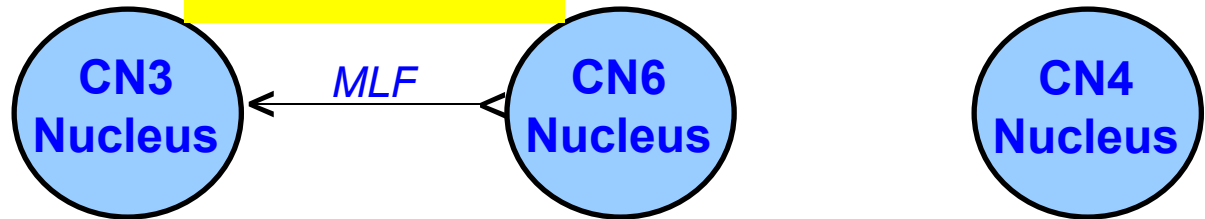


Supranuclear

This slide summarizes the basic organization of EOM control. When you encounter a pt with a motility issue, your first thought should be: *Is this issue **nuclear**, **supranuclear**, **internuclear**, or **infranuclear** in origin?*

Nuclear

Internuclear



Infranuclear

Extraocular muscle

Supranuclear

This slide summarizes the basic organization of EOM control. When you encounter a pt with a motility issue, your first thought should be: *Is this issue **nuclear**, **supranuclear**, **internuclear**, or **infranuclear** in origin?*

Internuclear

Nuclear

CN3

MLF

CN6

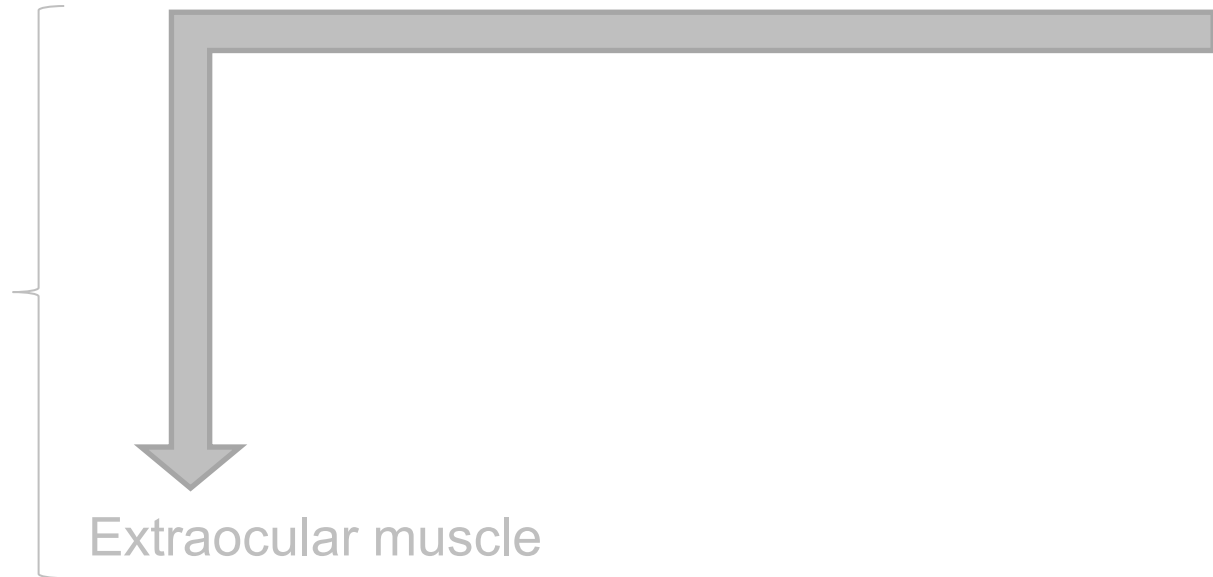
CN4

S

Next we will look at each level/pathway in more detail

Infranuclear

Extraocular muscle

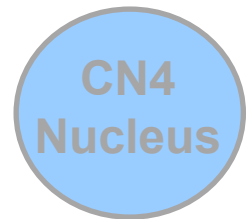
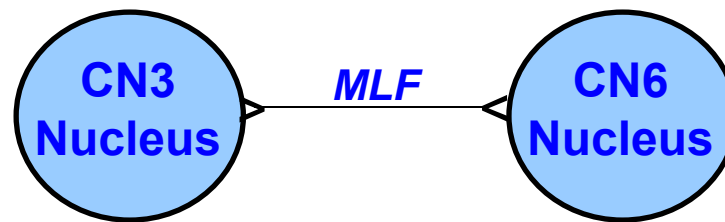




Motility Disorders: Overview

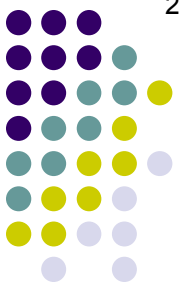
Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

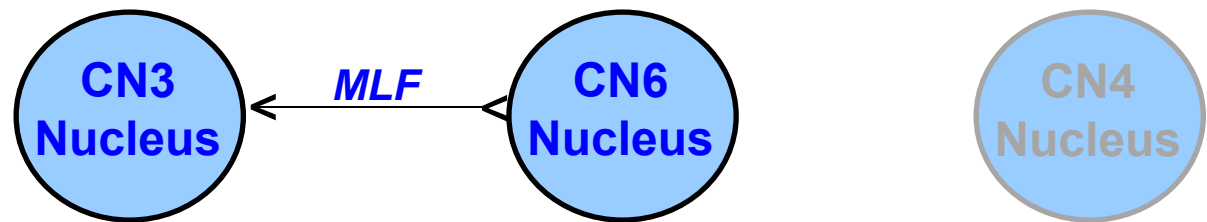
Infranuclear



Motility Disorders: Overview

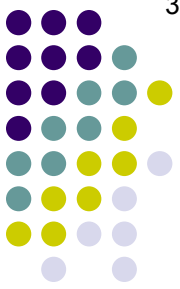
Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?
 From the CN6 nucleus to the ipsi- v contralateral CN3 nucleus

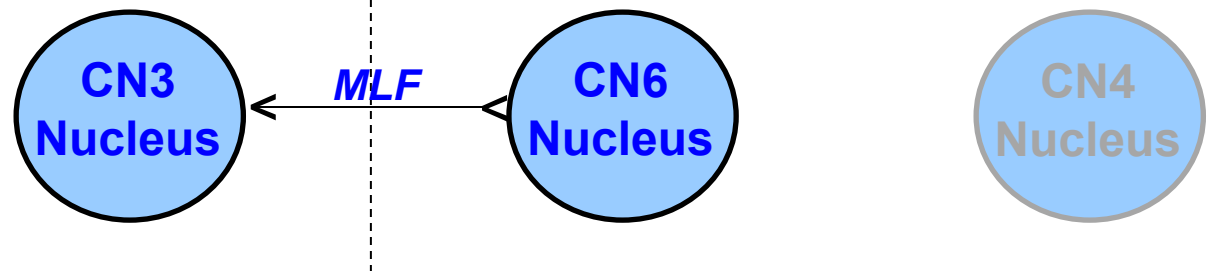
Infranuclear



Motility Disorders: Overview

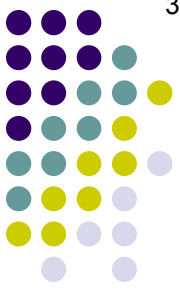
Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?
 From the CN6 nucleus to the contralateral CN3 nucleus

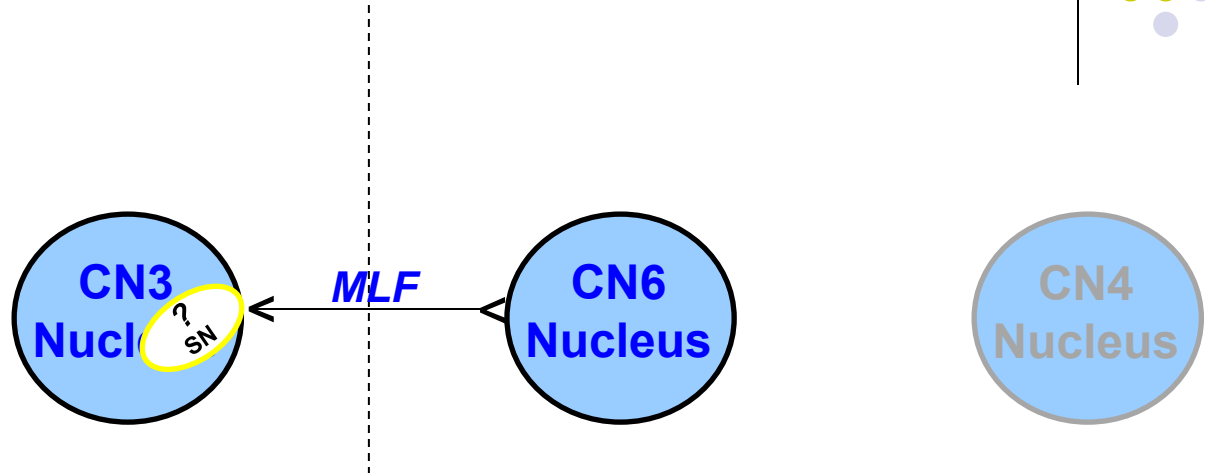
Infranuclear



Motility Disorders: Overview

Supranuclear

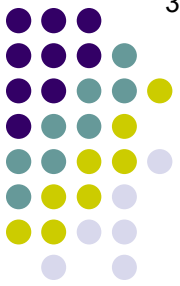
Nuclear



From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its EOM subnucleus

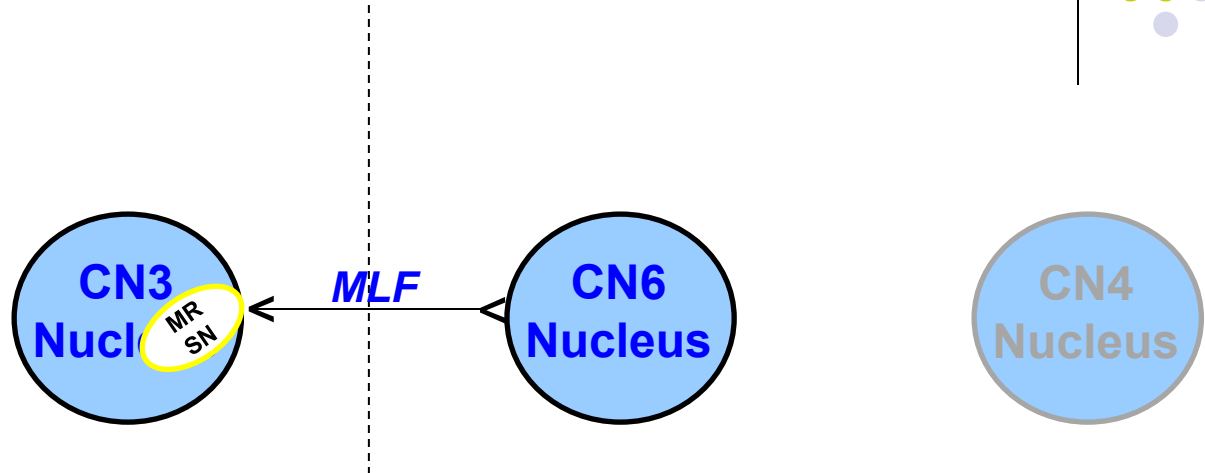
Infranuclear



Motility Disorders: Overview

Supranuclear

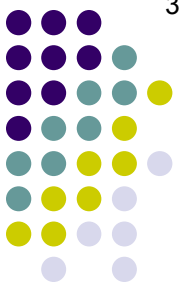
Nuclear



From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) subnucleus

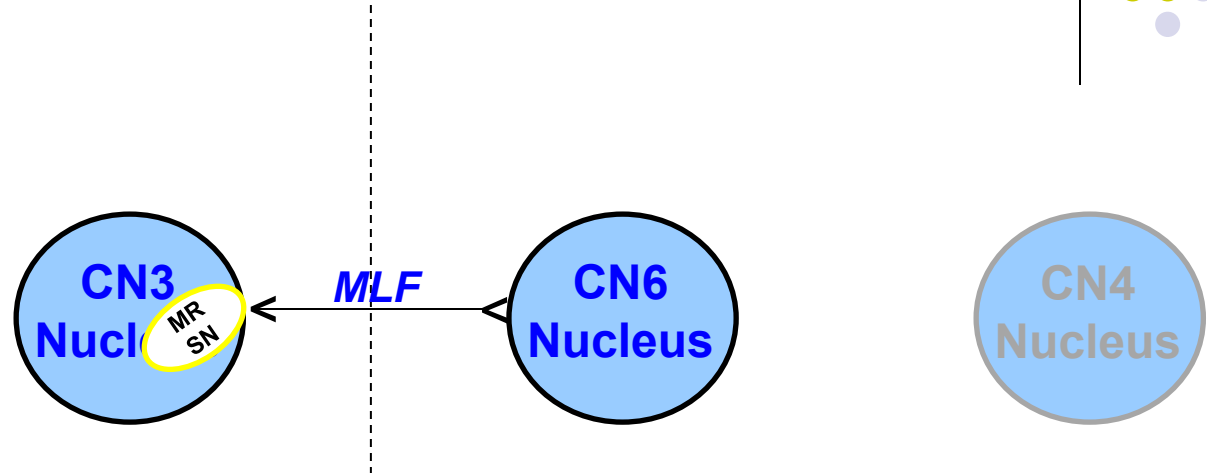
Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

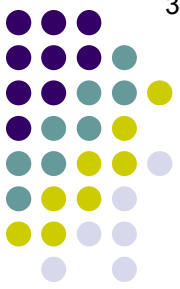


From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) subnucleus

Infranuclear

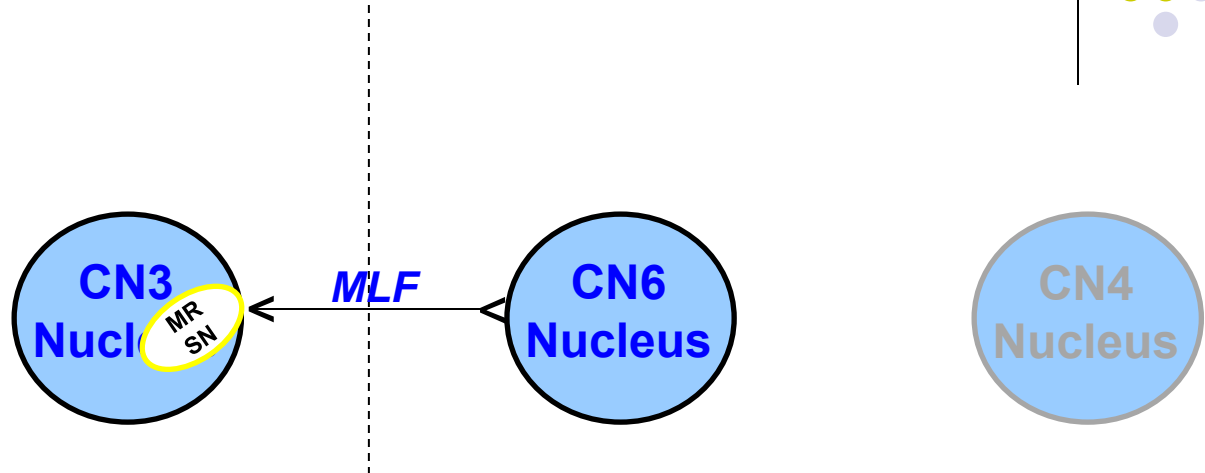
What purpose does the MLF serve?



Motility Disorders: Overview

Supranuclear

Nuclear



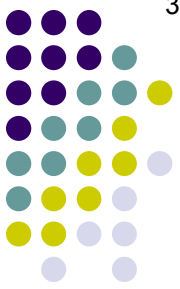
From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) subnucleus

Infranuclear

What purpose does the MLF serve?

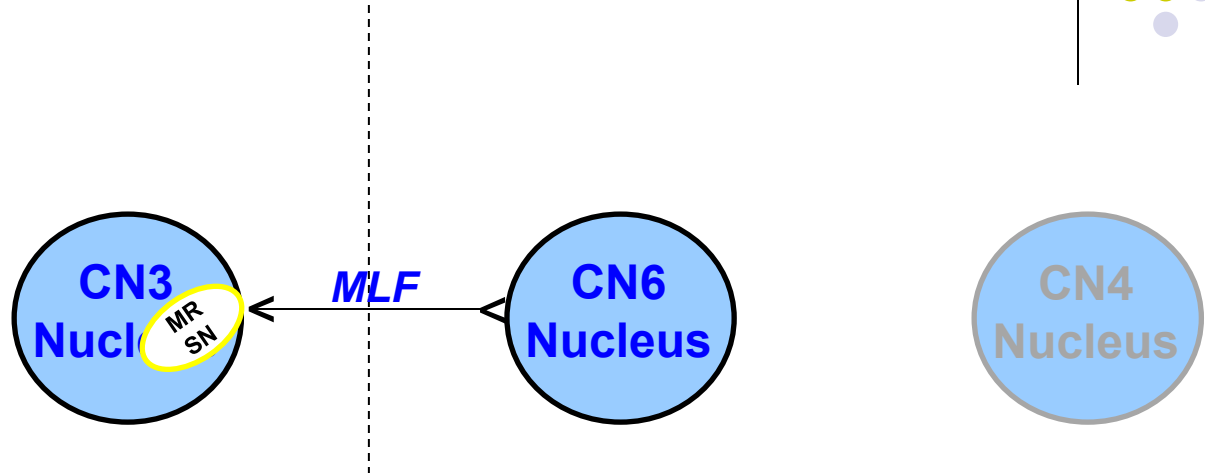
To allow coordinated lateral gaze of both eyes



Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

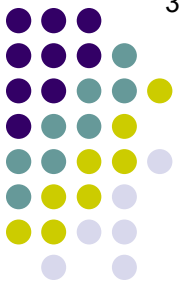
From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) subnucleus

Infranuclear

What purpose does the MLF serve?

To allow coordinated lateral gaze of both eyes

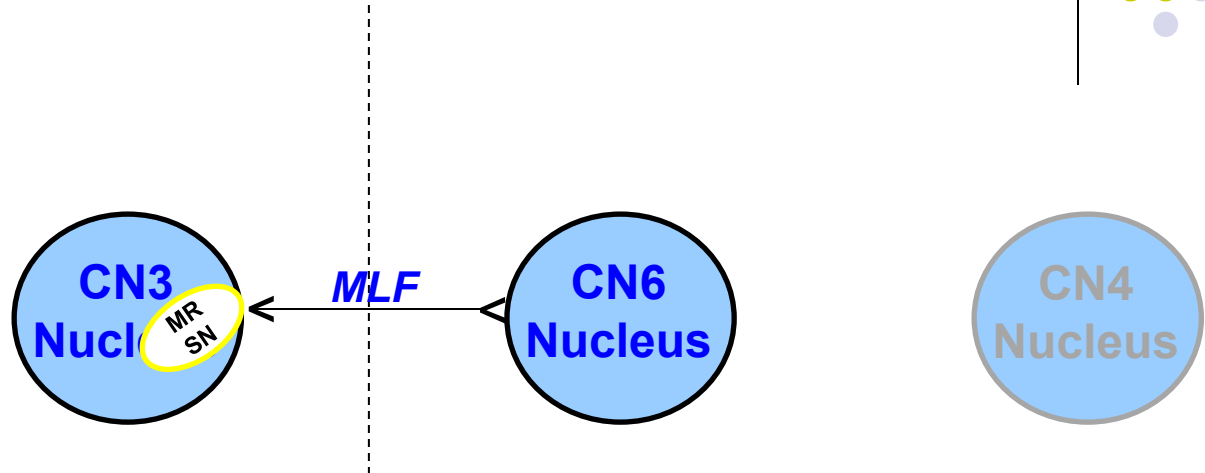
How does the MLF facilitate lateral gaze coordination?



Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) subnucleus

Infranuclear

What purpose does the MLF serve?

To allow coordinated lateral gaze of both eyes

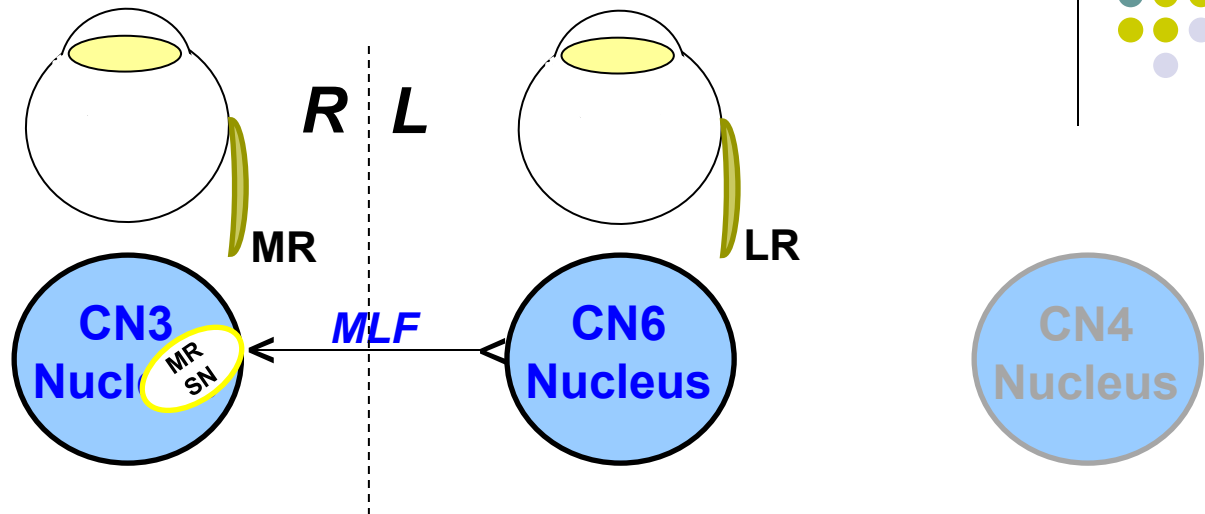
How does the MLF facilitate lateral gaze coordination?

By causing the contralateral MR to fire simultaneously with the ipsilateral lateral rectus (LR), thus ensuring both eyes turn into lateral gaze together

Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

So if the depicted CN6 nucleus is on a pt's left side, the depicted MLF runs to her right MR subnucleus.

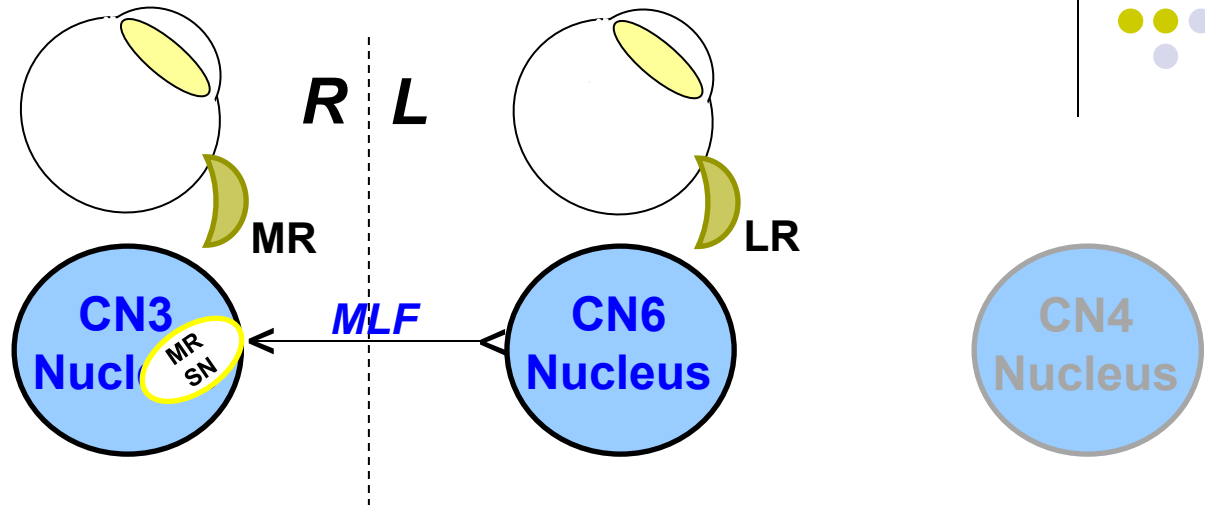
Infr

By causing the contralateral MR to fire simultaneously with the ipsilateral lateral rectus (LR), thus **ensuring both eyes turn into lateral gaze together**

Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

Infr

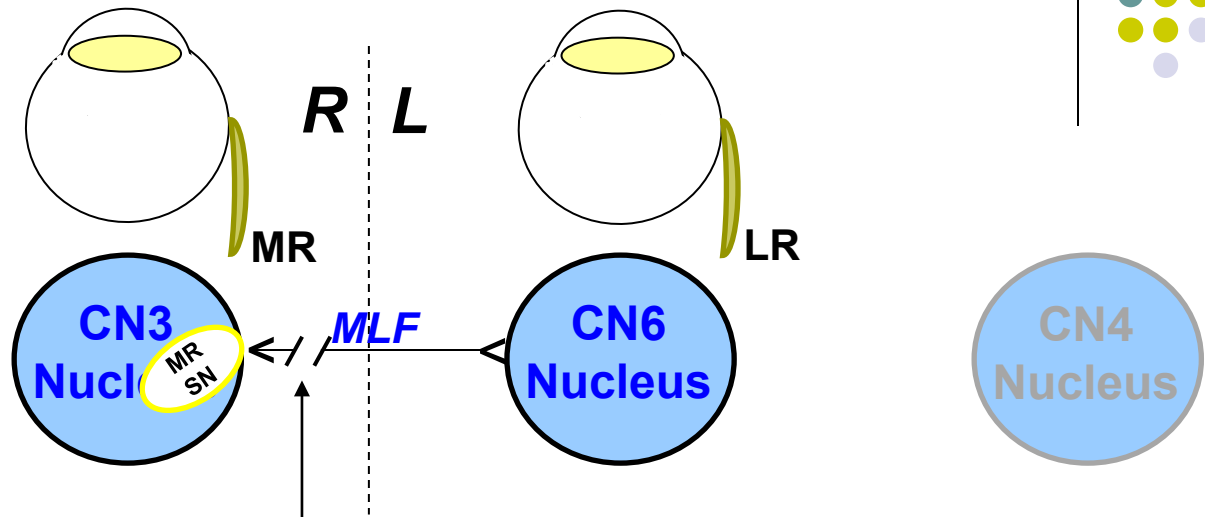
So if the depicted CN6 nucleus is on a pt's left side, the depicted MLF runs to her right MR subnucleus. When the pt endeavors to look to her left, the left CN6 nucleus causes the left LR to contract while also sending impulses (via the MLF) to her right MR subnucleus, which in turn causes the right MR to contract simultaneously—and both eyes shift into left gaze in coordinated fashion.

By causing the contralateral MR to fire simultaneously with the ipsilateral lateral rectus (LR), thus **ensuring both eyes turn into lateral gaze together**

Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

From the CN6 nucleus to the contralateral CN3 nucleus—specifically, to its medial rectus (MR) sub-nucleus.

What effect does a lesion of the MLF have on lateral gaze?

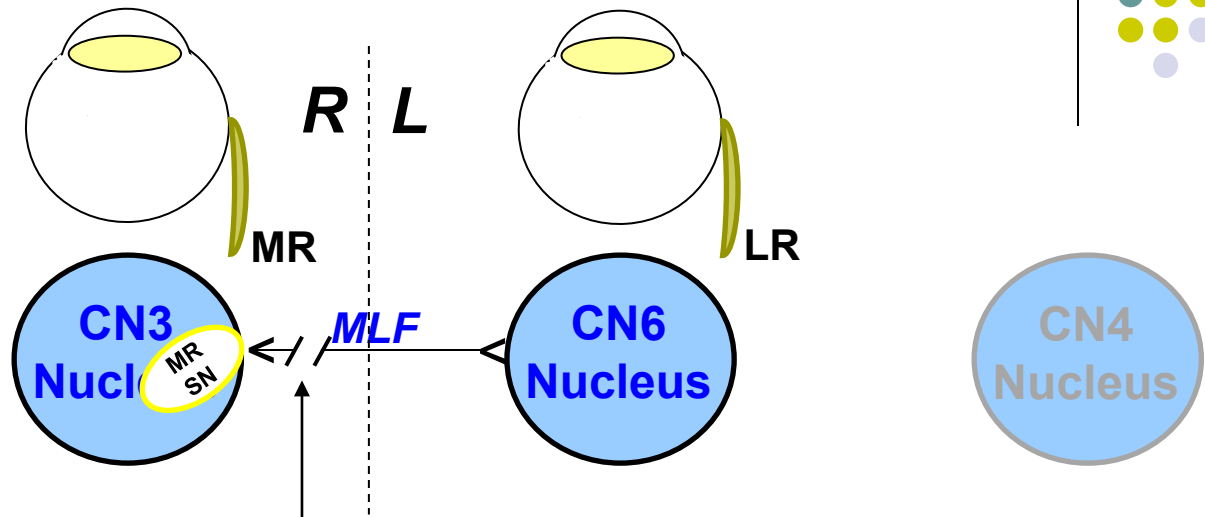
How does the MLF facilitate lateral gaze coordination?

By causing the contralateral MR to fire simultaneously with the ipsilateral lateral rectus (LR), thus ensuring both eyes turn into lateral gaze together

Motility Disorders: Overview

Supranuclear

Nuclear



From where to where do the fascicles of the MLF run?

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If the MLF is bagged, the impulse intended to result in the firing of the contralateral MR is affected...

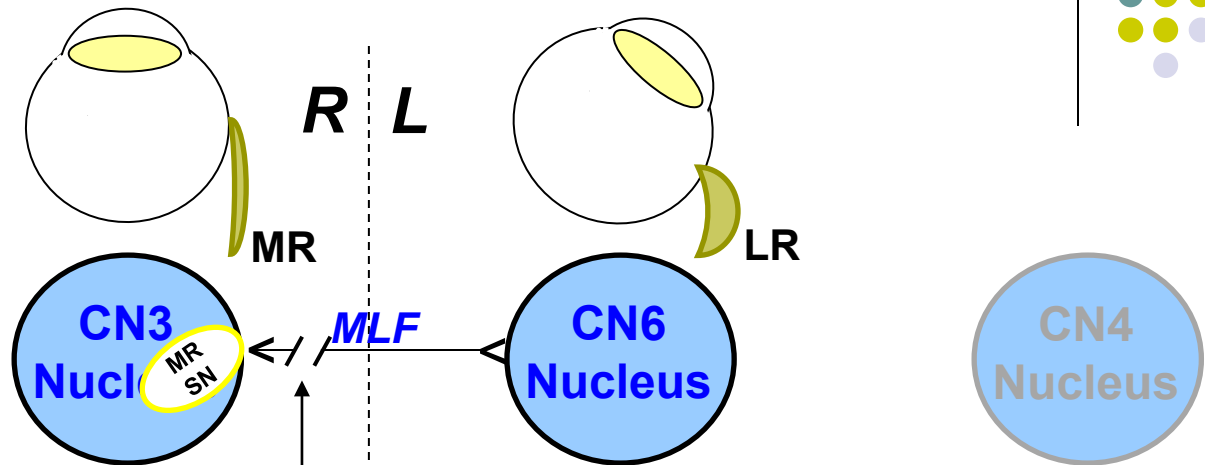
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Motility Disorders: Overview

Supranuclear

Nuclear



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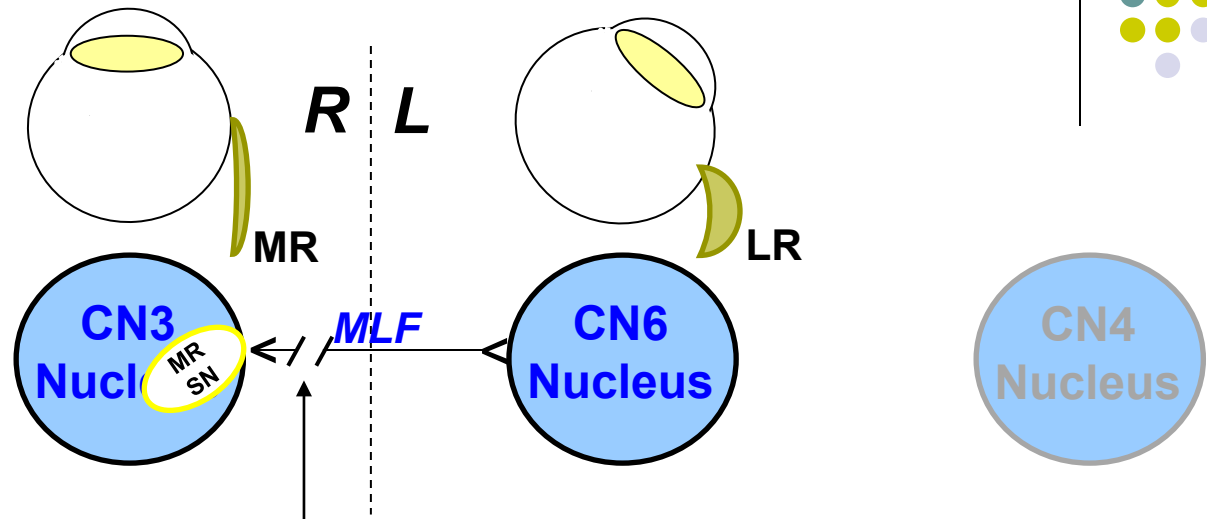
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Motility Disorders: Overview

Supranuclear

Nuclear



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Thus, attempted lateral gaze results in normal ABduction of the ipsilateral eye, but impaired ADduction of the contralateral eye.

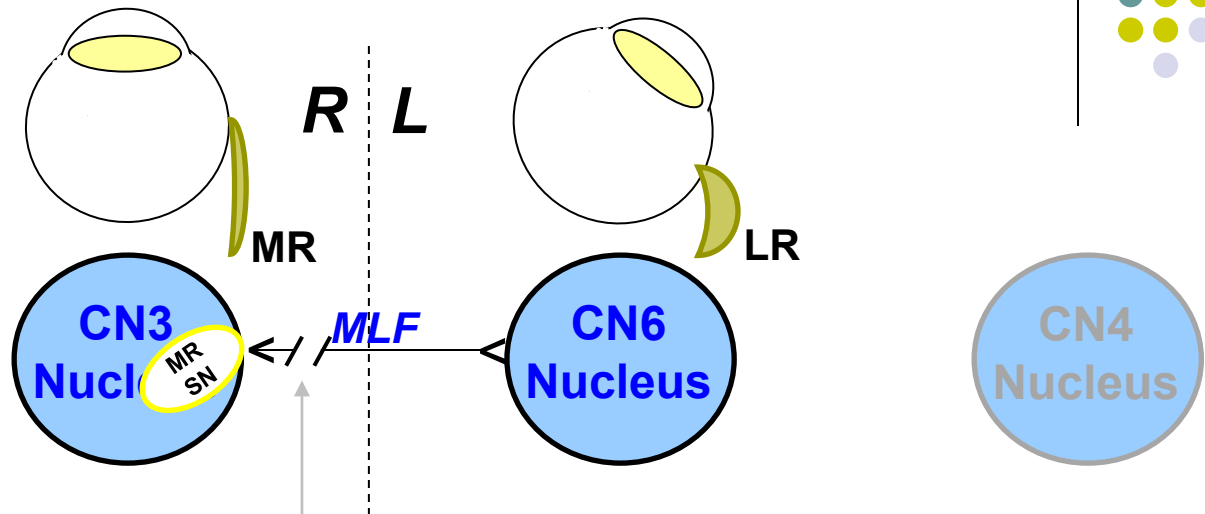
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Motility Disorders: Overview

Supranuclear

Nuclear



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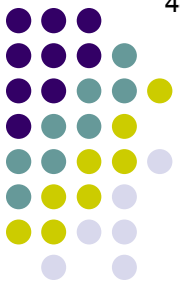
Thus, attempted lateral gaze results in normal ABduction of the ipsilateral eye, but impaired ADduction of the contralateral eye.

How does the MLF facilitate lateral gaze coordination?

By carrying the contralateral MR to fire simultaneously with the

This is an internuclear ophthalmoplegia (INO; see slide-set N20)

lateral gaze together



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Infranuclear

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

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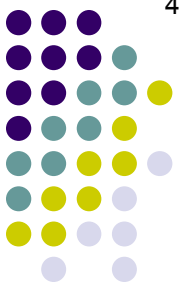
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← *The first portion of the nerve as it leaves the nucleus, but before leaving the substance of the brainstem*



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

The first portion of the nerve as it leaves the nucleus, but before leaving the substance of the brainstem

?

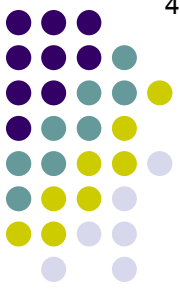
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Infranuclear



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

CN3
Nucleus

MLF

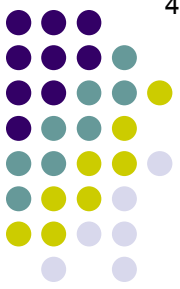
CN6
Nucleus

CN4
Nucleus

Fascicular

Infranuclear

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.



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

CN3
Nucleus

MLF

CN6
Nucleus

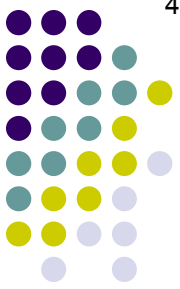
CN4
Nucleus

Fascicular

Infranuclear

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



Motility Disorders: Overview

Supranuclear

Internuclear

Nuclear

CN3
Nucleus

MLF

CN6
Nucleus

CN4
Nucleus

Fascicular

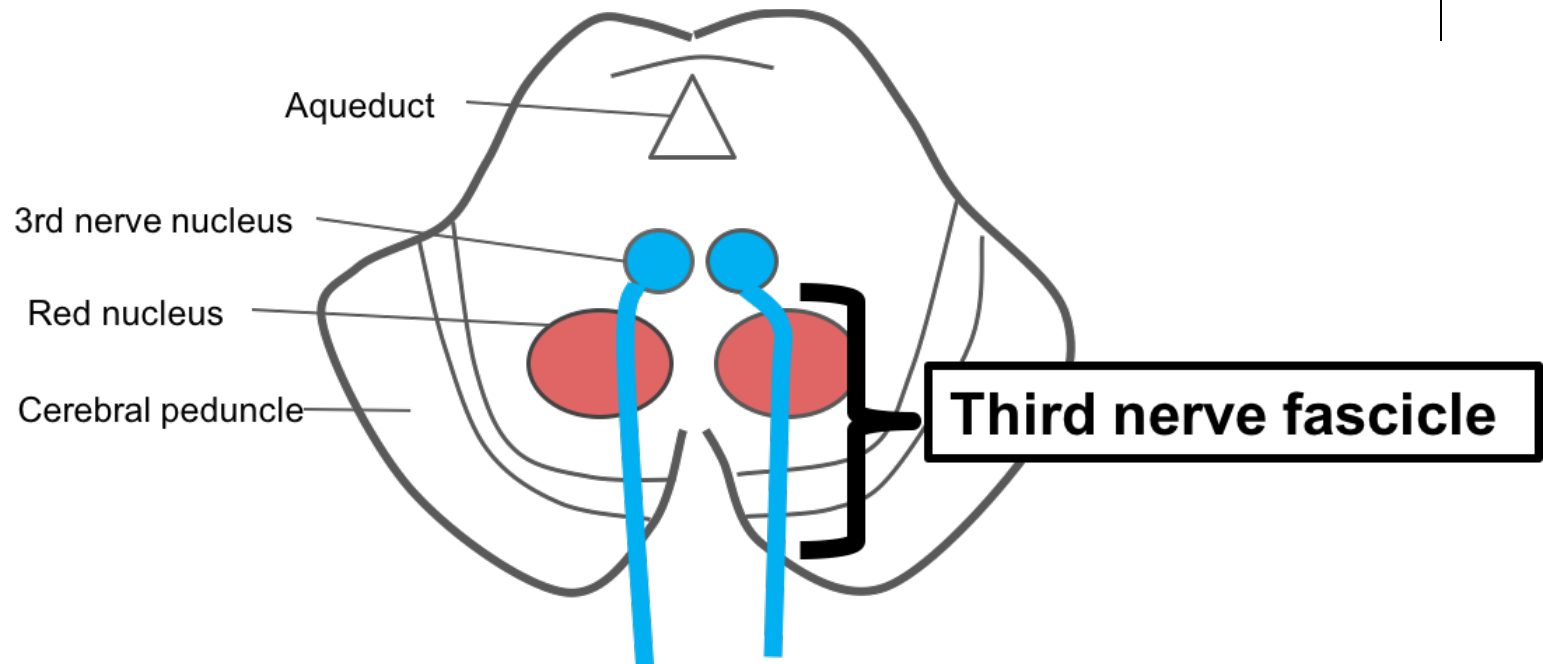
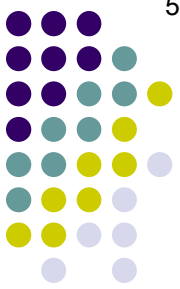
Infranuclear

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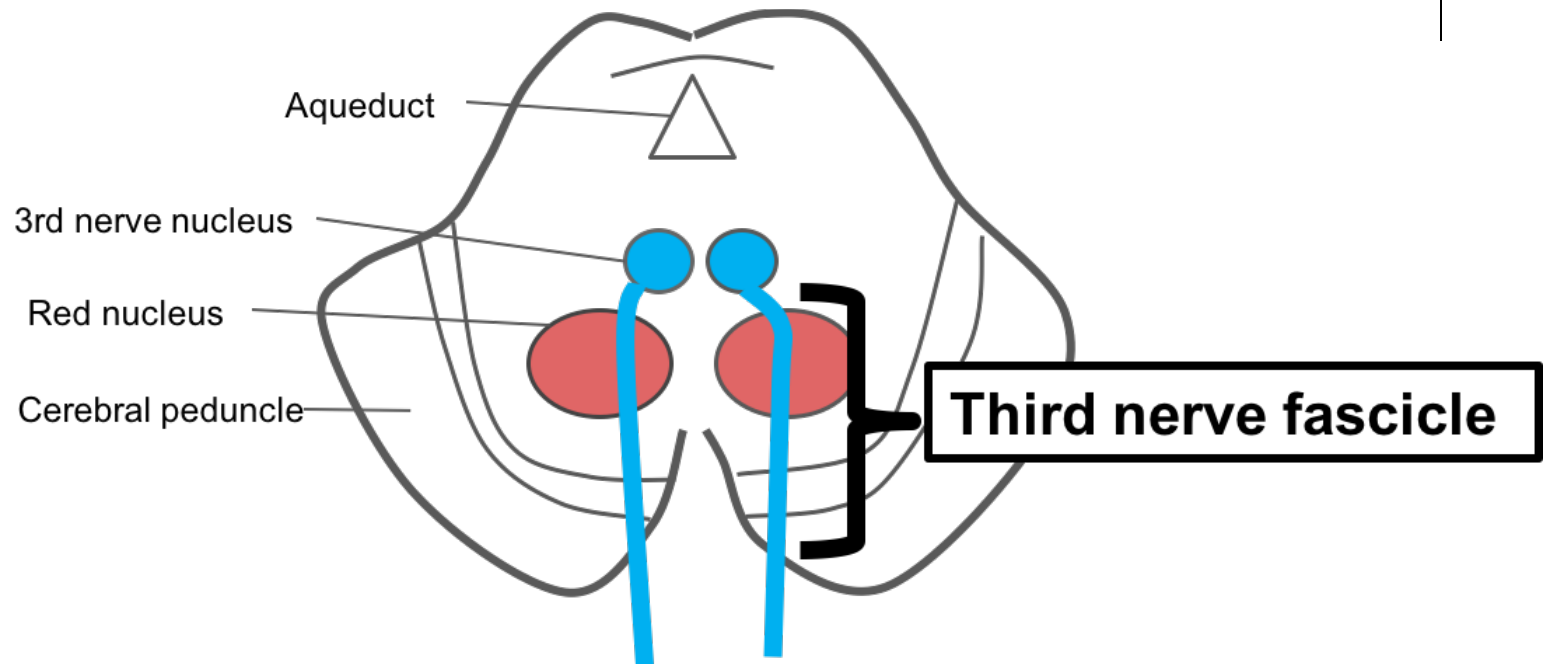
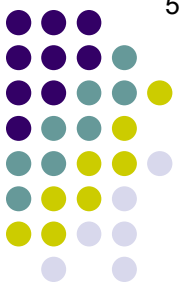
Fascicular syndrome

Motility Disorders: Overview



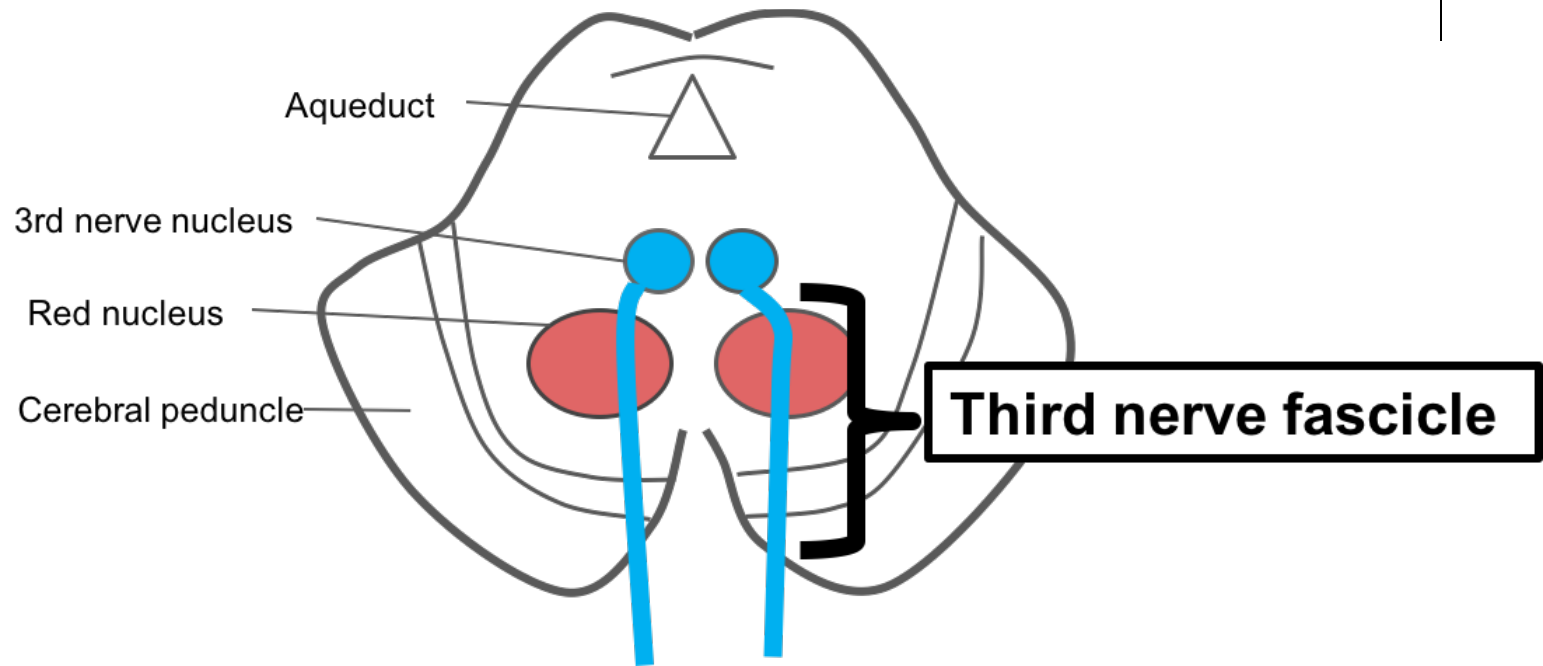
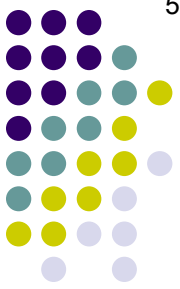
This is a rendering of a cranial nerve **fascicle** (in this case, CN3). Like the cranial-nerve proper, a fascicle is the bundle of axons that left their nucleus headed toward the target tissue; however, we don't start calling this bundle a 'nerve' until it breaks out of the substance of the brainstem and into the subarachnoid space.

Motility Disorders: Overview

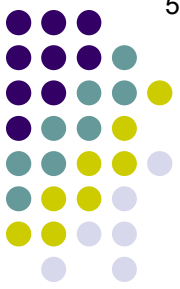


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Motility Disorders: Overview



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Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3
Nucleus

MLF

CN6
Nucleus

CN4
Nucleus

Fascicular

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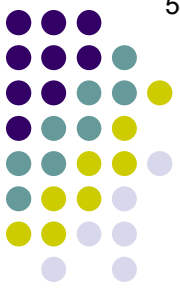
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Speaking of fascicular syndromes: The Neuro book describes four involving the CN3 fascicle

CN3 fascicular syndromes:

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Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

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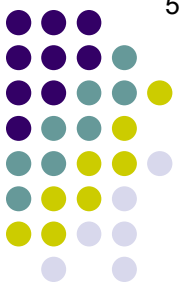
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Infranuclear

Speaking of fascicular syndromes: The Neuro book describes four involving the CN3 fascicle

CN3 fascicular syndromes:

- Weber syndrome
- Benedikt syndrome
- Claude syndrome
- Nothnagel syndrome



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

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Infranuclear

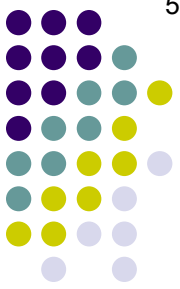
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CN3 fascicular syndromes:

- Weber syndrome
- Benedikt syndrome
- Claude syndrome
- Nothnagel syndrome

CN6 fascicular syndromes:

- ?
- ?
- ?



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

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Infranuclear

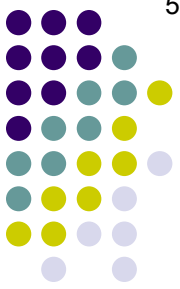
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- Benedikt syndrome
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CN6 fascicular syndromes:

- Foville syndrome
- Millard-Gubler syndrome
- Raymond syndrome



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear

CN3
Nucleus

MLF

CN6
Nucleus

CN4
Nucleus

Fascicular

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CN3 fascicular syndromes:

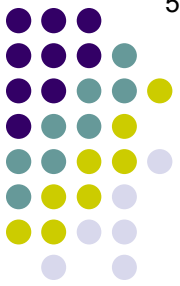
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- Benedikt syndrome
- Claude syndrome
- Nothnagel syndrome

The fascicular syndromes are addressed in detail in their own slide-set (N14)

CN6 fascicular syndromes:

- Foville syndrome
- Millard-Gubler syndrome
- Raymond syndrome

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

?

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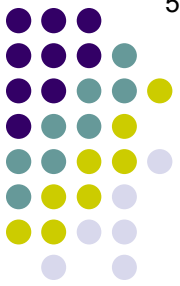
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*The next portion commences once the fascicles exit the brainstem--now they're a **nerve**. Named for the space in which the nerves travel.*

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



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.*

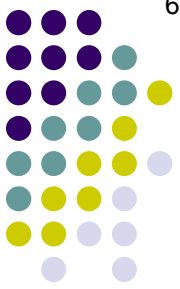
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Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

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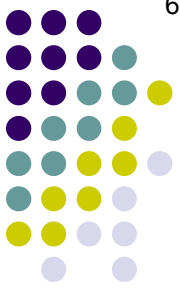
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Infranuclear

Which cause of ophthalmoparesis--common among vasculopathies--is attributed to damage occurring to the subarachnoid segments?



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

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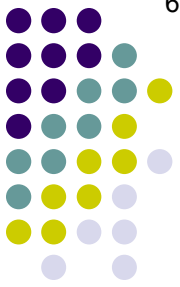
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Infranuclear

Which cause of ophthalmoparesis--common among vasculopathies--is attributed to damage occurring to the subarachnoid segments?

Ischemic palsies (ie, a so-called 'diabetic third' or 'diabetic sixth')



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

?

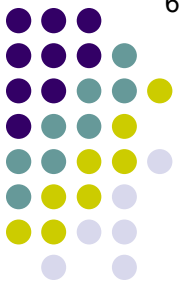
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Infranuclear

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.



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

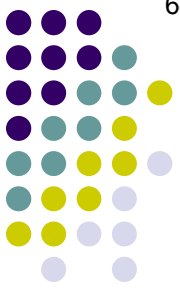
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Infranuclear

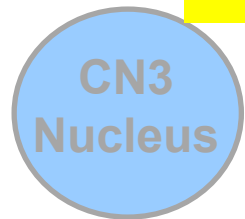


Motility Disorders: Overview

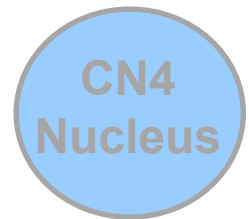
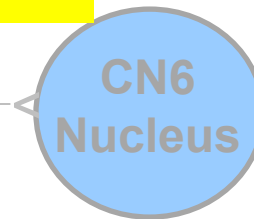
Supranuclear

Nuclear

Internuclear



MLF



Fascicular

Subarachnoid

Cavernous sinus

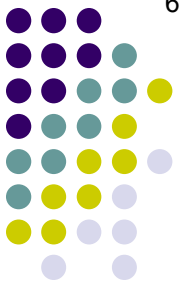
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What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

Infranuclear

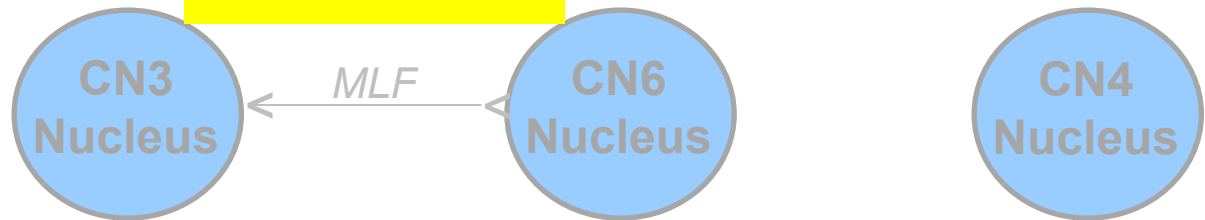


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

?

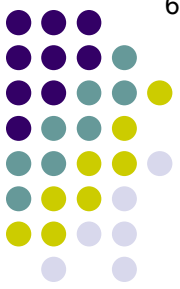
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What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously

Infranuclear

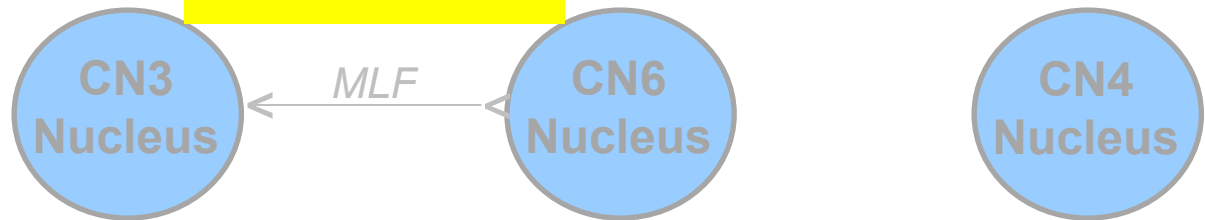


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

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What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously

Which nerves may be involved?

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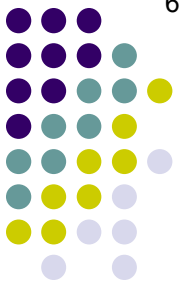
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Infranuclear

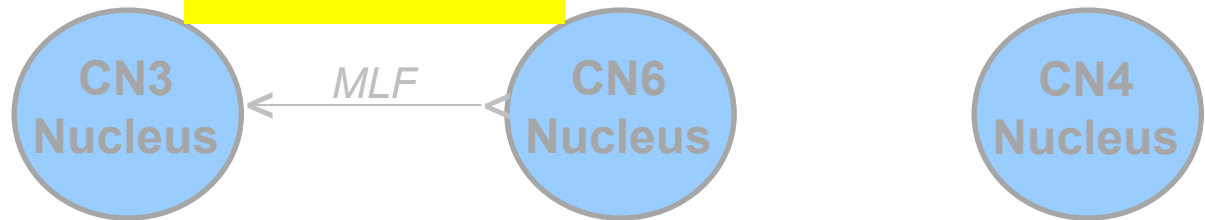


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

?

?

?

Infranuclear

What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously

Which nerves may be involved?

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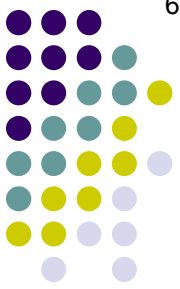
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Involvement manifests as

Involvement manifests as

← Involvement manifests as

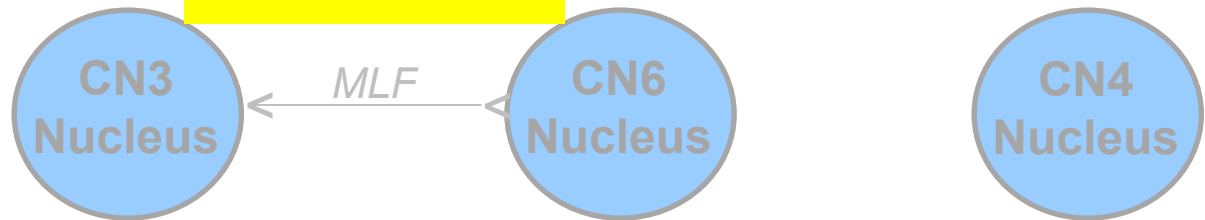


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

?

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What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously

Which nerves may be involved?

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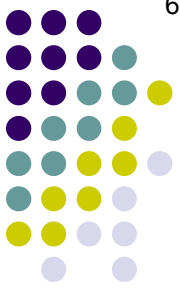
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Involvement manifests as ophthalmoplegia

Involvement manifests as facial hypoesthesia

← Involvement manifests as Horner's

Infranuclear

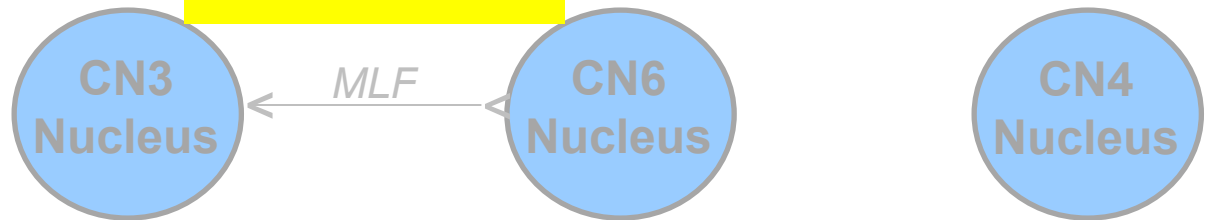


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

?

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?

What is the hallmark of ophthalmoplegia 2ndry to a cavernous sinus process?

The involvement of two or more cranial nerves simultaneously

Which nerves may be involved?

--CN3

--CN4

--CN6

--V₁

--V₂

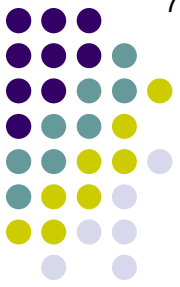
--Sympathetics

Involvement manifests as ophthalmoplegia

Involvement manifests as facial hypoesthesia

Involvement manifests as Horner's

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

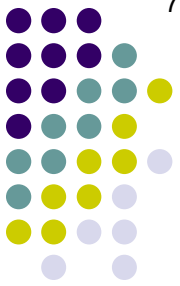
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Getting pretty close now. Post-cavernous sinus, another well-defined space.

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

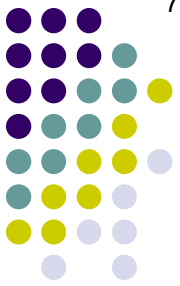
Orbital

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

?

?

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

Superior orbital fissure ←

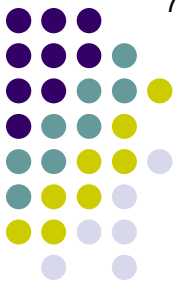
Orbital

?

?

Infranuclear

The answer *superior orbital fissure* is just as good here (if not better, as the *Neuro* book breaks out the fissure as a separate structure in the pathway)

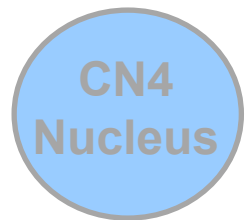


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

Superior orbital fissure
Orbital apex

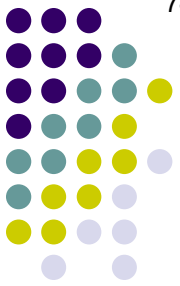
Orbital

?

?

Infranuclear

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

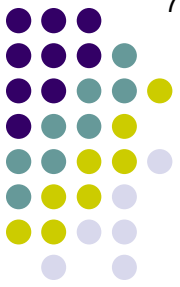
Orbital

?

?

Infranuclear

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

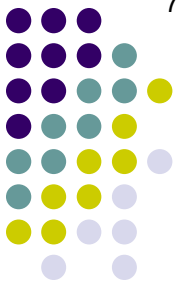
Orbital

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Infranuclear

← *Where the journey ends
for the nerves.*

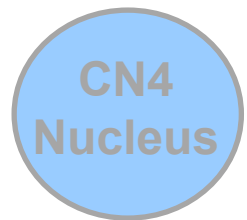


Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

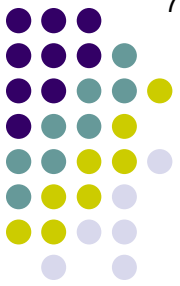
Orbital

Neuromuscular junction

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Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

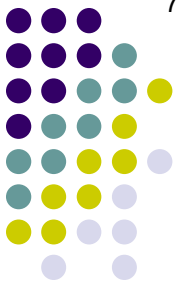
Orbital

Neuromuscular junction

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Per the Neuro book, what is the “prototypical” disease of the neuromuscular junction?

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

Subarachnoid

Cavernous sinus

Orbital

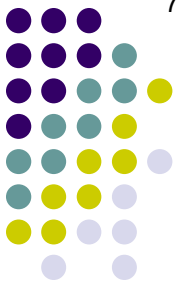
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Myasthenia gravis

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

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Fascicular

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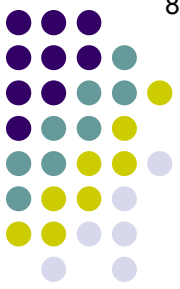
Orbital

Neuromuscular junction

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*And finally...Don't forget
pathology here when
evaluating motility disorders!*

Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

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Fascicular

Subarachnoid

Cavernous sinus

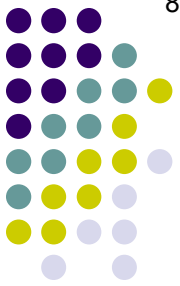
Orbital

Neuromuscular junction

Extraocular muscle

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Infranuclear



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



Fascicular

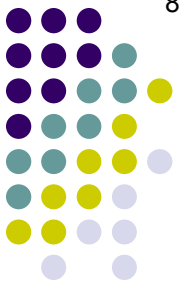
Subarachnoid

Cavernous sinus

Infranuclear

What sorts of conditions are included here?

Extraocular muscle



Motility Disorders: Overview

Supranuclear

Nuclear

Internuclear



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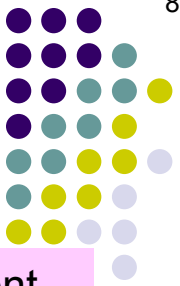
Cavernous sinus

Infranuclear

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

Before discussing **supranuclear lesions**, we need to define the role of the efferent (ie, motor) component of the visual system. But before we do *that*, we have to define the role of the *afferent* system.

Infranuclear

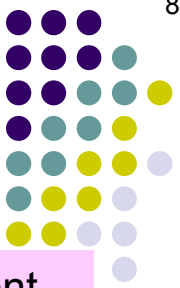
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Cavernous sinus

Orbital

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Extraocular muscle



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Infranuclear

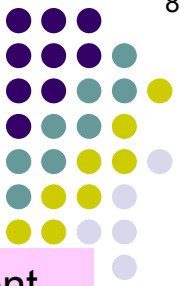
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Infranuclear

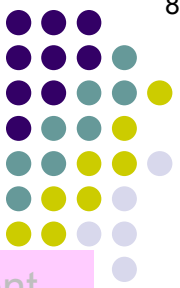
Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

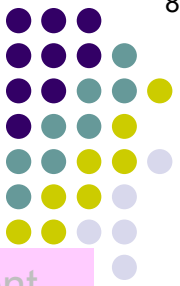
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Extracocular Muscles



Motility Disorders: Overview

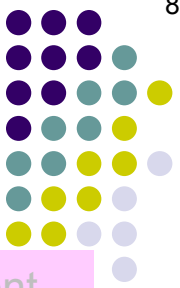
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Extracocular Muscles



Motility Disorders: Overview

Supranuclear

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Extracocular Muscles



Motility Disorders: Overview

Supranuclear

The *supranuclear pathways* consist of six systems in the primate CNS that deal with these fixation-related issues. Thus, lesions of a supranuclear pathway manifest as difficulties with either the **maintenance** or **acquisition** of bifixation.

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Extracocular Muscles



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal

with these fixation-related issues

Internuclear

1) The two words **system** is responsible for maintaining a high-quality image of a stationary object when the head is still.

Infranuclear

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when the head is still.

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Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal

with these fixation-related issues

Internuclear

1) The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when the head is still. It does this via continuous *microsaccadic refixation movements*, which produce a constant shifting among the PRs regarding which are responsible for the retinal image. This shifting prevents PR fatigue (and subsequent image loss) from occurring.

Infranuclear

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **two words system** is responsible for maintaining fixation on a moving object. When it is impaired pursuit movements may either lag behind the object or jump ahead of it.

Infranuclear

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system** is responsible for maintaining fixation on a moving object. When it is impaired pursuit movements may either lag behind the object or jump ahead of it.

Infranuclear

Fascicular

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Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system** is responsible for maintaining fixation on a moving object. When it is impaired pursuit movements may either lag behind the object or jump ahead of it. **Of note, that this is the only supranuclear pathway that is activated voluntarily.**

Infranuclear

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The ***ocular fixation system***

2) The ***smooth-pursuit system***

3) The ***vestibulo-ocular system*** is responsible for maintaining fixation on an object that is moving toward or away from the eyes, thus necessitating they converge or diverge.

Infranuclear

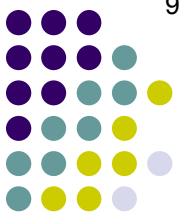
Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system**

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Infranuclear

Subarachnoid

Cavernous sinus

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Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system**

3) The **vergence system** is responsible for maintaining fixation on an object that is moving toward or away from the eyes, thus necessitating they converge or diverge. Many forms of vergence dysfunction can occur, including *convergence insufficiency*, *divergence insufficiency*, *accommodative esotropia*, and *spasm of the near*.

Infranuclear

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system**

3) The **vergence system**

4) The three words **system** and the 5) two words **system** are responsible for holding an image steady during head rotations—either brief and rapid (VOR) or slower and sustained (OKN).

Infranuclear

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

1) The **ocular fixation system**

2) The **smooth-pursuit system**

3) The **vergence system**

4) The **vestibulo-ocular reflex (VOR) system** and the 5) **optokinetic nystagmus (OKN) system** are responsible for holding an image steady during head rotations—either brief and rapid (VOR) or slower and sustained (OKN).

Infranuclear

Cavernous sinus

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Neuromuscular junction

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Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

Internuclear

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4) The **vestibulo-ocular reflex (VOR) system** and the 5) **optokinetic nystagmus (OKN) system** are responsible for holding an image steady during head rotations—either brief and rapid (VOR) or slower and sustained (OKN). The VOR is controlled by the vestibular labyrinth, ie, the semicircular canals and otoliths. In contrast, the OKN system is driven by images sweeping across the retina.

Infranuclear

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

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- 2) The **smooth-pursuit system**
- 3) The **vergence system**
- 4) The **vestibulo-ocular reflex (VOR) system** and the 5) **optokinetic nystagmus (OKN) system**
- 6) The **transsaccadic system** is responsible for rapidly shifting fixation from the current object of interest to a new one located in the visual periphery.

Infranuclear

Cavernous sinus
Orbital
Neuromuscular junction
Extraocular muscle



Motility Disorders: Overview

Supranuclear

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- 6) The **saccadic system** is responsible for rapidly shifting fixation from the current object of interest to a new one located in the visual periphery.

Infranuclear

Cavernous sinus
Orbital
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Extraocular muscle



Motility Disorders: Overview

Supranuclear

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- 6) The ***saccadic system***

An important rule-of-thumb can be stated regarding supranuclear motility disorders and diplopia—what is it?

Orbital

Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

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

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Neuromuscular junction

Extraocular muscle



Motility Disorders: Overview

Supranuclear

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Why don't most pts with supranuclear disorders have diplopia?



Motility Disorders: Overview

Supranuclear

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Orbital

Neuromuscular

Extraocular muscle

Why don't most pts with supranuclear disorders have diplopia?
Because most supranuclear disorders affect **both** eyes in a **symmetric** fashion



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

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1) The **ocular fixation system**

2) The **smooth-pursuit system**

3) The **vergence system**

4) The **vestibulo-ocular reflex (VOR) system** and the

6) The **saccadic system**

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

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*An important rule-of-thumb can be stated: **supranuclear disorders do not cause diplopia**—what is it?*

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Orbital

Neuromuscular

Extraocular muscle

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Motility Disorders: Overview

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4) The **vestibulo-ocular reflex (VOR) system** and the

6) The **saccadic system**

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

An important rule-of-thumb can be stated about diplopia—what is it?

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Orbital

Neuromuscular

Extraocular muscle

Why don't most pts with supranuclear disorders have diplopia?
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Motility Disorders: Overview

Supranuclear

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- Gaze palsies, eg, **Parinaud syndrome**
- Congenital ocular motor apraxia (COMA)**
- Progressive supranuclear palsy (PSP)**
- Saccadic disorders**

An important rule-of-thumb can be stated: In the absence of diplopia—what is it?

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

Orbital

Neuromuscular

*Why don't most pts with supranuclear disorders have diplopia? Because most supranuclear disorders affect **both** eyes in a **symmetric** fashion*

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



Motility Disorders: Overview

Supranuclear

six systems in the primate CNS that deal with these fixation-related issues

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