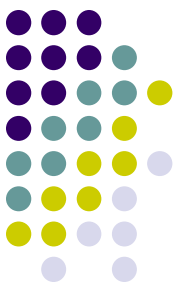


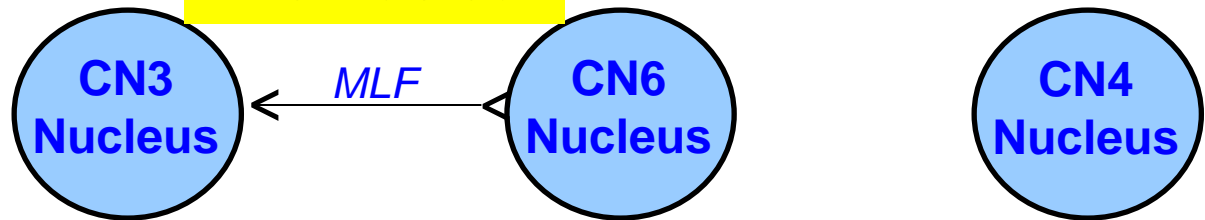
# Motility Disorders: *Supranuclear Syndromes*



*Supranuclear*

*Nuclear*

*Internuclear*



*Infranuclear*

Fascicular

Subarachnoid

Cavernous sinus

Orbital

Neuromuscular junction

Extraocular muscle

This slide captures one way to think about the motility disorders. If it is unfamiliar, I strongly suggest you review the slide-set entitled '***Motility disorders: Overview***' (N18) before proceeding.

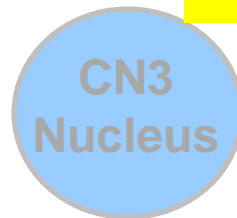


## *Supranuclear*

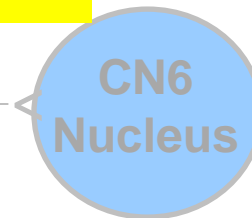
*In this slide-set, we'll take a look at supranuclear syndromes*

### *Internuclear*

### *Nuclear*



← MLF →



### *Infranuclear*

Fascicular

Subarachnoid

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## Motility Disorders: *Supranuclear Syndromes*



### ***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. (Get comfy, this is gonna take a minute.)

**Infratentorial**

Orbital

Neuromuscular junction

Extraocular muscle

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In primates, vision has two purposes: 1) to **detect** objects of interest (eg, things you may want to eat, or may want to eat you), and 2) to **scrutinize** objects of interest (ie, to determine definitively whether it's an eat-er vs an eat-ee).

Orbital

Neuromuscular junction

Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*



### ***Supranuclear***

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Orbital  
Neuromuscular junction  
Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*



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--A central, high-resolution channel for scrutinizing objects. Anatomically, this channel consists of the foveas, which provide high-resolution vision allowing objects to be examined in sharp detail and stereoscopic depth.

--

**Infraocular**

Orbital

Neuromuscular junction

Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*



### ***Supranuclear***

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- A peripheral, low-resolution channel for detecting objects. The peripheral channel is very sensitive to motion. Anatomically, this channel consists of the rest of the retina, ie, the parafoveal macula on out.

Orbital

Neuromuscular junction

Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*



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Reiterating for emphasis: The afferent system has two jobs: Scrutinize an object of regard (central channel), while also monitoring for other objects that may require scrutinization (peripheral channel).

Orbital

Neuromuscular junction

Extraocular muscle



## Motility Disorders: *Supranuclear Syndromes*



### *Supranuclear*

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Reiterating for emphasis: The afferent system has two jobs: Scrutinize an object of regard (central channel), while also monitoring for other objects that may require scrutinization (peripheral channel). It follows from this that the *efferent* visual system has two jobs: 1) Keep both foveas pointing at the current object of regard; and 2) rapidly redirect both foveas to a new object when one is detected in the periphery.

Orbital

Neuromuscular junction

Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*

### ***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. (Get comfy, this is gonna take a minute.)

**But let's consider what it takes to accomplish these tasks.**

In the visual system, we have two main channels: the central channel and the peripheral channel. The central channel is responsible for high-resolution vision, while the peripheral channel is responsible for low-resolution vision. The peripheral channel is very sensitive to motion. Anatomically, this channel consists of the rest of the retina, ie, the parafoveal macula on out.

Reiterating for emphasis: The afferent system has two jobs: Scrutinize an object of regard (central channel), while also monitoring for other objects that may require scrutinization (peripheral channel). It follows from this that the *efferent* visual system has two jobs: 1) Keep both foveas pointing at the current object of regard; and 2) rapidly redirect both foveas to a new object when one is detected in the periphery.

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Neuromuscular junction

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# Motility Disorders: *Supranuclear Syndromes*

## Supranuclear

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Keep both foveas pointing at the current object of regard;

Orbital

Neuromuscular junction

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Keep both foveas pointing at the current object of regard;

Orbital

Neuromuscular junction

Extraocular muscle

## Motility Disorders: *Supranuclear Syndromes*

### *Supranuclear*



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rapidly redirect both foveas to a new object when one is detected in the periphery.

Orbital

In order to rapidly refixate both foveas on a peripheral image, the efferent system must first produce just enough torque to overcome inertia and rotate the eyes to this image, then 'ramp down' the amount of torque to the level needed to maintain gaze in this new direction.

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At long last, the payoff for all this backstory: The *supranuclear pathways* consist of six systems in the primate CNS that deal with these fixation-related issues.

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*Next, we will drill down on the six supranuclear pathways/control systems. They will be presented in an order that makes sense (to me), but that should not be (mis)interpreted as reflecting some sort of intrinsic order of importance.*

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# Motility Disorders: *Supranuclear Syndromes*

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Thus, lesions of a supranuclear pathway manifest as difficulties with either the **maintenance** or **acquisition** of bifi

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

## Motility Disorders: *Supranuclear Syndromes*

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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 three words, 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.

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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.

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Dysregulation of this system manifests as either visually disruptive larger refixation movements (ie, saccadic, not **microsaccadic**) or visually disruptive slower refixation movements (nystagmus, not saccades).

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The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when the head is still. It does this by shifting among the PRs. 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.

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Thus, lesions of a supranuclear pathway manifest as difficulties with either the **maintenance** or **acquisition** of bifixation.

The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when the head is still. It does this by shifting among the PRs. 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.

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# Motility Disorders: *Supranuclear Syndromes*

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# Motility Disorders: *Supranuclear Syndromes*

## Supranuclear



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Thus, lesions of a supranuclear pathway manifest as difficulties with either the **maintenance** or **acquisition** of bifixation.

The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when

The **two words** is responsible for maintaining fixation on an object that is moving toward or away from the eyes, thus necessitating they converge or diverge.

# Motility Disorders: *Supranuclear Syndromes*

## Supranuclear

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But let's consider what it takes to accomplish these tasks. Scrutinizing an object requires steady bifixation—but not *too* steady, or the photoreceptors (PRs) will fatigue and the image will disappear. Further, the object might be moving closer to/farther from the system has to precisely track it. Further still, the primate's *head* might be moving the eyes, necessitating object-tracking.

--A peripheral, low-resolution channel for detecting objects. The peripheral channel is very sensitive to motion.

At long last, the payoff for all this back story: The *supranuclear pathways* consist

**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.

The **ocular fixation system** is responsible for maintaining a high-quality image of a stationary object when

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.



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two words

two different words

two still different words

, and

four words

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The **VOR** is controlled by the vestibular labyrinth, ie, the two words and . Rapid rotation of the head in a given plane leads to an excitatory burst from the canal of the same plane on the side of the head toward which the rotation occurred, while simultaneously leading to an inhibitory burst from the canal of the same plane on the opposite side.

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That said, the majority of VOR dysfunction cases stem from peripheral issues, ie, disorders of the semicircular canals or otoliths. Of these, semicircular canal issues are the most common, usually manifesting with  .

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# Motility Disorders: *Supranuclear Syndromes*

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-- two words

-- three words

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--Ocular tilt reaction : **two words** accompanied by a **two diff words**

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--A peripheral, low-resolution channel for detecting objects. The peripheral channel is very sensitive to motion.

At long last, the payoff for all this backstory: The *supranuclear pathways* consist

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

In contrast, the **OKN** system is driven by phrase

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

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Thus, lesions of a supranuclear pathway manifest as difficulties with either the **maintenance** or **acquisition** of bifixation.

In order to rapidly refixate both foveas on a peripheral image, the efferent system must first produce just enough torque to overcome inertia and rotate the eyes to this image, then 'ramp down' the amount of torque to the level needed to maintain gaze in this new direction.

## Motility Disorders: *Supranuclear Syndromes*

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The **saccadic system** is responsible for rapidly shifting fixation from the current object of interest to a new one located in the visual periphery. These refixation movements are the result of a two-stage process. First there is the **burst**, which is the burst of innervation that is needed to overcome inertia and get the eyes rapidly rotating into position to fixate the new object. The magnitude of the burst (and thus the speed of the rotation) is proportional to how far the eyes need to go, ie, the farther the rotational distance, the bigger the burst.

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## Motility Disorders: *Supranuclear Syndromes*



### ***Supranuclear***

Disorders of the saccadic system can take many forms: Saccadic intrusions (unwanted saccades pulling gaze off its intended target); alterations in saccade speed or accuracy.

with necessitating object-tracking. A peripheral, low-resolution channel for detecting objects. The peripheral channel is very sensitive to motion.

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Orbi

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## Motility Disorders: *Supranuclear Syndromes*



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 At long last, the payoff for all this backstory: 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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In order to rapidly refixate both foveas on a peripheral image, the efferent system must first produce just enough torque to overcome inertia and rotate the eyes to this image, then 'ramp down' the amount of torque to the level needed to maintain gaze in this new direction.

## Motility Disorders: *Supranuclear Syndromes*

### *Supranuclear*

Disorders of the saccadic system can take many forms: Saccadic intrusions (unwanted saccades pulling gaze off its intended target); alterations in saccade speed or accuracy. The inability to initiate saccades is the hallmark of **congenital ocular motor apraxia (COMA)**. Infants with COMA appear to be blind for the first several months of life, because their inability to initiate saccades gives the impression they have no interest in visual stimuli. However, once the infants gain control over head movements (around age 2 months), they learn to induce horizontal versions by action and thereby inducing a VOR response.

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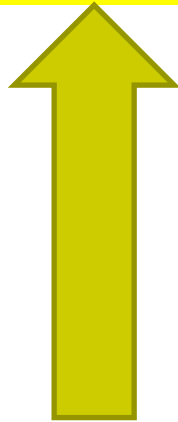
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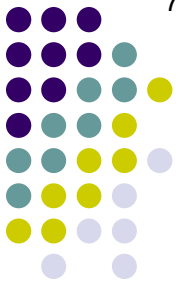
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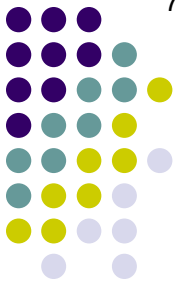


Summary slides coming up...



## Motility Disorders: *Supranuclear Syndromes*

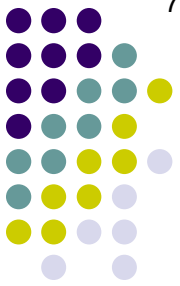
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## Motility Disorders: *Supranuclear Syndromes*

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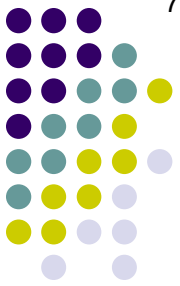


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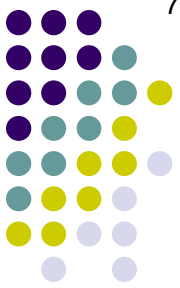
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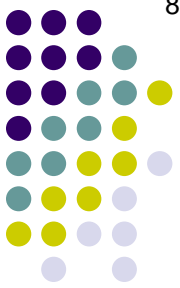
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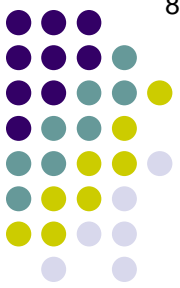
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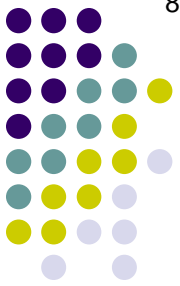
4) The **optokinetic (OKN) system** is responsible for maintaining fixation on a moving object. An important rule-of-thumb can be stated regarding supranuclear motility disorders and diplopia—what is it?

The OKN system, by images sweeping across the retina.

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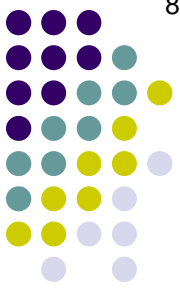
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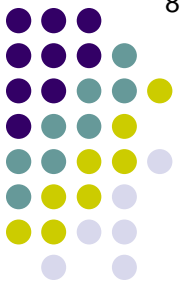
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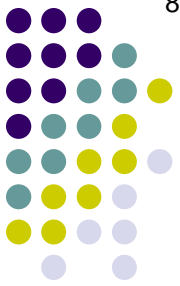
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*Why don't most pts with supranuclear disorders have diplopia?*  
Because most supranuclear disorders affect **both** eyes in a **symmetric** fashion

# Motility Disorders: *Supranuclear Syndromes*

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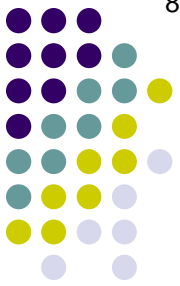
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- Congenital ocular motor apraxia (COMA)
- Progressive supranuclear palsy (PSP)
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ding

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## Motility Disorders: *Supranuclear Syndromes*

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4) The **responsible system** for maintaining single vision is the **vergence system**. An important rule-of-thumb can be stated: **Supranuclear disorders do not typically present with diplopia—what is it?** It is this: With four important exceptions, **supranuclear pts do not complain of diplopia**. OKN system, by images sweeping across the retina.

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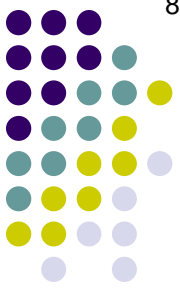
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*Each of these is addressed in detail in other slide-sets—check the ToC*

## Motility Disorders: *Supranuclear Syndromes*



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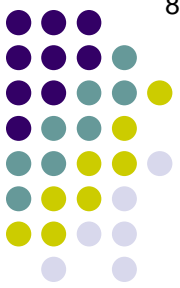
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- Skew deviation
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- Convergence insufficiency
- Convergence spasm

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