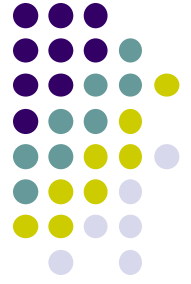


Nystagmus



What is Nystagmus ?

Nystagmus



What is Nystagmus ?

An involuntary repetitive eye movement that initiates with a speed movement off the visual target followed by a purpose of movement movement

Nystagmus



What is Nystagmus ?

An involuntary repetitive eye movement that initiates with a slow movement off the visual target followed by a refixation movement

Nystagmus

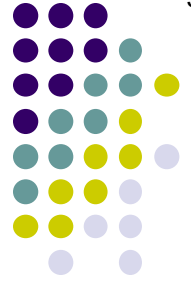


What is Nystagmus ?

An involuntary repetitive eye movement that initiates with a slow movement off the visual target followed by a **refixation movement**

Is the refixation movement fast or slow?

Nystagmus



What is Nystagmus ?

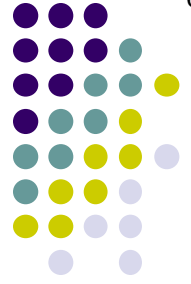
An involuntary repetitive eye movement that initiates with a slow movement off the visual target followed by a **refixation movement**

Is the refixation movement fast or slow?

It can be either.

--If it is fast, the pattern is called a nystagmus

Nystagmus



What is Nystagmus ?

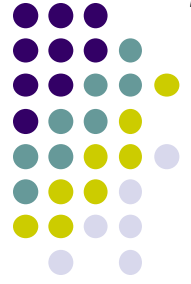
An involuntary repetitive eye movement that initiates with a slow movement off the visual target followed by a **refixation movement**

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Nystagmus



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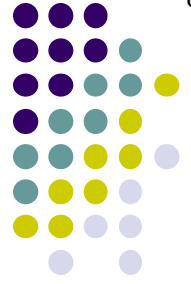
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--If it is slow, it is known as a nystagmus

Nystagmus



What is Nystagmus ?

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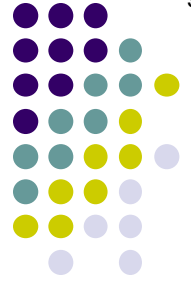
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Nystagmus



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It's important to note that nystagmus can change as a function of **direction of gaze**. One classic example of this is a pendular nystagmus that transforms into a jerk nystagmus in lateral gaze.

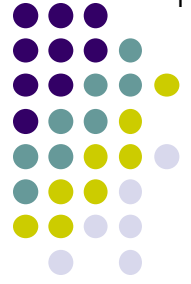
Nystagmus



What is Nystagmus ?

An involuntary repetitive eye movement that **initiates** with a *fast* **movement** off the visual target? followed by a refixation movement

*Can a nystagmus initiate with a **fast** movement?*



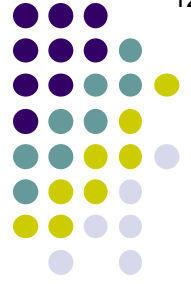
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Nystagmus

same word

Intrusion/

Oscillation

What is ~~Nystagmus~~ ?

An involuntary repetitive eye movement that **initiates with a fast movement off the visual target** followed by a refixation movement

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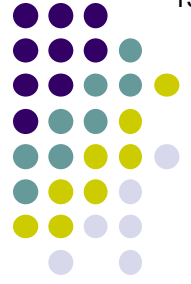
No! By definition, nystagmus commences with a slow movement. If a disordered eye-movement pattern commences with a *fast* movement, it is a

same word

intrusion (if intermittent) or a

same word

oscillation (if sustained).



Nystagmus

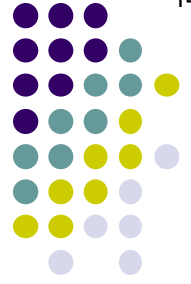
*Saccadic Intrusion/
Oscillation*

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Nystagmus

*Saccadic Intrusion/
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What is ~~Nystagmus~~ ?

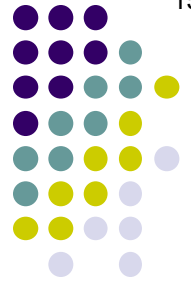
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What does it mean to say that saccadic intrusions are 'intermittent' and saccadic oscillations are 'sustained'?

Nystagmus



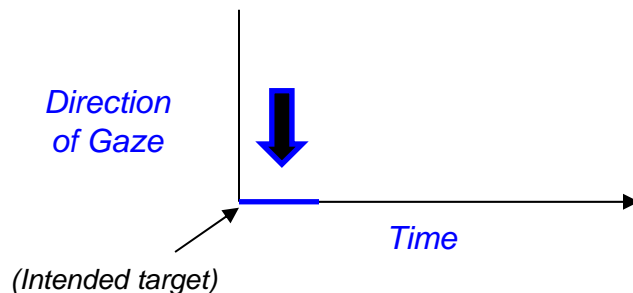
Saccadic Intrusion/ Oscillation What is Nystagmus?

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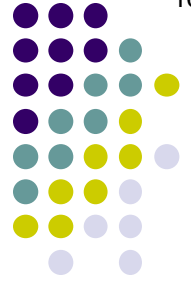
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In *saccadic intrusions*, the eyes fixate the target for a period of time...



Nystagmus

*Saccadic Intrusion/
Oscillation*

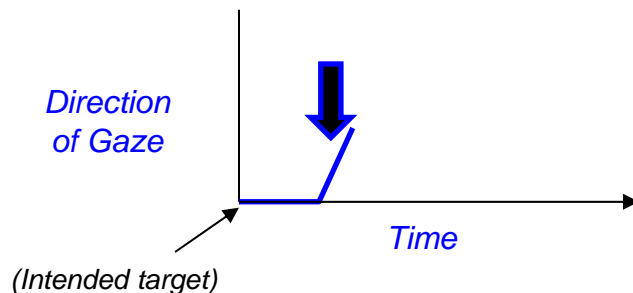
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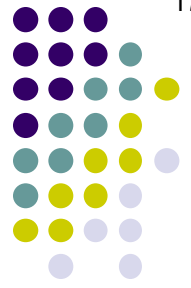
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Nystagmus

*Saccadic Intrusion/
Oscillation*

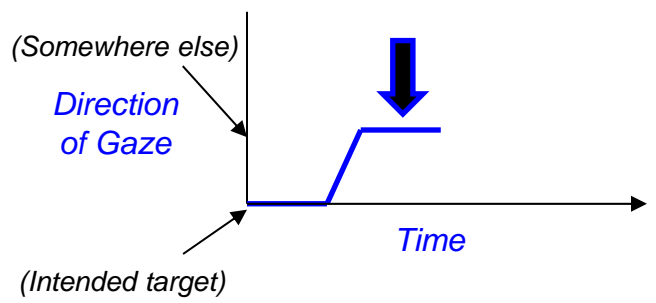
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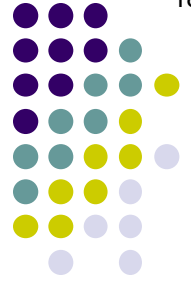
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In *saccadic intrusions*, the eyes fixate the target for a period of time... Then they saccade, and... Fixate a **new** point for a period of time, until they...



Nystagmus

*Saccadic Intrusion/
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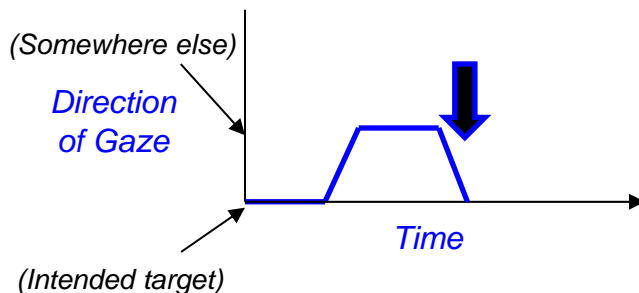
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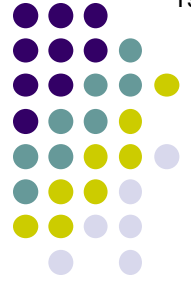
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Saccade again, and...



Nystagmus

*Saccadic Intrusion/
Oscillation*

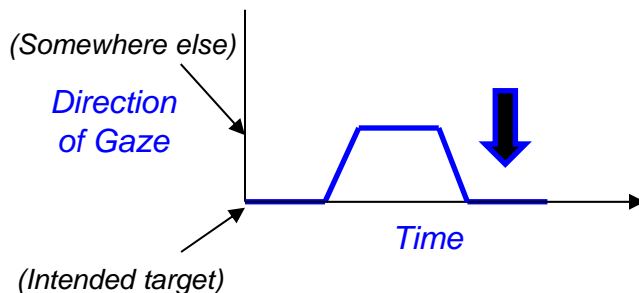
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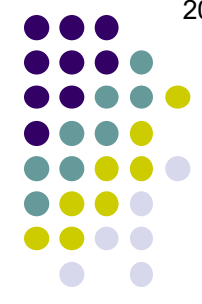
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Fixate a **new** point for a period of time, until they...
Saccade again, and...
Fixate somewhere else for a period of time (note: it doesn't have to be the original fixation target), until they...



Nystagmus

*Saccadic Intrusion/
Oscillation*

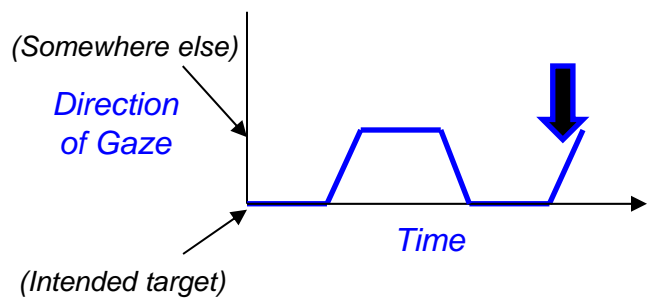
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Nystagmus

*Saccadic Intrusion/
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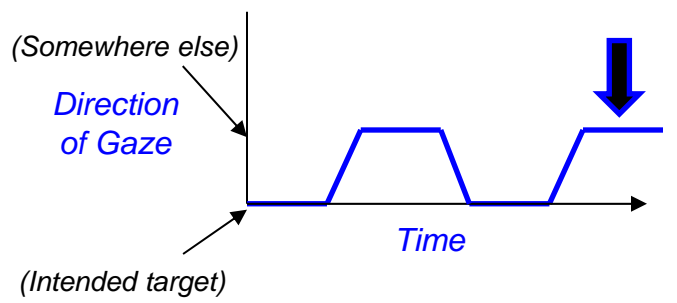
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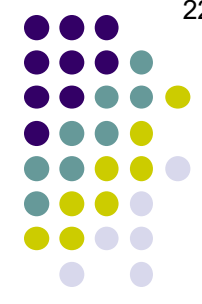
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Nystagmus

*Saccadic Intrusion/
Oscillation*

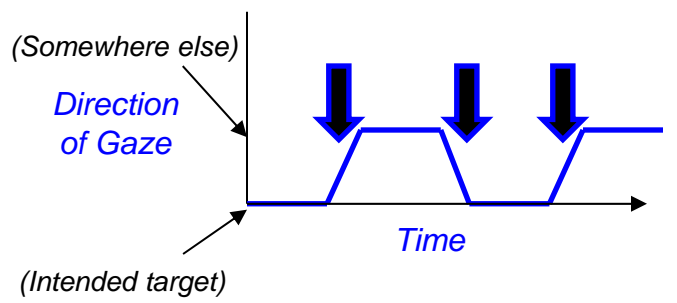
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Nystagmus

*Saccadic Intrusion/
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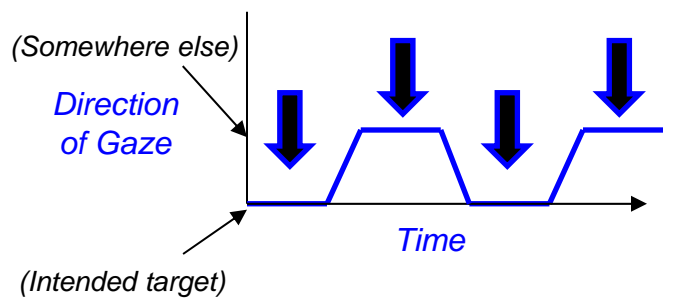
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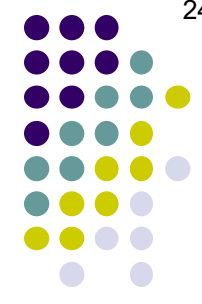
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What does it mean to say that saccadic intrusions are 'intermittent' and saccadic oscillations are 'sustained'?



In short, in saccadic intrusions the saccades are intermittent in the sense that... They are separated by periods in which the eyes are fixated (ie, *not saccading*).



Nystagmus

*Saccadic Intrusion/
Oscillation*

What is ~~Nystagmus~~ ?

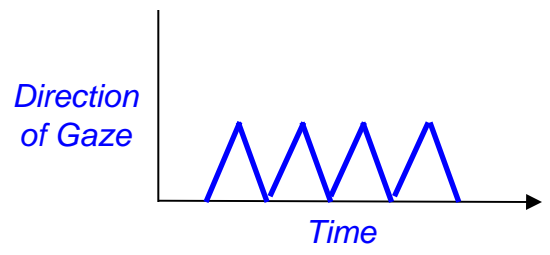
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What does it mean to say that saccadic intrusions are 'intermittent' and saccadic oscillations are 'sustained'?

In contrast, in *saccadic oscillations*, the eyes **never** fixate a target; each saccade is followed immediately by another one. (Note that the saccades need not be of uniform size and pattern as depicted.)





Nystagmus

*Saccadic Intrusion/
Oscillation*

What is ~~Nystagmus~~ ?

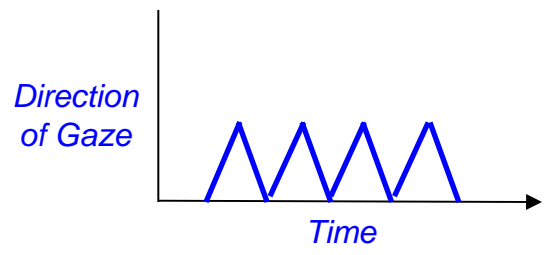
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What does it mean to say that saccadic intrusions are 'intermittent' and saccadic oscillations are 'sustained'?

So, in saccadic oscillations the saccades are sustained in the sense that there is no 'down time,' ie, no time when the eyes are not saccading.





Nystagmus

*Saccadic Intrusion/
Oscillation*

What is ~~Nystagmus~~ ?

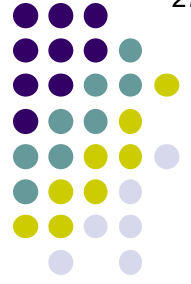
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Give an example of a saccadic intrusion:



Nystagmus

*Saccadic Intrusion/
Oscillation*

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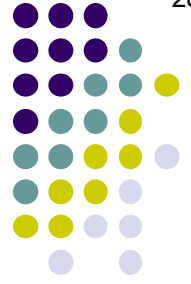
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Give an example of a saccadic intrusion:

Square wave jerks



Nystagmus

*Saccadic Intrusion/
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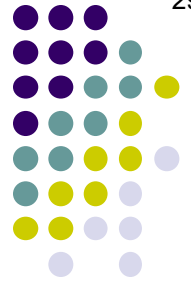
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Give an example of a saccadic intrusion:
Square wave jerks



*Give **two** examples of a saccadic oscillation:*
--
--



Nystagmus

*Saccadic Intrusion/
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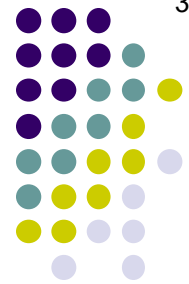
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Give an example of a saccadic intrusion:
Square wave jerks



*Give **two** examples of a saccadic oscillation:*
--**Opsoclonus**
--**Convergence-retraction nystagmus**



Nystagmus

*Saccadic Intrusion/
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Give an example of a saccadic intrusion:

Square w **The Peds book refers to these as “nystagmus-like disorders”**



*Give **two** examples of a saccadic oscillation:*

{ --**Opsoclonus**
--**Convergence-retraction nystagmus**



Nystagmus

*Saccadic Intrusion/
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Give an example of a saccadic intrusion.
 Note: Despite the fact that opsoclonus and convergence-retraction nystagmus are not nystagmus, they will be addressed in this slide-set



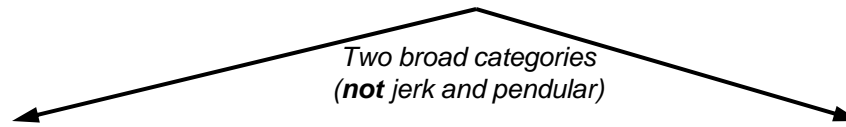
Give two examples of a saccadic oscillation:

- Opsoclonus
- Convergence-retraction nystagmus

Nystagmus

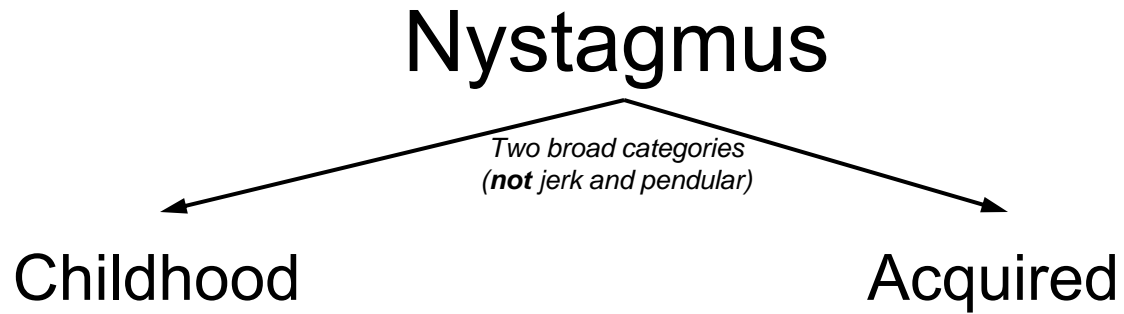


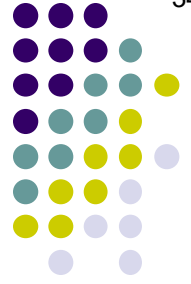
Nystagmus



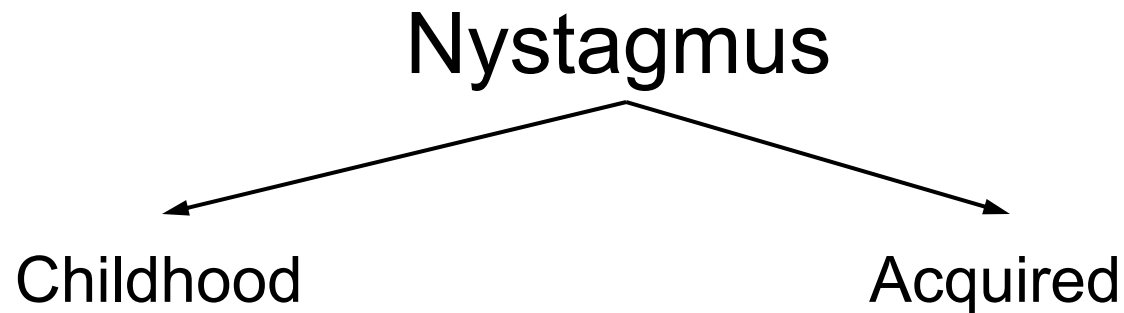
*Two broad categories
(**not** jerk and pendular)*

Nystagmus

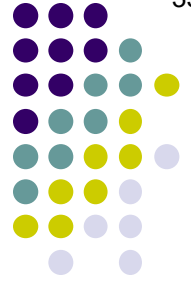




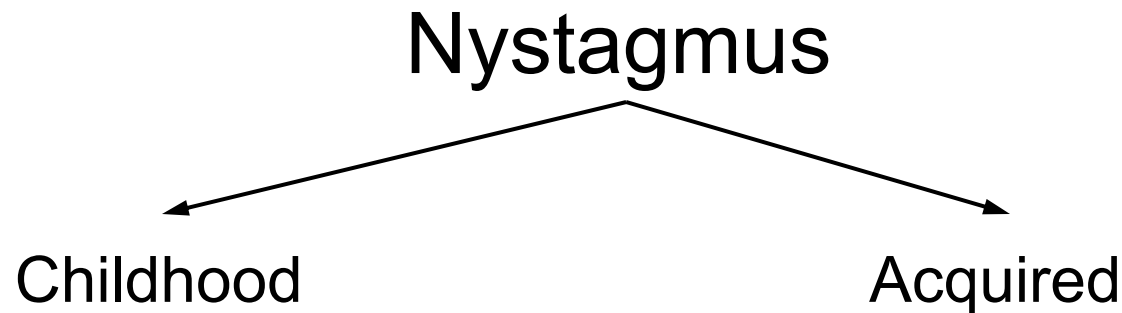
Nystagmus



What one question can be asked, the answer to which will let you know whether you're dealing with a congenital vs an acquired nystagmus (other than 'Have you had this your whole life?'--duh)?

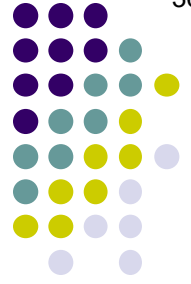


Nystagmus

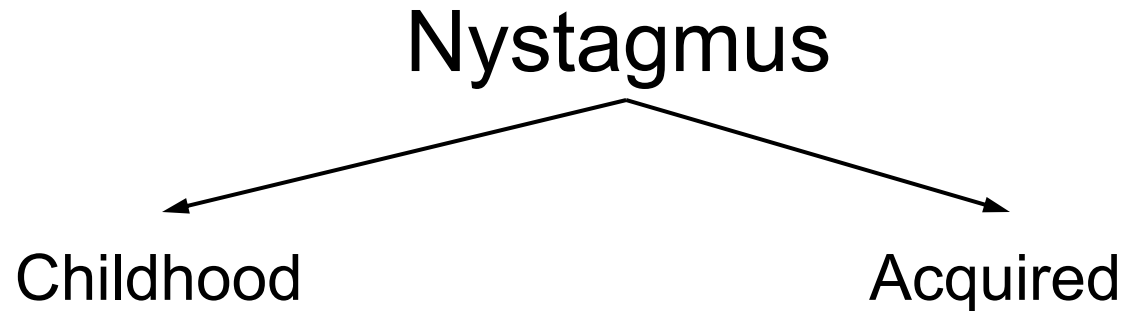


What one question can be asked, the answer to which will let you know whether you're dealing with a congenital vs an acquired nystagmus (other than 'Have you had this your whole life?'--duh)?

The question is, **'Does it look to you as if the world is jumping around?'** If the answer is 'No,' the nystagmus is most likely congenital; if 'Yes,' it is likely acquired.



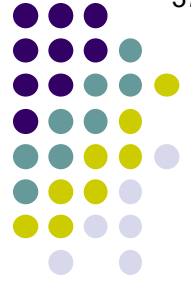
Nystagmus



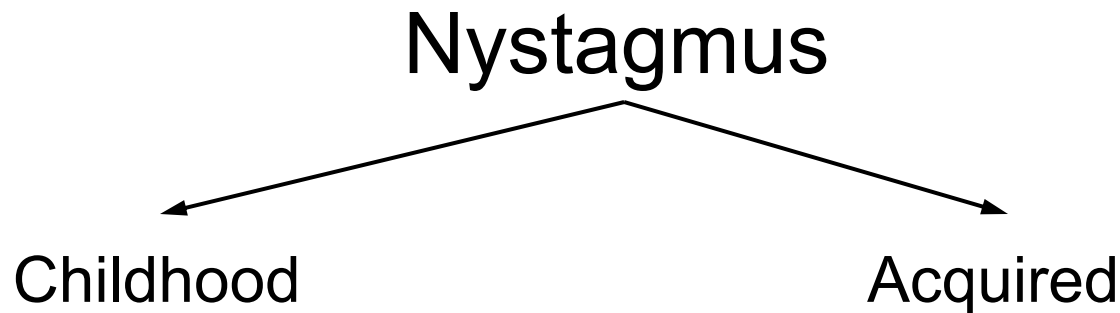
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What is the formal term for the visual experience of 'the world jumping around,' ie, of illusory movement of a stationary world?



Nystagmus

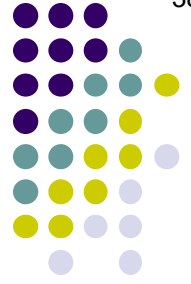


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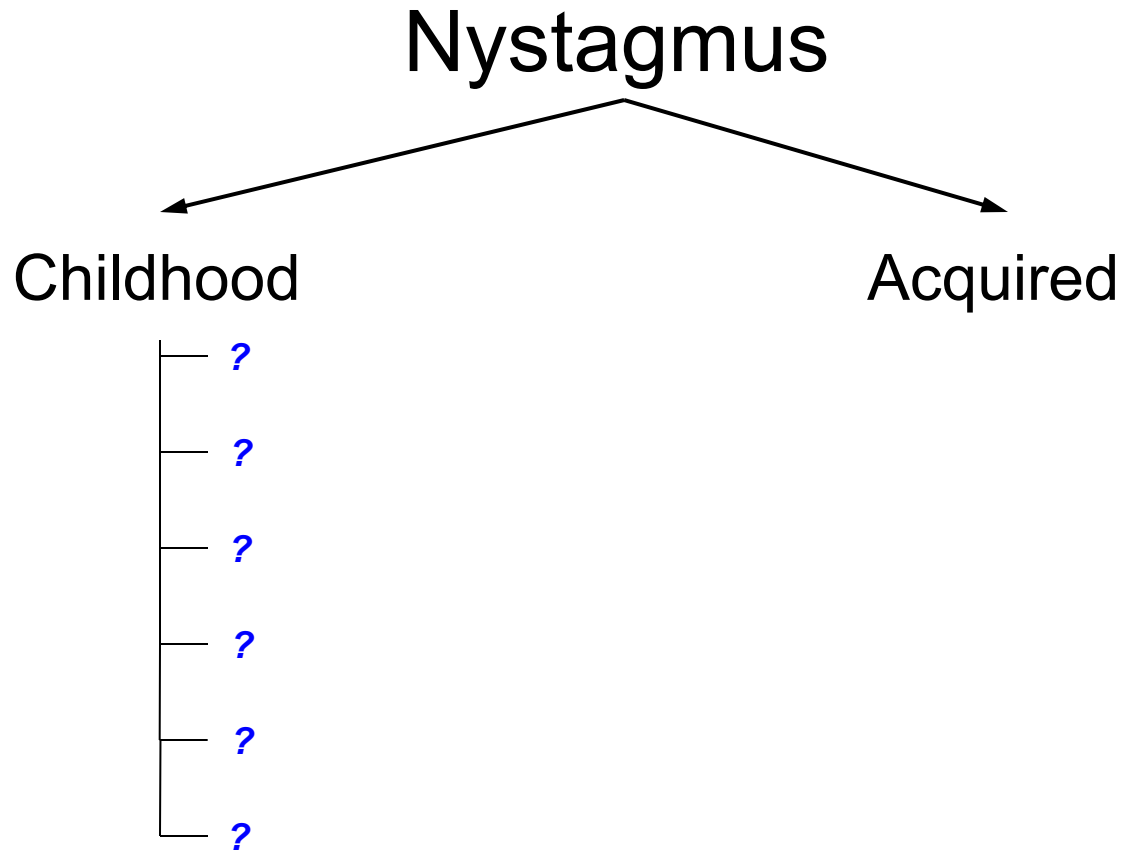
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Oscillopsia

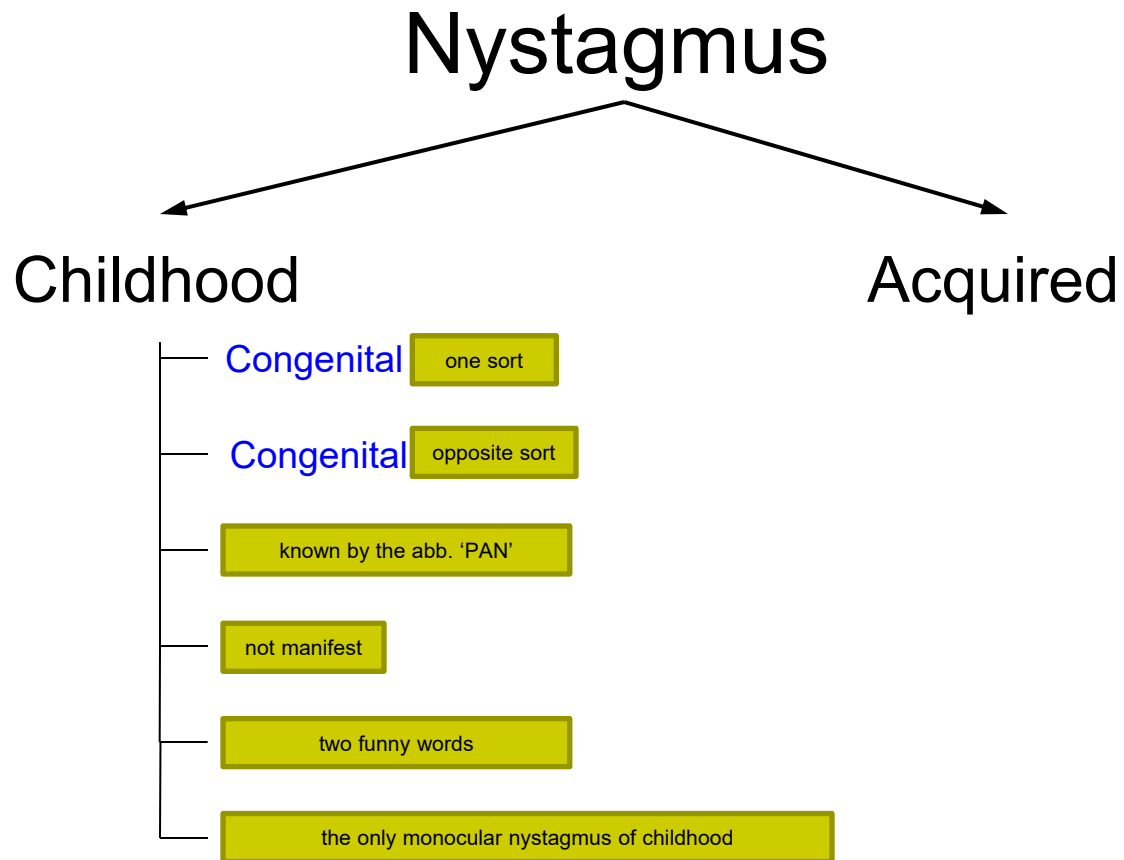


Nystagmus



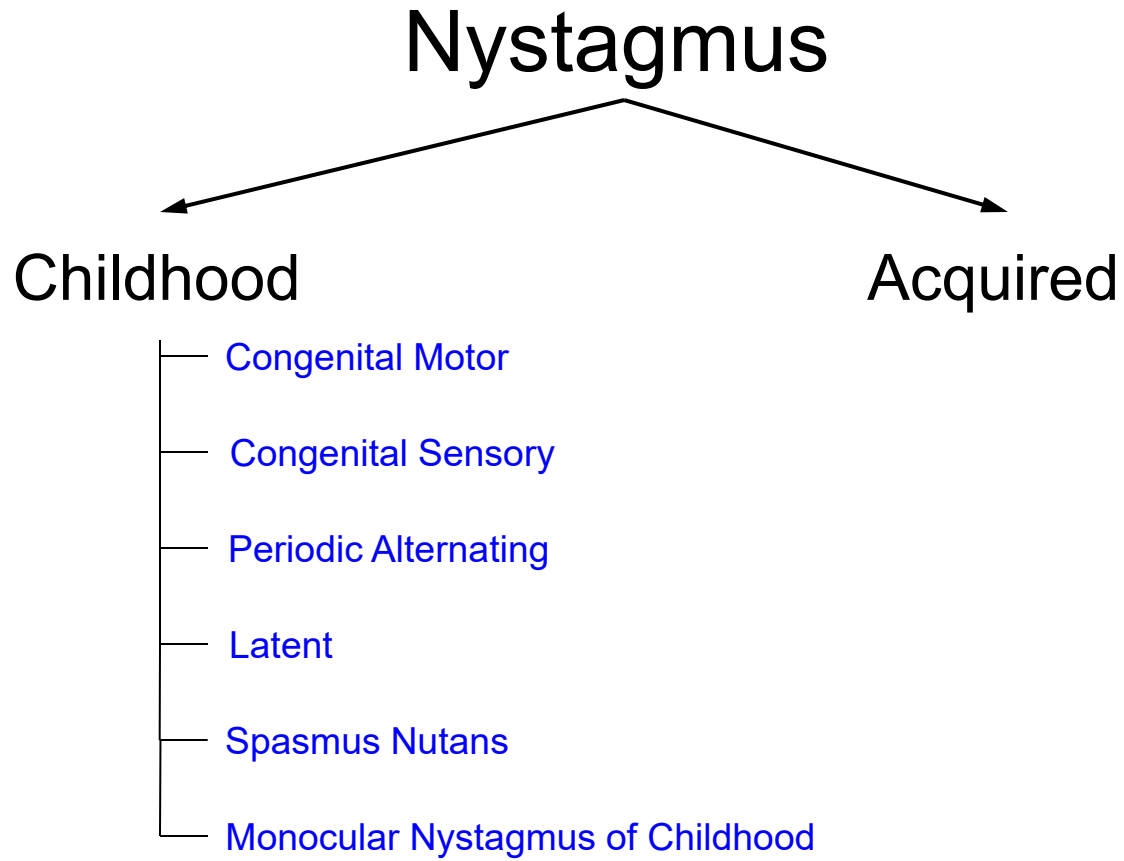


Nystagmus



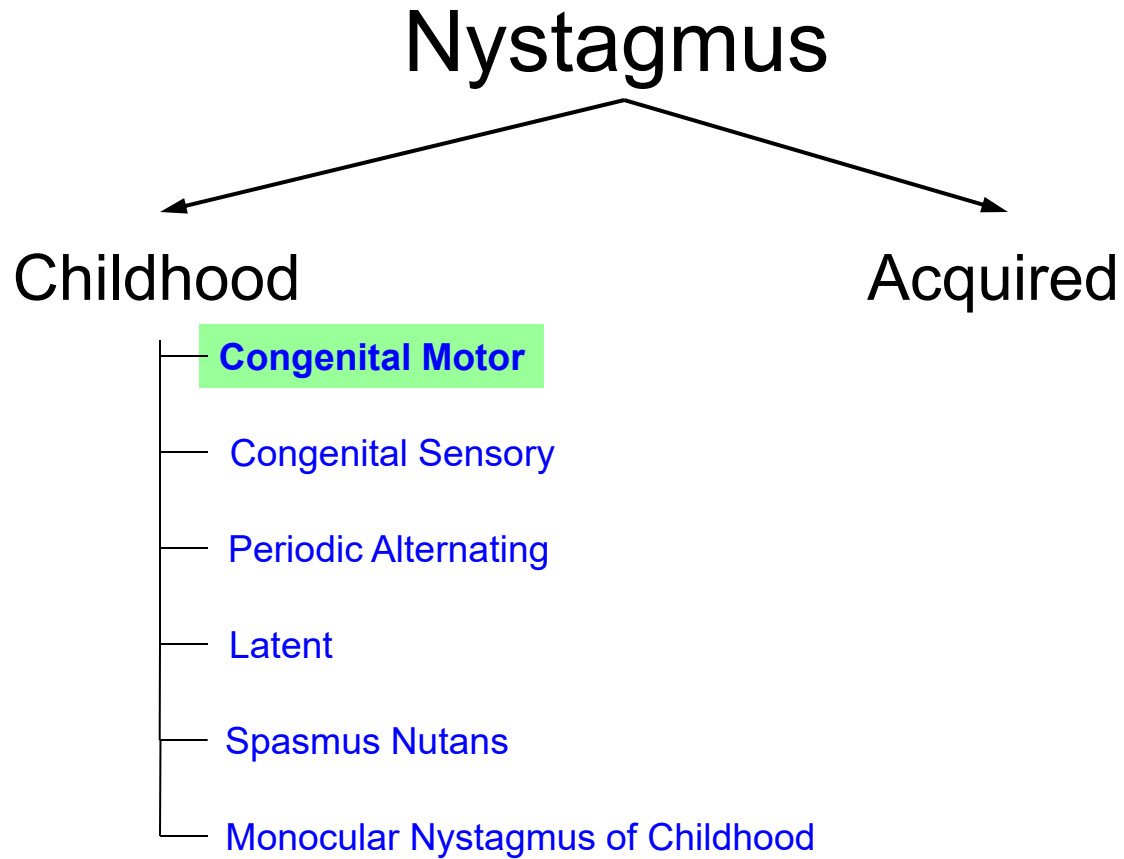


Nystagmus



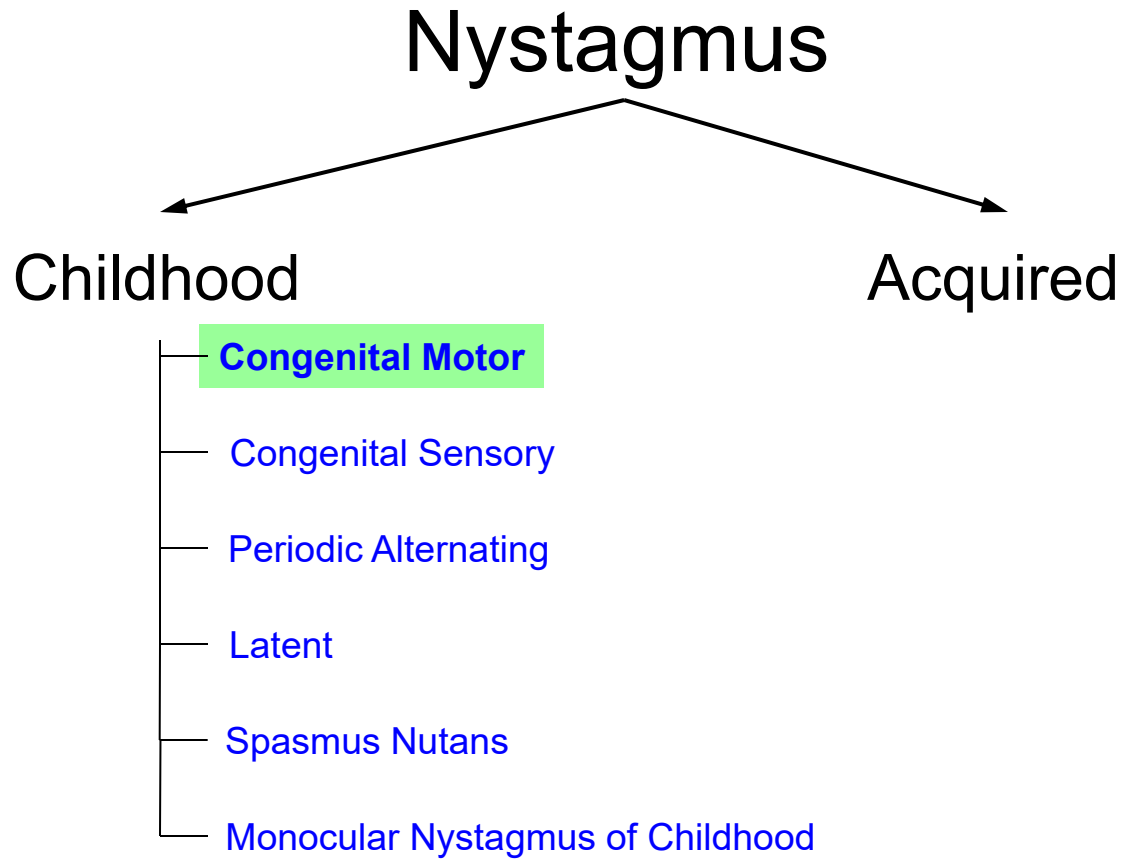
Nystagmus

Congenital motor nystagmus
--Usually...[directionality]



Nystagmus

Congenital motor nystagmus
--Usually...horizontal

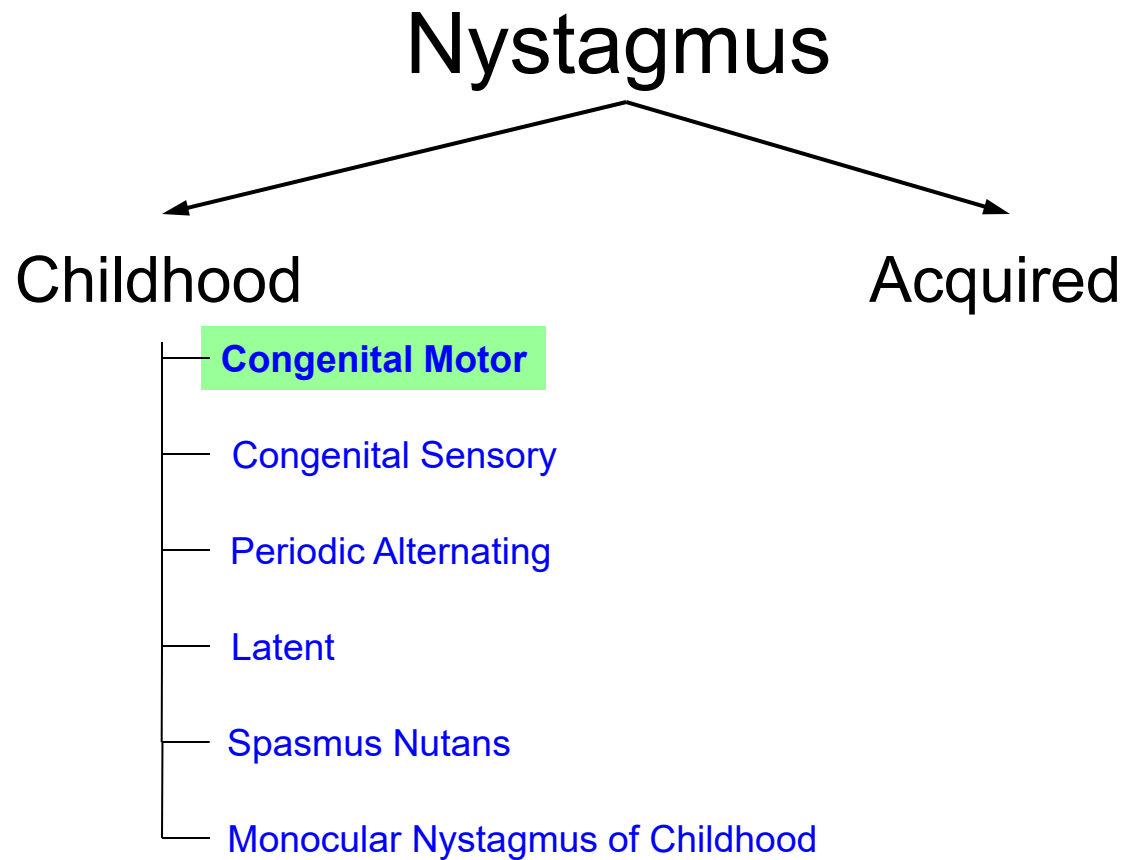


Nystagmus

Congenital motor nystagmus

--Usually...**horizontal**

--Remains horizontal in up/downgaze



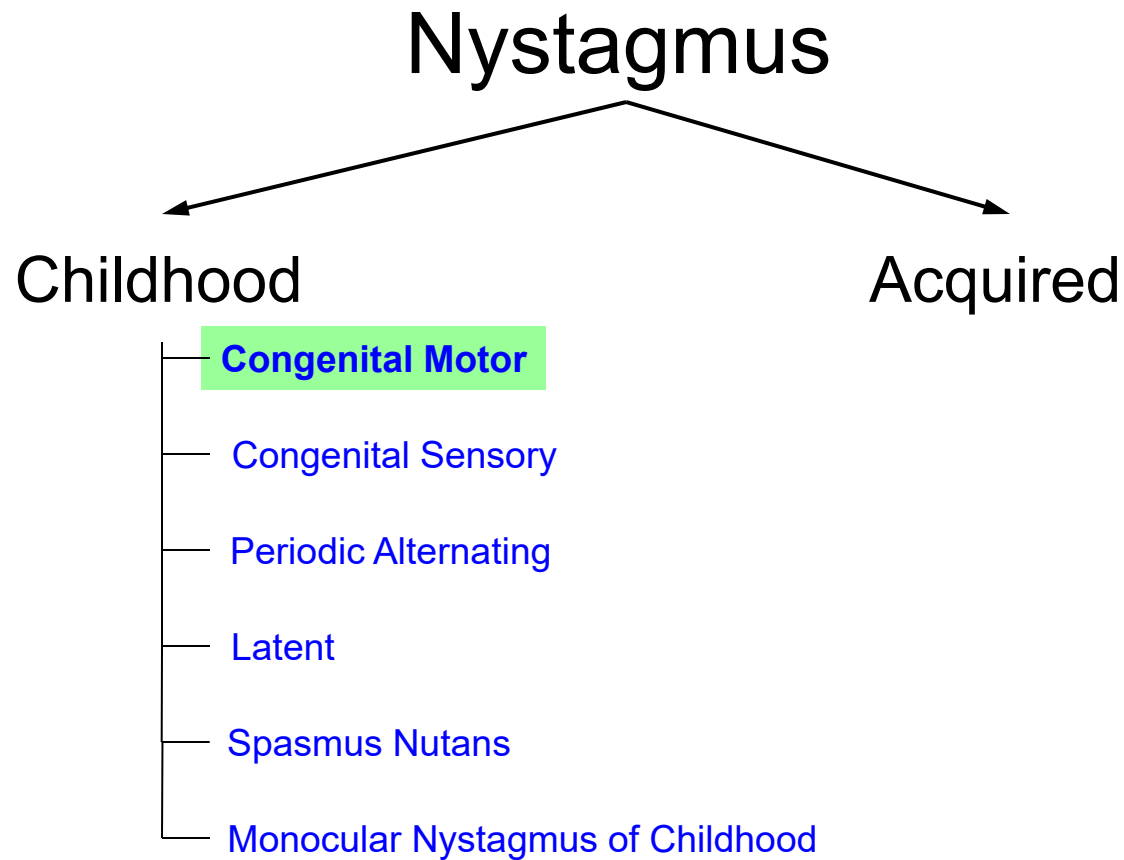
Nystagmus

Congenital motor nystagmus

--Usually...**horizontal**

--Remains horizontal in up/downgaze

--Vision usually... [*good vs bad*]



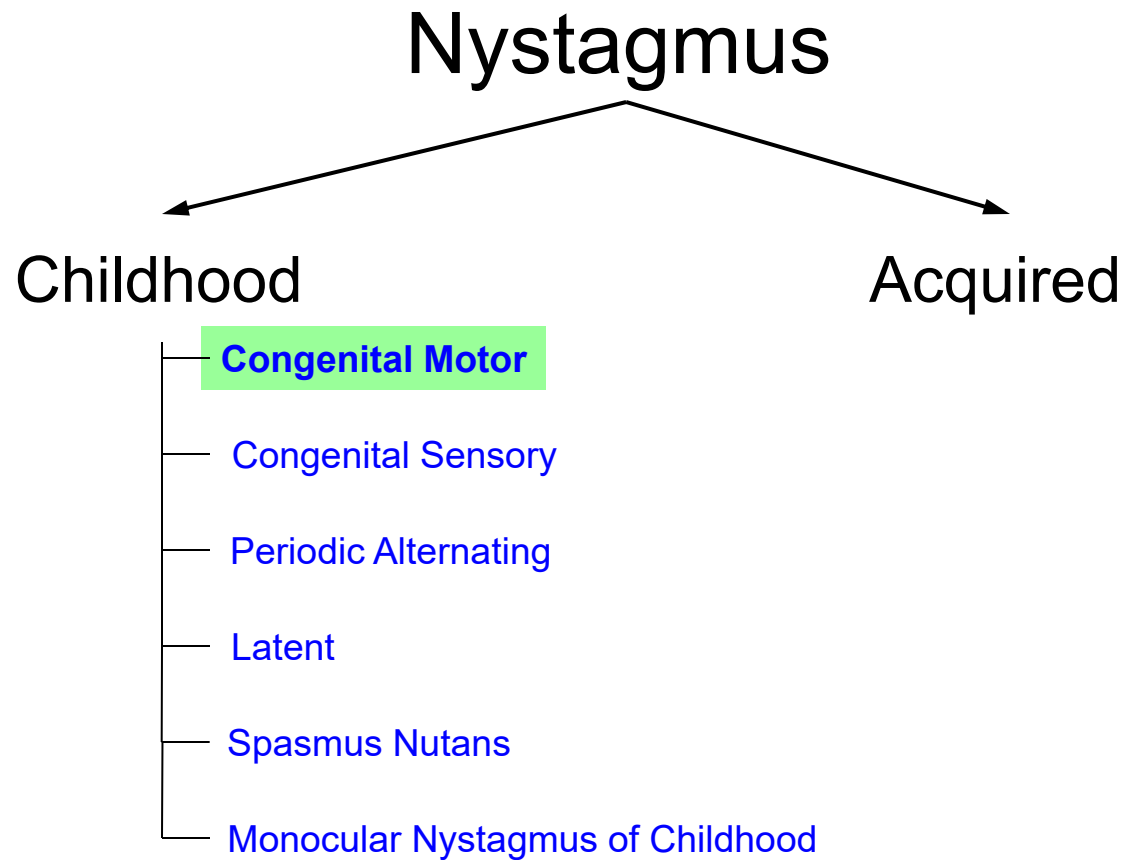
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Nystagmus

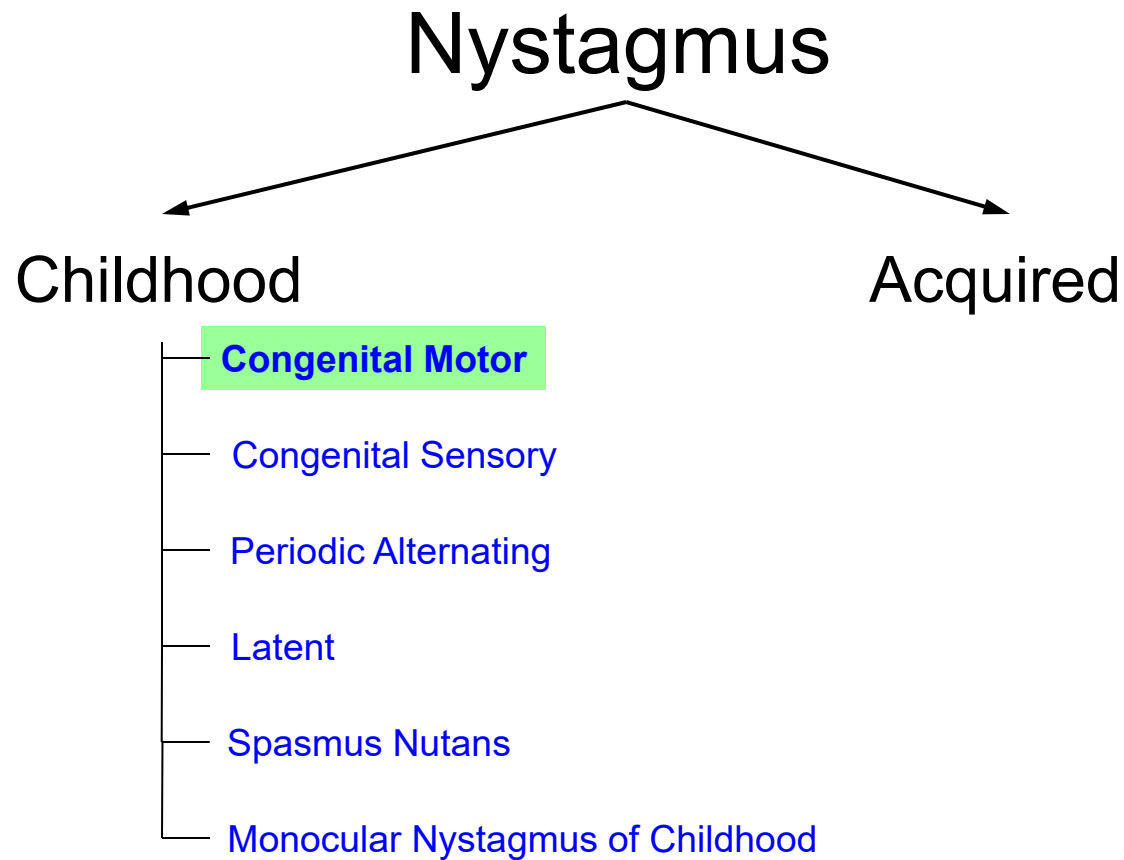
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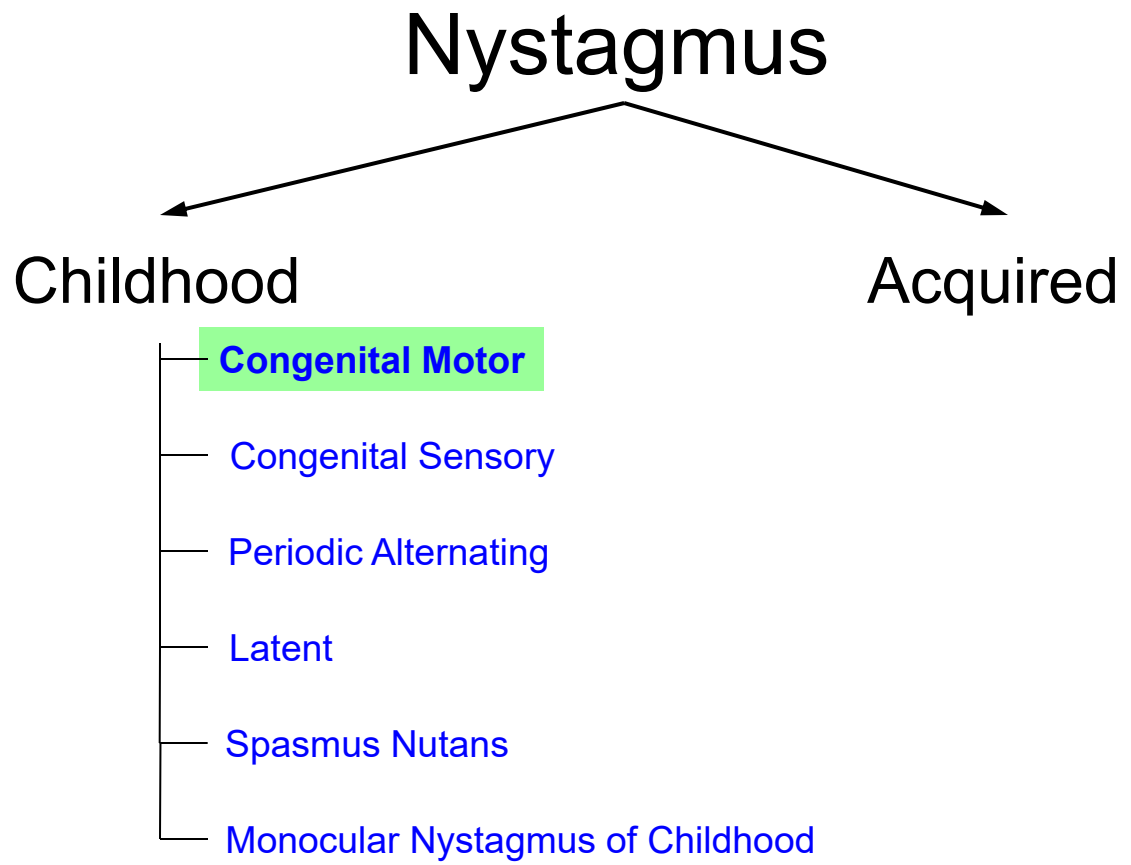
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Nystagmus

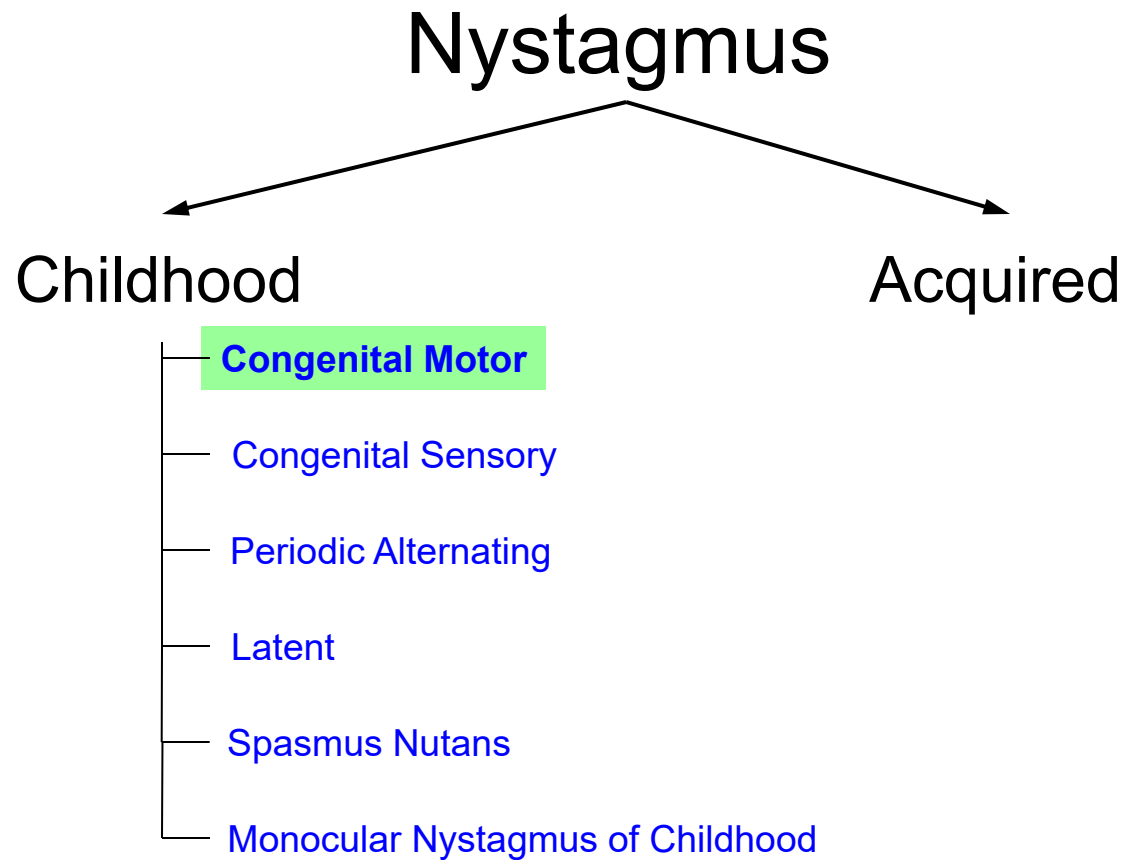
Congenital motor nystagmus
--Usually...**horizontal**
--Remains horizontal in up/downgaze
--Vision usually...**good**
--Nystagmus + good VA = *congenital motor*
--Only form with...[*interesting phenomenon*]



Nystagmus

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
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What is a paradoxical OKN response?

Childhood

Acquired

Congenital Motor

Congenital Sensory

Periodic Alternating

Latent

Spasmus Nutans

Monocular Nystagmus of Childhood

Nystagmus

Congenital motor nystagmus

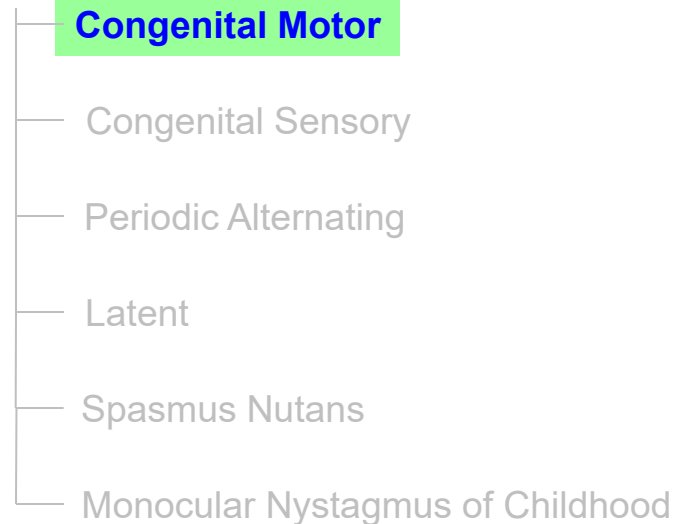
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What is a paradoxical OKN response?

The optokinetic nystagmus (OKN) drum is spun in the direction congruent with the already-present nystagmus. This would be expected to amplify the nystagmus. However, in in congenital motor nystagmus, presentation of congruent OKN movement produces a dampening or even reversal of the nystagmus—hence the term *paradoxical OKN response*.

Childhood

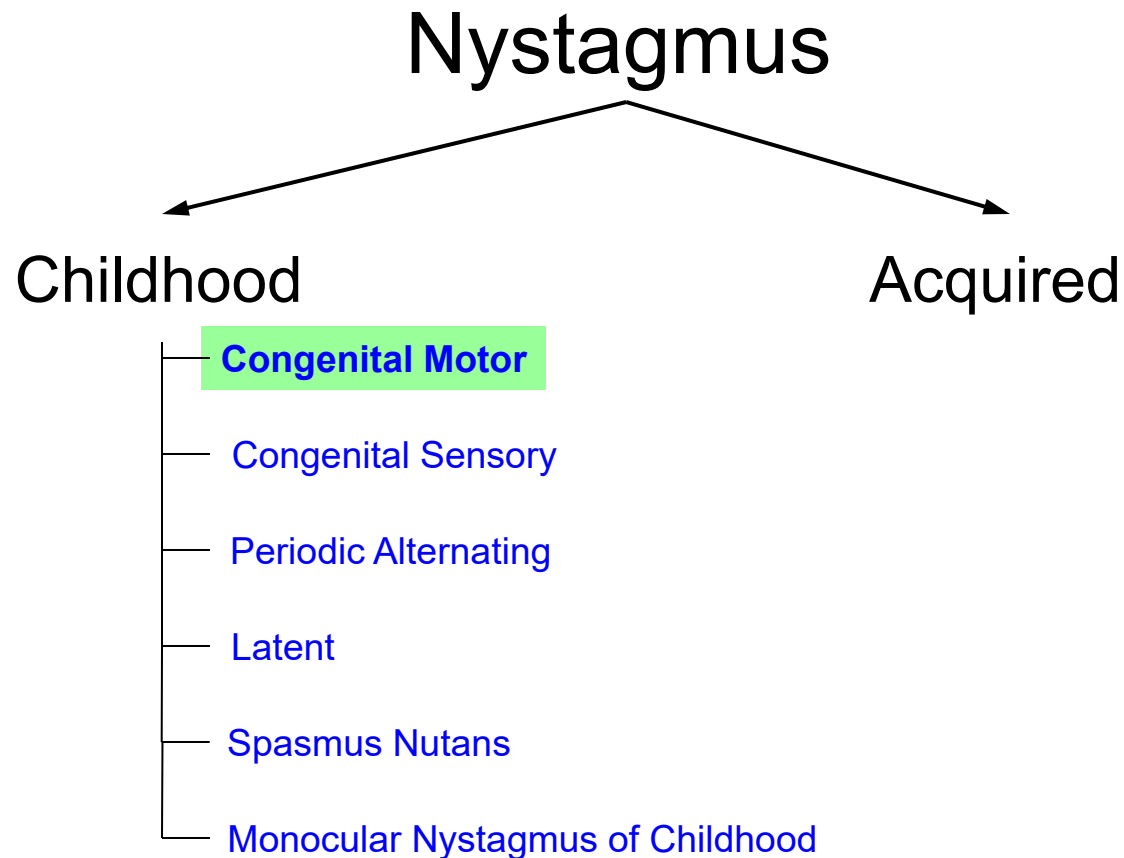
Acquired



Nystagmus

Congenital motor nystagmus

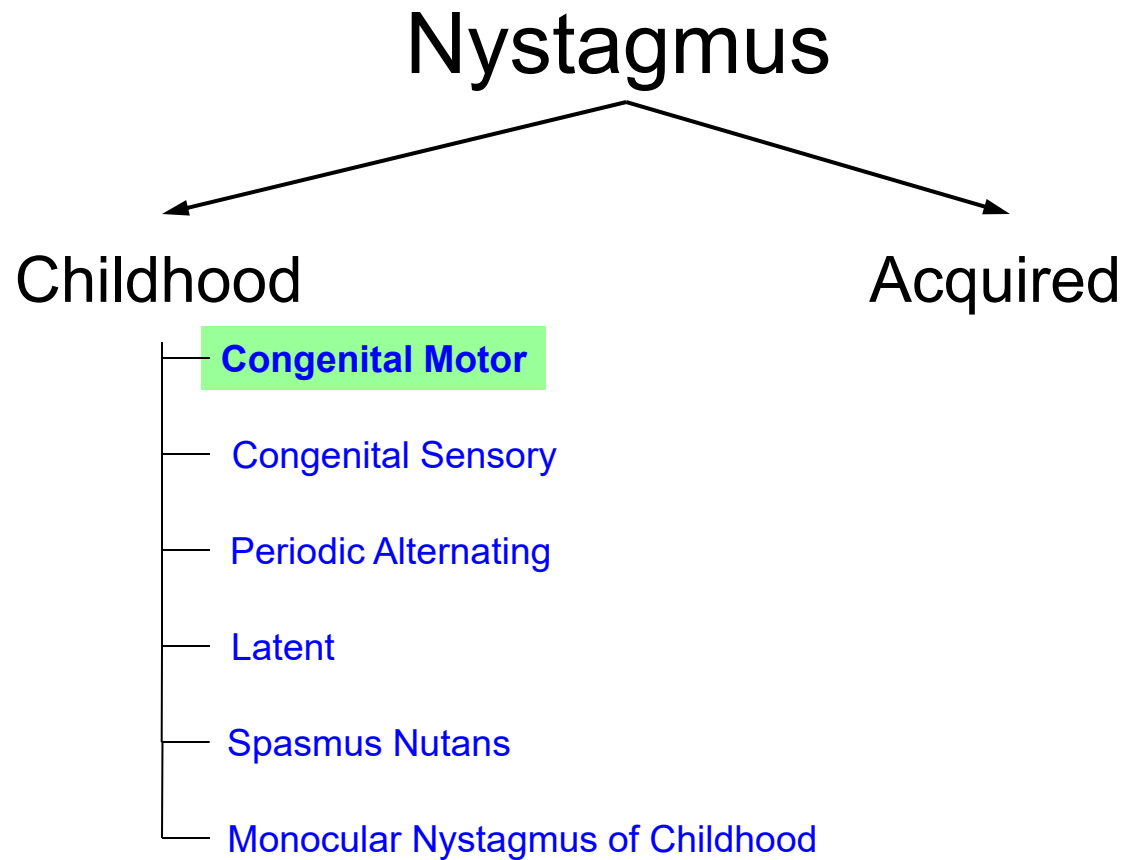
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- Only form with...***paradoxical OKN response***
- Likely to have a...[*nystagmus characteristic*]



Nystagmus

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What is a null point?

- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Nystagmus

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A face turn, i.e., the patient will turn his/her head such that the null point is 'aimed' at an object of regard

- Periodic Alternating
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Is a face turn problematic?

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Is a face turn problematic?

Yes, for multiple reasons--including socialization, cosmesis, and facial development (face turn and/or head tilt at an early age will lead to facial asymmetry)

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Monocular Nystagmus of Childhood

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The slow phase. So for example, in a left-beating jerk nystagmus (ie, fast movement to the **RvL**, slow to the **RvL**), the null point will be located in **RvL** gaze. (Note that this means the pt will adopt a **RvL** face turn in order to place the null point directly in front of her.)

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Monocular Nystagmus of Childhood

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That it gets worse

Spasmus Nutans

Monocular Nystagmus of Childhood

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This observation is ubiquitous enough to have earned the title of 'law.' What is the eponymous name of this law?

Monocular Nystagmus of Childhood

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Alexander's law, which states that "nystagmus increases in intensity (amplitude and frequency) as the eyes are moved in the direction of the fast phase" (per the BCSC *Neuro* book)

Monocular Nystagmus of Childhood

Nystagmus

Congenital motor nystagmus

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*If a jerk
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Note: Strictly speaking, Alexander's law applies only to nystagmuses secondary to vestibular dysfunction (which is not the case in congenital motor nystagmus). However, the concept provides a useful analogy for understanding why null points exist.

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Monocular Nystagmus of Childhood

Nystagmus

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What is a null point?
A direction of gaze in which the intensity of the nystagmus is minimized

In congenital motor nystagmus, there is another 'direction of gaze' in which nystagmus intensity is minimized. What 'direction' is that?

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Nystagmus

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In convergence

*If we'
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Congenital motor nystagmus children often maintain an overconverged state in order to block their nystagmus, and thus will present with an apparent esotropia. What is the name for this syndrome?

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Nystagmus blocking syndrome

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Nystagmus

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Nystagmus blocking syndrome

What two exam findings indicate you may be dealing with a CMN pt exhibiting nystagmus blocking syndrome?

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Nystagmus

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Nystagmus blocking syndrome

What two exam findings indicate you may be dealing with a CMN pt exhibiting nystagmus blocking syndrome?

- There is an inverse relationship between the degree of esotropia and the intensity of the nystagmus; ie, the more crossed their eyes are, the less intense is their nystagmus; and
- During attempts to measure their esotropia, the child 'eats up' prism; ie, the more prism you put in front of them, the more esotropic they become

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three-word term

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In one word, what sort of condition is A-T?

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This implies that the oculomotor findings in A-T change over time. Do they get better, or worse?

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Much worse. Vertical movements become involved in childhood; eventually, the impairment progresses to total ophthalmoplegia.

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Nystagmus

Sensory nystagmus
 --2ndry to early laterality poor vision

Childhood

- Congenital Motor
- **Congenital Sensory**
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

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Weirdly, many of the causes of bilateral poor vision are associated with the letter 'A':

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Anterior segment issues (eg, congenital glaucoma, cataracts)

Aniridia

Leber's congenital **A**maurosis

Achromatopsia

Albinism

Optic nerve **A**trophy

Aicardi syndrome

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Highly convenient mnemonic forthcoming...

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What are the findings of the **Aicardi** syndrome?

- A**icardi syndrome
- I**s...
- C** ← Eye issue
- A** ← CNS issue
- R** ← Cognitive issue
- D** ← Eye issue
- I** ← CNS issue

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- A**genesis of the corpus callosum ← *CNS issue*
- R**etardation ← *Cognitive issue*
- D**epigmented chorioretinal lacunae ← *Eye issue*
- I**nfantile seizures ← *CNS issue*

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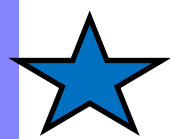
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Optic nerve Atrophy

Aicardi syndrome



Aicardi syndrome is a rare disorder that presents in infancy with seizures (usually infantile spasms). DFE may be requested to assess for chorioretinal lacunae, which are considered pathognomonic for the dz. Other reported ocular associations include PHPV, microphthalmos, cataract and iris abnormalities. Facial dysmorphia can occur. The retardation is usually severe.



(Review slide—no questions)

Nystagmus

Congenital motor nystagmus
 --Usually...**horizontal**
 --Remains horizontal in up/downgaze
 --Vision usually...**good**
 --Nystagmus + good VA = *congenital motor*
 --Only form with...**paradoxical OKN response**
 --Likely to have a...**null point**

Nystagmus

Sensory nystagmus
 --2ndry to early **bilateral** poor vision
 --Waveform depends on visual acuity:
 --20/60 – 20/100:

Childhood

- Congenital Motor
- **Congenital Sensory**
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

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Congenital motor nystagmus

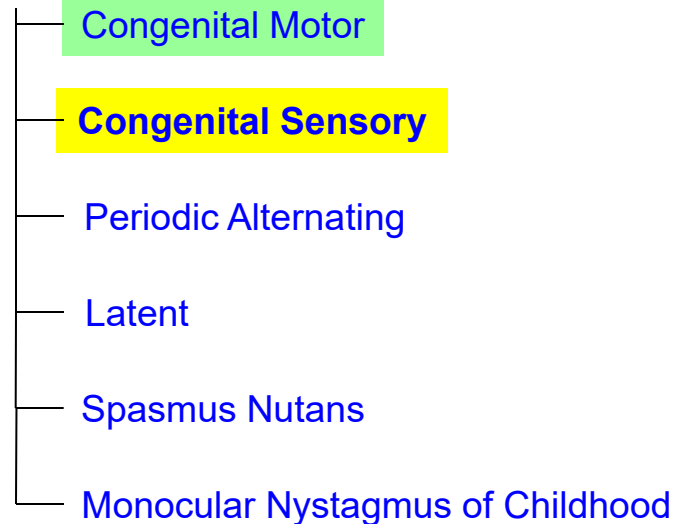
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Childhood



Nystagmus

Congenital motor nystagmus

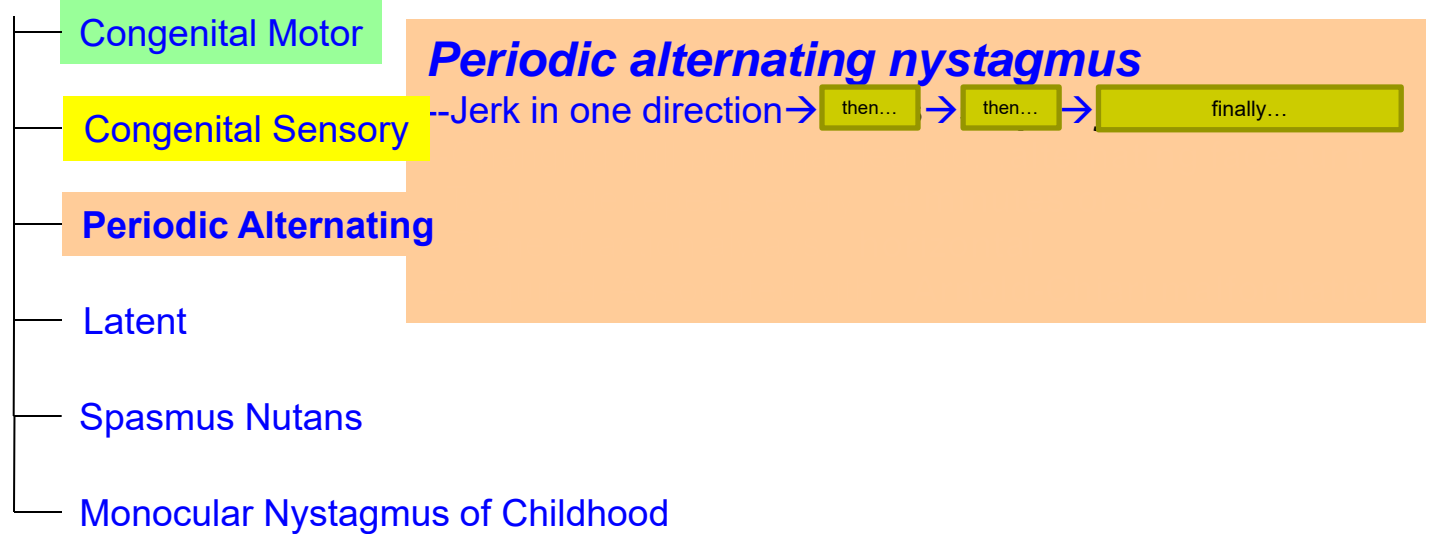
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Childhood



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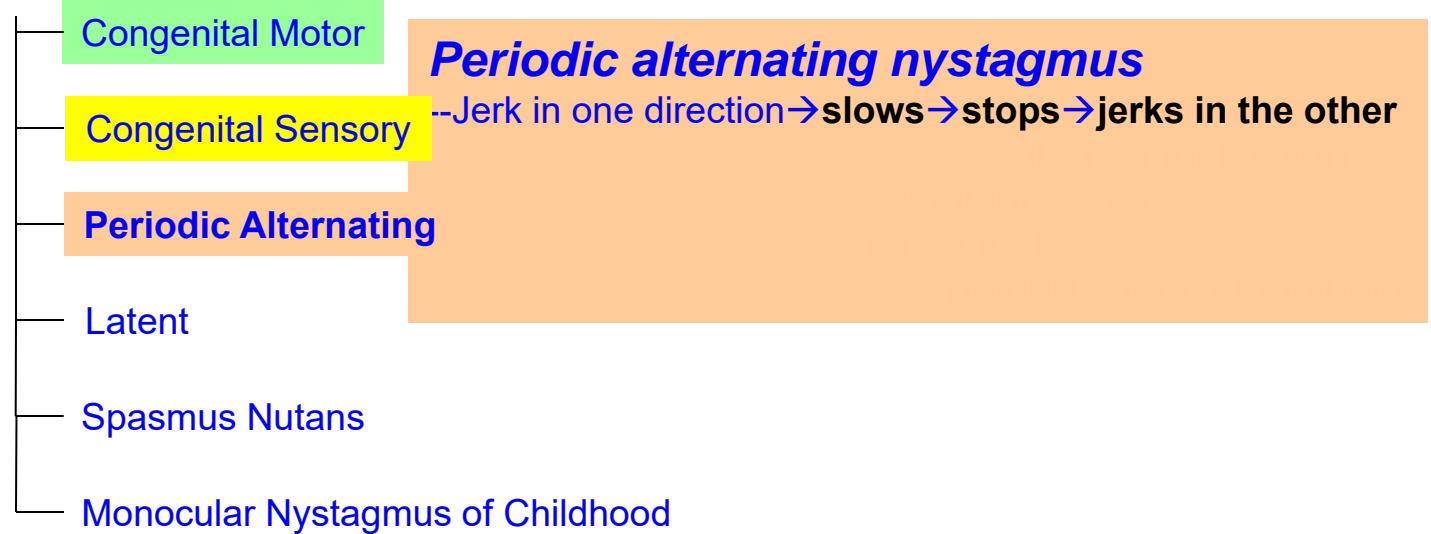
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Childhood



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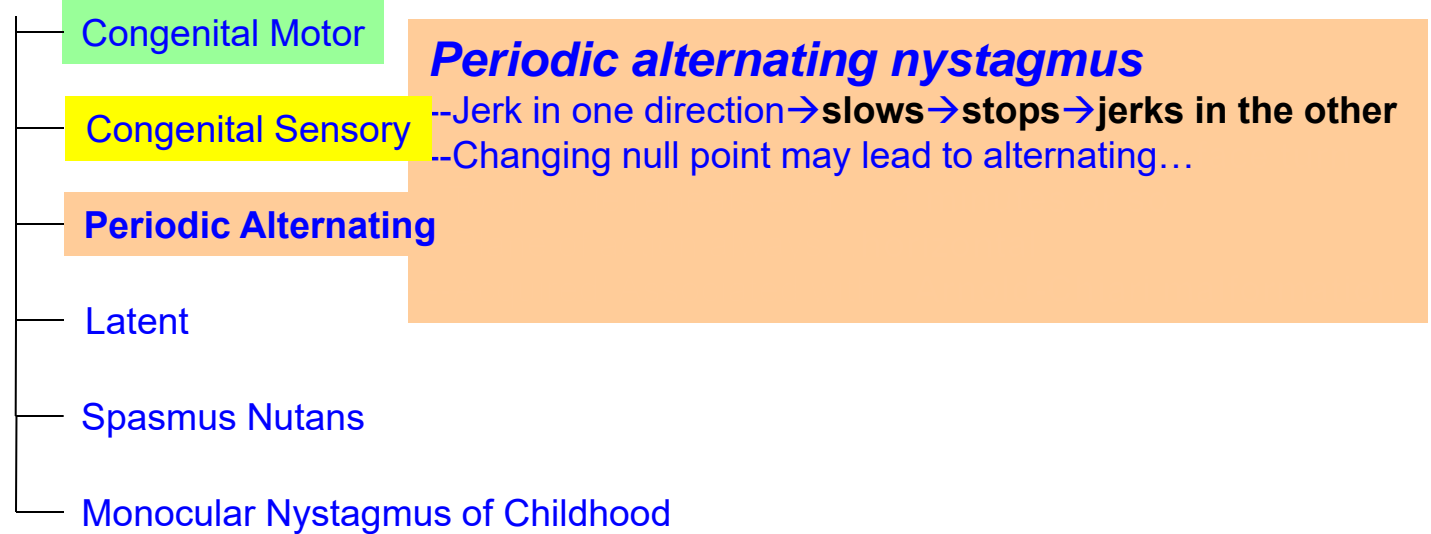
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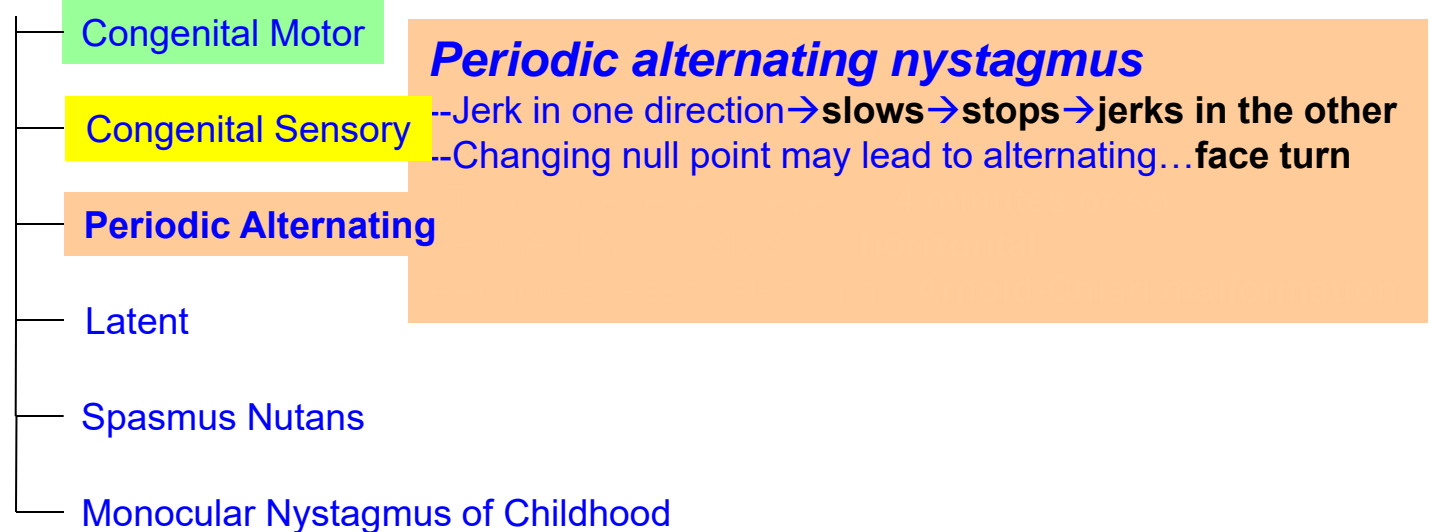
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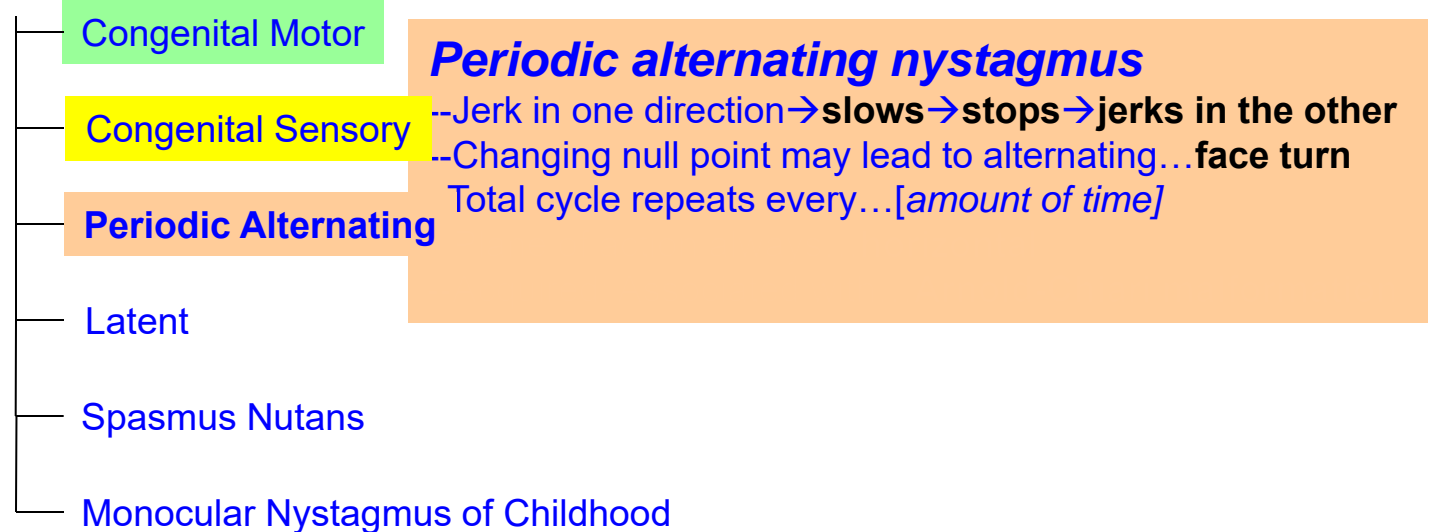
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Childhood



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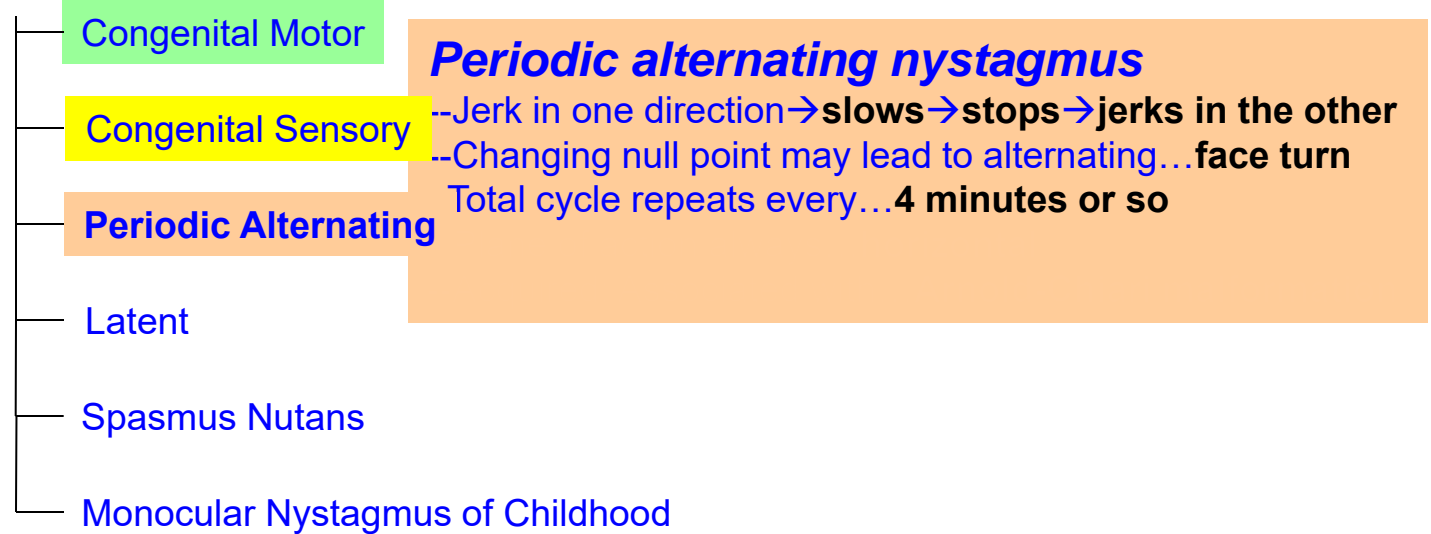
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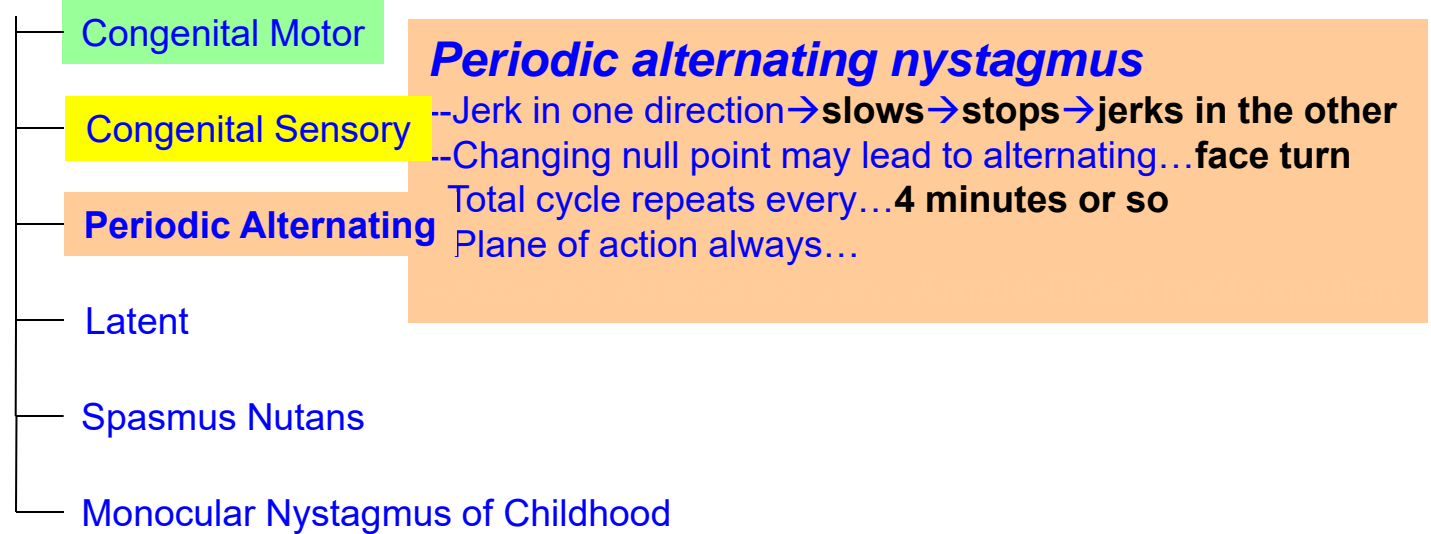
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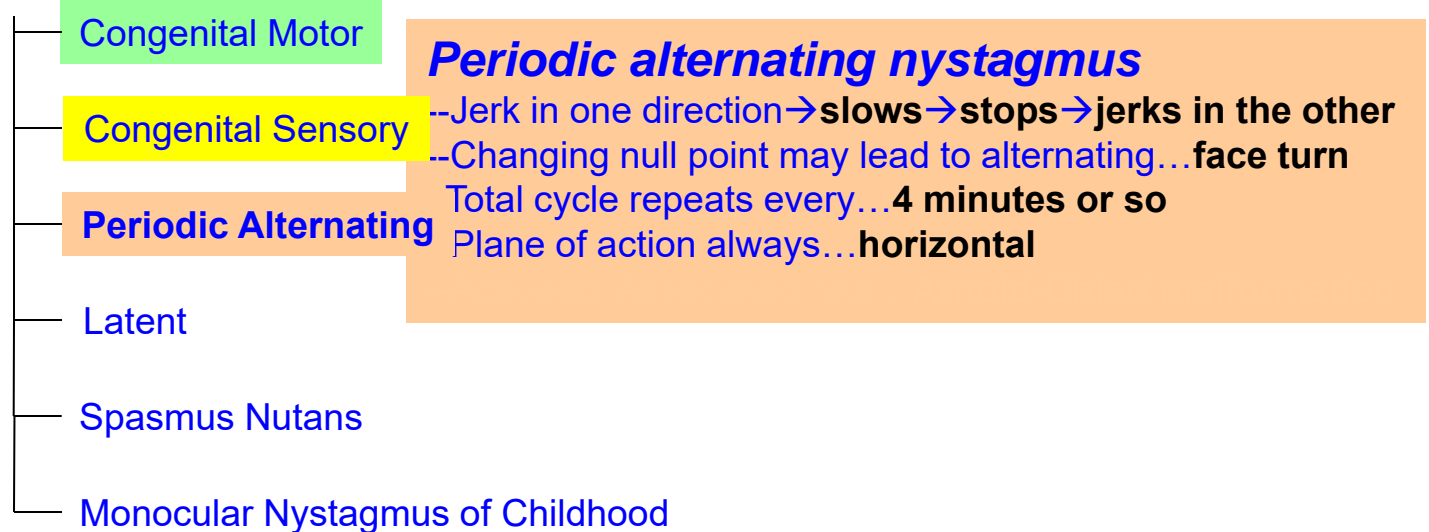
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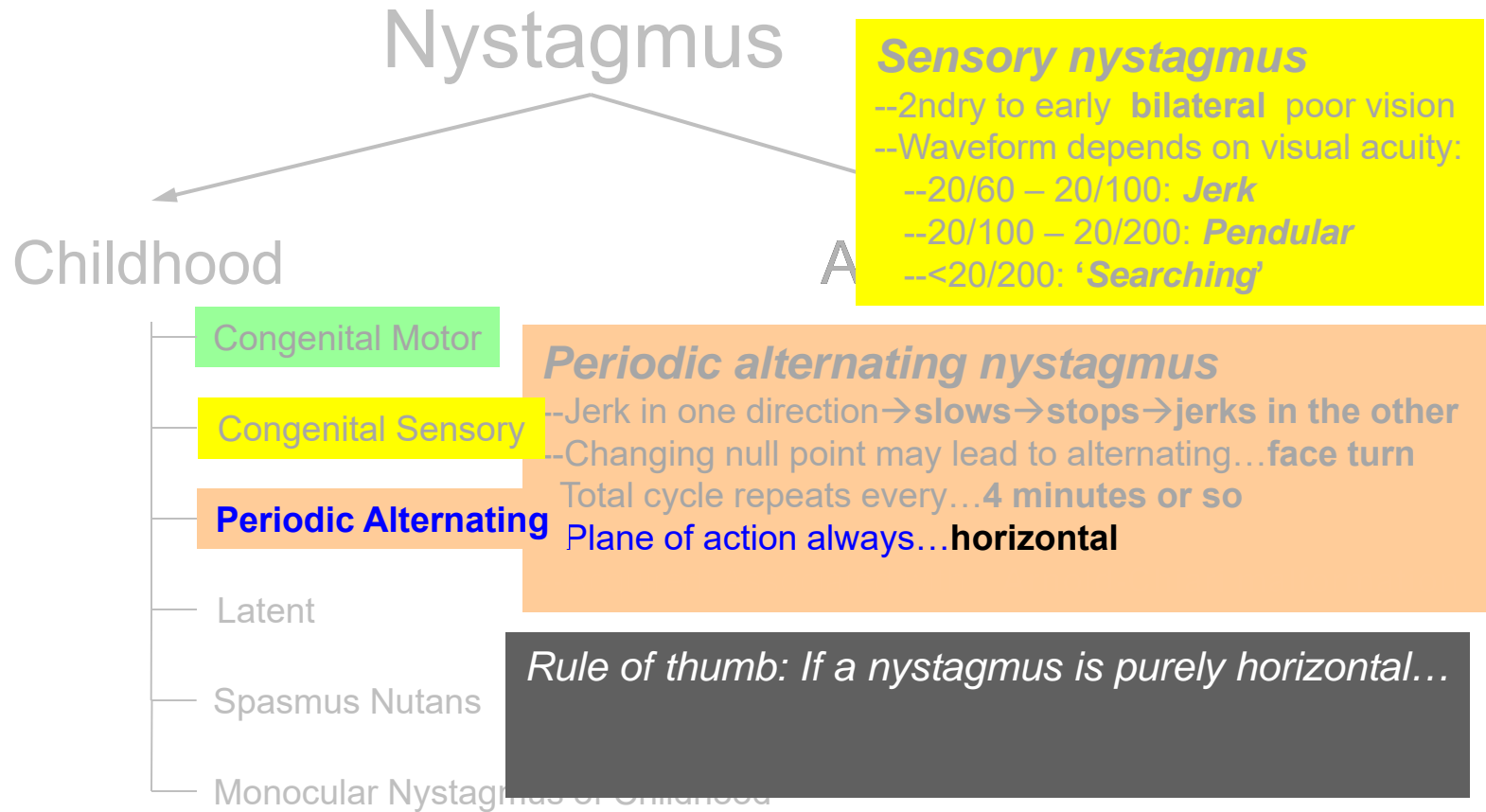


Periodic alternating nystagmus

- Jerk in one direction → **slows** → **stops** → **jerks** in the other
- Changing null point may lead to alternating... **face turn**
- Total cycle repeats every... **4 minutes or so**
- Plane of action always... **horizontal**

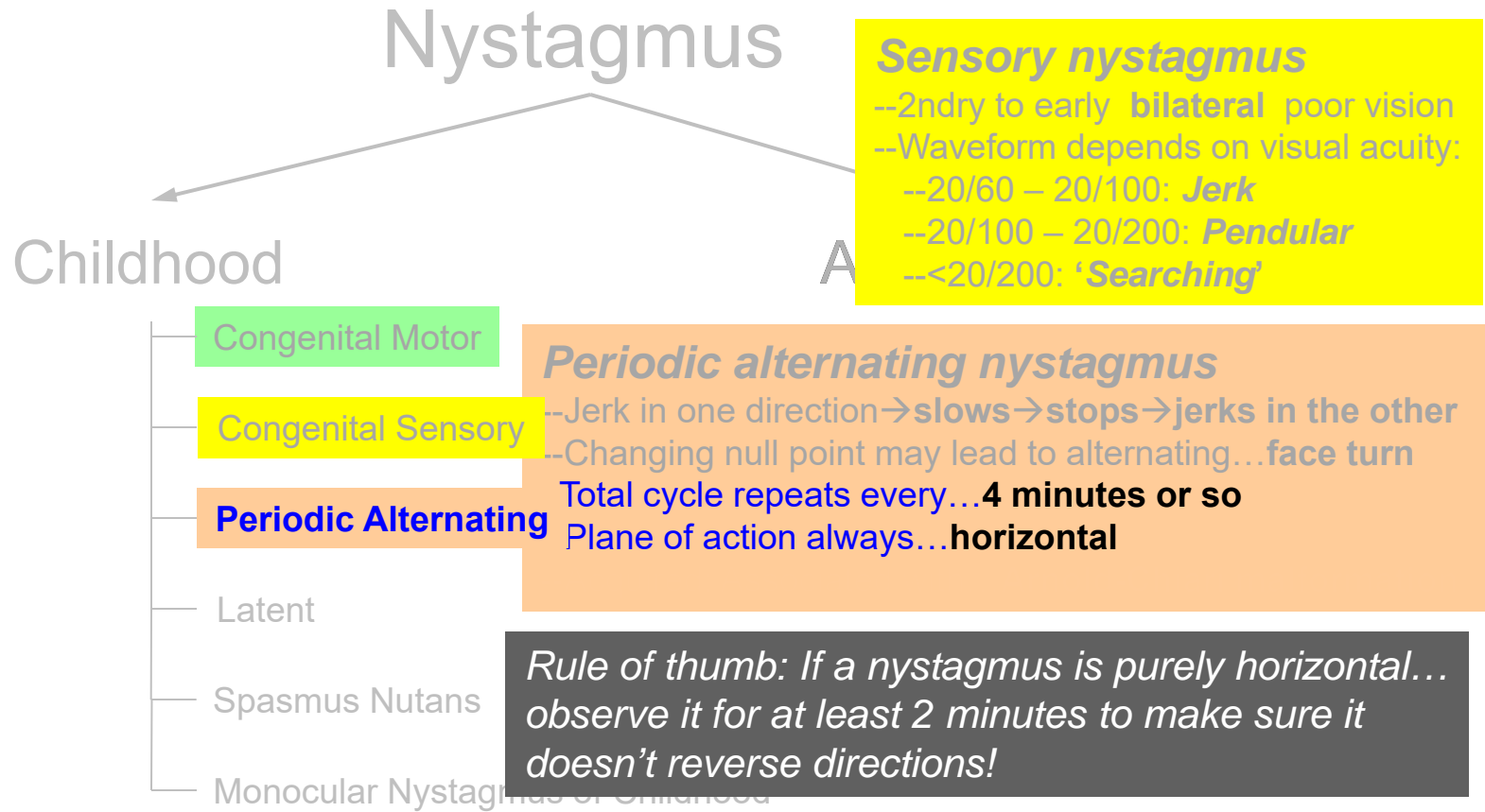
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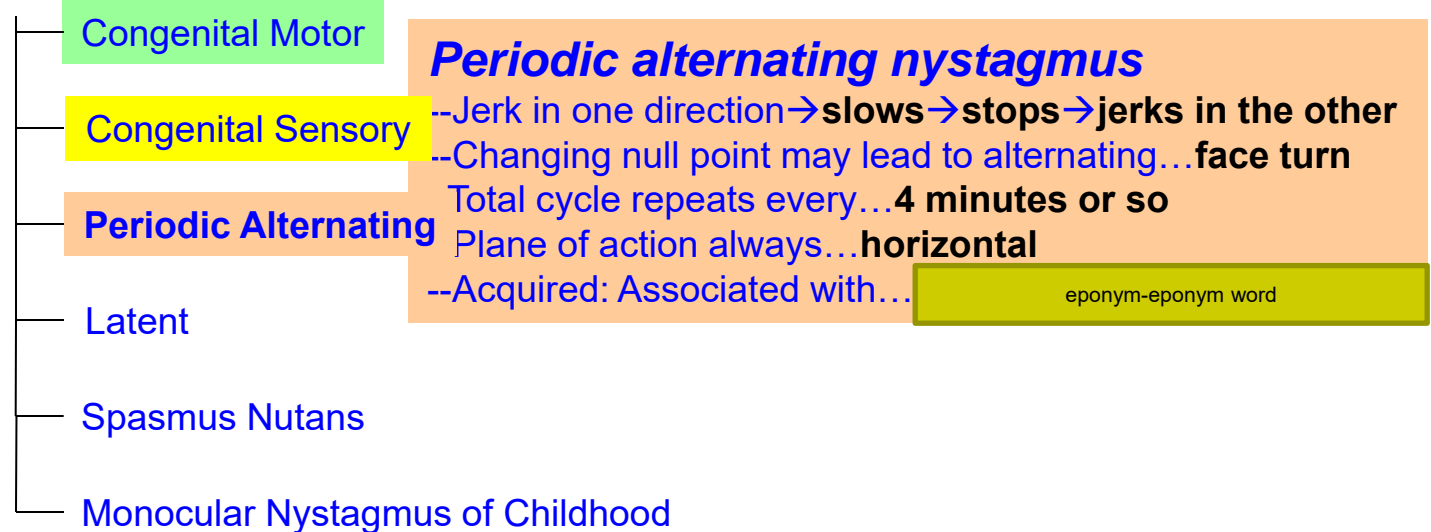
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Nystagmus

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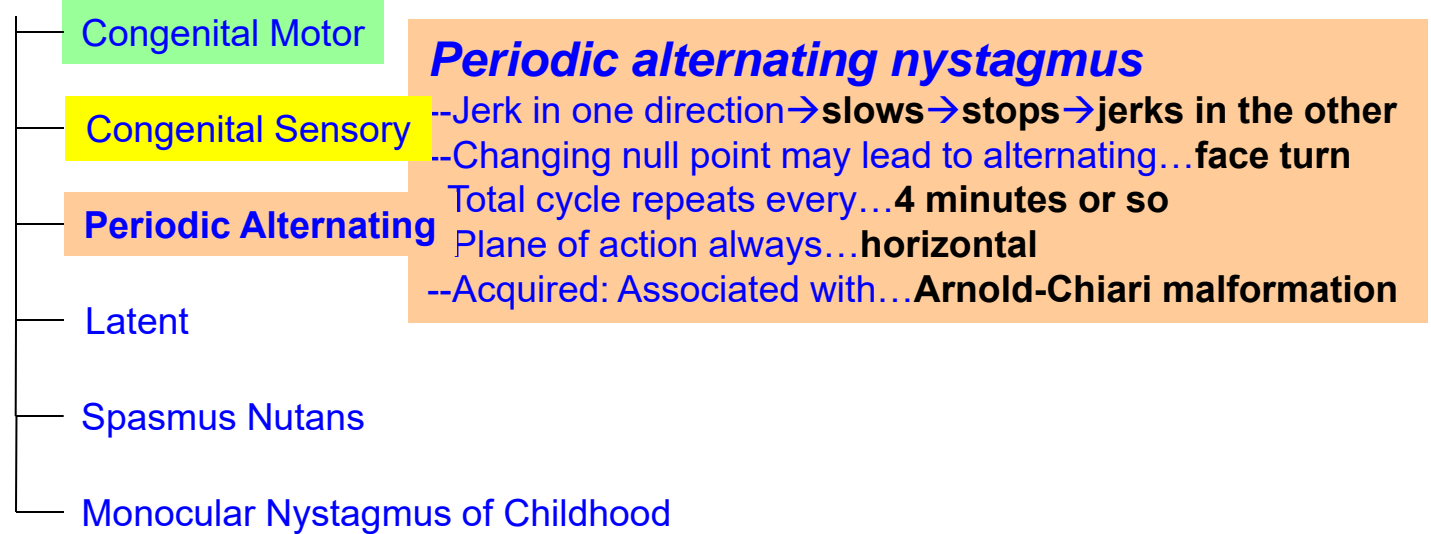
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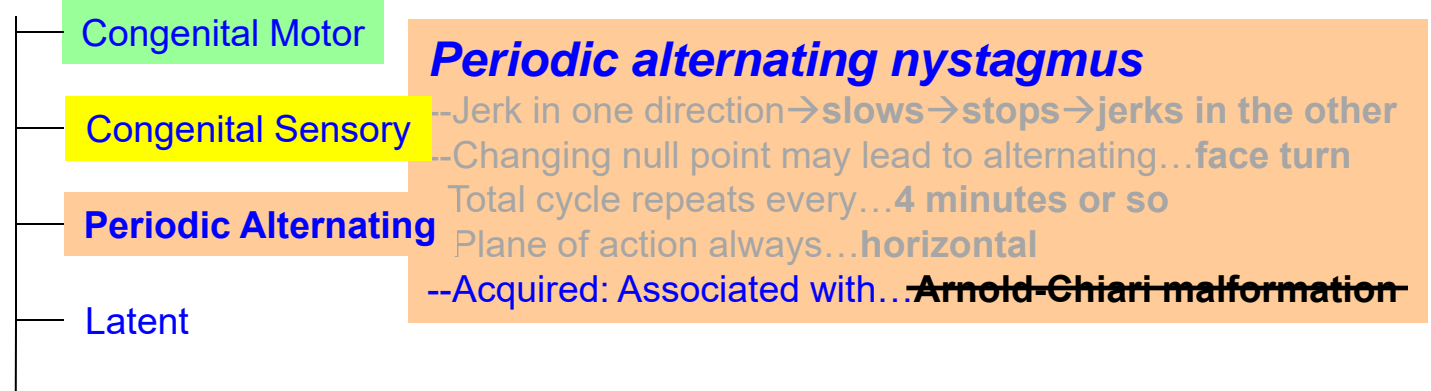
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Childhood



What if it presents as a new sign/symptom in a young adult female--what disease should you consider?

— Monocular Nystagmus of Childhood

Nystagmus

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Childhood

A

Congenital Motor

Periodic alternating nystagmus

Congenital Sensory

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- Changing null point may lead to alternating...**face turn**
- Total cycle repeats every...**4 minutes or so**

Periodic Alternating

Plane of action always...**horizontal**

Latent

--Acquired: Associated with...**Arnold-Chiari malformation**

What if it presents as a new sign/symptom in a young adult female--what disease should you consider?
Multiple sclerosis

Monocular Nystagmus of Childhood

Nystagmus

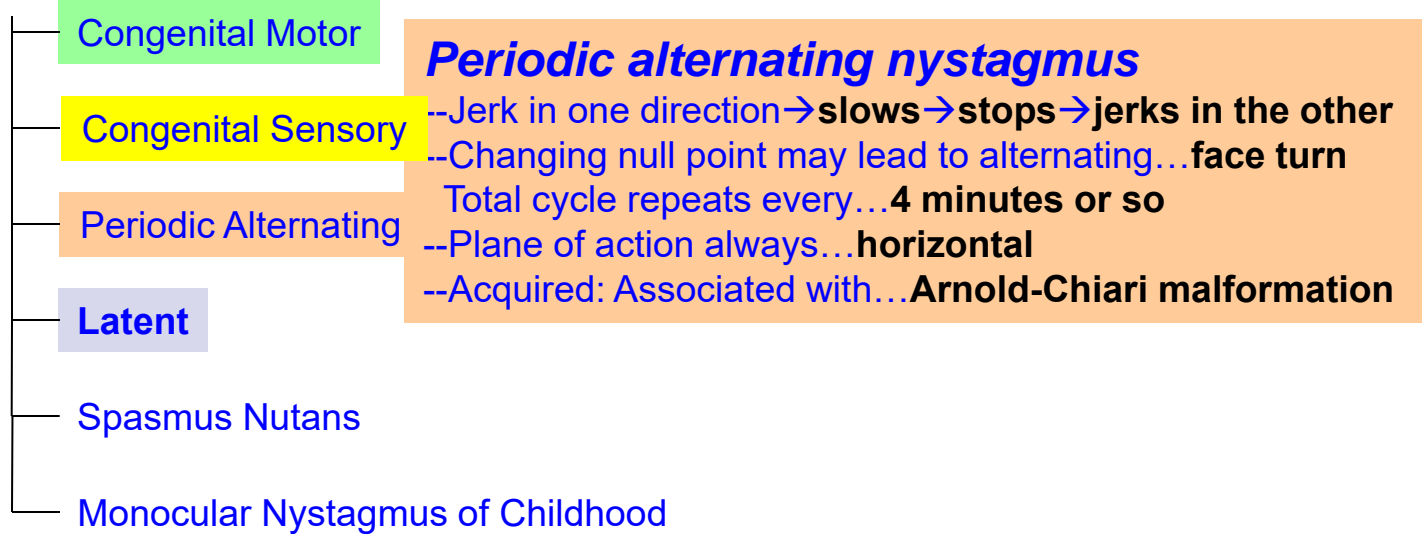
Latent nystagmus
 --Occurs when one eye is...*[vision status]*

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Nystagmus

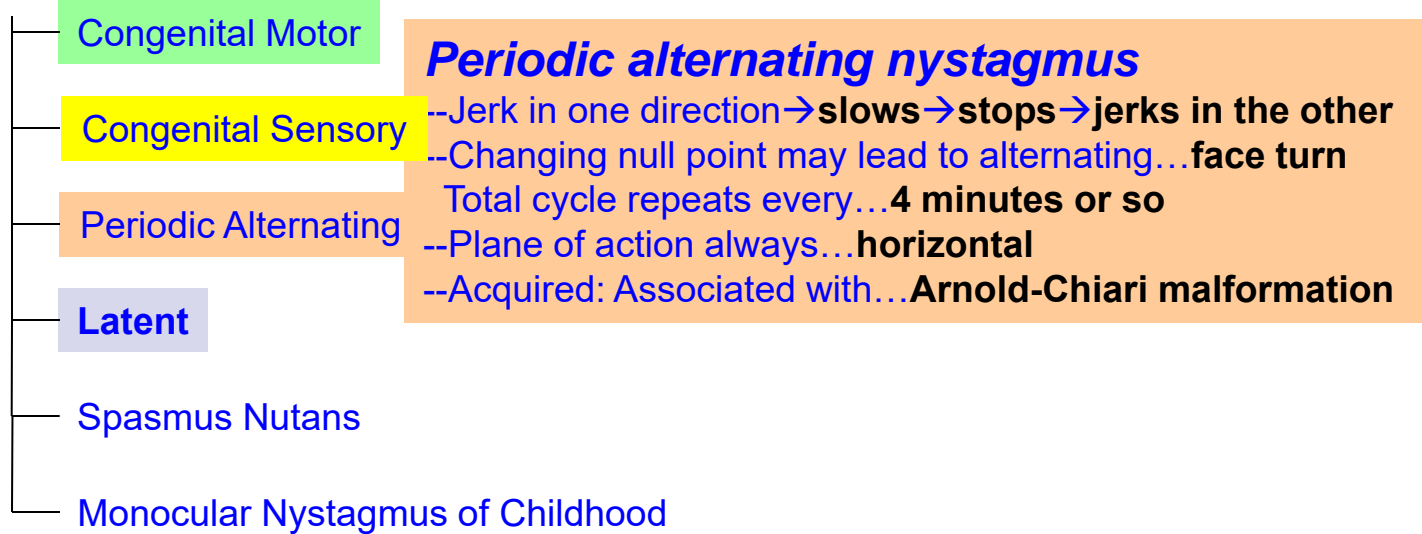
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Childhood



Nystagmus

Latent nystagmus

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Congenital motor nystagmus

- Usually...horizontal
- Remains horizontal in up/downgaze
- Vision usually...good
- Nystagmus + good VA = congenital motor
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Sensory nystagmus

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Periodic alternating nystagmus

What ubiquitous exam component is likely to be spuriously poor in latent nystagmus?

in the other face turn

malformation

Childhood

Congenital Motor

Spasmus Nutans

Monocular Nystagmus of Childhood

agmus

A

Nystagmus

Latent nystagmus

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Sensory nystagmus

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- Waveform depends on visual acuity:
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Periodic alternating nystagmus

What ubiquitous exam component is likely to be spuriously poor in latent nystagmus?
 Occluding one eye to assess visual acuity will induce nystagmus in the eye getting the VA check, resulting in an inaccurately low Snellen acuity

in the other face turn

malformation

Childhood

Congenital Motor

Spasmus Nutans

Monocular Nystagmus of Childhood

Nystagmus

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Periodic alternating nystagmus

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How can you get around this problem?

Childhood

Congenital Motor

Spasmus Nutans

Monocular Nystagmus of Childhood

in the other face turn

malformation

Nystagmus

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Congenital Motor

Periodic alternating nystagmus
 ...in the other
 face turn
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 Occluding one eye to assess visual acuity will induce nystagmus in the eye getting the VA check, resulting in an inaccurately low Snellen acuity

How can you get around this problem?
 Have the patient keep both eyes open, but use plus lenses to fog the fellow eye

- Spasmus Nutans
- Monocular Nystagmus of Childhood

Nystagmus

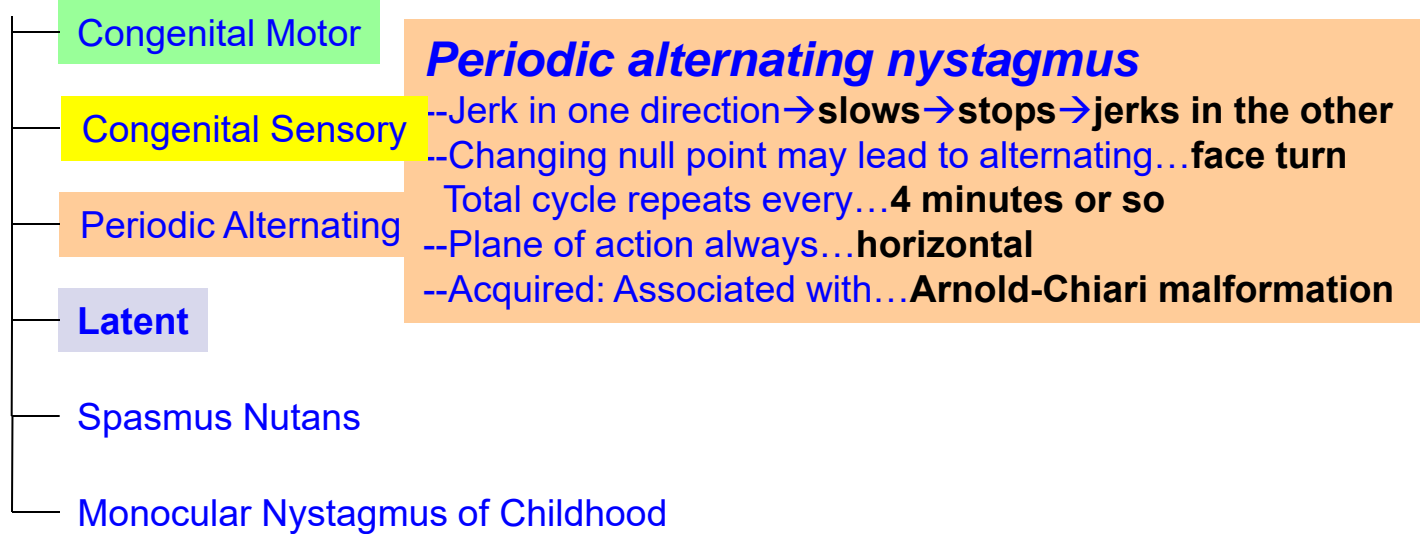
Latent nystagmus
 --Occurs when one eye is... **occluded**
 --Jerk nystagmus toward... [direction]

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Latent nystagmus

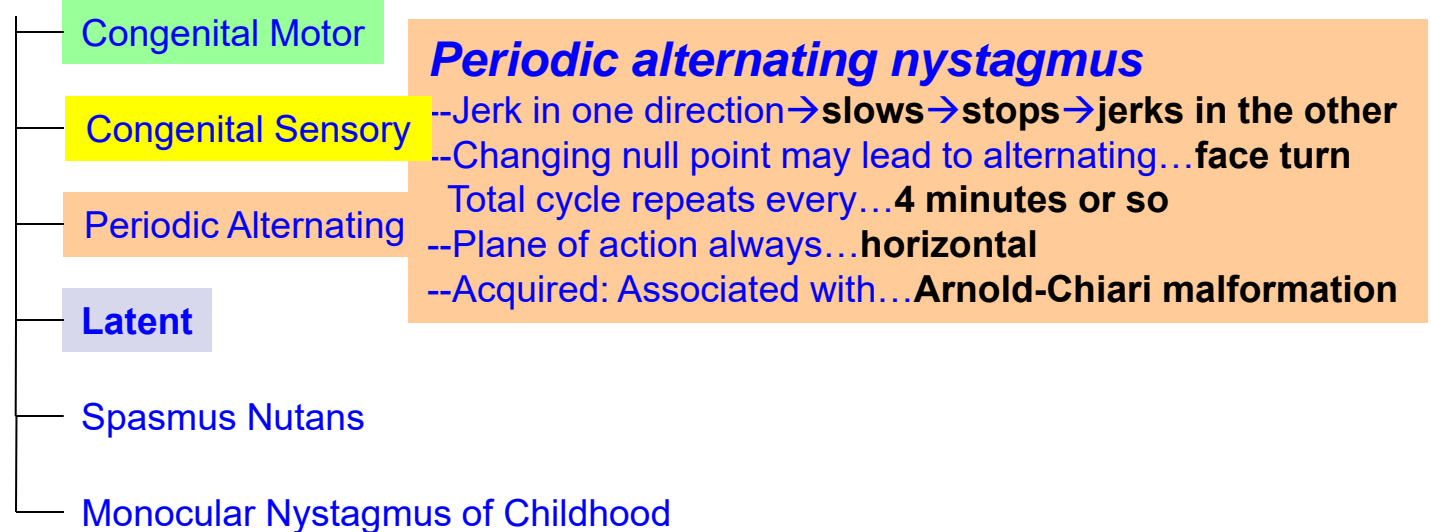
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Nystagmus

Sensory nystagmus

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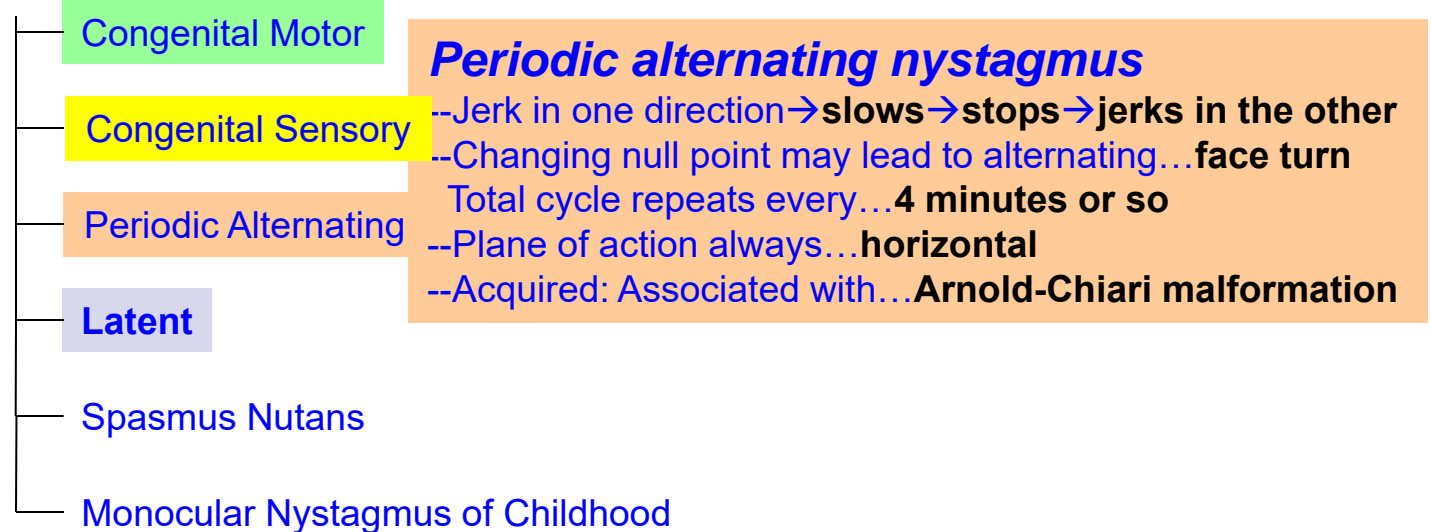
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agmus

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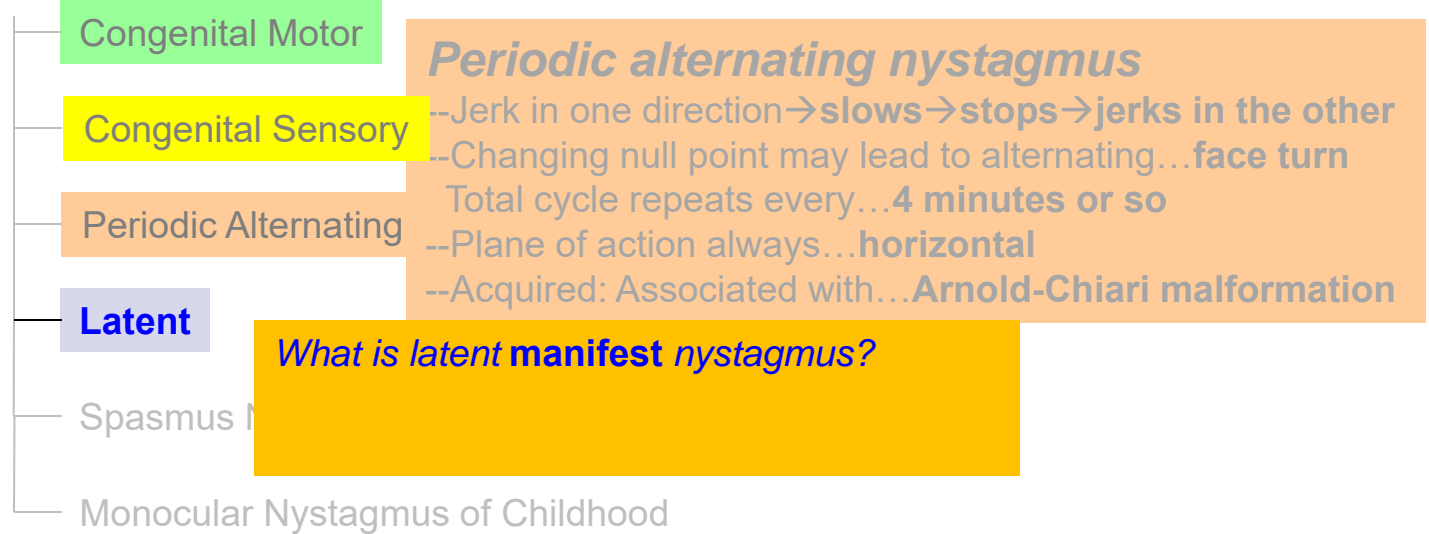
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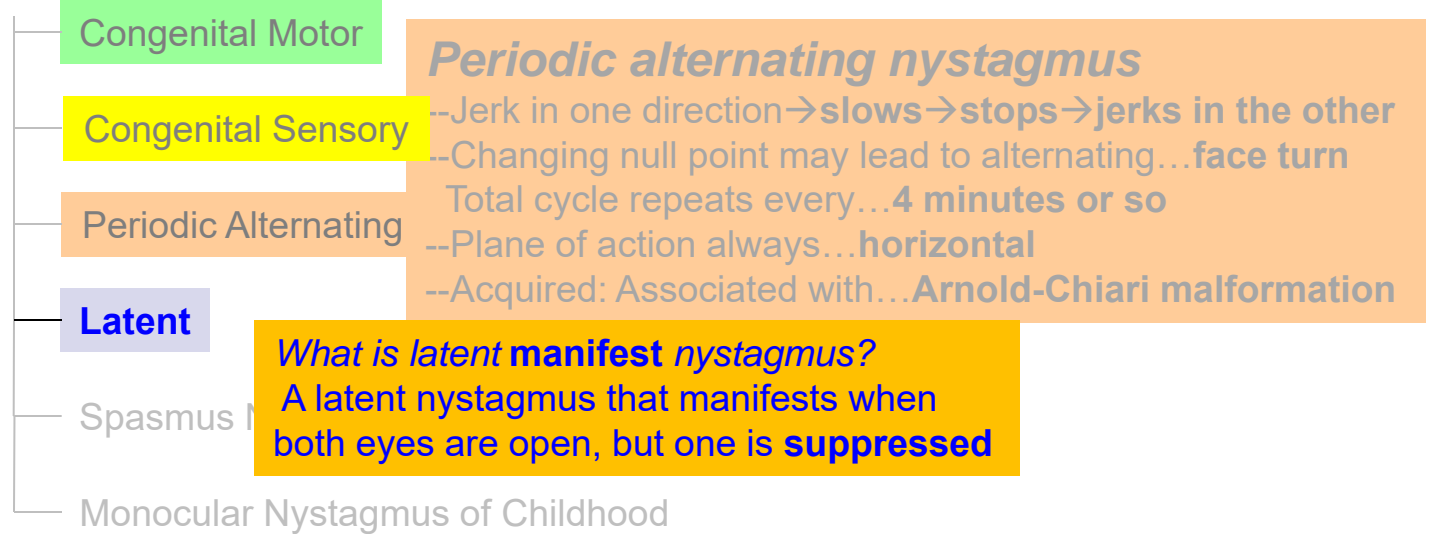
Latent nystagmus
 --Occurs when one eye is...occluded
 --Jerk nystagmus toward...fixating eye
 --Only nystagmus to change direction with fixation

Nystagmus

Sensory nystagmus
 --2ndry to early bilateral poor vision
 --Waveform depends on visual acuity:
 --20/60 – 20/100: **Jerk**
 --20/100 – 20/200: **Pendular**
 --<20/200: **'Searching'**

Childhood

A



Nystagmus

Congenital motor nystagmus

- Usually...horizontal
- Remains horizontal in up/downgaze
- Vision usually...good
- Nystagmus + good VA = *congenital motor*
- Only form with...*paradoxical OKN response*
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Nystagmus

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Childhood

Adulthood

Congenital Motor

Periodic alternating nystagmus

What is 'suppression' in this context?

Spasmus

What is latent manifest nystagmus?

A latent nystagmus that manifests when both eyes are open, but one is suppressed

Monocular Nystagmus of Childhood

Other
rn
ation

Nystagmus

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Childhood

Adulthood

Congenital Motor

Periodic alternating nystagmus

What is 'suppression' in this context?

It is the prevention of an image in one eye from reaching conscious awareness

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Spasmus

Monocular Nystagmus of Childhood

Other
rn
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Nystagmus

Sensory nystagmus

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Childhood

A

Congenital Motor

Periodic alternating nystagmus

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How does the phenomenon of suppression come about?

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Spasmus l

Monocular Nystagmus of Childhood

other
rn
ation

Nystagmus

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Latent nystagmus

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Nystagmus

Sensory nystagmus

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Childhood

Adulthood

Congenital Motor

Periodic alternating nystagmus

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Spasmus I

Monocular Nystagmus of Childhood

Other
rn
ation

Nystagmus

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Nystagmus

Sensory nystagmus

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Childhood

A

Visual confusion *and* diplopia? Aren't those the same thing?

it is one of three sensory adaptations the visual system employs to avoid the occurrence of **visual confusion and/or diplopia**

What is latent manifest nystagmus?
A latent nystagmus that manifests when both eyes are open, but **one is suppressed**

Spasmus I
Monocular Nystagmus of Childhood

Nystagmus

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Nystagmus

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Childhood

A

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 No, not at all

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Monocular Nystagmus of Childhood

Nystagmus

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Childhood

A

Visual confusion **and** diplopia? Aren't those the same thing?

No, not at all

OK, what are they?

Visual confusion is...

Diplopia is...

it is one of three sensory adaptations the visual system employs to avoid the occurrence of **visual confusion and/or diplopia**

What is latent manifest nystagmus?

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Spasmus I
Monocular Nystagmus of Childhood

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Nystagmus

Sensory nystagmus

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Childhood

Adulthood

Visual confusion **and** diplopia? Aren't those the same thing?
 No, not at all

OK, what are they?

Visual confusion is...the visual impression of **two** objects occupying a **single** location in visual space

Diplopia is...the visual impression of **one** object occupying **two** locations in visual space

It is one of three sensory adaptations the visual system employs to avoid the occurrence of **visual confusion and/or diplopia**

What is latent manifest nystagmus?

A latent nystagmus that manifests when both eyes are open, but **one is suppressed**

Monocular Nystagmus of Childhood

Nystagmus

Congenital motor nystagmus
 --Usually...horizontal
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 --Only form with...paradoxical OKN response
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Latent nystagmus
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Nystagmus

Sensory nystagmus
 --2ndry to early bilateral poor vision
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Childhood

What are the other two sensory adaptations the visual system employs to avoid the occurrence of visual confusion and/or diplopia?
 --Suppression
 --?
 --?
 Mnemonic is...

How does the phenomenon of suppression come about?
 It is one of three sensory adaptations the visual system employs to avoid the occurrence of visual confusion and/or diplopia

What is latent manifest nystagmus?
 A latent nystagmus that manifests when both eyes are open, but one is suppressed

- Spasmus I
- Monocular Nystagmus of Childhood

Nystagmus

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Nystagmus

Sensory nystagmus

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Childhood

What are the other two sensory adaptations the visual system employs to avoid the occurrence of visual confusion and/or diplopia?

- S**uppression
- A**
- M**

Mnemonic is...SAM

How does the phenomenon of suppression come about?

It is one of **three sensory adaptations** the visual system employs to avoid the occurrence of visual confusion and/or diplopia

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Spasmus

Monocular Nystagmus of Childhood

Nystagmus

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Nystagmus

Sensory nystagmus

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Childhood

What are the other two sensory adaptations the visual system employs to avoid the occurrence of visual confusion and/or diplopia?

- Suppression
- Anomalous retinal correspondence (ARC)
- Monofixation syndrome

How does the phenomenon of suppression come about?

It is one of three sensory adaptations the visual system employs to avoid the occurrence of visual confusion and/or diplopia

What is latent manifest nystagmus?

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Spasmus

Monocular Nystagmus of Childhood

Nystagmus

Latent nystagmus

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- ### Congenital motor nystagmus
- Usually...horizontal
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- ### Sensory nystagmus
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 - 20/60 – 20/100: **Jerk**
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Childhood

Adulthood

Congenital Motor

- ### Periodic alternating nystagmus
- Jerk in one direction → slows → stops → jerks in the other
 - big null point may lead to alternating...face turn
 - cycle repeats every...4 minutes or so
 - action always...horizontal
 - Acquired: Associated with...Arnold-Chiari malformation

Latent nystagmus and manifest latent nystagmus are sometimes referred to by what single name?

Latent

What is latent manifest nystagmus?
A latent nystagmus that manifests when both eyes are open, but one is suppressed

Spasmus

Monocular Nystagmus of Childhood

Nystagmus

Latent nystagmus

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Childhood

Adulthood

Congenital Motor

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 - Cycle repeats every...4 minutes or so
 - Action always...horizontal
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Latent nystagmus and manifest latent nystagmus are sometimes referred to by what single name?
Fusion maldevelopment nystagmus syndrome (FMNS)

Latent

What is **latent manifest nystagmus**?
 A latent nystagmus that manifests when both eyes are open, but one is **suppressed**

Spasmus

Monocular Nystagmus of Childhood

Spasmus nutans

--Triad of duh + two words + one word

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
- Likely to have a...**null point**

Latent nystagmus

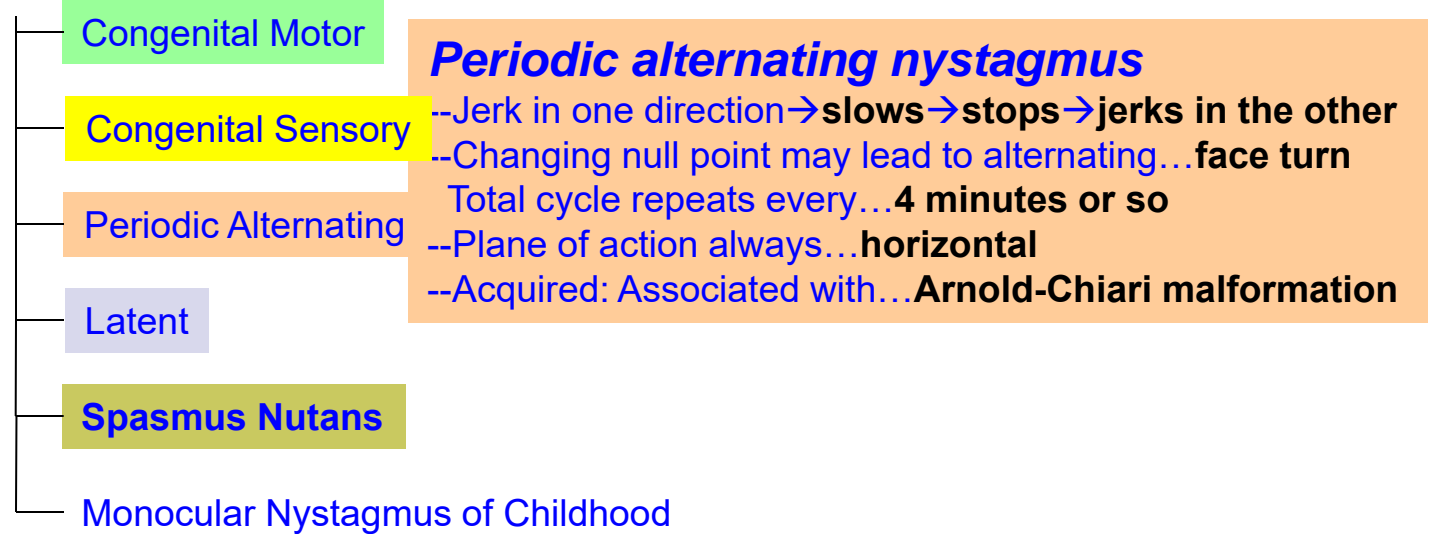
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Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
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Childhood



Spasmus nutans

--Triad of **nystagmus + head nodding + torticollis**

Congenital motor nystagmus

--Usually...**horizontal**
 --Remains horizontal in up/downgaze
 --Vision usually...**good**
 --Nystagmus + good VA = *congenital motor*
 --Only form with...**paradoxical OKN response**
 --Likely to have a...**null point**

Latent nystagmus

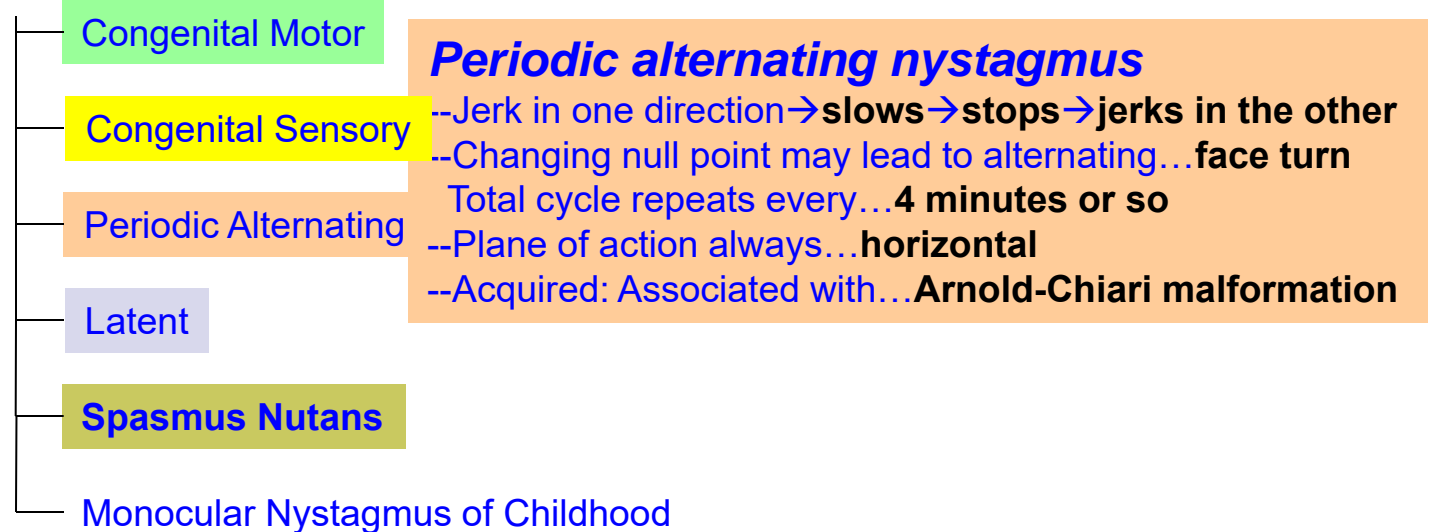
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Nystagmus

Sensory nystagmus

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 --Waveform depends on visual acuity:
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very... classic description

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
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- Only form with...**paradoxical OKN response**
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Latent nystagmus

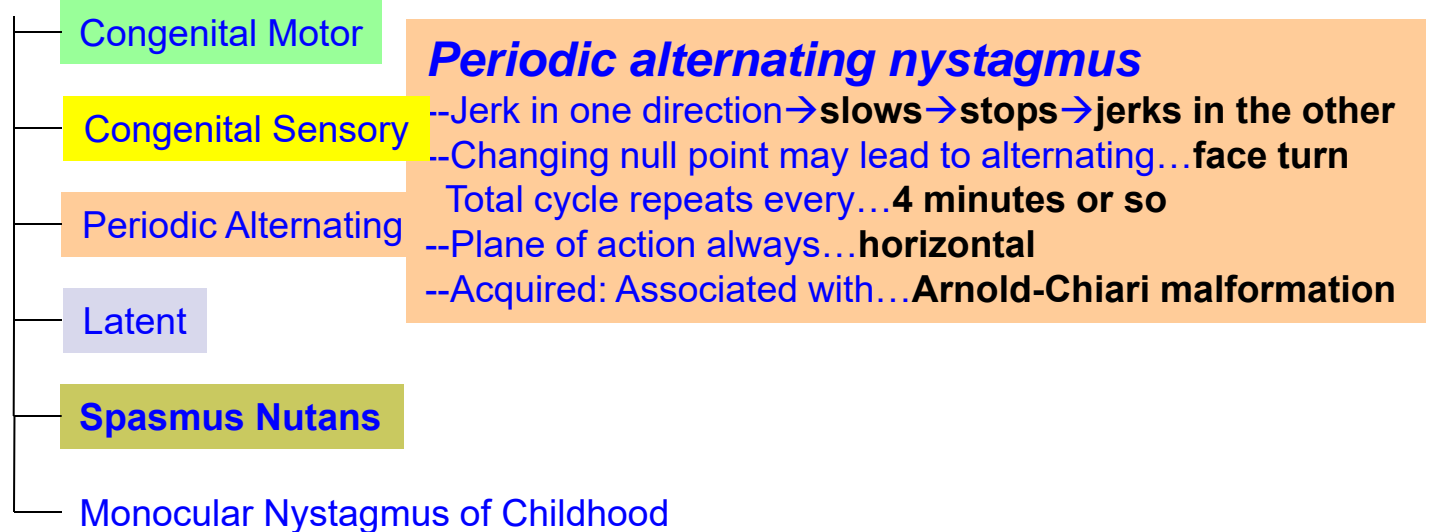
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Nystagmus

Sensory nystagmus

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- Waveform depends on visual acuity:
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...**small** ('shimmer')

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
- Likely to have a...**null point**

Latent nystagmus

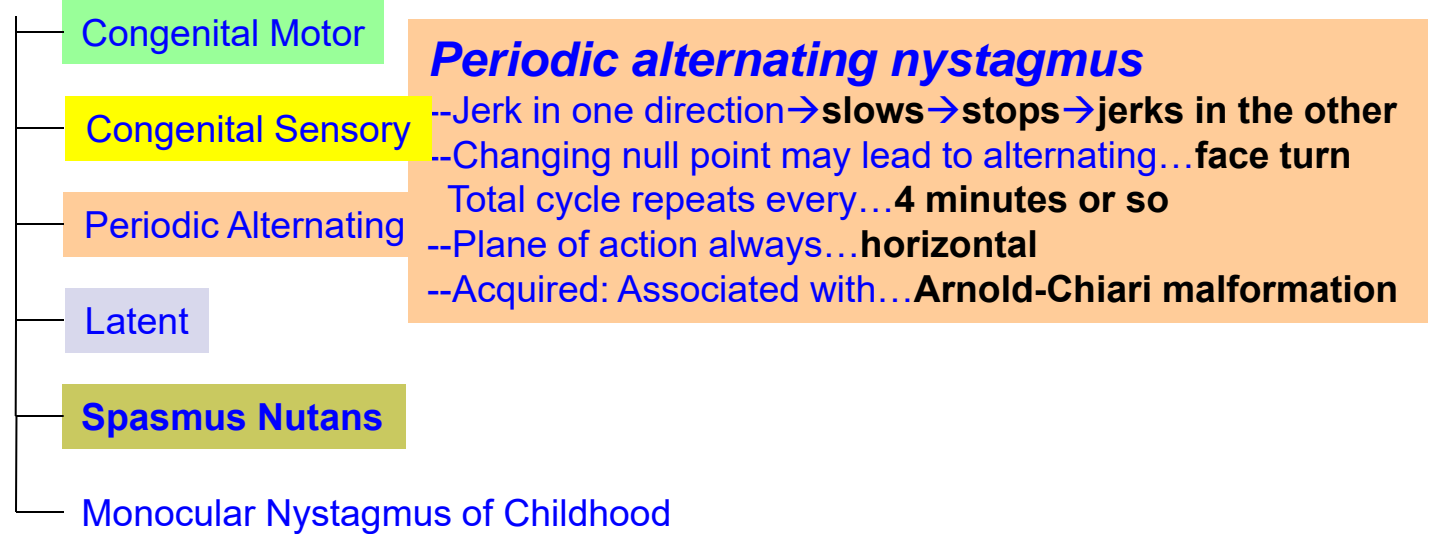
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Nystagmus

Sensory nystagmus

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- Waveform depends on visual acuity:
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very... **small** ('shimmer')
- Usually bilateral vs unilateral but can seem bilateral vs unilateral

Congenital motor nystagmus

- Usually... **horizontal**
- Remains horizontal in up/downgaze
- Vision usually... **good**
- Nystagmus + good VA = *congenital motor*
- Only form with... **paradoxical OKN response**
- Likely to have a... **null point**

Latent nystagmus

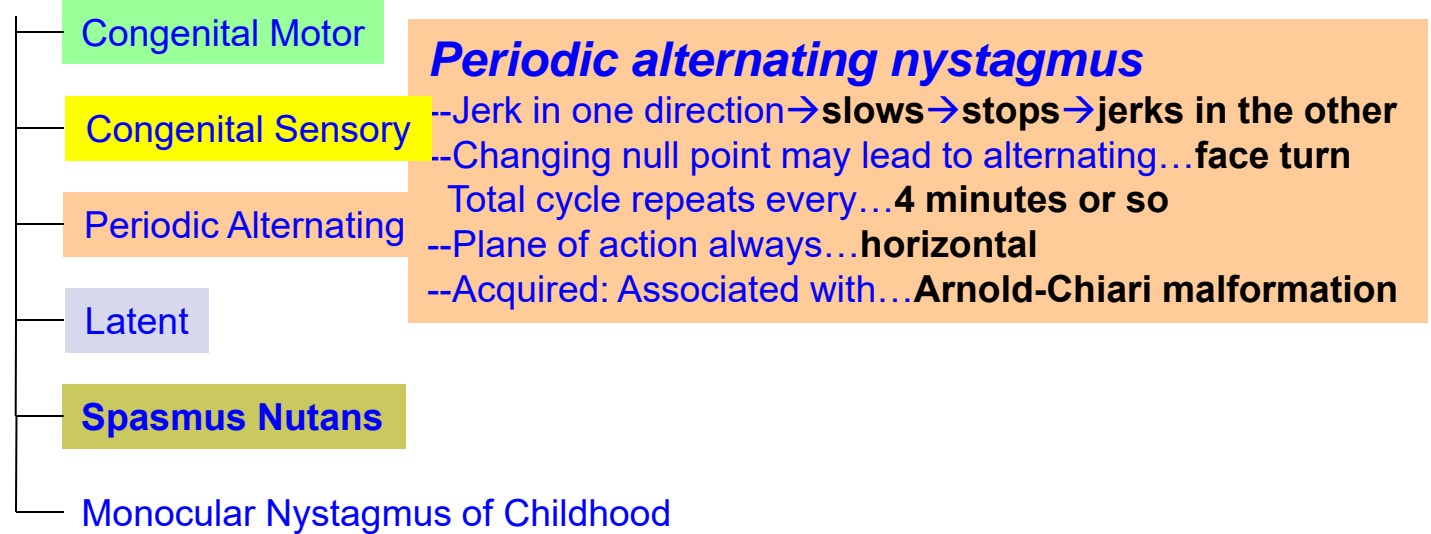
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Nystagmus

Sensory nystagmus

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- Waveform depends on visual acuity:
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...**small** ('shimmer')
- Usually **bilateral** , but can seem **unilateral**

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
- Likely to have a...**null point**

Latent nystagmus

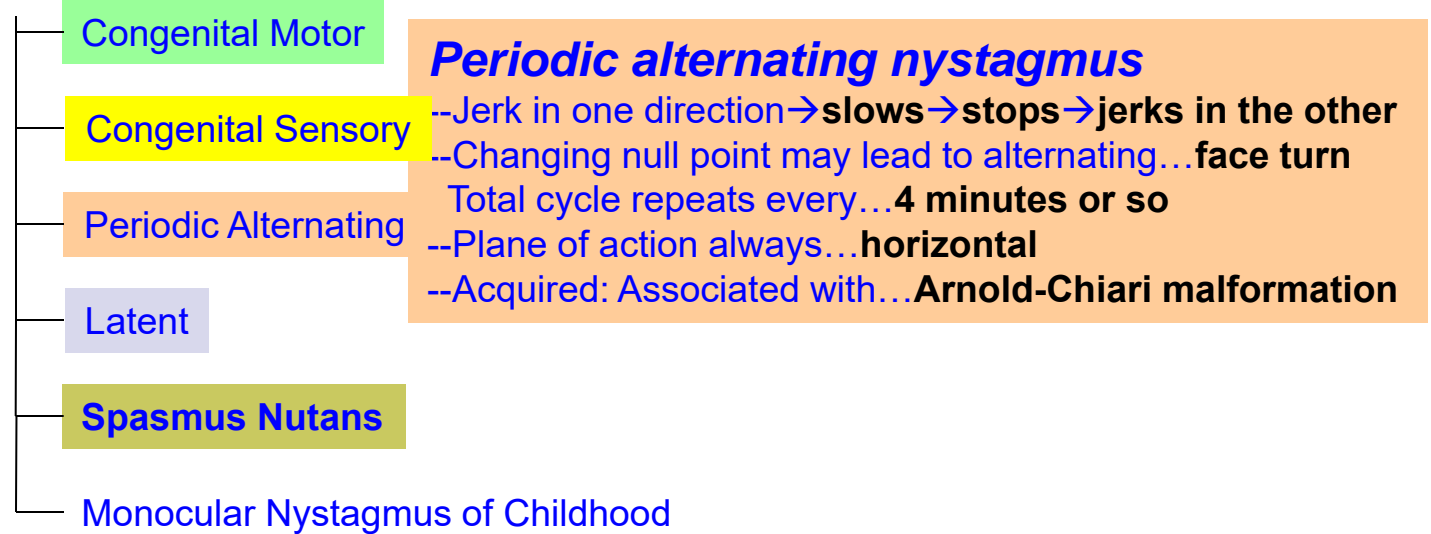
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Nystagmus

Sensory nystagmus

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- Waveform depends on visual acuity:
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...small ('shimmer')
- Usually **bilateral**, but **can seem unilateral**

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- paradoxical OKN response*
- ...null point*

Why the qualifier about it sometimes 'seeming' to be unilateral?

Latent

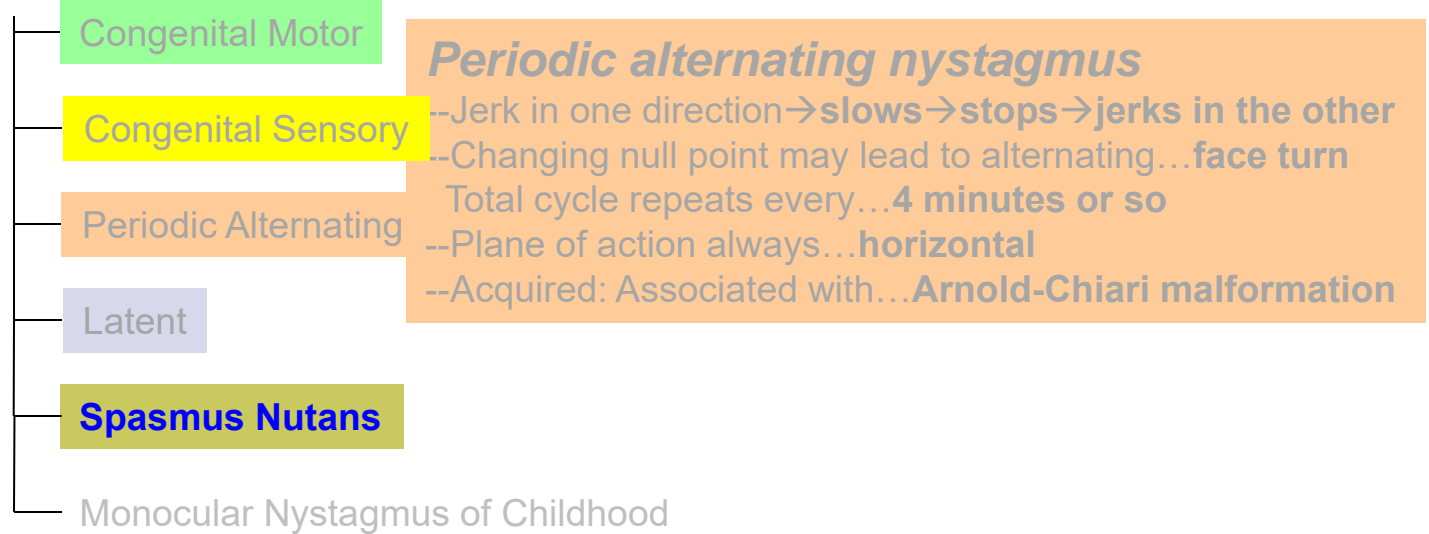
- Occurs
- Jerk ny
- Only nystagmus to change direction with fixation

Sensory nystagmus

- Due to early **bilateral** poor vision
- Waveform depends on visual acuity:
 - 20/60 – 20/100: **Jerk**
 - 20/100 – 20/200: **Pendular**
 - <20/200: '**Searching**'

Childhood

A



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...small ('shimmer')
- Usually **bilateral**, but **can seem unilateral**

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor paradoxical OKN response*
- ...**null point**

Why the qualifier about it sometimes 'seeming' to be unilateral?

Because, while it is a bilateral condition, spasmus nutans can present in highly asymmetric fashion--so much so that involvement of one eye cannot be reliably detected clinically (Note: The importance of this factoid will be made clear in a few slides)

Latent

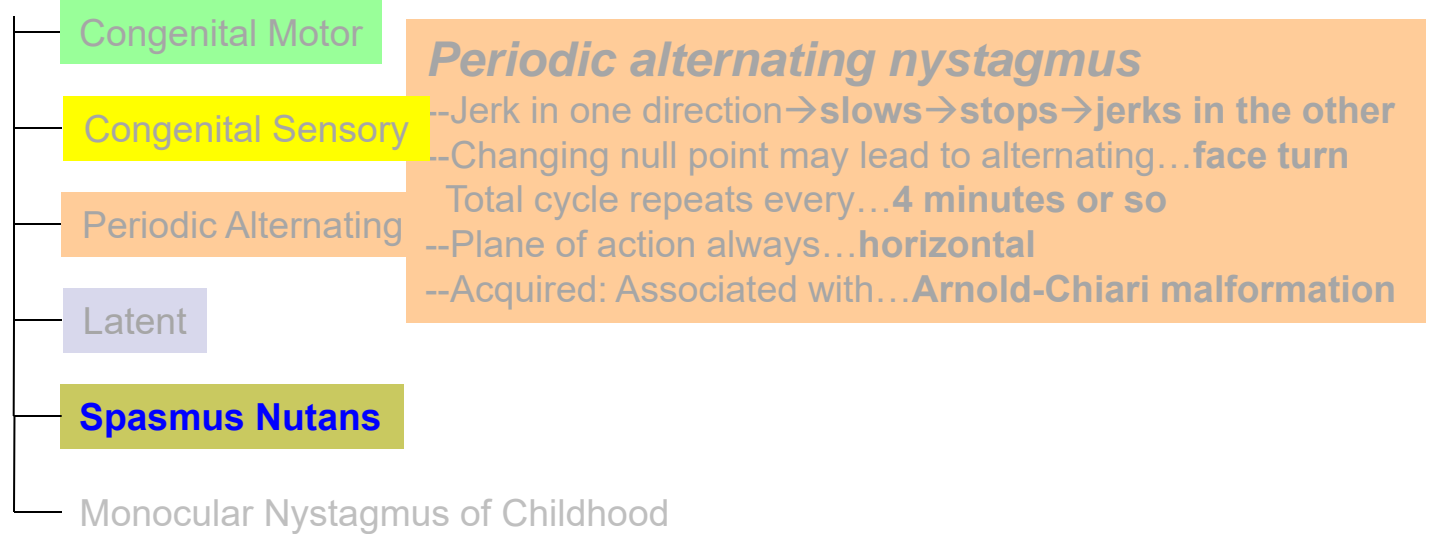
- Occurs
- Jerk ny
- Only nystagmus to change direction with fixation

Acquired sensory nystagmus

- ...to early **bilateral** poor vision
- Waveform depends on visual acuity:
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- <20/200: '**Searching**'

Childhood

A



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...**small** ('shimmer')
- Usually **bilateral**, but can seem **unilateral**

benign vs terrible

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
- Likely to have a...**null point**

Latent nystagmus

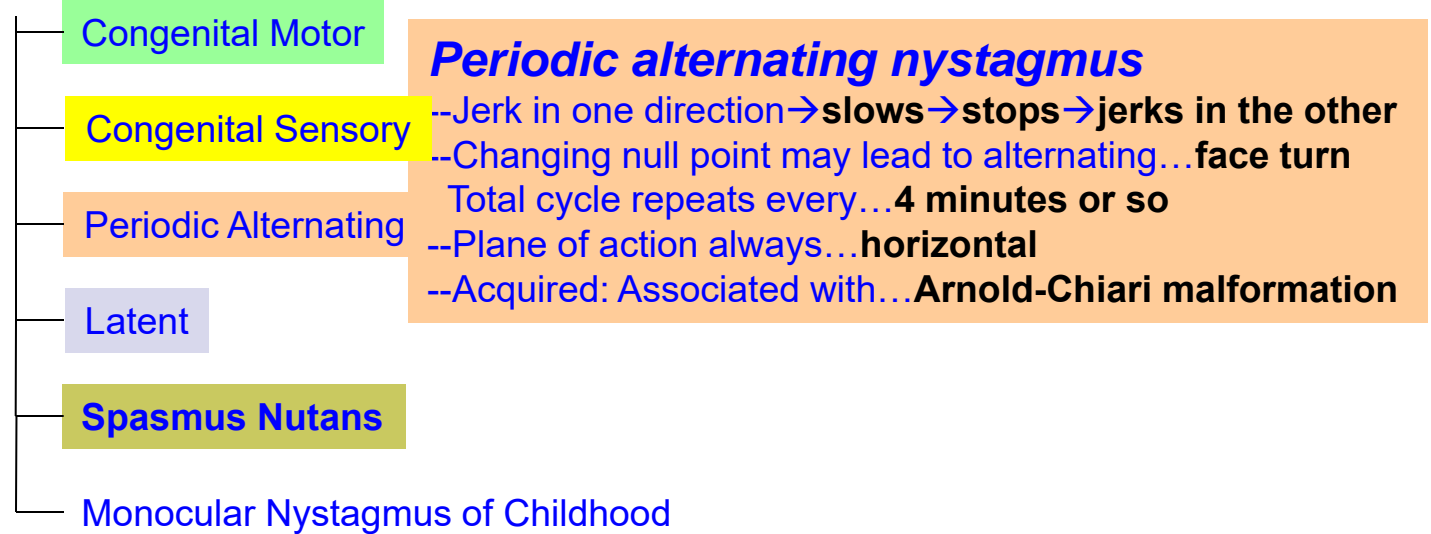
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Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
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Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...**small** ('shimmer')
- Usually **bilateral** , but can seem **unilateral**
- **Benign** .

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
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Latent nystagmus

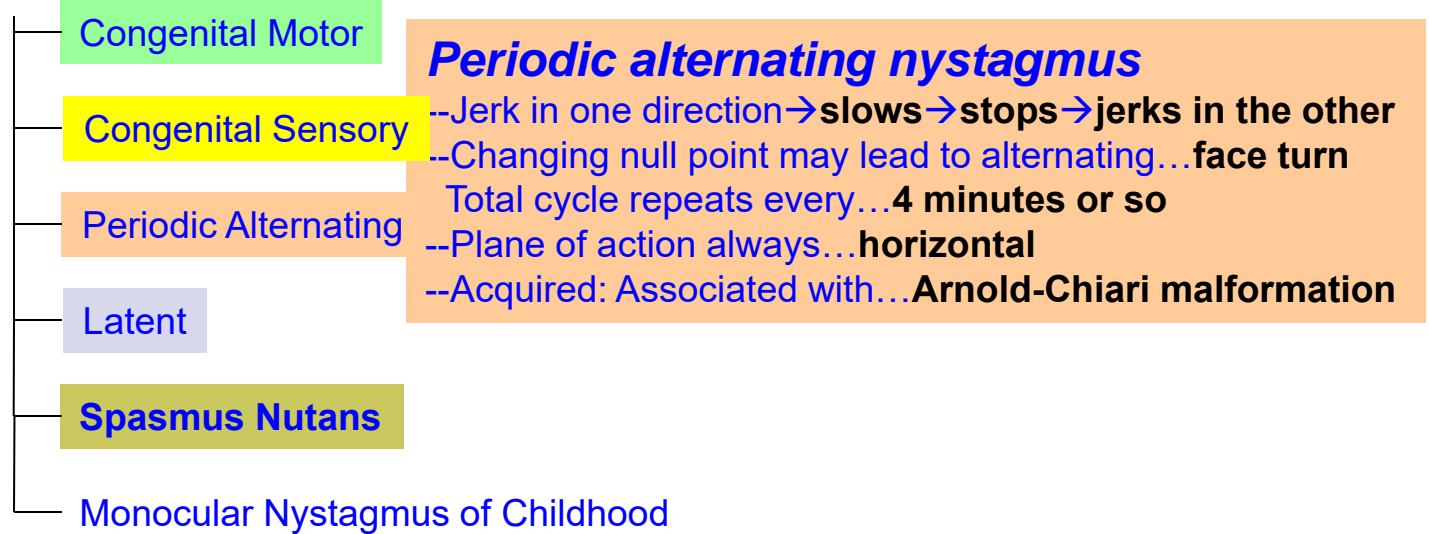
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Nystagmus

Sensory nystagmus

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 - 20/100 – 20/200: **Pendular**
 - <20/200: '**Searching**'

Childhood



Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very...**small** ('shimmer')
- Usually **bilateral**, but can seem **unilateral**
- **Benign**. Resolves by age range.

Congenital motor nystagmus

- Usually...**horizontal**
- Remains horizontal in up/downgaze
- Vision usually...**good**
- Nystagmus + good VA = *congenital motor*
- Only form with...**paradoxical OKN response**
- Likely to have a...**null point**

Latent nystagmus

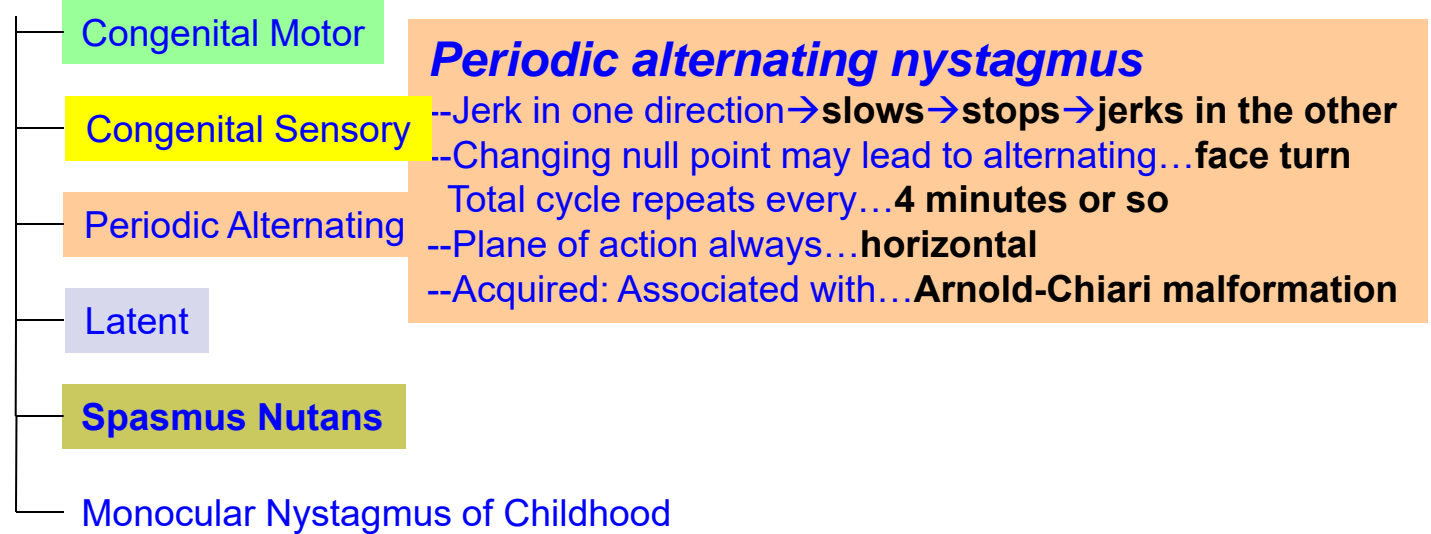
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Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
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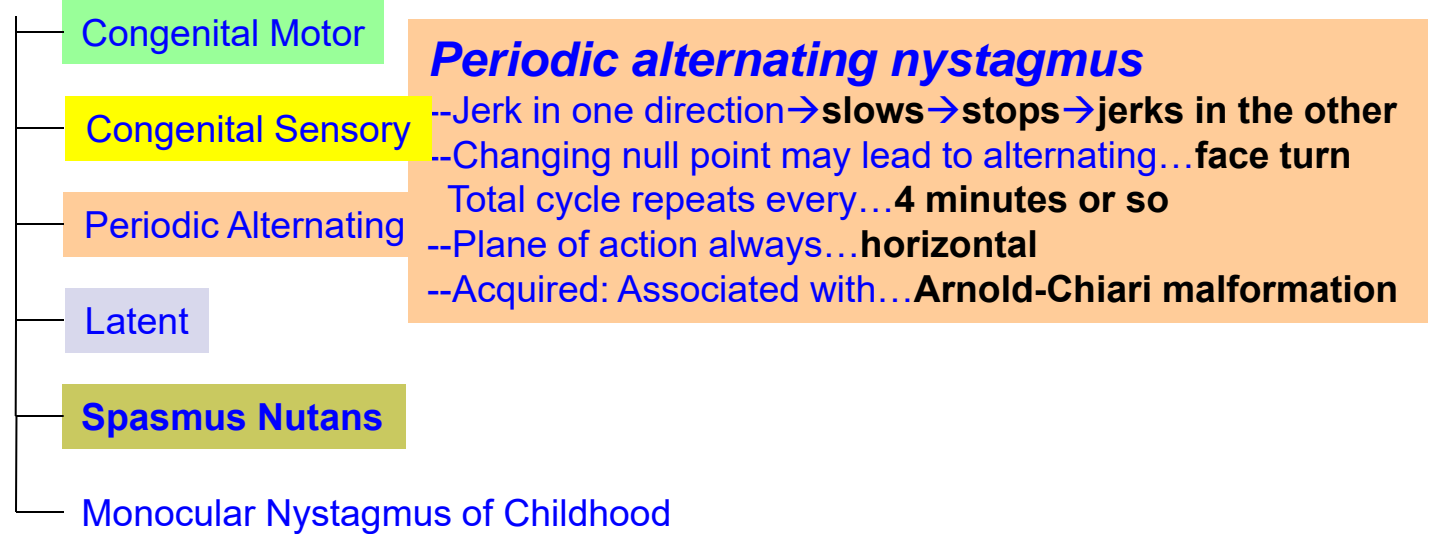
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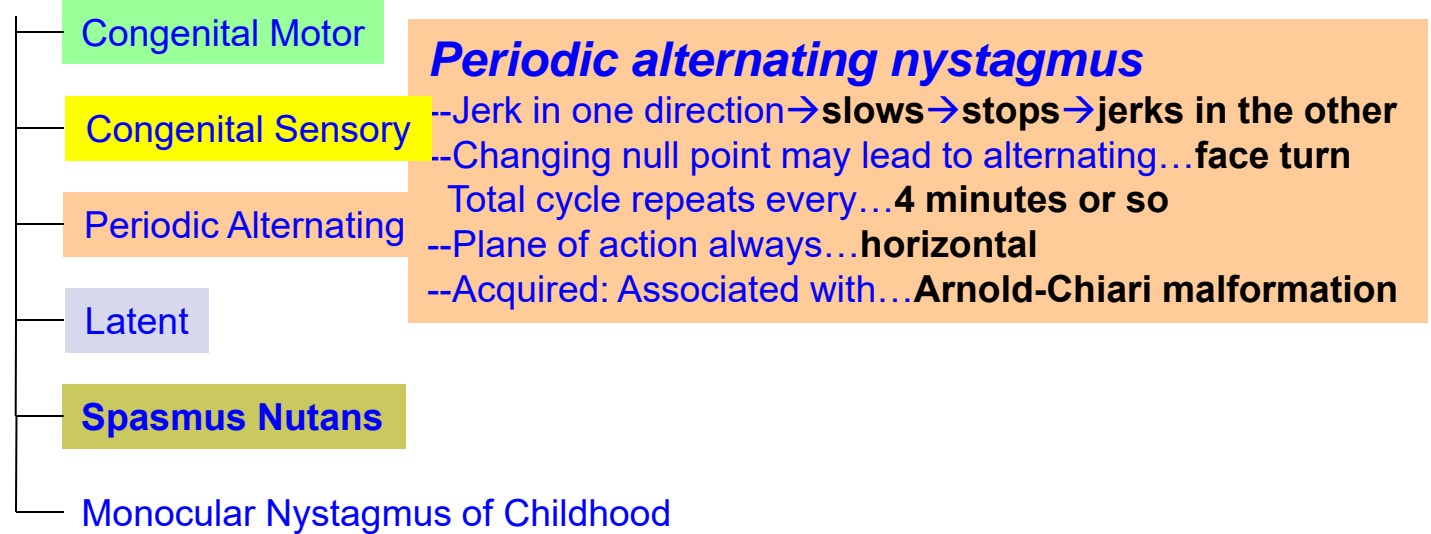
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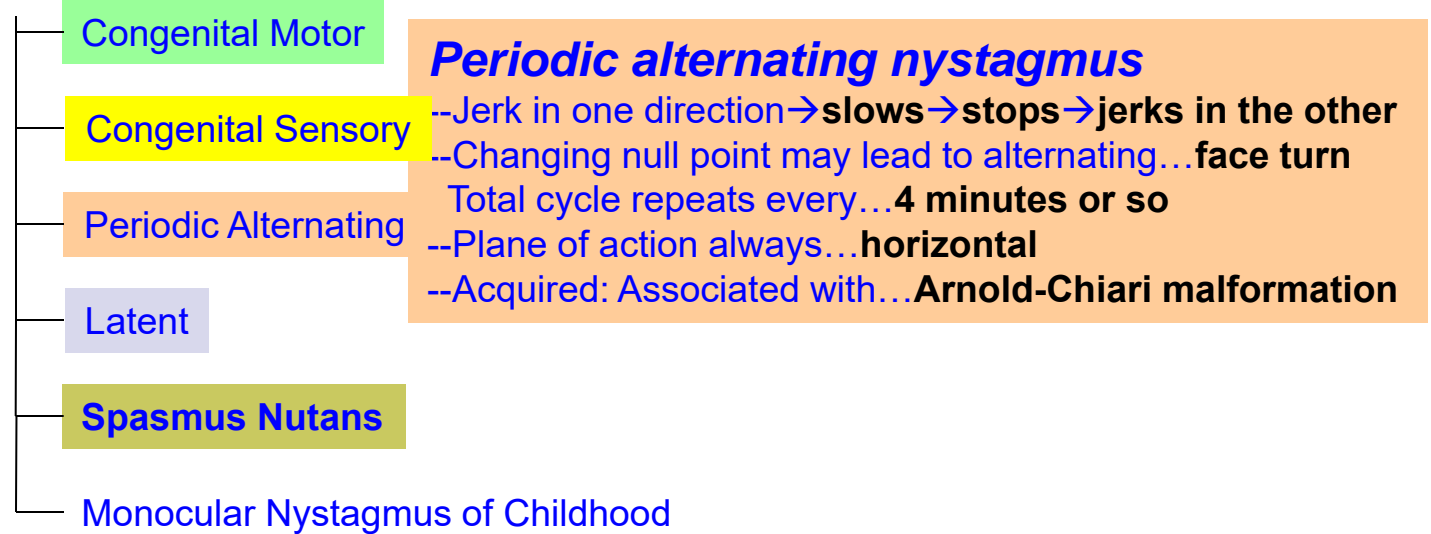
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Nystagmus

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Where might such a glioma be located?

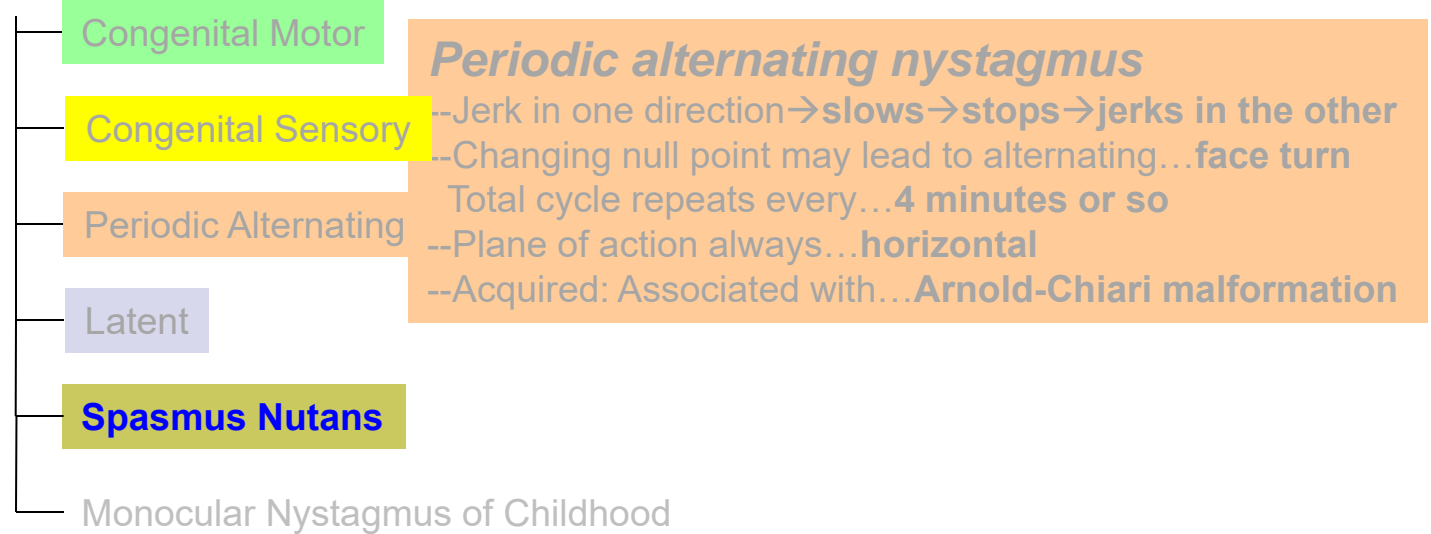
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Where might such a glioma be located?

In the anterior visual pathway, ie, the **two words** or **agnumus**

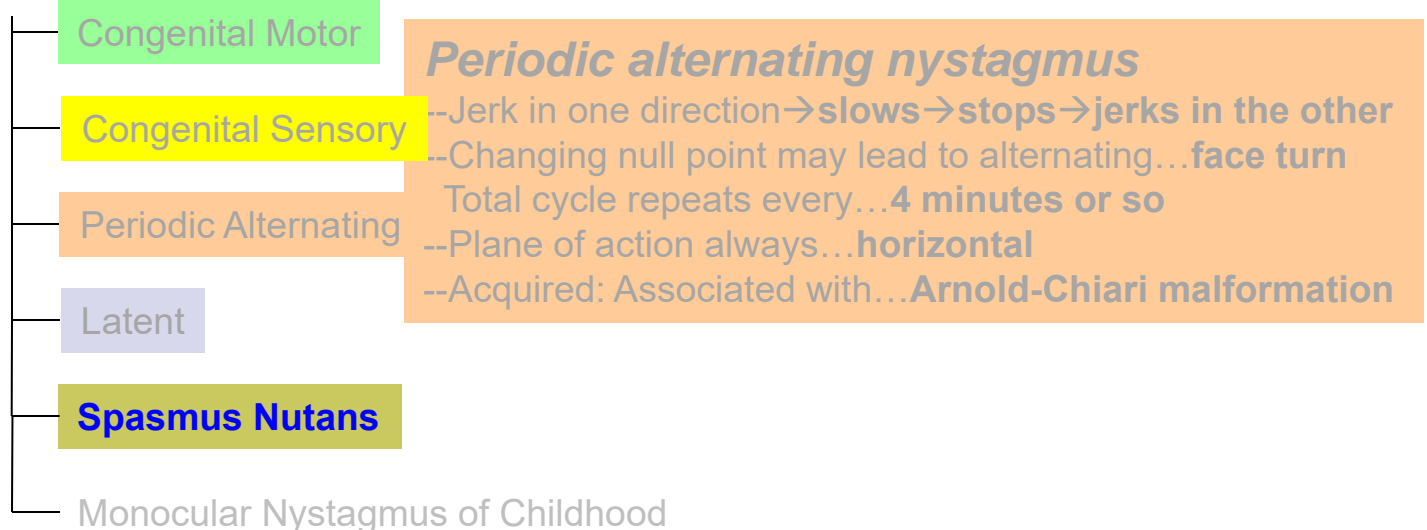
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Childhood

A



- ### Periodic alternating nystagmus
- Jerk in one direction → **slows** → **stops** → jerks in the other
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 - Total cycle repeats every... **4 minutes or so**
 - Plane of action always... **horizontal**
 - Acquired: Associated with... **Arnold-Chiari malformation**

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Where might such a glioma be located?
 In the anterior visual pathway, ie, the optic nerve or chiasm

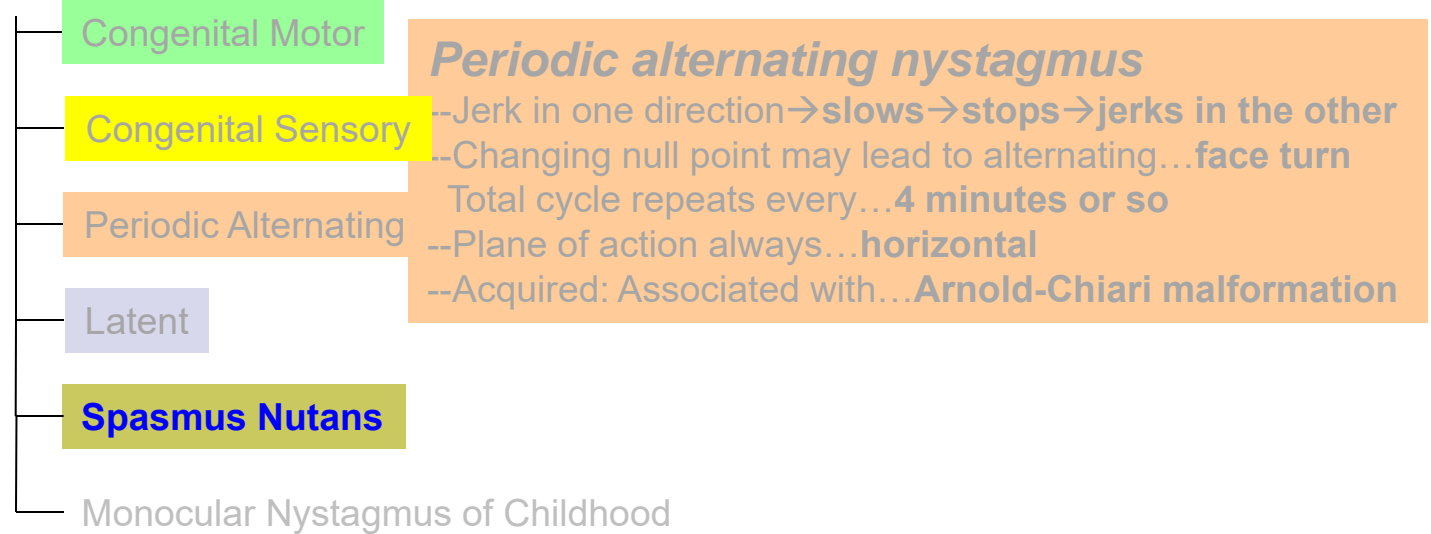
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Latent nystagmus

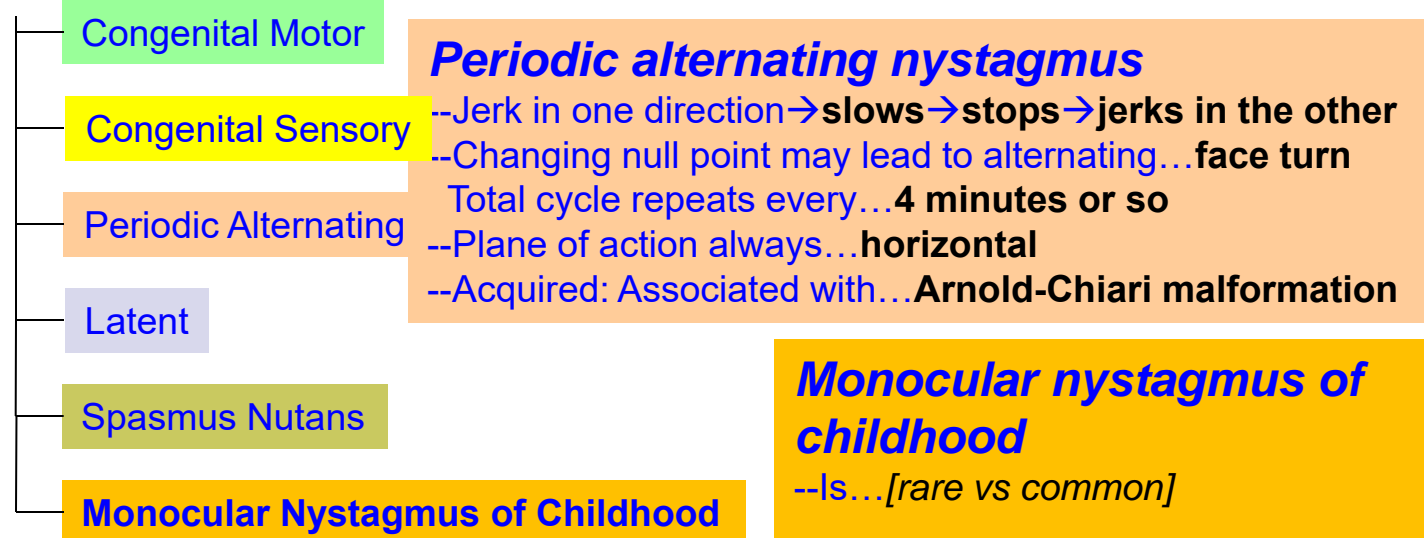
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Monocular nystagmus of childhood

- Is... [rare vs common]

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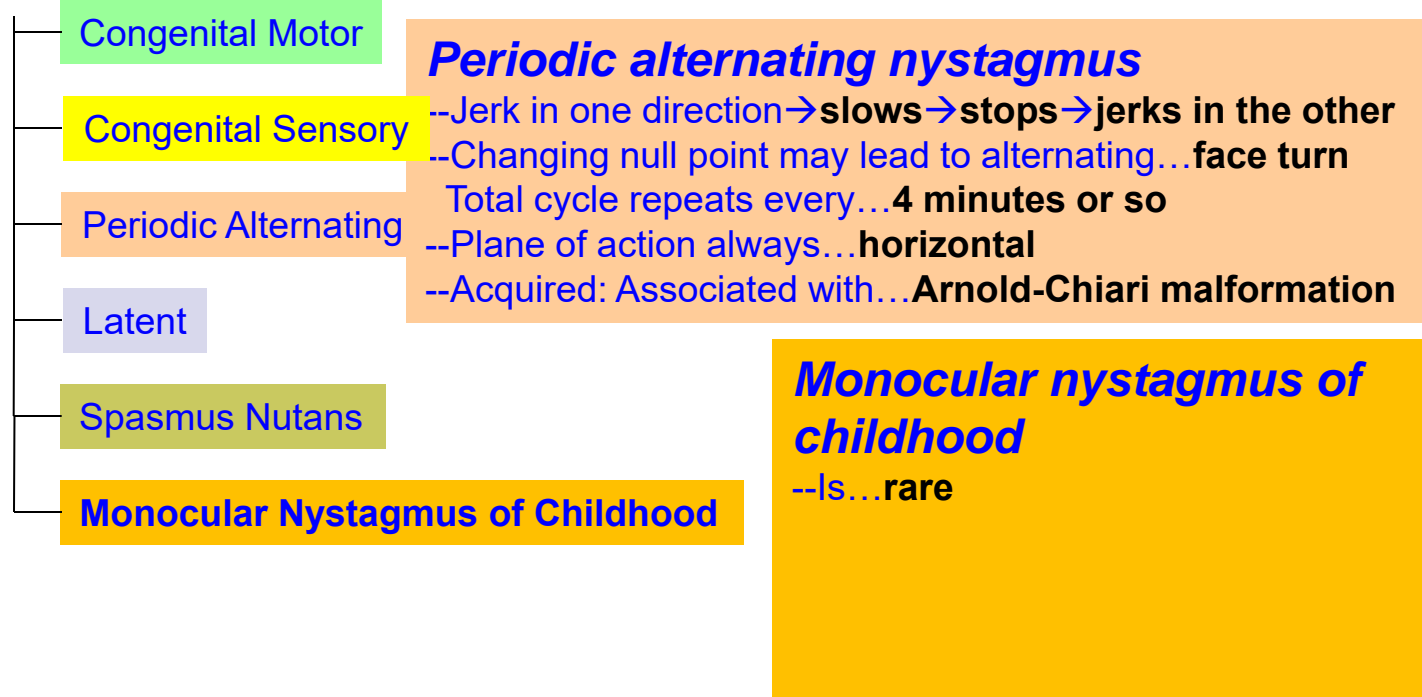
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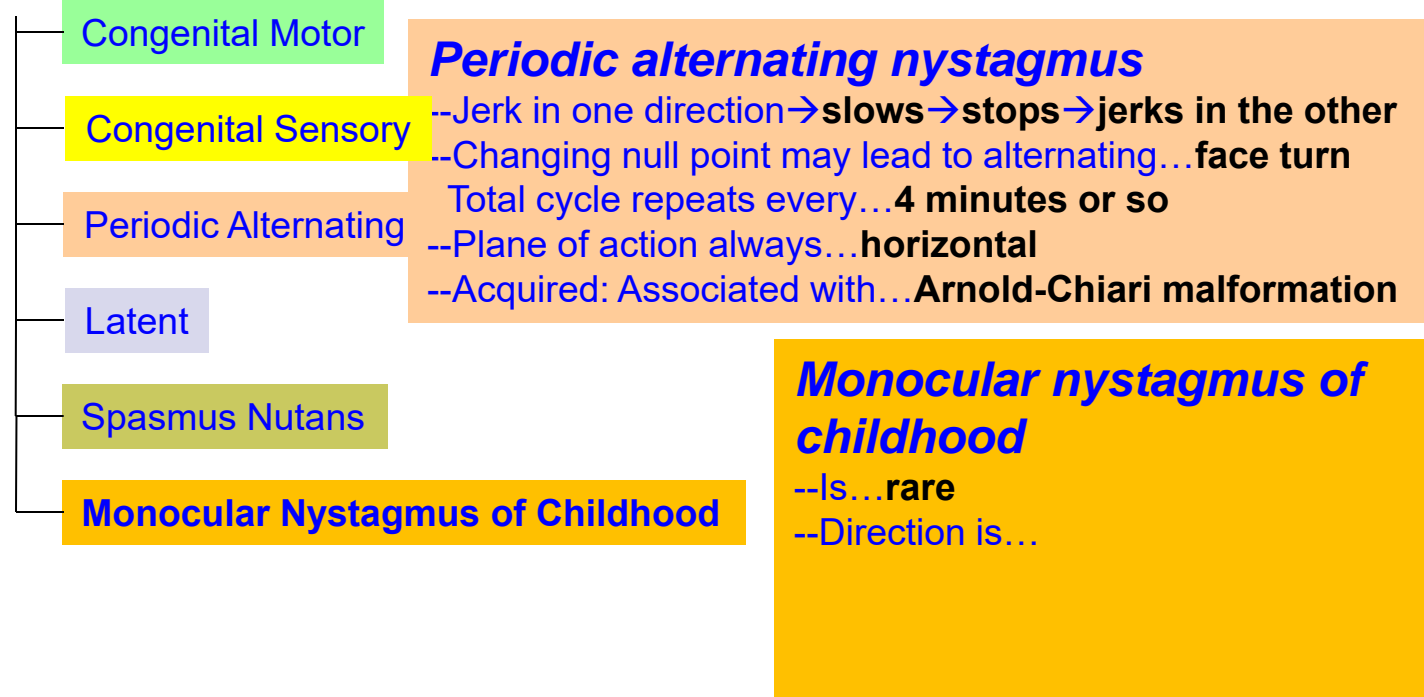
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Latent nystagmus

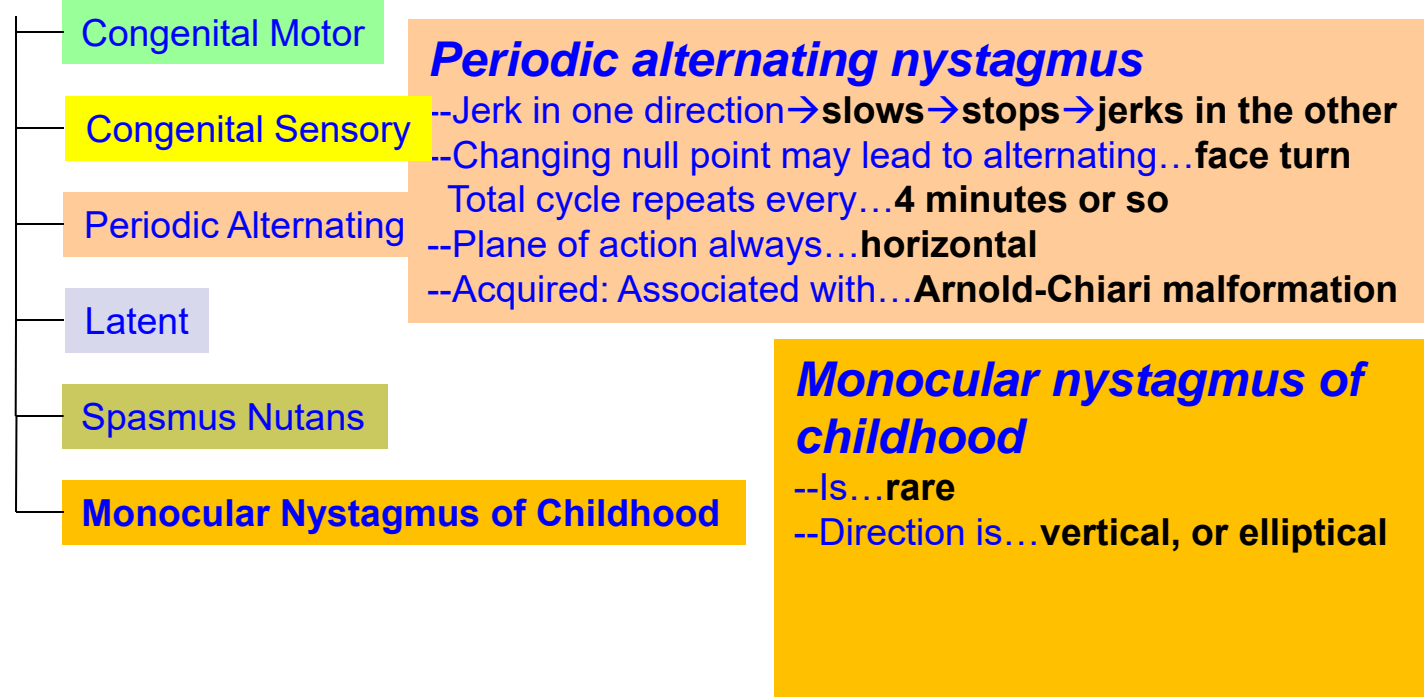
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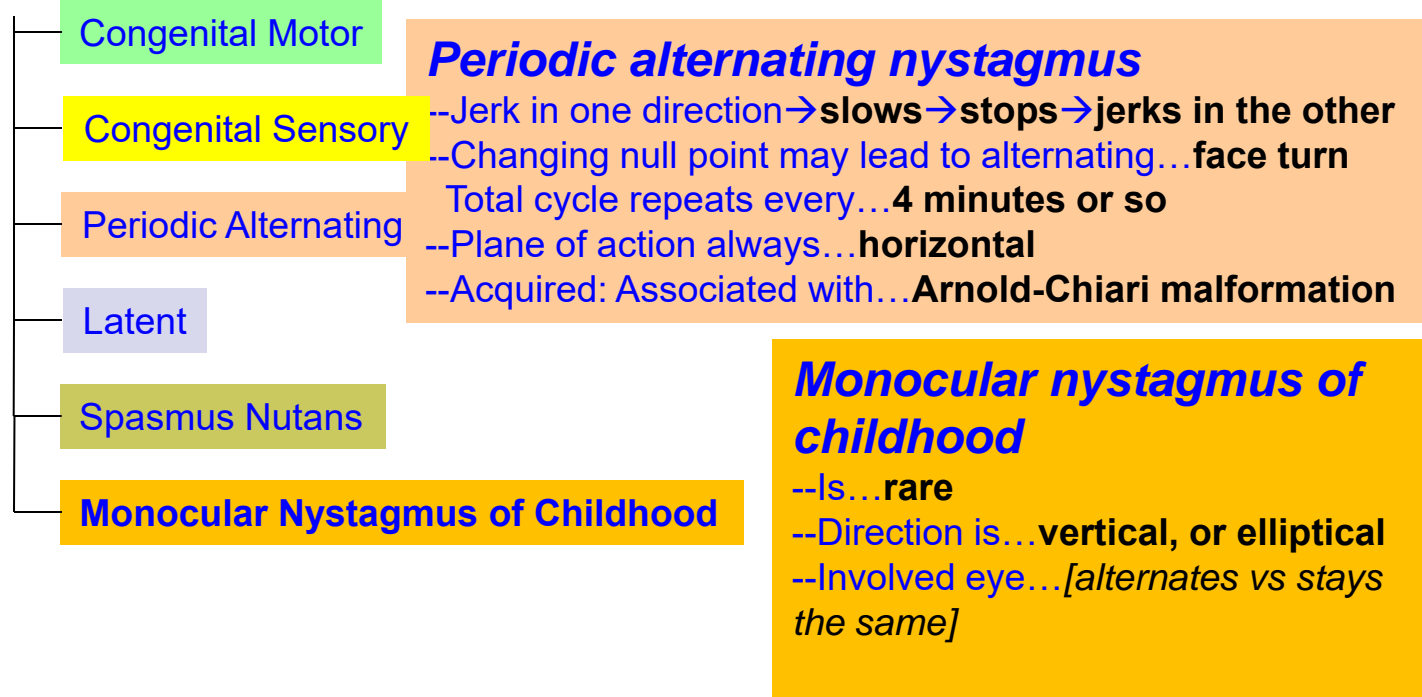
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nystagmus

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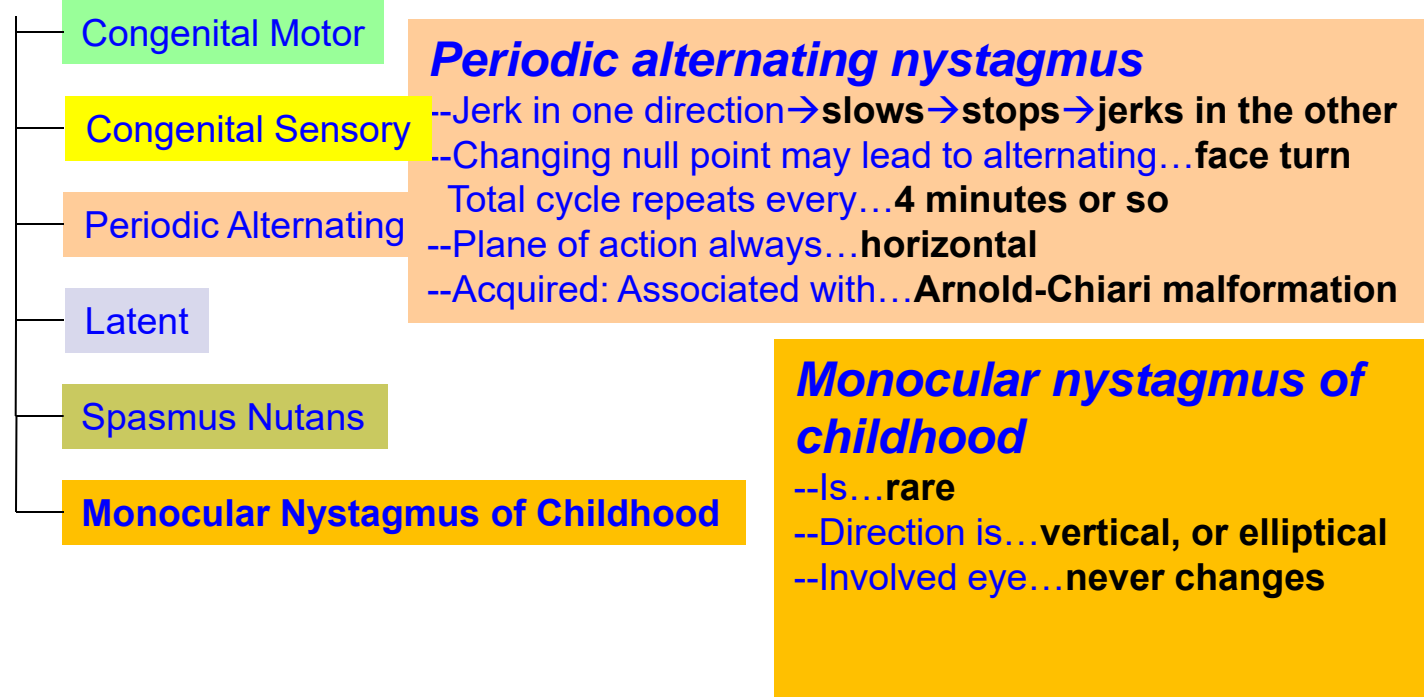
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nystagmus

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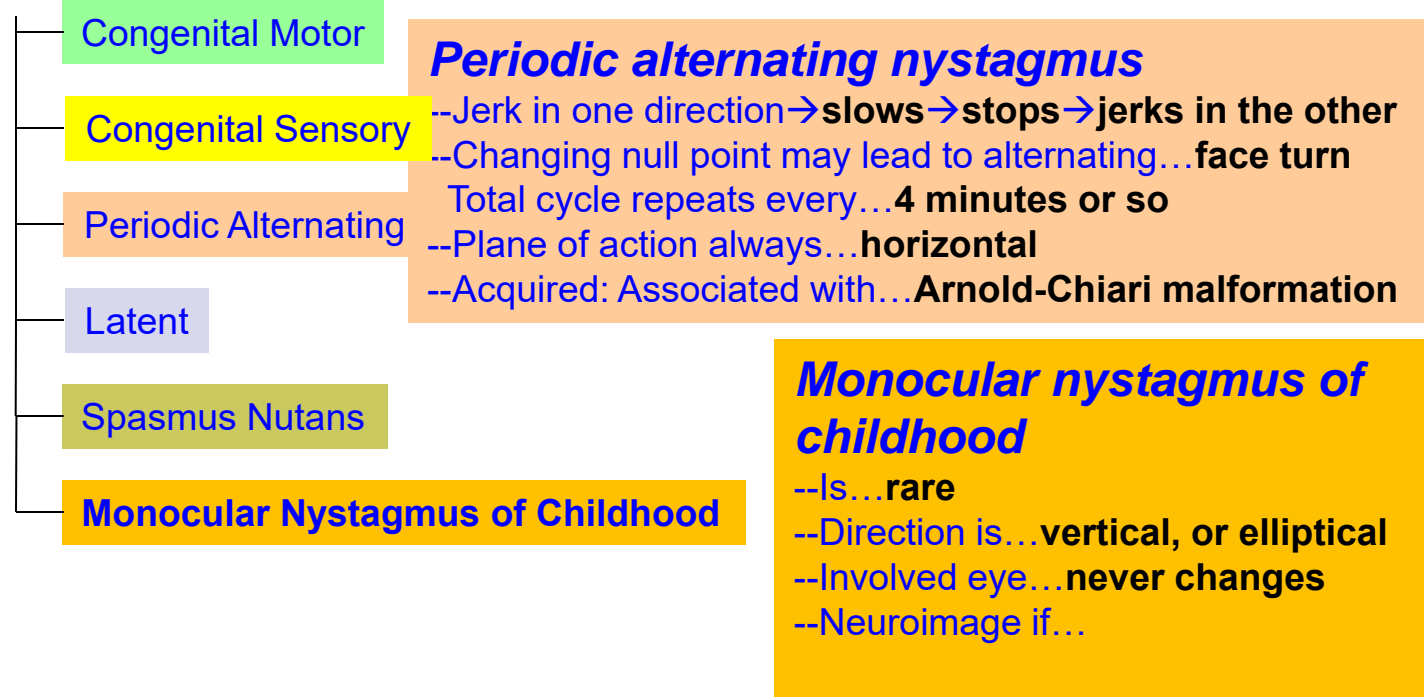
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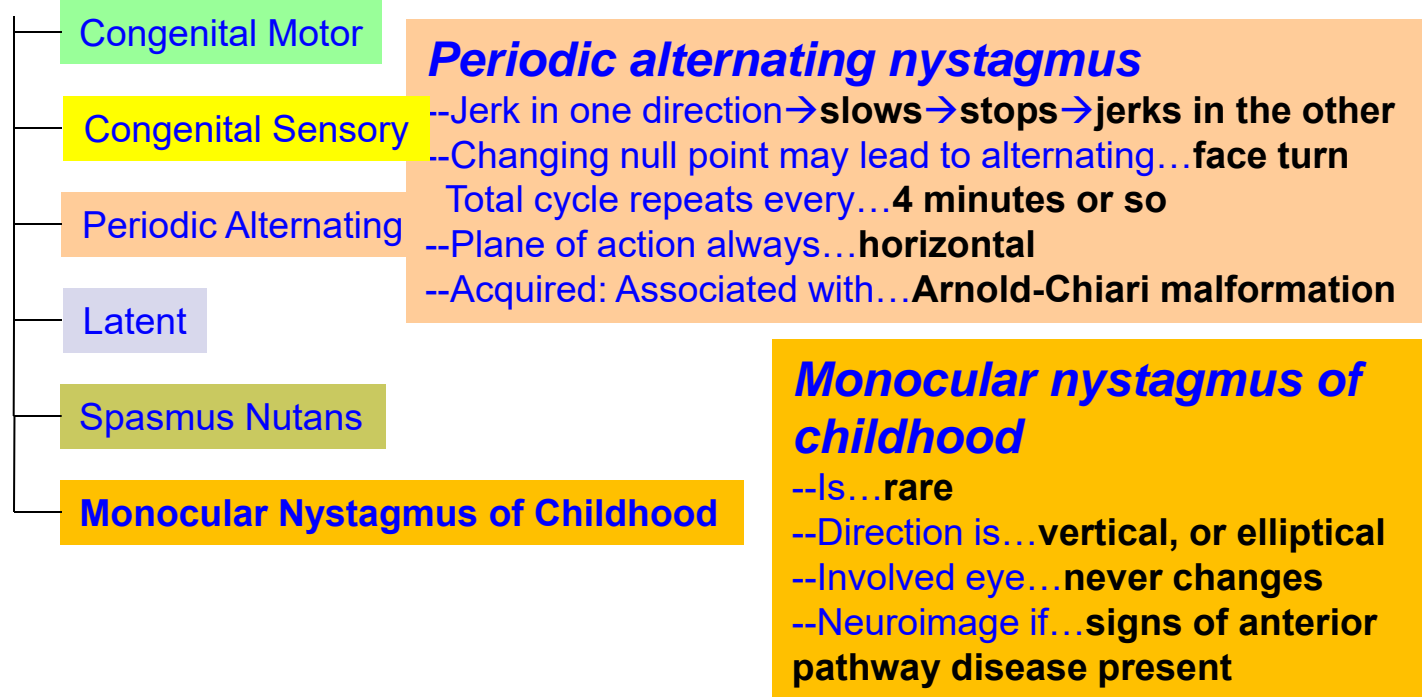
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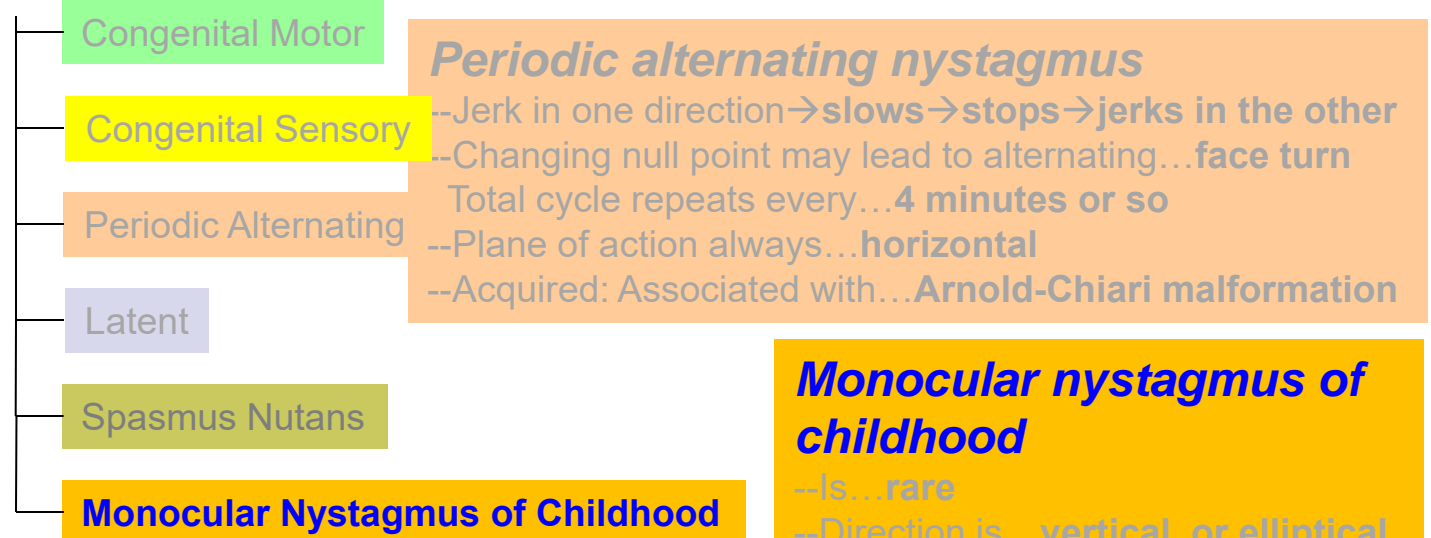
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Childhood



Periodic alternating nystagmus

- Jerk in one direction → **slows** → **stops** → jerks in the other
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- Total cycle repeats every... **4 minutes or so**
- Plane of action always... **horizontal**
- Acquired: Associated with... **Arnold-Chiari malformation**

Monocular nystagmus of childhood

- Is... **rare**
- Direction is... **vertical, or elliptical**
- Involved eye... **never changes**
- Neuroimage if... **signs of anterior pathway disease present**

What might these signs be?

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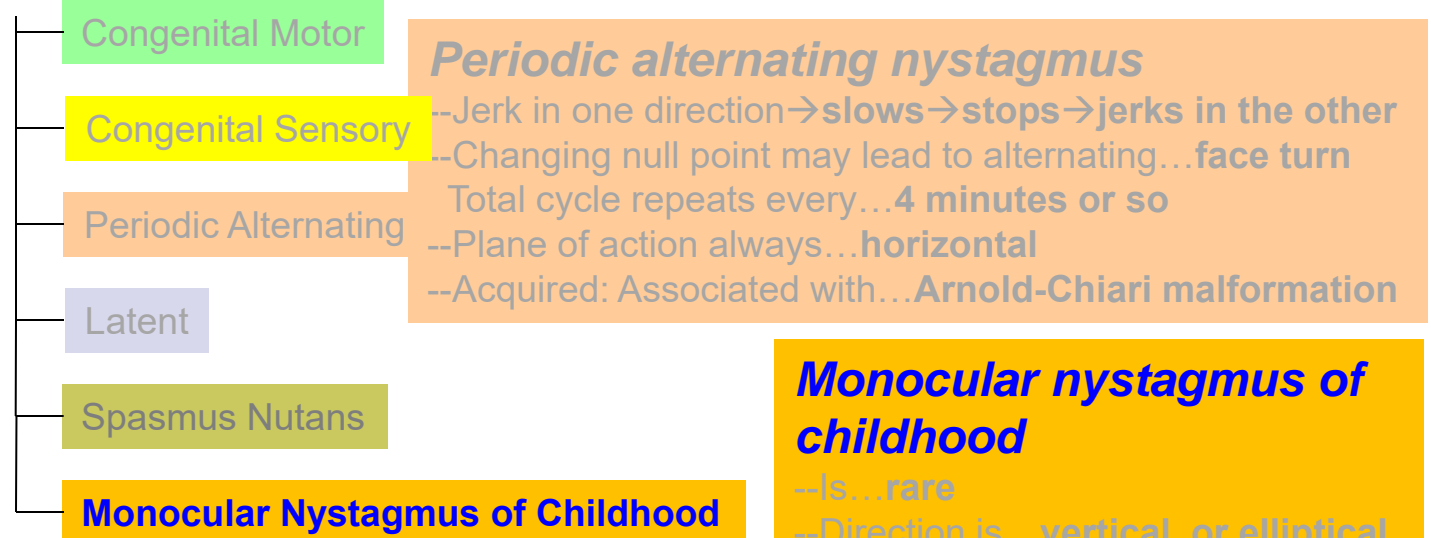
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- Involved eye... **never changes**
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An APD; ONH atrophy on DFE

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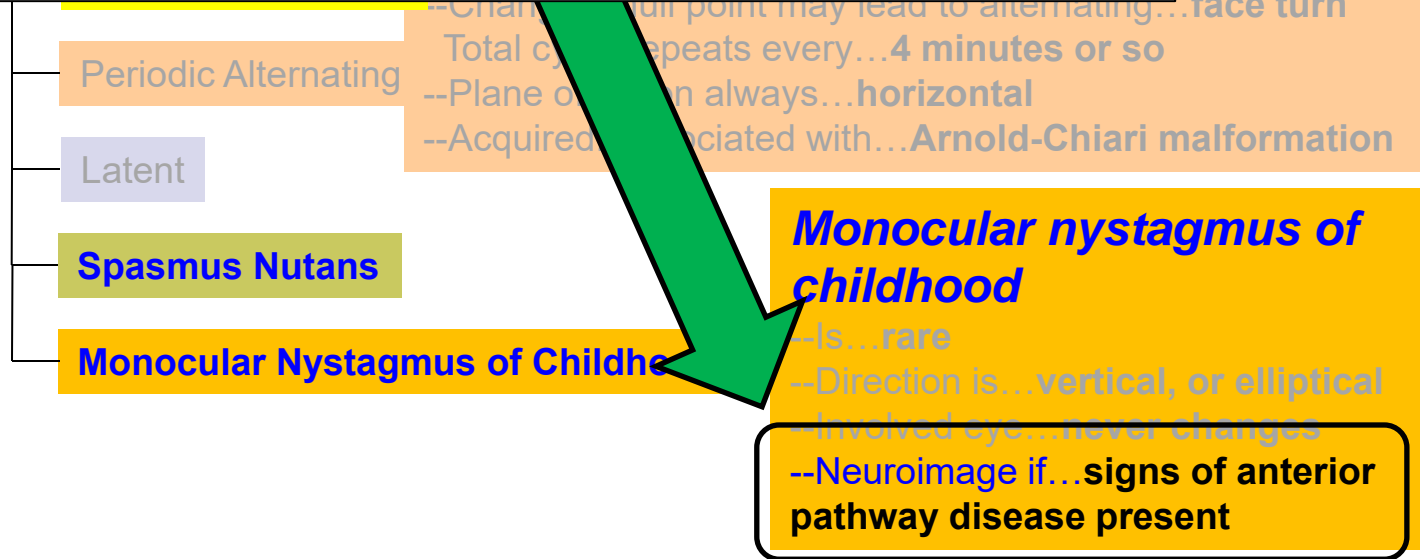
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This is why 'benign' Spasmus Nutans must be imaged (especially if seems monocular)--on the chance that it is actually *Monocular Nystagmus of Childhood*



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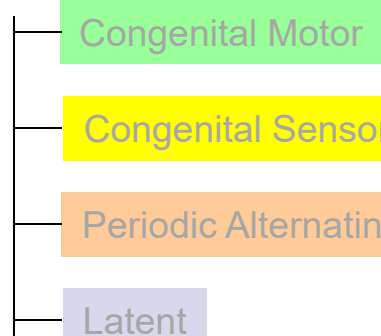
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- Plane of action always...**horizontal**
- Acquired: Associated with...**Arnold-Chiari malformation**

Can this pattern of nystagmus develop in adulthood?

Monocular nystagmus of childhood

- Is...**rare**
- Direction is...**vertical, or elliptical**
- Involved eye...**never changes**
- Neuroimage** if...**signs of anterior pathway disease present**

Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very... **small** ('shimmer')
- Usually **bilateral**, but can seem **unilateral**
- **Benign**. Resolves by age **3-4 years**. *But...*
- **Glioma** can present similarly, so image

Congenital motor nystagmus

- Usually... **horizontal**
- Remains horizontal in up/downgaze
- Vision usually... **good**
- Nystagmus + good VA = *congenital motor*
- Only form with... **paradoxical OKN response**
- Likely to have a... **null point**

Latent nystagmus

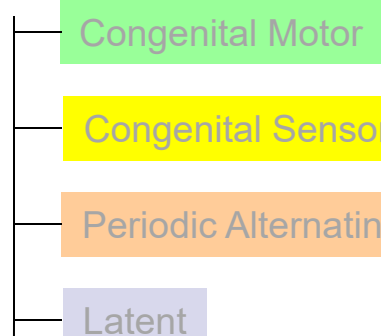
- Occurs when one eye is... **occluded**
- Jerk nystagmus toward... **fixating eye**
- Only nystagmus to change direction with fixation

Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
- Waveform depends on visual acuity:
 - 20/60 – 20/100: **Jerk**
 - 20/100 – 20/200: **Pendular**
 - <20/200: **'Searching'**

Childhood



Periodic alternating nystagmus

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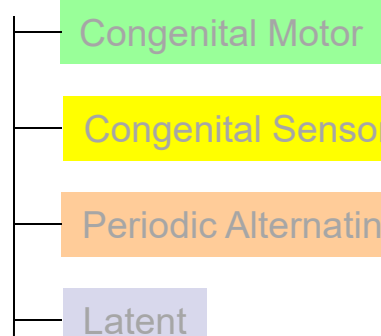
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Can this pattern of nystagmus develop in adulthood?

Yes

What is the adult version called (assuming it's not called something goofy like 'adult-onset monocular nystagmus of childhood')?

???????

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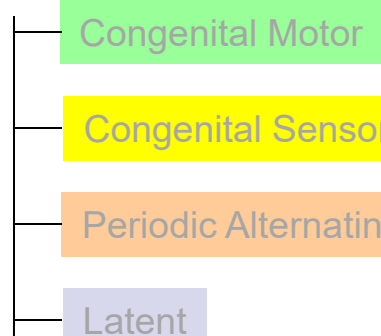
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Can this pattern of nystagmus develop in adulthood?

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It is referred to as 'the Heimann-Bielschowsky phenomenon'

The Heimann-Bielschowsky Phenomenon

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Childhood

Congenital Motor

Periodic alternating nystagmus

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Can
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Childhood

Adulthood

Congenital Motor

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- Step 3:**

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v
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Step 3: Tilt the head toward each **bodypart**, and see which direction **worsens v improves** the deviation

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- Vision usually... **good**
- Nystagmus + good VA = *congenital motor*
- Only form with... **paradoxical OKN response**
- Likely to have a... **null point**

Latent nystagmus

- Occurs when one eye is... **occluded**
- Jerk nystagmus toward... **fixating eye**
- Only nystagmus to change direction with fixation

Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
- Waveform depends on visual acuity:
 - 20/60 – 20/100: **Jerk**
 - 20/100 – 20/200: **Pendular**
 - <20/200: **'Searching'**

Childhood

Congenital Motor

Periodic alternating nystagmus

Bielschowsky... That name sounds familiar. Where have I heard it before?
 You are likely thinking of the **Parks-Bielschowsky 3-step test**

Uh, yeah, that's it. Remind me, what is that test used for?
 To identify the **cyclovertical** muscle responsible for a vertical deviation

Oh yeah, now I remember. Say, can you remind me of the steps in that test?
Step 1: Determine which eye is higher via cover testing
Step 2: Identify the gaze (ie, right vs left) in which the deviation gets worse
Step 3: Tilt the head toward each shoulder, and see which direction worsens the deviation

Can
Yes
What
good

It is referred to as 'the Heimann-**Bielschowsky** phenomenon'

stops → jerks in the other
 to alternating... **face turn**
 minutes or so
 ontal
 rdnold-Chiari malformation

Heimann-Bielschowsky phenomenon

- ... is... **vertical**, or elliptical
- Involved eye... **never changes**
- Neuroimage if... **signs of anterior pathway disease present**

Spasmus nutans

- Triad of **nystagmus + head nodding + torticollis**
- Nystagmus amplitude very... **small** ('shimmer')
- Usually **bilateral**, but can seem **unilateral**
- **Benign**. Resolves by age **3-4 years**. *But...*
- **Glioma** can present similarly, so image

Congenital motor nystagmus

- Usually... **horizontal**
- Remains horizontal in up/downgaze
- Vision usually... **good**
- Nystagmus + good VA = *congenital motor*
- Only form with... **paradoxical OKN response**
- Likely to have a... **null point**

Latent nystagmus

- Occurs when one eye is... **occluded**
- Jerk nystagmus toward... **fixating eye**
- Only nystagmus to change direction with fixation

Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
- Waveform depends on visual acuity:
 - 20/60 – 20/100: **Jerk**
 - 20/100 – 20/200: **Pendular**
 - <20/200: **'Searching'**

Childhood

Adulthood

Congenital Motor

Periodic alternating nystagmus

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 You are likely thinking of the **Parks-Bielschowsky 3-step test**

Uh, yeah, that's it. Remind me, what is that test used for?
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Oh yeah, now I remember. Say, can you remind me of the steps in that test?
Step 1: Determine which eye is higher via cover testing
Step 2: Identify the gaze (ie, right vs left) in which the deviation gets worse
Step 3: Tilt the head toward each shoulder, and see which direction worsens the deviation (note that this step is called the **head-tilt test**)

dat name again

Can
Yes
What
good

It is referred to as 'the Heimann-**Bielschowsky** phenomenon'

stops → jerks in the other
 alternating... **face turn**
 minutes or so
 ontal
 Arnold-Chiari malformation

Heimann-Bielschowsky phenomenon

- is... **vertical**, or elliptical
- involved eye... **never changes**
- Neuroimage if... **signs of anterior pathway disease present**

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Congenital motor nystagmus

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- Remains horizontal in up/downgaze
- Vision usually... **good**
- Nystagmus + good VA = *congenital motor*
- Only form with... **paradoxical OKN response**
- Likely to have a... **null point**

Latent nystagmus

- Occurs when one eye is... **occluded**
- Jerk nystagmus toward... **fixating eye**
- Only nystagmus to change direction with fixation

Nystagmus

Sensory nystagmus

- 2ndry to early **bilateral** poor vision
- Waveform depends on visual acuity:
 - 20/60 – 20/100: **Jerk**
 - 20/100 – 20/200: **Pendular**
 - <20/200: **'Searching'**

Childhood

Congenital Motor

Periodic alternating nystagmus

Bielschowsky... That name sounds familiar. Where have I heard it before?
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Step 3: Tilt the head toward each shoulder, and see which direction worsens the deviation (note that this step is called the *Bielschowsky head-tilt test*)

Can
 Yes
 What
 good

It is referred to as 'the Heimann-**Bielschowsky** phenomenon'

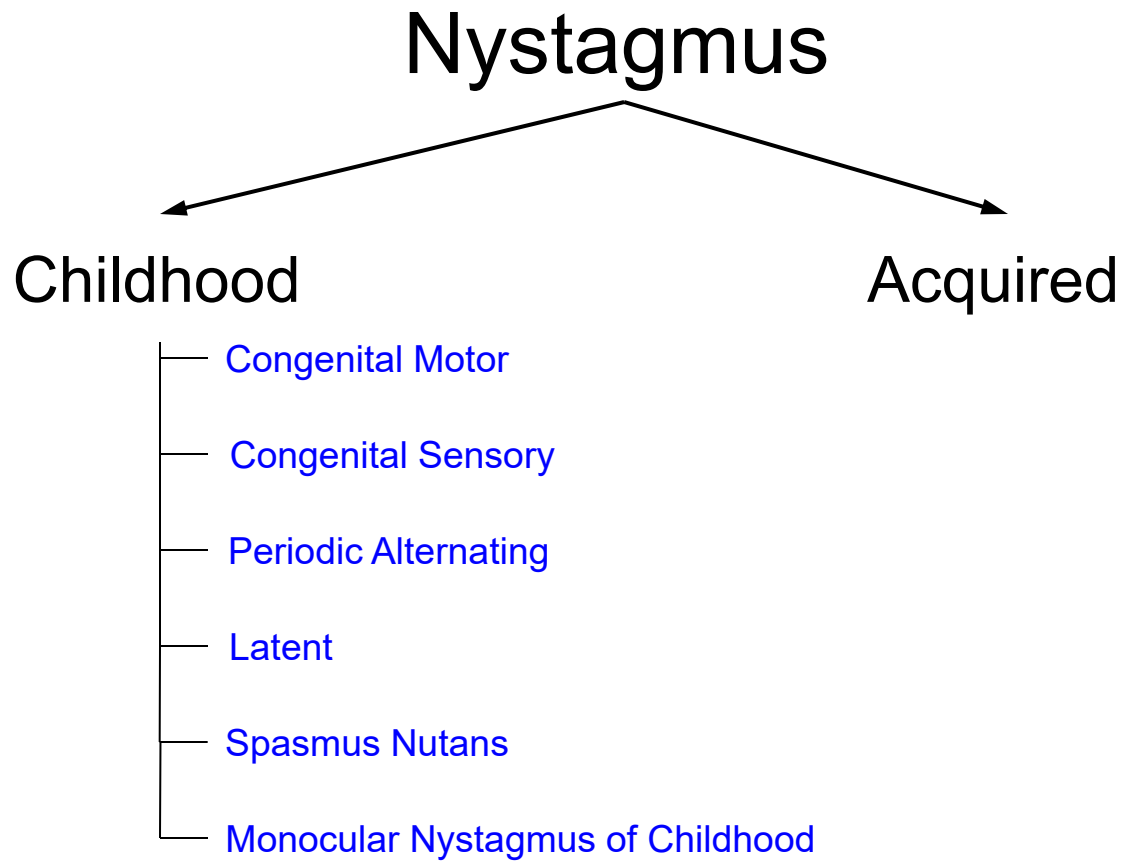
stops → jerks in the other
 to alternating... face turn
 minutes or so
 ontal
 rbold-Chiari malformation

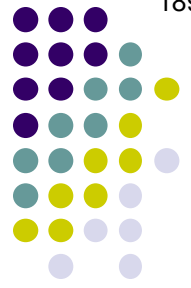
Heimann-Bielschowsky phenomenon

- ... is... **vertical**, or elliptical
- Involved eye... **never changes**
- Neuroimage if... **signs of anterior pathway disease present**

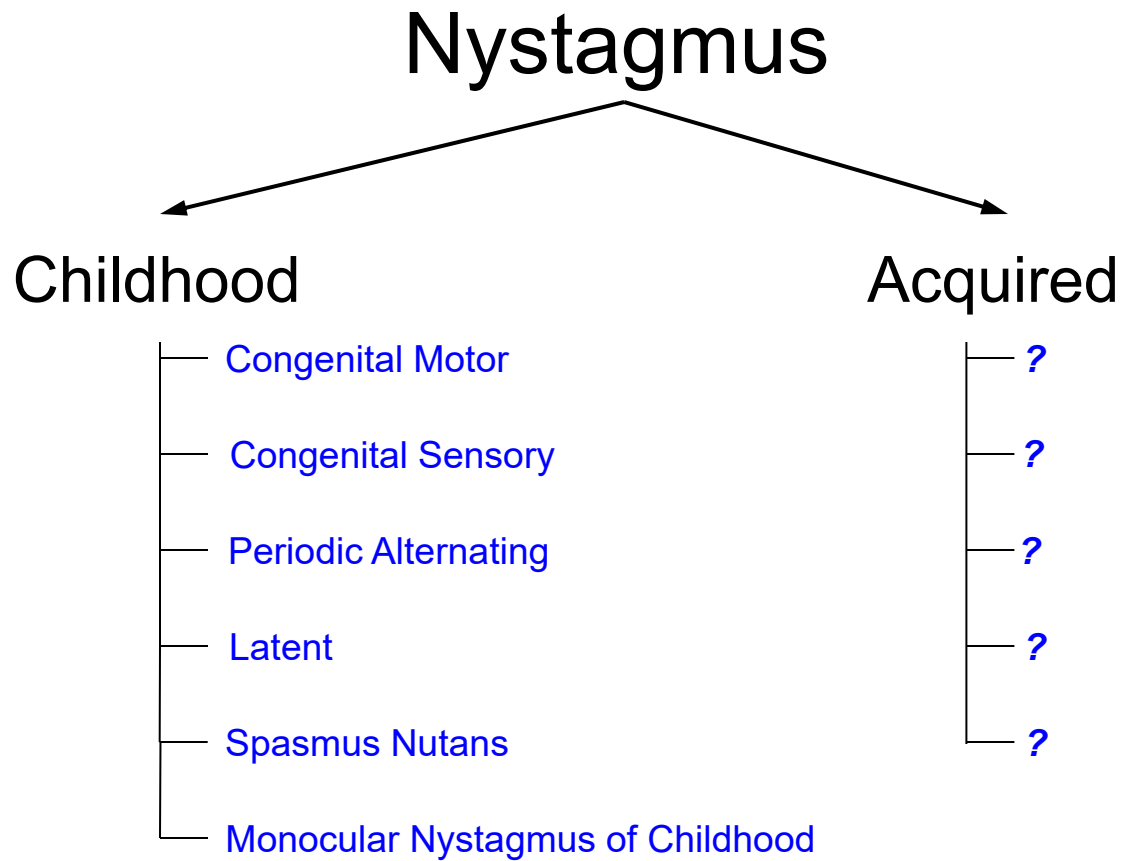


Nystagmus



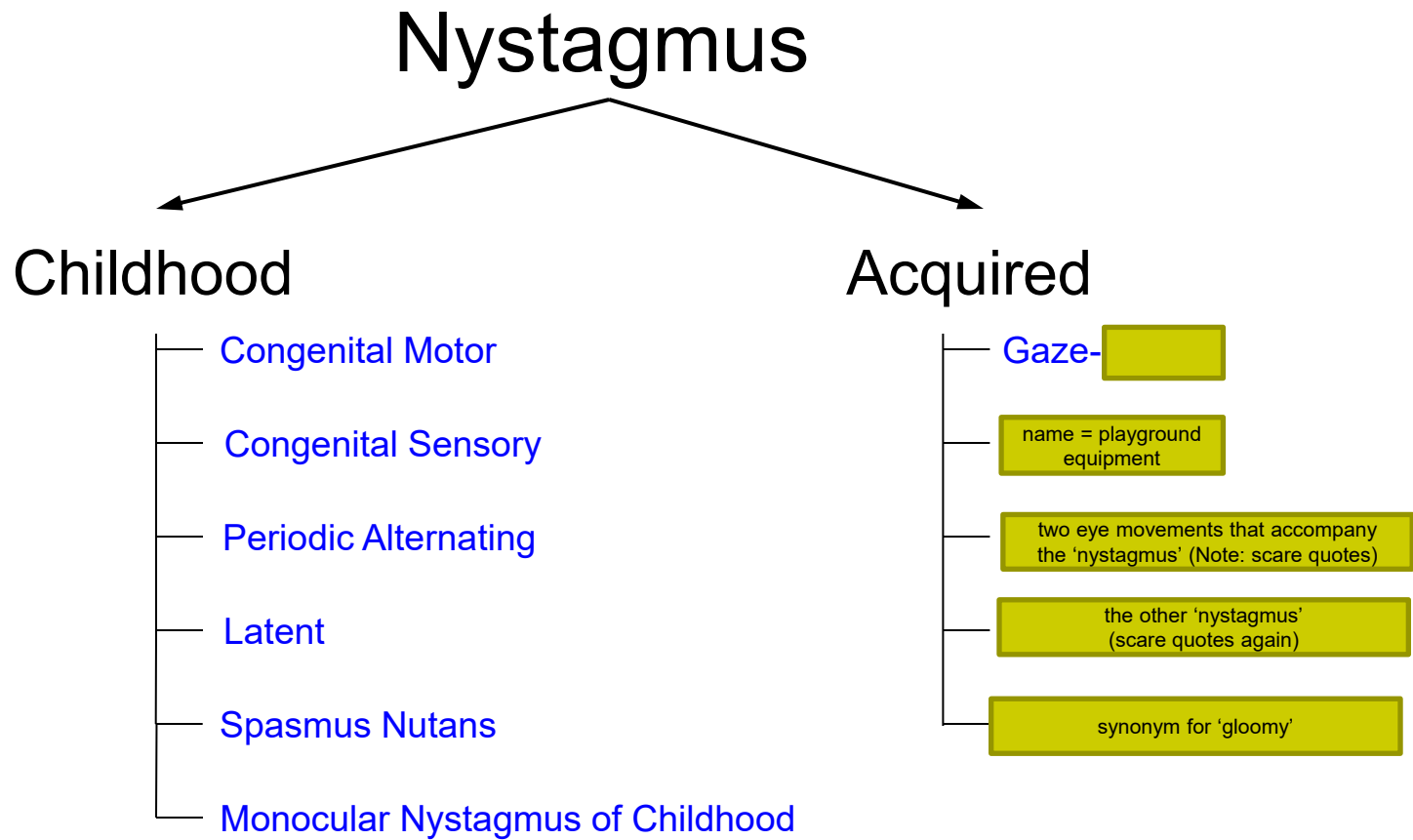


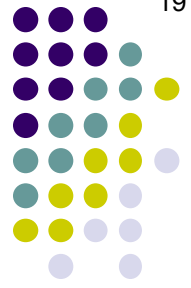
Nystagmus



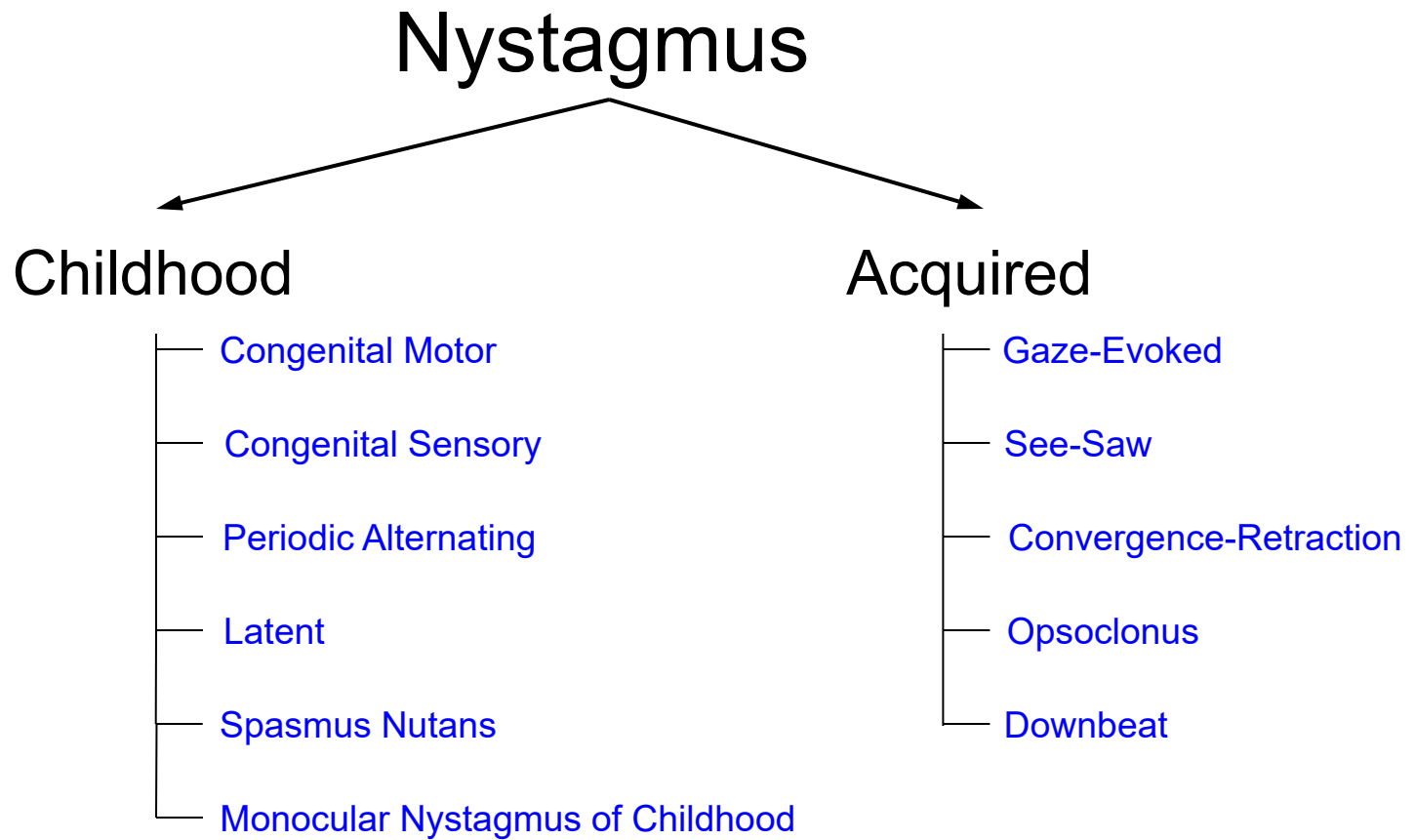


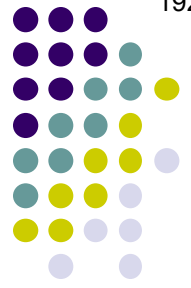
Nystagmus





Nystagmus

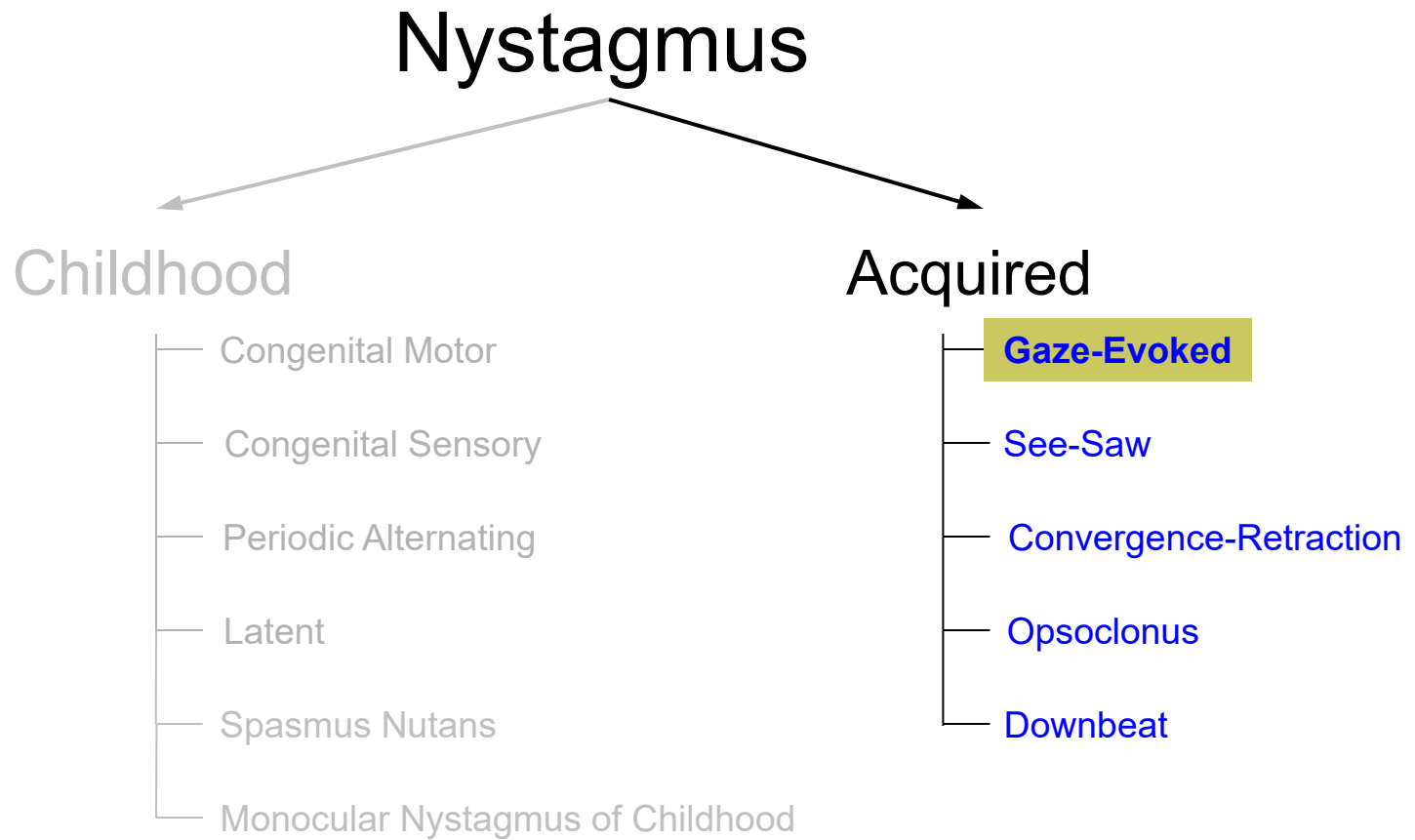


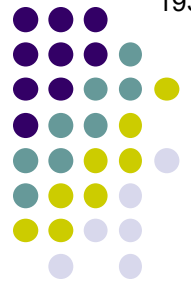


Nystagmus

Gaze-evoked nystagmus

--2° to inability of eyes to maintain...['direction' of fixation]





Nystagmus

Gaze-evoked nystagmus

--2° to inability of eyes to maintain...eccentric fixation

Nystagmus

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- **Gaze-Evoked**
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat

Nystagmus

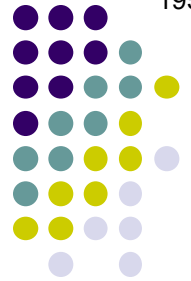
Gaze-evoked nystagmus

--2° to inability of eyes to maintain... **eccentric fixation**



Why might an eye be unable to maintain eccentric fixation?

- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

--2° to inability of eyes to maintain... **eccentric fixation**

Why might an eye be unable to maintain eccentric fixation?

The eye is suspended in place by an elaborate web of soft tissue arising in the orbit. This 'suspensory system' can be thought of as a set of rubber bands extending from the walls of the orbit to enmesh the globe. And like rubber bands, the tissues suspending the globe resist being stretched--if you pull on them, they pull back. When they're not being stretched by ocular rotations, the summation of all these rubber-band forces want to keep the globe in (or close to) primary gaze. Thus, *eccentric fixation requires the continuous generation of the force needed to overcome this elastic countertraction.*

- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...[toward, or away from, direction of gaze]

Nystagmus

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- **Gaze-Evoked**
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze

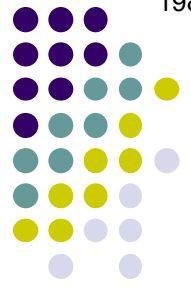
Nystagmus

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- **Gaze-Evoked**
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



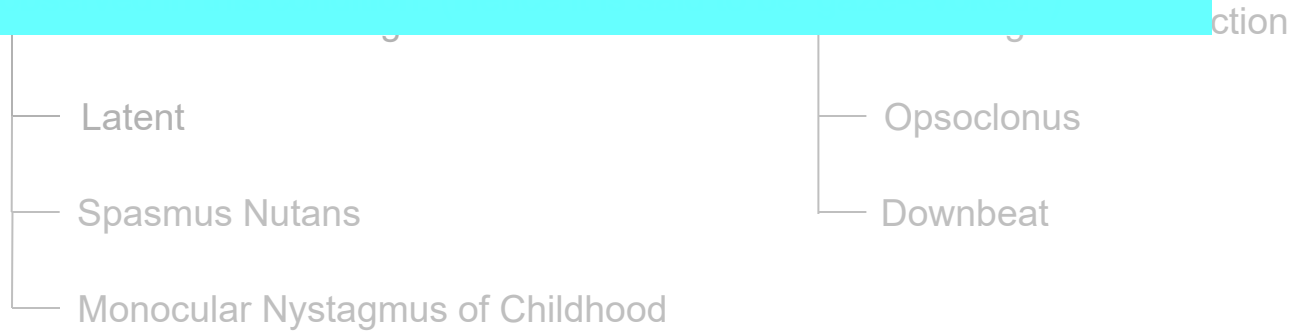
Nystagmus

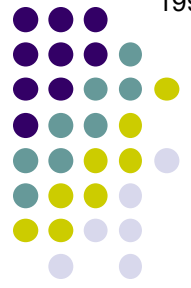
Gaze-evoked nystagmus

-- due to inability of eyes to maintain...eccentric fixation

--Fast phase always...toward direction of gaze

As stated previously, eccentric fixation requires the continuous generation of enough force to overcome the inherent countertraction produced by the elastic properties of the globe's suspensory system. Generating this force is the task of a two words --a CNS nucleus responsible for producing the graded signal needed to keep the globe in eccentric fixation.



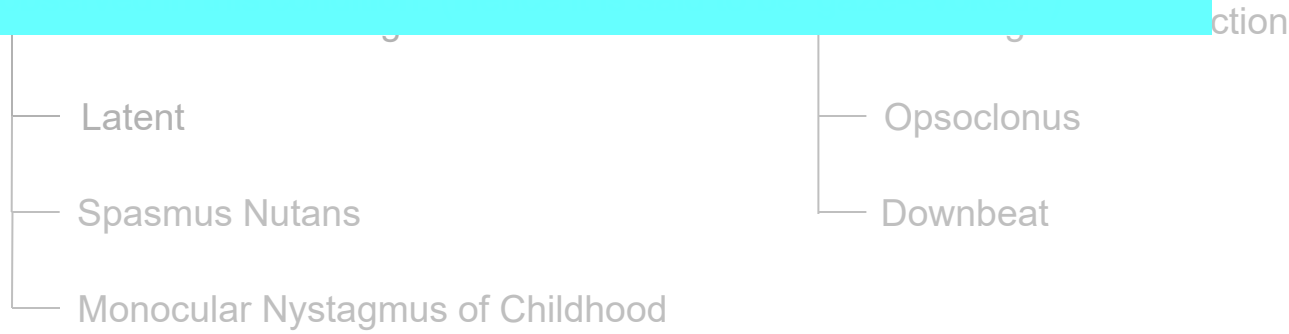


Nystagmus

Gaze-evoked nystagmus

- due to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze

As stated previously, eccentric fixation requires the continuous generation of enough force to overcome the inherent countertraction produced by the elastic properties of the globe's suspensory system. Generating this force is the task of a *neural integrator* --a CNS nucleus responsible for producing the graded signal needed to keep the globe in eccentric fixation.





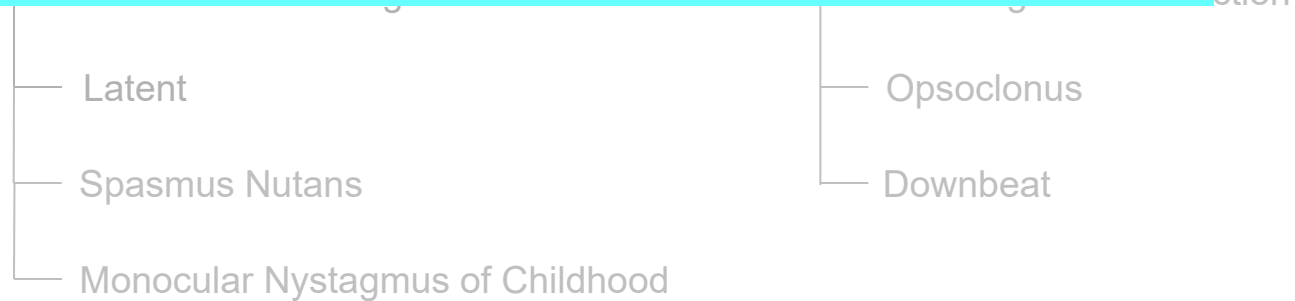
Nystagmus

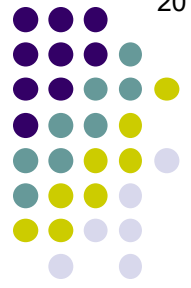
Gaze-evoked nystagmus

- due to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze

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In gaze-evoked nystagmus, the neural integrator becomes 'leaky' in that it fails to reliably produce the signal required to maintain the needed force generation. Thus, after the eye saccades into eccentric gaze, leakiness by the neural integrator allows the elastic forces of the orbit to pull the eye back towards primary gaze. At some point during this drift the eye re-saccades back out to its intended eccentric position, only to start drifting primary-ward again. A 'loop' of these two events--the saccade into eccentric gaze, and the slow drift back toward primary--comprise the nystagmus observed in this condition. (Hence it is said to be 'gaze-evoked.')





Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...[what was the name of that law again?]

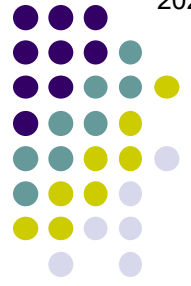
Nystagmus

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

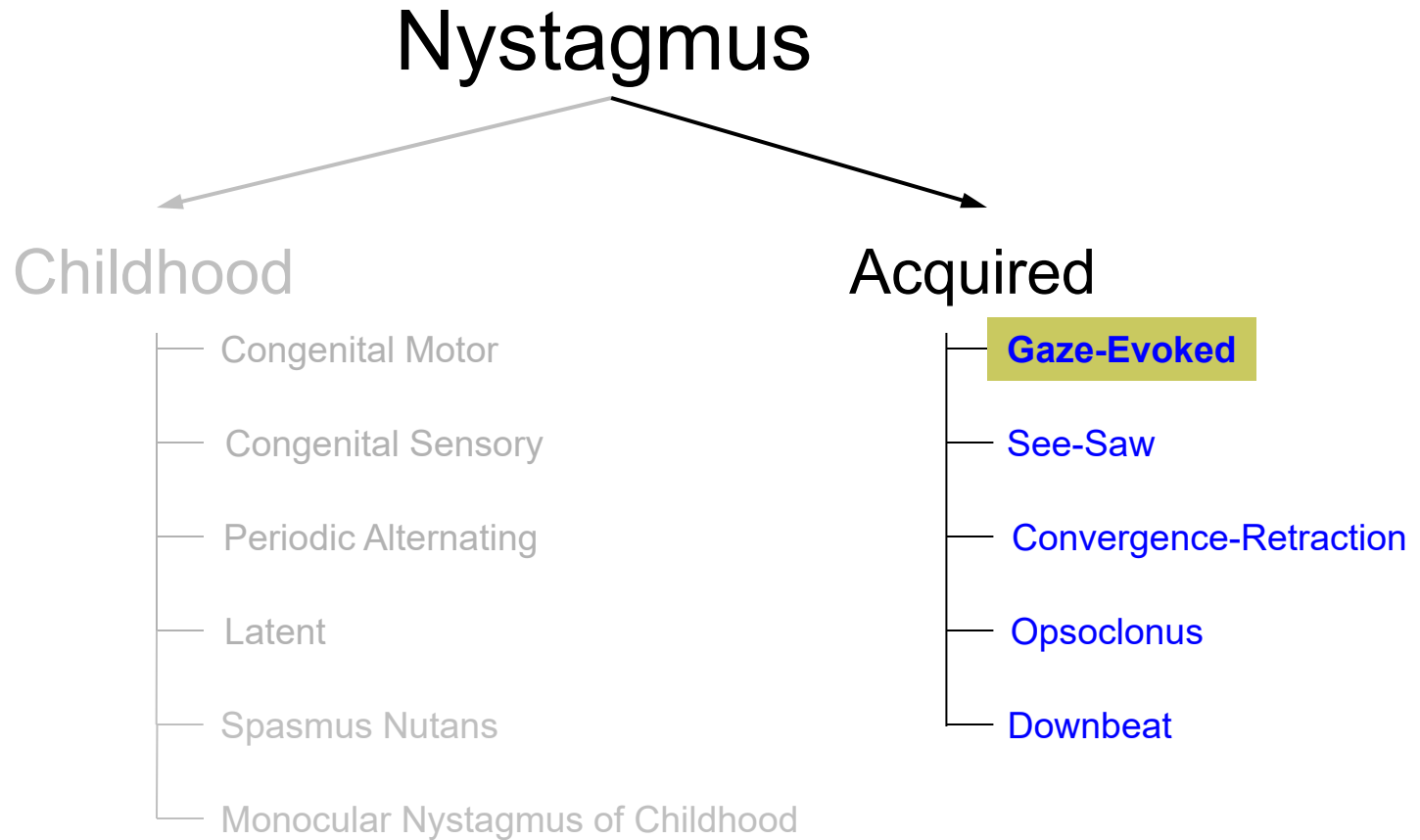
- **Gaze-Evoked**
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat

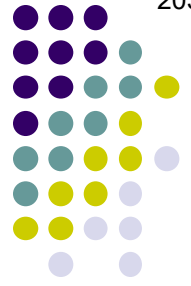


Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law





Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

It makes sense that gaze-evoked nystagmus would obey Alexander's law--the more extreme the eccentric gaze, the greater will be the elastic force pulling the globe back towards primary. (The farther you stretch a rubber band, the greater the force it produces.) This will increase both the rate at which the phases cycle, and the speed with which the movements occur.

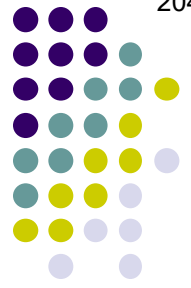
Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- **Gaze-Evoked**
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat

(No question—proceed when ready)



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

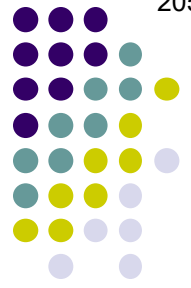
--Eyes alternate movement + movement with movement + movement

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...[specific tumor type]

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

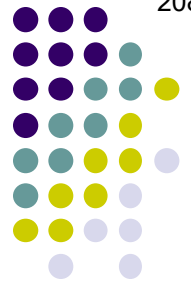
- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

- Congenital Motor
- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
- See-Saw
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Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Gaze-Evoked

Convergence-retraction nystagmus

What is convergence-retraction nystagmus?



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

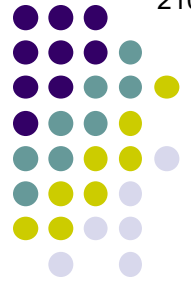
Acquired

Convergence-retraction nystagmus

Gaze-Evoked

What is convergence-retraction nystagmus?

A phenomena in which attempted upgaze causes the globes to retract (ie, sink deeper into the orbit), converge, and 'shimmy' (for lack of a better word; it is not a true nystagmus)



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Convergence-retraction nystagmus

Gaze-Evoked

What is convergence-retraction nystagmus?

A phenomena in which attempted upgaze causes the globes to retract (ie, sink deeper into the orbit), converge, and 'shimmy' (for lack of a better word); **it is not a true nystagmus**

Why isn't it a true nystagmus?



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Convergence-retraction nystagmus

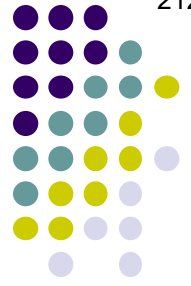
Gaze-Evoked

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Why isn't it a true nystagmus?

Because it doesn't initiate with a slow movement



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Convergence-retraction nystagmus

Gaze-Evoked

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Why isn't it a true nystagmus?

Because it doesn't initiate with a slow movement

If it's not a nystagmus, what is it?



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Convergence-retraction nystagmus

Gaze-Evoked

What is convergence-retraction nystagmus?

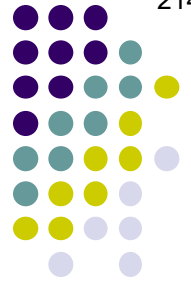
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Why isn't it a true nystagmus?

Because it doesn't initiate with a slow movement

If it's not a nystagmus, what is it?

A saccadic intrusion (remember?)



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Acquired

Convergence-retraction nystagmus

Gaze-Evoked

What is convergence-retraction nystagmus?

A phenomena in which attempted upgaze causes the globes to retract (ie, sink deeper into the orbit), converge, and 'shimmy' (for lack of a better word; it is not a true nystagmus)

What causes the eyes to retract?



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...**eccentric fixation**
- Fast phase always...**toward direction of gaze**
- Obeys...**Alexander's law**

Nystagmus

See-saw nystagmus

- Eyes alternate **elevation + intorting** with **depression + extorting**
- Associated with...**craniopharyngioma**

Childhood

Acquired

Convergence-retraction nystagmus

Gaze-Evoked

What is convergence-retraction nystagmus?

A phenomena in which attempted upgaze causes the globes to retract (ie, sink deeper into the orbit), converge, and 'shimmy' (for lack of a better word; it is not a true nystagmus)

What causes the eyes to retract?

Attempted elevation causes the medial and lateral recti muscles to fire simultaneously, the net result of which is the globes being pulled back into the orbits--retracting, in other words



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...**eccentric fixation**
- Fast phase always...**toward direction of gaze**
- Obeys...**Alexander's law**

Nystagmus

See-saw nystagmus

- Eyes alternate **elevation + intorting** with **depression + extorting**
- Associated with...**craniopharyngioma**

Childhood

Acquired

Gaze-Evoked

Convergence-retraction nystagmus

What is convergence-retraction nystagmus?

A phenomena in which attempted upgaze causes the globes to retract (ie, sink deeper into the orbit), converge, and 'shimmy' (for lack of a better word; it is not a true nystagmus)

What causes the eyes to retract?

Attempted elevation causes the medial and lateral recti muscles to fire simultaneously, the net result of which is the globes being pulled back into the orbits--retracting, in other words

OK, but if the MR and LR are both firing, why do the eyes converge?



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...**eccentric fixation**
- Fast phase always...**toward direction of gaze**
- Obeys...**Alexander's law**

Nystagmus

See-saw nystagmus

- Eyes alternate **elevation + intorting** with **depression + extorting**
- Associated with...**craniopharyngioma**

Childhood

Acquired

Gaze-Evoked

Convergence-retraction nystagmus

What is convergence-retraction nystagmus?

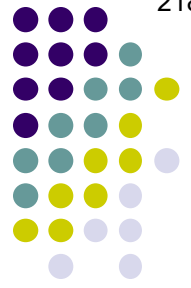
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What causes the eyes to retract?

Attempted elevation causes the medial and lateral recti muscles to fire simultaneously, the net result of which is the globes being pulled back into the orbits--retracting, in other words

OK, but if the MR and LR are both firing, why do the eyes converge?

Because the medial recti are the strongest EOMs. Thus, in a battle royale among the recti, the MR are going to cause both eyes to adduct--to converge, in other words.



Nystagmus

Gaze-evoked nystagmus

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Nystagmus

See-saw nystagmus

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Childhood

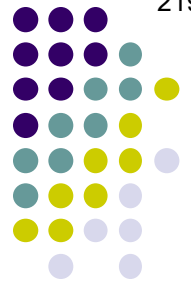
Convergence-retraction nystagmus

--Part of **eponym-word-word** syndrome

- Congenital Sensory
- Periodic Alternating
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
- See-Saw
- Convergence-Retraction
- Opsoclonus
- Downbeat



Nystagmus

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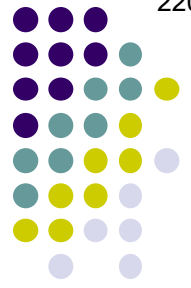
Convergence-retraction nystagmus

- Part of Parinaud's dorsal midbrain syndrome

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- Periodic Alternating
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Childhood

Convergence-retraction nystagmus

--Part of Parinaud's dorsal midbrain syndrome

What are the findings in Parinaud's dorsal midbrain syndrome?

- 1) Convergence-retraction nystagmus (duh)
- 2)
- 3)
- 4)

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

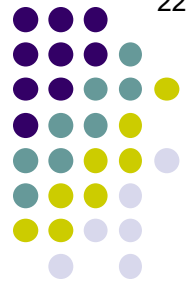
Gaze-Evoked

See-Saw

Convergence-Retraction

Opsoclonus

Downbeat



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See-saw nystagmus

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Childhood

Convergence-retraction nystagmus

--Part of Parinaud's dorsal midbrain syndrome

What are the findings in Parinaud's dorsal midbrain syndrome?

- 1) Convergence-retraction nystagmus
- 2) Impaired upgaze
- 3) Lid retraction
- 4) Light-near dissociation

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

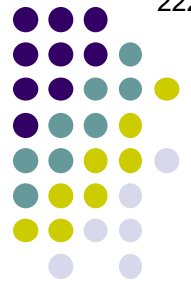
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Childhood

Convergence-retraction nystagmus

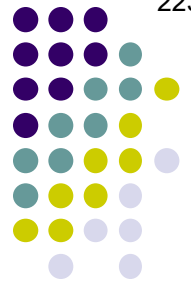
- Part of Parinaud's dorsal midbrain syndrome

The etiology of a Parinaud syndrome is often a function of who the pt is.
 For each of these pts with Parinaud's, state the most likely cause:
 --A child:

- Spasmus Nutans
- Monocular Nystagmus of Childhood

Acquired

- Gaze-Evoked
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 For each of these pts with Parinaud's, state the most likely cause:
 --A child: **Hydrocephalus**

Spasmus Nutans

Monocular Nystagmus of Childhood

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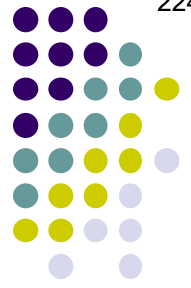
Gaze-Evoked

See-Saw

Convergence-Retraction

Opsoclonus

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 --A young man:

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

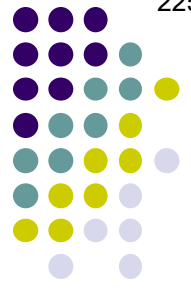
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The etiology of a Parinaud syndrome is often a function of who the pt is. For each of these pts with Parinaud's, state the most likely cause:

- A child: **Hydrocephalus**
- A young man: **A pineal tumor**

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

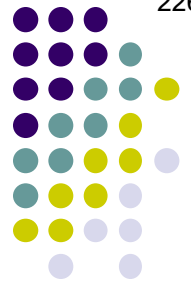
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- A young woman:

Spasmus Nutans

Monocular Nystagmus of Childhood

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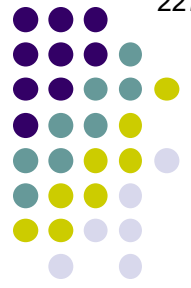
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- A child: **Hydrocephalus**
- A young man: **A pineal tumor**
- A young woman: **MS**

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

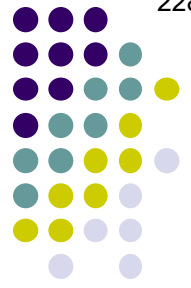
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- A child: **Hydrocephalus**
- A young man: **A pineal tumor**
- A young woman: **MS**
- An **older** adult:

Spasmus Nutans

Monocular Nystagmus of Childhood

Acquired

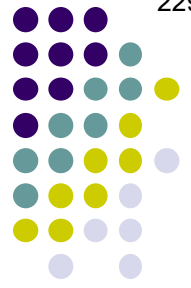
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- A young man: **A pineal tumor**
- A young woman: **MS**
- An **older** adult: **CVA**

Spasmus Nutans

Monocular Nystagmus of Childhood

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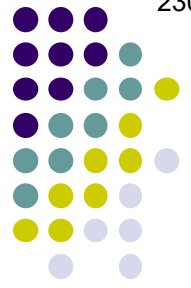
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Childhood

Convergence-retraction nystagmus

- Part of Parinaud's dorsal midbrain syndrome

Opsoclonus

- Not a true nystagmus—is a two words

Monocular Nystagmus of Childhood

Acquired

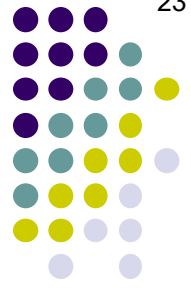
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Childhood

Convergence-retraction nystagmus

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Opsoclonus

- Not a true nystagmus—is a saccadic oscillation

Monocular Nystagmus of Childhood

Acquired

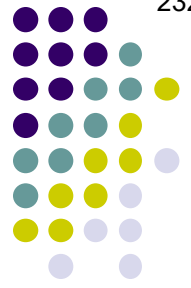
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Childhood

Convergence-retraction nystagmus

- Part of Parinaud's dorsal midbrain syndrome

Opsoclonus

- Not a true nystagmus—is a saccadic oscillation
- Movements are velocity and directionality

Monocular Nystagmus of Childhood

Acquired

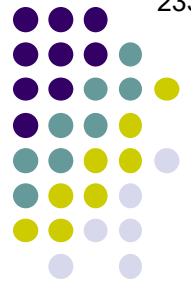
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Childhood

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- Part of Parinaud's dorsal midbrain syndrome

Opsoclonus

- Not a true nystagmus—is a saccadic oscillation
- Movements are rapid and multivectorial

Monocular Nystagmus of Childhood

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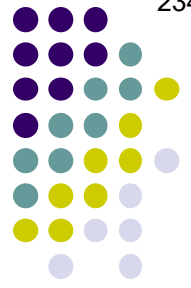
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 --Part of Parinaud's dorsal midbrain syndrome

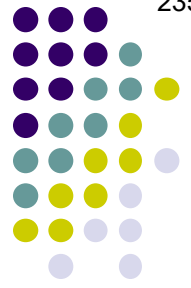
Opsoclonus
 --Not a true nystagmus—is a **saccadic oscillation**
 --Movements are **rapid and multivectorial**

In opsoclonus, do the movements persist during...
 --eyelid closure?
 --sleep?

Monocular Nystagmus of Childhood

Acquired

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In opsoclonus, do the movements persist during...

- eyelid closure? **Yes**
- sleep? **Yes**

Monocular Nystagmus of Childhood

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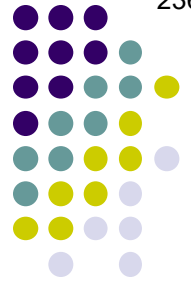
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Convergence-retraction nystagmus

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Opsoclonus

- Not a true nystagmus—is a saccadic oscillation
- Movements are rapid and multivectorial
- 4 main associations:
 - 1) A paraneoplastic syndrome in specific tumor type
 - 2)
 - 3)
 - 4)

Monocular Nystagmus of Childhood

Acquired

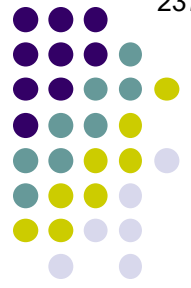
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- Movements are rapid and multivectorial
- 4 main associations:
 - 1) A paraneoplastic syndrome in neuroblastoma
 - 2)
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Is opsoclonus a good or bad prognostic sign in neuroblastoma?

Monocular Nystagmus of Childhood

Acquired

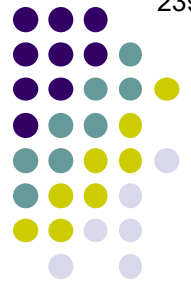
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- Movements are rapid and multivectorial
- 4 main associations:

- 1) A paraneoplastic syndrome in neuroblastoma
- 2)
- 3) *Is opsoclonus a good or bad prognostic sign in neuroblastoma?*
- 4) A good sign

Acquired

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Monocular Nystagmus of Childhood



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Neuroblastoma is a pediatric cancer. Can opsoclonus be associated with cancer in adults?

quired

Gaze-Evoked

See-Saw

Convergence-Retraction

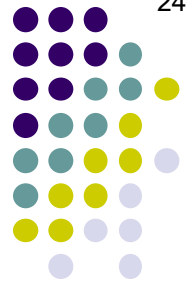
Opsoclonus

Downbeat

--4 main associations:

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- 2)
- 3)
- 4)

Monocular Nystagmus of Childhood



Nystagmus

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quired

Gaze-Evoked

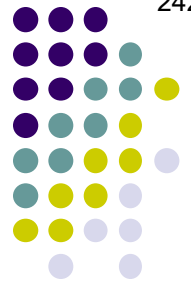
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Monocular Nystagmus of Childhood



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Neuroblastoma is a pediatric cancer. Can opsoclonus be associated with cancer in adults?

Yes

With what three adult cancers is opsoclonus most commonly associated?

-
- (Mnemonic forthcoming...)
-

--4 main associations:

- 1) A paraneoplastic syndrome in **neuroblastoma**
- 2)
- 3)
- 4)

quired

Gaze-Evoked

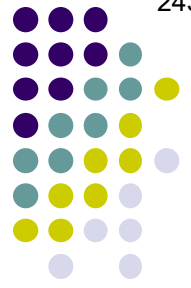
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Downbeat

Monocular Nystagmus of Childhood



Nystagmus

Gaze-evoked nystagmus

- 2° to inability of eyes to maintain...eccentric fixation
- Fast phase always...toward direction of gaze
- Obeys...Alexander's law

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Neuroblastoma is a pediatric cancer. Can opsoclonus be associated with cancer in adults?

Yes

With what three adult cancers is opsoclonus most commonly associated?

- S
- O Cancer-associated opsoclonus in adults is so sad, it'll make you SOB
- B

--4 main associations:

- 1) A paraneoplastic syndrome in neuroblastoma
- 2)
- 3)
- 4)

quired

Gaze-Evoked

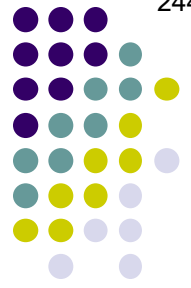
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Convergence-Retraction

Opsoclonus

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Neuroblastoma is a pediatric cancer. Can opsoclonus be associated with cancer in adults?

Yes

With what three adult cancers is opsoclonus most commonly associated?

- Small-cell lung** (most common--remember this one for sure!)
- Ovarian**
- Breast**

--4 main associations:

- 1) A paraneoplastic syndrome in **neuroblastoma, small-cell lung CA, ovarian CA, breast CA**
- 2)
- 3)
- 4)

quired

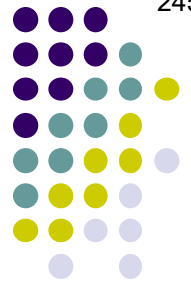
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- Part of Parinaud's dorsal midbrain syndrome

Opsoclonus

- Not a true nystagmus—is a saccadic oscillation
- Movements are rapid and multivectorial
- 4 main associations:
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 - 2) two words ataxia
 - 3)
 - 4)

Acquired

Gaze-Evoked

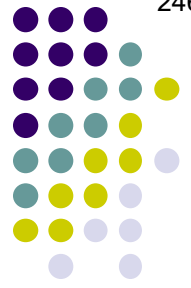
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 - 2) Acute cerebellar ataxia
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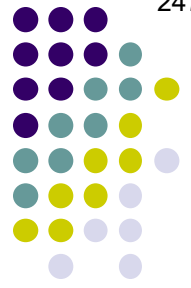
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- 2) **Acute cerebellar ataxia**
- 3)
- 4)

What is the classic description of acute cerebellar ataxia?

Acquired

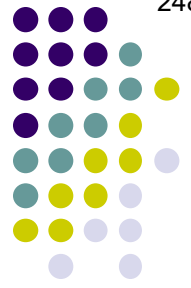
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What is the classic description of acute cerebellar ataxia?
'Dancing eyes and dancing feet'

Acquired

Gaze-Evoked

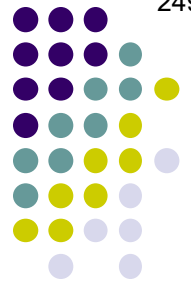
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What childhood syndrome consist of opsoclonus, myoclonus and ataxia?

See-saw

- Eyes alt
- Associa

Converge

- Part of Par

Opsoclo

- Not a true
- Movemen

--4 main associations:

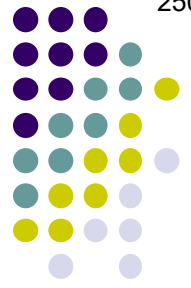
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Opsoclonus

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Note: The BCSC *Neuro* book calls it *Opsoclonus-Myoclonus Syndrome*. The name *opsoclonus-myoclonus-ataxia syndrome* is more descriptive, however.

What childhood syndrome consist of opsoclonus, myoclonus and ataxia?
Opsoclonus-myoclonus-ataxia syndrome (OMAS)

See-saw

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- Associa

Convergent

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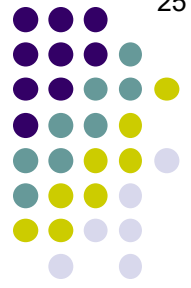
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What other symptoms/signs may be present?

-
-
-

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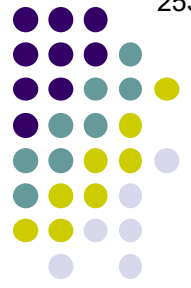
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What percentage of OMAS pts have neuroblastoma?

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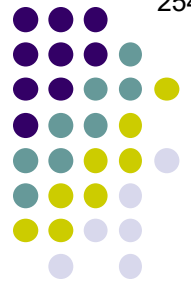
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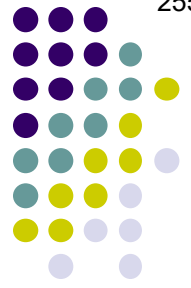
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Does the presence of OMAS in neuroblastoma convey a good, or poor prognosis?

--4 main associations:

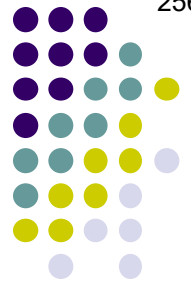
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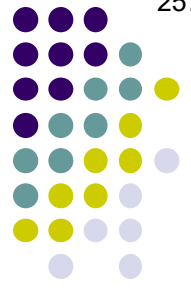
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 - 2) Acute cerebellar ataxia
 - 3) Post...[type of infection]
 - 4)

Acquired

Gaze-Evoked

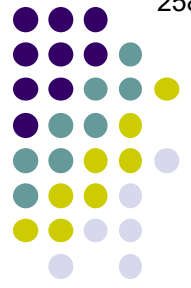
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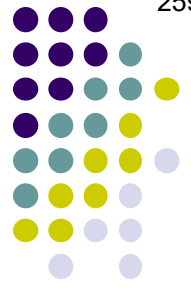
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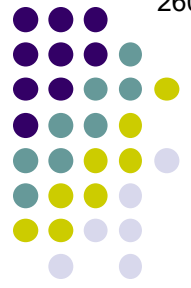
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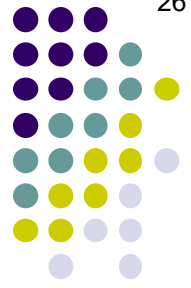
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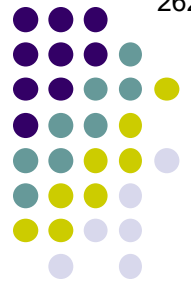
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Opsoclonus

Rule of thumb for etiology of opsoclonus:

- In children:
- In young adults:
- In older adults:

Monocul



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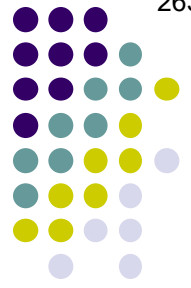
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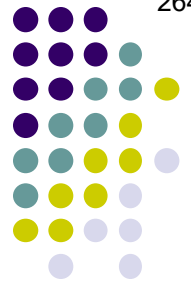
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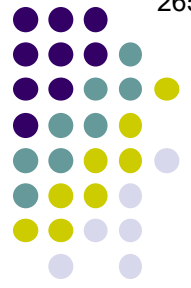
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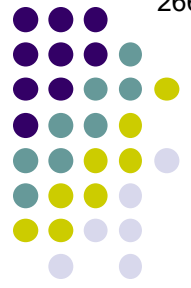
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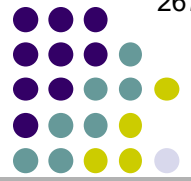
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Downbeat nystagmus

- Associated with eponym-eponym malformation

Nystagmus

See-saw nystagmus

- Eyes alternate elevation + intorting with depression + extorting
- Associated with...craniopharyngioma

Childhood

Convergence-retraction nystagmus

- Part of Parinaud's dorsal midbrain syndrome

Opsoclonus

- Not a true nystagmus—is a saccadic oscillation
- Movements are rapid and multivectorial
- 4 main associations:
 - 1) A paraneoplastic syndrome in neuroblastoma
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 - 3) Post...viral
 - 4) Post...encephalitis

Acquired

Gaze-Evoked

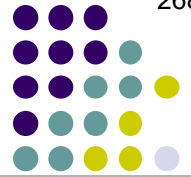
See-Saw

Convergence-Retraction

Opsoclonus

Downbeat

Monocular Nystagmus of Childhood



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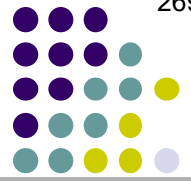
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Previously, it was mentioned that the acquired form of one of the childhood nystagmus was associated with Arnold-Chiari malformation. Which childhood nystagmus was it?

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 Period-alternating nystagmus

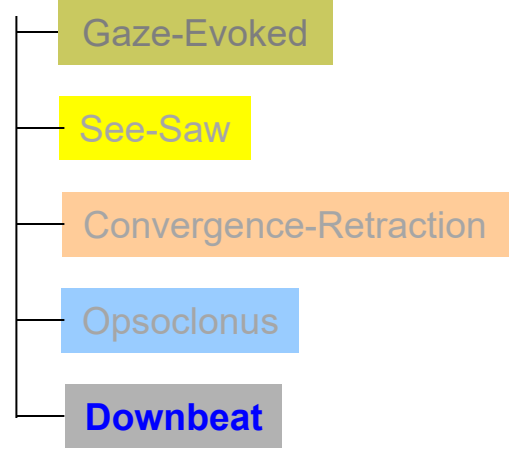
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Co

--Pa

O

--N

--M

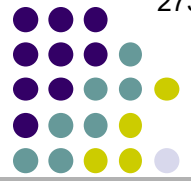
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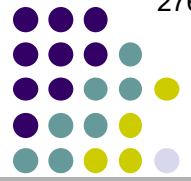
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Monocular Nystagmus of Childhood



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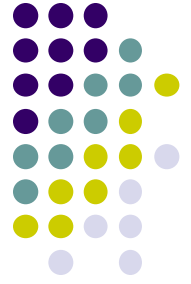
Downbeat

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Nystagmus

- **Nystagmus: Management**
 - Two main treatment goals:
 - *Goal 1: Decrease...*
 - *Goal 2: Improve...*





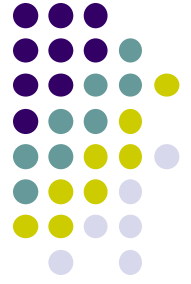
Nystagmus

- **Nystagmus: Management**

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- *Goal 1: Decrease...nystagmus intensity*

- *Goal 2: Improve...head position (if a face turn is present)*



Nystagmus

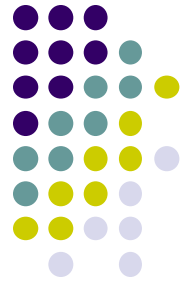
- **Nystagmus: Management**

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- Give...[optical device]
- If no face turn: [surgery]

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Nystagmus

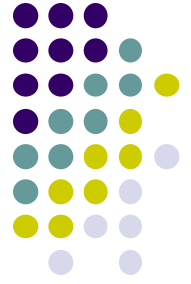
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- *Goal 1: Decrease...nystagmus intensity*

- Give...BO prism (will dampen the nystagmus)
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Nystagmus

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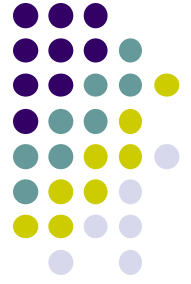
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- Use prisms to...



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- Use prisms to...shift null point to primary



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 - Consider [surgical procedure]

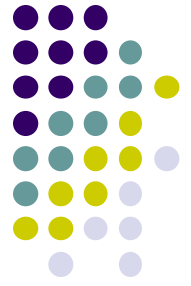


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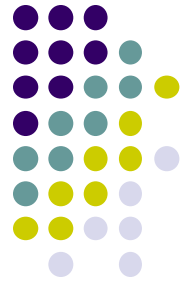


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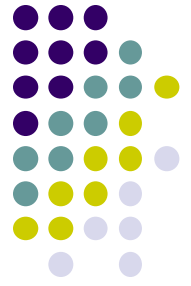
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 - Consider ***Kestenbaum-Anderson procedure***
 - **Procedure in a nutshell: Bilateral recessions/resections to make it harder to look in preferred direction**

e.g., consider a patient who has a right face turn (i.e., the null point is in left gaze). To treat the face turn surgically, recess-resect the horizontal muscles OU to make it more difficult to reach left gaze:
 --**recess** the left LR and right MR, and
 --**resect** the left MR and the right LR