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
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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Is the refixation movement fast or slow?
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Is the refixation movement fast or slow?
It can be either.
--If it is fast, the pattern is called a jerk nystagmus.
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--If it is fast, the pattern is called a \textit{jerk nystagmus}
--If it is slow, it is known as a \textit{pendular nystagmus}
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--If it is fast, the pattern is called a *jerk nystagmus*.
--If it is slow, it is known as a *pendular nystagmus*.

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.
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?
Nystagmus

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Can a nystagmus initiate with a fast movement?
No! By definition, nystagmus commences with a slow movement.
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Can a nystagmus initiate with a fast movement?
No! By definition, nystagmus commences with a slow movement. If a disordered eye-movement pattern commences with a fast movement, it is an intrusion (if intermittent) or a oscillation (if sustained).

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**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**?
No! By definition, nystagmus commences with a slow movement. If a disordered eye-movement pattern commences with a **fast movement**, it is a saccadic intrusion (if **intermittent**) or a saccadic oscillation (if **sustained**).

*What does it mean to say that saccadic intrusions are ‘intermittent’ and saccadic oscillations are ‘sustained’?*
Nystagmus

Saccadic Intrusion / Oscillation

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*What does it mean to say that saccadic intrusions are ‘intermittent’ and saccadic oscillations are ‘sustained’?*

In *saccadic intrusions*, the eyes fixate the target for a period of time...
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*What does it mean to say that saccadic intrusions are ‘intermittent’ and saccadic oscillations are ‘sustained’?*

In *saccadic intrusions*, the eyes fixate the target for a period of time… Then they saccade, and…
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What does it mean to say that saccadic intrusions are ‘intermittent’ and saccadic oscillations are ‘sustained’?
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…
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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… Saccade again, and… Fixate somewhere else for a period of time (note: it doesn’t have to be the original fixation target), until they…

(Somewhere else)  Direction of Gaze

(Intended target)  Time
**Nystagmus**

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**What does it mean to say that saccadic intrusions are ‘intermittent’ and saccadic oscillations are ‘sustained’?**

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…
Saccade again, and…

In *saccadic oscillations*, the eyes fixate somewhere else for a period of time (note: it doesn’t have to be the original fixation target), until they…
Saccade again, and…
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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… Saccade again, and… Fixate somewhere else for a period of time (note: it doesn’t have to be the original fixation target), until they… Saccade again, and… Etc.
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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…
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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**

**What is Nystagmus?**
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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 saccadic intrusion (if intermittent) or a saccadic oscillation (if sustained).

**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.)
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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,’ i.e., no time when the eyes are not saccading.
Nystagmus

Saccadic Intrusion/
Oscillation

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Give an example of a saccadic intrusion:
Nystagmus

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Give an example of a saccadic intrusion:
Square wave jerks
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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---

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

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

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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 / Oscillation

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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

The Peds book refers to these as “nystagmus-like disorders”
Nystagmus

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Note: Despite the fact that opsoclonus and convergence-retraction nystagmus are not nystagmuses, they will be addressed in this slide-set.

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

Two broad categories (not jerk and pendular)
Nystagmus

Two broad categories (not jerk and pendular)

Childhood

Acquired
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

Nystagmus

Childhood

Acquired

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.
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.

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

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

Oscillopsia
Nystagmus

Childhood

Acquired

?
Nystagmus

Childhood
- Congenital one sort
- Congenital opposite sort
  - known by the abb. ‘PAN’
  - not manifest
  - two funny words
  - the only monocular nystagmus of childhood

Acquired
Nystagmus

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

Acquired
Nystagmus

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

- 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

Congenital motor nystagmus
--Usually...horizontal

Nystagmus

Childhood

Congenital Motor

Congenital Sensory

Periodic Alternating

Latent

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Monocular Nystagmus of Childhood

Acquired

Nystagmus

- Congenital motor nystagmus:
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  - Remains horizontal in up/downgaze
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- Childhood
  - Congenital Motor
    - Congenital Sensory
    - Periodic Alternating
    - Latent
    - Spasmus Nutans
    - Monocular Nystagmus of Childhood

- Acquired
Congenital motor nystagmus
--Usually... horizontal
--Remains horizontal in up/downgaze
--Vision usually... [good vs bad]
Nystagmus

**Congenital motor nystagmus**
-- Usually **horizontal**
-- Remains horizontal in up/downgaze
-- Vision usually **good**

Nystagmus

Childhood

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

Acquired
**Nystagmus**

- **Congenital motor nystagmus**
  - Usually horizontal
  - Remains horizontal in up/downgaze
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---

**Nystagmus**

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

Childhood

**Congenital Motor**

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

Acquired
Nystagmus

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

**Nystagmus**

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

- Congenital Motor
  - Congenital Sensory
  - Periodic Alternating
  - Latent
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- Acquired

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

*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 congenital motor nystagmus, presentation of congruent OKN movement produces a dampening or even reversal of the nystagmus—hence the term **paradoxical OKN response**.
Nystagmus

**Congenital motor nystagmus**
-- Usually **horizontal**
-- Remains horizontal in up/downgaze
-- Vision usually **good**
-- Nystagmus + good VA = **congenital motor**
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*What is a paradoxical OKN response?*
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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 paradoxical OKN response
  - Likely to have a [nystagmus characteristic]

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

Childhood

Acquired
Nystagmus

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

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Nystagmus

Childhood

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

Acquired
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**

*What is a null point?*

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

---

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

---

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

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

*If a null point is not in primary (i.e., the straight ahead position), what will result?*
Nystagmus

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

*If a null point is not in primary (i.e., the straight ahead position), what will result?*
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
- Latent
- Spasmus Nutans
- Monocular Nystagmus of Childhood
Nystagmus

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

If a null point is not in primary (i.e., the straight ahead position), what will result?
A face turn, i.e., the patient will turn his/her head such that the null point is ‘aimed’ at an object of regard.

Is a face turn problematic?

---

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

Periodic Alternating

Latent

Spasmus Nutans

Monocular Nystagmus of Childhood
Nystagmus

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  --Remains horizontal in up/downgaze
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**What is a null point?**
A direction of gaze in which the intensity of the nystagmus is minimized

**If a null point is not in primary (i.e., the straight ahead position), what will result?**
A face turn, i.e., the patient will turn his/her head such that the null point is 'aimed' at an object of regard

**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)
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**

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

If a null point is not in primary (i.e., the straight ahead position), what will result?
A face turn, i.e., the patient will turn his/her head such that the null point is 'aimed' at an object of regard

If we’re talking about a jerk nystagmus, in which direction (ie, the fast phase, or the slow) is the null point typically found?

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

If a jerk nystagmus gets better in ‘slow phase gaze,’ what does this imply about its intensity in ‘fast phase gaze’?
That it gets worse
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

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

If a null point is not in primary (i.e., the straight ahead position), what will result?
A face turn, i.e., the patient will turn her head such that the null point is 'aimed' at an object of regard.

If we're talking about a jerk nystagmus, in which direction (i.e., the fast phase, or the slow) is the null point typically found?
The slow phase. So for example, in a left-beating jerk nystagmus (i.e., fast movement to the left, slow to the right), the null point will be located in right gaze. (Note that this means the patient will adopt a left face turn in order to place the null point directly in front of her.)
Nystagmus

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

*If a null point is not in primary (i.e., the straight ahead position), what will result?*
A face turn, i.e., the patient will turn their head such that the null point is 'aimed' at an object of regard.

*Is a face turn problematic?*
Yes, for multiple reasons— including socialization, cosmesis, and facial development.

*If we’re talking about a jerk nystagmus, in which direction (i.e., the fast phase, or the slow) is the null point typically found?*
The slow phase. So, for example, in a left-beating jerk nystagmus (i.e., fast movement to the left, slow to the right), the null point will be located in right gaze. (Note that this means the patient will adopt a left face turn in order to place the null point directly in front of them.)

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-- Remains horizontal in up/downgaze
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-- Nystagmus + good VA = congenital motor
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If a jerk nystagmus gets better in ‘slow phase gaze,’ what does this imply about its intensity in ‘fast phase gaze’?
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This observation is ubiquitous enough to have earned the title of 'law.' What is the eponymous name of this law?
Alexander's law.
Nystagmus

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In convergence

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?
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;
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**Congenital ocular motor apraxia (COMA)**

*What is the primary oculomotor deficit in COMA?*
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**Congenital ocular motor apraxia (COMA)**

*What is the primary oculomotor deficit in COMA?*
An inability to initiate horizontal saccades, which impedes horizontal pursuit (pursuit movements in infancy are actually a series of small saccades)
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You see a 3 y.o. toddler you previously diagnosed with COMA. At this visit, you notice her gait is poor, and that she has markedly dilated and tortuous conjunctival vessels in the interpalpebral zone (a new finding). At this point, you realize you have misdiagnosed this pt. What does she have?

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Ataxia-telangiectasia, aka Louis-Bar syndrome. The initial oculomotor findings in A-T involve an inability to initiate saccades, and thus are similar to COMA.

Reason, the infant seems to null-point in both directions.

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They’re not--their inability to follow objects with their eyes (prior to gaining control of head movements) makes them seem so.
Nystagmus

You see an infant who has been diagnosed with congenital motor nystagmus because he seems to engage in null-point viewing. Mom reports the infant seemed blind for the first several months of his life because he wouldn’t fixate on anything. Then, at around age 2 months, he started looking at things, but “only out of the side of his eyes” (ie, with a pronounced face-turn). On your exam, the child does face-turn; however, he face turns to the left when you move an object of regard to his right, and face-turns to the right when you move the object to the left. Additionally, you fail to appreciate any nystagmus. What is your diagnosis?

Congenital ocular motor apraxia (COMA)

You see a 3 y.o. toddler you previously diagnosed with COMA. At this visit, you notice her gait is poor, and that she has markedly dilated and tortuous conjunctival vessels in the interpalpebral zone (a new finding). At this point, you realize you have misdiagnosed this pt. What does she have?
Ataxia-telangiectasia, aka Louis-Bar syndrome. The initial oculomotor findings in A-T involve an inability to initiate saccades, and thus are similar to COMA.

reason, the infant seems to null-point in both directions.

Why are these infants blind before age 2 months?
They’re not--their inability to follow objects with their eyes (prior to gaining control of head movements) makes them seem so
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*In one word, what sort of condition is A-T?*

reason, the infant seems to null-point in both directions.

*Why are these infants blind before age 2 months?*

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A phakomatosis

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

A phakomatosis

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

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

Why are these infants blind before age 2 months?

They’re not--their inability to follow objects with their eyes (prior to gaining control of head movements) makes them seem so.
You see an infant who has been diagnosed with congenital motor nystagmus because he seems to engage in null-point viewing. Mom reports the infant seemed blind for the first several months of his life because he wouldn’t fixate on anything. Then, at around age 2 months, he started looking at things, but “only out of the side of his eyes” (ie, with a pronounced face-turn). On your exam, the child does face-turn; however, he face turns to the left when you move an object of regard to his right, and face-turns to the right when you move the object to the left. Additionally, you fail to appreciate any nystagmus. What is your diagnosis?

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Why are these infants blind before age 2 months?

They’re not–their inability to follow objects with their eyes (prior to gaining control of head movements) makes them seem so.
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*

**Sensory nystagmus**
-- 2ndry to early *laterality* poor vision

---

Childhood

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

**Congenital motor nystagmus**
-- Usually **horizontal**
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-- Only form with **paradoxical OKN response**
-- Likely to have a **null point**

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

---

**Nystagmus**

**Childhood**

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

---

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Nystagmus

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

**Sensory nystagmus**
--2ndry to early...bilateral...poor vision

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

A
A
A
A
A

Spasmus Nutans

Monocular Nystagmus of Childhood
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

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

*Weirdly, many of the causes of bilateral poor vision are associated with the letter ‘A’*
- Anterior segment issues (eg, congenital glaucoma, cataracts)
- Aniridia
- Leber’s congenital amaurosis
- Achromatopsia
- Albinism
- Optic nerve atrophy
- Aicardi syndrome

Spasmus Nutans
Monocular Nystagmus of Childhood

Optic nerve atrophy
Leber’s congenital amaurosis
Achromatopsia
Albinism
Optic nerve atrophy
Aicardi syndrome

Weirdly, many of the causes of bilateral poor vision are associated with the letter ‘A’:
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**Spasmus Nutans**

**Monocular Nystagmus of Childhood**

**Optic nerve atrophy**
Nystagmus

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-- Usually... horizontal
-- Remains horizontal in up/downgaze
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-- Likely to have a... *null point*

**Sensory nystagmus**
-- 2ndry to early... bilateral... poor vision

---

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

---

*Highly convenient mnemonic forthcoming...*
Nystagmus

**Congenital motor nystagmus**
-- Usually horizontal
-- Remains horizontal in up/down gaze
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**Sensory nystagmus**
-- 2ndry to early bilateral poor vision

---What are the findings of the Aicardi syndrome?
--- **Aicardi syndrome**
--- I's...
--- C:
--- A:
--- R:
--- D:
--- I:

--- Eye issue
--- CNS issue

---Cognitive issue
---Eye issue

---CNS issue

--- Optic nerve Atrophy
--- Spasmus Nutans
--- Monocular Nystagmus of Childhood

**Why are many of the causes of bilateral poor vision are associated with the letter ‘A’?**

--- Optic nerve
--- Achromatopsia
--- Aniridia
--- Albinism
--- Amaurosis
--- Anterior segment issues (eg, congenital glaucoma, cataracts)

--- Weirdly, many of the causes of bilateral poor vision are associated with the letter ‘A’:

--- Aicardi syndrome
--- Is...
--- C:
--- A:
--- R:
--- D:
--- I:

--- Eye issue
--- CNS issue

--- Cognitive issue
--- Eye issue

--- CNS issue

--- Aicardi syndrome
Nystagmus

What are the findings of the Aicardi syndrome?
-- Aicardi syndrome
-- Is...
-- Coloboma of the optic nerve
-- Agenesis of the corpus callosum
-- Retardation
-- Depigmented chorioretinal lacunae
-- Infantile seizures

Nystagmus

Sensory nystagmus
-- 2ndry to early bilateral poor vision

Congenital motor nystagmus
-- Usually... horizontal
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-- Likely to have a... null point

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**Nystagmus**

*Congenital motor nystagmus*
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-- Only form with... *paradoxical OKN response*
-- Likely to have a... *null point*

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

---

What are the findings of the *Aicardi syndrome*?
--- *Aicardi syndrome*
--- Is...
--- *Coloboma of the optic nerve*
--- *Agenesis of the corpus callosum*
--- *Retardation*
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*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

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

---**Congenital Motor**

---**Congenital Sensory**

---Periodic Alternating

---Latent

---Spasmus Nutans

---Monocular Nystagmus of Childhood
**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

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

**Childhood**

- Congenital Motor
  - Congenital Sensory
    - Periodic Alternating
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Nystagmus

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

---

**Childhood Nystagmus**

- **Congenital Motor**
- **Congenital Sensory**
- Periodic Alternating
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Nystagmus

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-- Secondary to early bilateral poor vision
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  -- 20/100 – 20/200: Pendular

---

**Childhood Spasmus Nutans**

**Monocular Nystagmus of Childhood**

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**
Nystagmus

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**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:

**Childhood Nystagmus**

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- Congenital Sensory
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**Sensory nystagmus**
-- Secondary to early bilateral poor vision
-- Waveform depends on visual acuity:
  -- 20/60 – 20/100: Jerk
  -- 20/100 – 20/200: Pendular
  -- <20/200: ‘Searching’

---

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**

**Monocular Nystagmus of Childhood**

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**Childhood**

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**Nystagmus**

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**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**’

**Periodic alternating nystagmus**
-- Jerk in one direction then... finally...
Nystagmus

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**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’**

**Periodic alternating nystagmus**
--Jerk in one direction → **slows** → **stops** → jerks in the other

**Monocular Nystagmus of Childhood**
--Jerk in one direction → **slows** → **stops** → jerks in the other
Nystagmus

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**Sensory nystagmus**
-- 2ndry to early bilateral poor vision
-- Waveform depends on visual acuity:
  -- 20/60 – 20/100: Jerk
  -- 20/100 – 20/200: Pendular
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**Periodic alternating nystagmus**
-- Jerk in one direction → slows → stops → jerks in the other
-- Changing null point may lead to alternating…

**Monocular Nystagmus of Childhood**

**Congenital Motor**

**Congenital Sensory**

**Latent**

**Spasmus Nutans**

**Childhood**
Nystagmus

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

**Periodic alternating nystagmus**
-- Jerk in one direction → slows → stops → jerks in the other
-- Changing null point may lead to alternating... face turn

---

Monocular Nystagmus of Childhood

Congenital Motor

Congenital Sensory

Periodic Alternating

Latent

Spasmus Nutans

Childhood

---

---
Nystagmus

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

**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... [**amount of time**]

---

**Childhood**

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

**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

**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

**Sensory nystagmus**
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
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--Usually... horizontal
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**Monocular Nystagmus of Childhood**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**
Nystagmus

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…

Congenital motor nystagmus
-- Usually… horizontal
  -- Remains horizontal in up/downgaze
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Sensory nystagmus
-- 2ndry to early bilateral poor vision
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Congenital Motor

Congenital Sensory

Periodic Alternating

Latent

Spasmus Nutans

Monocular Nystagmus of Childhood
Nystagmus

Childhood

Congenital Motor

Congenital Sensory

Periodic Alternating

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Monocular Nystagmus of Childhood

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**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*’

**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

---

111

---

Monocular Nystagmus of Childhood
Periodic alternating nystagmus
--Jerk in one direction → slows → stops → jerks in the other
--Changing null point may lead to alternating...
--Plane of action always...horizontal

Congenital motor nystagmus
--Usually...horizontal
--Remains horizontal in up/downgaze
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Sensory nystagmus
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
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Rule of thumb: If a nystagmus is purely horizontal...
Nystagmus

**Congenital motor nystagmus**
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**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'

**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**

**Rule of thumb:** If a nystagmus is purely horizontal...
observe it for at least 2 minutes to make sure it
doesn’t reverse directions!

---

**Monocular Nystagmus of Childhood**

**Acquired**

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**
Nystagmus

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
--Acquired: Associated with Arnold-Chiari malformation

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’

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Congenital Motor

Congenital Sensory

Periodic Alternating

Latent

Spasmus Nutans

Monocular Nystagmus of Childhood
Nystagmus

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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
- Acquired: Associated with… Arnold-Chiari malformation

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**Congenital Motor**

**Congenital Sensory**

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

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**Childhood**

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Nystagmus

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

**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’

**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

**Congenital motor nystagmus**
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  - 20/60 – 20/100: Jerk
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  - <20/200: ‘Searching’

**Latent**

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

**Monocular Nystagmus of Childhood**
Nystagmus

Periodic alternating nystagmus
--Jerk in one direction → slows → stops → jerks in the other
--Changing null point may lead to alternating...
--Plane of action always...horizontal
--Acquired: Associated with...Arnold-Chiari malformation

Congenital motor nystagmus
--Usually...horizontal
--Remains horizontal in up/downgaze
--Vision usually...good
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--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
  --<20/200: ‘Searching’

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]*

**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

**Congenital sensory nystagmus**
-- Secondary to early bilateral poor vision
-- Waveform depends on visual acuity:
  -- 20/60 – 20/100: *Jerk*
  -- 20/100 – 20/200: *Pendular*
  -- <20/200: *Searching*

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

**Spasmus Nutans**
-- Jerk nystagmus toward fixating eye
-- Only nystagmus to change direction with fixation

**Monocular Nystagmus of Childhood**
Nystagmus

**Latent nystagmus**
--Occurs when one eye is occluded

**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

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--Secondary to early bilateral poor vision
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  --20/60 – 20/100: Jerk
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--2ndry to early bilateral poor vision
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  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
  --<20/200: ‘Searching’

**Monocular Nystagmus of Childhood**

**Spasmus Nutans**
--Jerk nystagmus toward fixating eye
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--Usually horizontal
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Nystagmus

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

**Periodic alternating nystagmus**
-- In the other eye: face turn
-- Associated with Arnold-Chiari malformation

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

Monocular Nystagmus of Childhood

Spasmus Nutans
Nystagmus

Latent nystagmus
-- Occurs when one eye is occluded

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

Periodic alternating nystagmus
-- In the other eye, face turn
-- Plane of action always horizontal

Acquired

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

Monocular Nystagmus of Childhood

Congenital Motor

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
Nystagmus

**Latent nystagmus**
--Occurs when one eye is occluded

**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

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

**Periodic alternating nystagmus**
--Jerk in one direction
→ slows
→ stops
→ jerks in the other
--Changing null point may lead to alternating face turn
--Plane of action always horizontal

**Acquired nystagmus**

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**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.

**How can you get around this problem?**
Have the patient keep both eyes open, but fog the fellow eye with plus lenses.
Nystagmus

**Latent nystagmus**
--Occurs when one eye is...**occluded**

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**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**

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**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'**

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

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What ubiquitous exam component is likely to be spuriously poor in latent nystagmus?
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Have the patient keep both eyes open, but use plus lenses to fog the fellow eye
Periodic alternating nystagmus
--Jerk in one direction → slows → stops → jerks in the other
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--Plane of action always... horizontal
--Total cycle repeats every... 4 minutes or so
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--Only form with... paradoxical OKN response
--Likely to have a... null point

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’

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

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

**Latent nystagmus**
--Occurs when one eye is occluded
--Jerk nystagmus toward fixating eye

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--2ndry to early bilateral poor vision
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Childhood

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

Spasmus Nutans

Monocular Nystagmus of Childhood
**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

- **Sensory nystagmus**
  - 2ndry to early bilateral poor vision
  - Waveform depends on visual acuity:
    - 20/60 – 20/100: Jerk
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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
  - Acquired: Associated with Arnold-Chiari malformation

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

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

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

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

**What is latent manifest nystagmus?**

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Childhood

---

Monocular Nystagmus of Childhood

---

Acquired

---

Congenital Motor

---

Congenital Sensory

---

Periodic Alternating

---

Latent
Nystagmus

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- Jerk nystagmus toward... fixating eye
- Only nystagmus to change direction with fixation

**Sensory nystagmus**
- Secondary to early bilateral poor vision
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  - 20/60 – 20/100: *Jerk*
  - 20/100 – 20/200: *Pendular*
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**What is latent manifest nystagmus?**
A latent nystagmus that manifests when both eyes are open, but one is suppressed
Nystagmus

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

**Congenital motor nystagmus**
--Usually **horizontal**
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--Only form with **paradoxical OKN response**
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**Sensory nystagmus**
--Secondary to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: **Jerk**
  --20/100 – 20/200: **Pendular**
  --<20/200: ‘**Searching**’

**Periodic alternating nystagmus**
--Nystagmus that changes direction
  --Plane of action always **horizontal**
  --Changing null point may lead to alternating face turn
  --Total cycle repeats every 4 minutes or so

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

**What is ‘suppression’ in this context?**
It is the prevention of an image in one eye from reaching conscious awareness

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
Nystagmus

**Latent nystagmus**
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- Jerk nystagmus toward fixating eye
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**Acquired nystagmus**
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What is latent manifest nystagmus?
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- Changing null point may lead to alternating... face turn
- Total cycle repeats every... 4 minutes or so
- Plane of action always... horizontal
- Acquired: Associated with... Arnold-Chiari malformation

**Monocular Nystagmus of Childhood**

- **Spasmus Nutans**
  - Acquired
  - 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

**Sensory nystagmus**

- 2ndry to early bilateral poor vision
- Waveform depends on visual acuity:
  - 20/60 – 20/100: Jerk
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**What is latent manifest nystagmus?**

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

**What is ‘suppression’ in this context?**

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**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.
Nystagmus

Periodic alternating nystagmus
--Changes null point may lead to alternating...
--Plane of action always...
--Acquired: Associated with...

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

Congenital motor nystagmus
--Usually horizontal
--Remains horizontal in up/downgaze
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--Secondary to early bilateral poor vision
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What is 'suppression' in this context?
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What is latent manifest nystagmus?
A latent nystagmus that manifests when both eyes are open, but one is suppressed
**Nystagmus**

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

**Congenital motor nystagmus**
--Usually horizontal
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--Secondary to early bilateral poor vision
--Waveform depends on visual acuity:
  – 20/60 – 20/100: Jerk
  – 20/100 – 20/200: Pendular
  – <20/200: 'Searching'

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.

Latent nystagmus that manifests when both eyes are open, but one is suppressed.

**Spasmus Nutans**

**Monocular Nystagmus of Childhood**
Nystagmus

**Latent nystagmus**
-- Occurs when one eye is occluded
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**Visual confusion and diplopia? Aren’t those the same thing?**
No, not at all

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

**Latent nystagmus**
-- Occurs when one eye is **occluded**
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-- Usually **horizontal**
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-- Vision usually **good**
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-- 2ndry to early bilateral poor vision
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  -- 20/60 – 20/100: **Jerk**
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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?
A latent nystagmus that manifests when both eyes are open, but **one is suppressed**
Nystagmus

Latent nystagmus
--Occurs when one eye is occluded
--Jerk nystagmus toward fixating eye
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--Usually horizontal
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Sensory nystagmus
--Secondary to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
  --<20/200: 'Searching'

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

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

**Latent nystagmus**
-- Occurs when one eye is occluded
-- Jerk nystagmus toward fixating eye
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-- Secondary to early bilateral poor vision
-- Waveform depends on visual acuity:
  -- 20/60 – 20/100: Jerk
  -- 20/100 – 20/200: Pendular
  -- <20/200: 'Searching'

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 the 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
Nystagmus

Latent nystagmus
-- Occurs when one eye is... occluded
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  - 20/60 – 20/100: Jerk
  - 20/100 – 20/200: Pendular
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What is latent manifest nystagmus?
A latent nystagmus that manifests when both eyes are open, but one is suppressed

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.

Mnemonic is... SAM

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

three sensory adaptations

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

**Latent nystagmus**
--Occurs when one eye is...occluded
--Jerk nystagmus toward...fixating eye
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**Congenital motor nystagmus**
--Usually...horizontal
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**Sensory nystagmus**
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
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**What is latent nystagmus?**
A latent nystagmus that manifests when both eyes are open, but one is suppressed

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

**Latent nystagmus**
- Occurs when one eye is occluded
- Jerk nystagmus toward fixating eye
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**Sensory nystagmus**
- Secondary to early bilateral poor vision
- Waveform depends on visual acuity:
  - 20/60 – 20/100: Jerk
  - 20/100 – 20/200: Pendular
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**Periodic alternating nystagmus**
- Jerk in one direction → slows → stops → jerks in the other
- Null point may lead to alternating face turn
- Cycle repeats every 4 minutes or so
- Plane of action always horizontal
- Acquired: Associated with Arnold-Chiari malformation

**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

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

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

**Monocular Nystagmus of Childhood**

**Spasmus Nautans**

**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

**Congenital sensory nystagmus**
- Jerk toward fixating eye
- Only nystagmus to change direction with fixation
- Null point

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

**Fusion maldevelopment nystagmus syndrome (FMNS)**

**Childhood**
**Nystagmus**

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

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

**Periodic alternating nystagmus**
- Jerk in one direction slows stops jerks in the other
- Null point may lead to alternating face turn
- Plane of action always horizontal
- Associated with Arnold-Chiari malformation

**Congenital motor nystagmus**
- Usually horizontal
- Remains horizontal in up/down gaze
- Vision usually good
- Nystagmus + good VA = congenital motor
- Likely to have a null point

**Fusion maldevelopment nystagmus syndrome (FMNS)**

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

**Fusion maldevelopment nystagmus syndrome (FMNS)**

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

- **Congenital 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’

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

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

- **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 Spasmus Nutans**

- Triad of: **dumb**, **two words**, **one word**
**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis

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

**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

**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’

**Periodic alternating nystagmus**
--Jerk in one direction slows stops jerks in the other
--Changing null point may lead to alternating face turn
--Plane of action always horizontal
--Acquired: Associated with Arnold-Chiari malformation

**Monocular Nystagmus of Childhood**

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**
**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very small

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

**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

**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’

**Periodic alternating nystagmus**
--Jerk in one direction \( \rightarrow \) slows \( \rightarrow \) stops \( \rightarrow \) jerks in the other
--Changing null point may lead to alternating…face turn
--Plane of action always horizontal
--Acquired: Associated with Arnold-Chiari malformation

**Monocular Nystagmus of 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  

**Congenital 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**’  

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

**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**’  

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

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

**Sensory nystagmus**
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
  --20/100 – 20/200: Pendular
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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
--Acquired: Associated with...Arnold-Chiari malformation

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

**Sensory nystagmus**
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
-20/60 – 20/100: Jerk
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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
--Plane of action always...horizontal
--Total cycle repeats every...4 minutes or so
--Acquired: Associated with...Arnold-Chiari malformation

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**

**Monocular Nystagmus of 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

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

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

Latent

Monocular Nystagmus of Childhood

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)
Monocular Nystagmus of Childhood

Spasmus nutans
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very small (‘shimmer’)
--Usually bilateral, but **can seem unilateral**

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)

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

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

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

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

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

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

Congenital Sensory

Latent

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

**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’

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

**Monocular Nystagmus of Childhood**

---

**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

**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’

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

---

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

---

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

Latent nystagmus
→ Jerk nystagmus toward fixating eye
→ Only nystagmus to change direction with fixation

Monocular Nystagmus of Childhood

Congenital Sensory Nystagmus
-- Short bursts
-- Occurs when one eye is occluded
-- Jerk nystagmus toward fixating eye

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

Congenital motor nystagmus
-- Only form with paradoxical OKN response
-- Likely to have a null point
-- Plane of action always horizontal

Congenital motor nystagmus
-- Usually horizontal in up/down gaze
-- Vision usually good

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

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

Congenital Alternating Sensory Nystagmus
-- Plane of action always horizontal
**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very small (‘shimmer’)
--Usually bilateral, but can seem unilateral
--Benign. Resolves by age

**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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--Usually horizontal
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--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

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

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

**Monocular Nystagmus of Childhood**

**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very small (‘shimmer’)
--Usually bilateral, but can seem unilateral
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--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

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

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

**Monocular Nystagmus of Childhood**
**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...

**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

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

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

--Monocular Nystagmus of Childhood
**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...* 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

**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*’

**Periodic alternating nystagmus**
--Jerk in one direction→slows→stops→jerks in the other
--Changing null point may lead to alternating... face turn
--Plane of action always... horizontal
--Acquired: Associated with... Arnold-Chiari malformation

**Monocular Nystagmus of Childhood**

**Childhood**

**Congenital Motor**

**Congenital Sensory**

**Periodic Alternating**

**Latent**

**Spasmus Nutans**
**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

**Sensory nystagmus**
--2ndry to early bilateral poor vision
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Total cycle repeats every...4 minutes or so
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Monocular Nystagmus of Childhood
Spasmus nutans
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very...small ("shimmer")
--Usually bilateral, but can seem unilateral
--Resolves by age 3-4 years. But...

Glioma

Where might such a glioma be located?
--Jerk nystagmus toward...fixating eye
--Only nystagmus to change direction with fixation

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

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

Sensory nystagmus
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
--20/60 – 20/100: Jerk
--20/100 – 20/200: Pendular
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Monocular Nystagmus of Childhood
Spasmus nutans
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very small ('shimmer')
--Usually bilateral, but can seem unilateral
--Resolves by age 3-4 years. But…
--Can present similarly, so image

Glioma

Where might such a glioma be located?
In the anterior visual pathway, ie, the optic nerve or chiasm
--Jerk nystagmus toward fixating eye
--Only nystagmus to change direction with fixation

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

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

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

Sensory nystagmus
--2ndry to early bilateral poor vision
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--Jerk in one direction → slows → stops → jerks in the other
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Monocular Nystagmus of Childhood
Spasmus nutans
--Triad of nystagmus + head nodding + torticollis
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--- Congenital sensory nystagmus
--Secondary to early bilateral poor vision
--Waveform depends on visual acuity:
  - 20/60 – 20/100: Jerk
  - 20/100 – 20/200: Pendular
  - <20/200: 'Searching'

--- Sensory nystagmus

--- Congenital motor nystagmus

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

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

--- Latent nystagmus

--- Monocular Nystagmus of Childhood

--- Spasmus Nutans

--- Congenital motor nystagmus

--- Acquired nystagmus

--- Periodic alternating nystagmus

--- Childhood nystagmus

--- Latent nystagmus

--- Sensory nystagmus

--- Glioma
**Spasmus nutans**

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

**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’

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

**Monocular nystagmus of childhood**

--Is...[rare vs common]
**Periodic alternating nystagmus**
--Jerk in one direction → slows → stops → jerks in the other
--Changing null point may lead to alternating... face turn
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**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very... small ('shimmer')
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**Monocular nystagmus of childhood**
--Is... rare

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

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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
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**Sensory nystagmus**
--2ndry to early bilateral poor vision
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**Monocular nystagmus of childhood**
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--Direction is vertical, or elliptical
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--Neuroimage if…
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Sensory nystagmus
--2ndry to early... bilateral... poor vision
--Waveform depends on visual acuity:
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Monocular Nystagmus of Childhood

What might these signs be?
**Periodic alternating nystagmus**
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- 2ndry to early...bilateral poor vision
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**Monocular nystagmus of childhood**
- Is...rare
- Direction is...vertical, or elliptical
- Involved eye...never changes
- Neuroimage if...signs of anterior pathway disease present

**Congenital motor nystagmus**
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**Congenital 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'

**Latent nystagmus**
- Occurs when one eye is...occluded
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*What might these signs be?*
An APD; ONH atrophy on DFE
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**Sensory nystagmus**
--2ndry to early...bilateral...poor vision
--Waveform depends on visual acuity:
--20/60 – 20/100: **Jerk**
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--Is...rare
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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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**Monocular nystagmus of childhood**
--Is rare
--Direction is vertical, or elliptical
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**Sensory nystagmus**
--Secondary to early bilateral poor vision
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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
--Acquired: Associated with Arnold-Chiari malformation

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’)?

It is referred to as the Heimann-Bielschowsky phenomenon
**Spasmus nutans**
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  --<20/200: 'Searching'

**Periodic alternating nystagmus**
--Steps→jerks in the other
--Changing null point may lead to alternating...face turn
--Total cycle repeats every...minutes or so
--Plane of action always...horizontal
--Acquired: Associated with...Arnold-Chiari malformation

**Bielschowsky...That name sounds familiar. Where have I heard it before?**

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')? It is referred to as ‘the Heimann-Bielschowsky phenomenon’
**Periodic alternating nystagmus**
--Jerk in one direction
  --slows
  --stops
  --jerks in the other
--Changing null point may lead to alternating...
--Total cycle repeats every...
  --4 minutes or so
--Plane of action always...
--Horizontal
--Acquired: Associated with...
  --Arnold-Chiari malformation

---

**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

---

**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

---

**Monocular nystagmus of childhood**
Acquired

---

**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'

---

**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’)?**
It is referred to as ‘the Heimann-Bielschowsky phenomenon’
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
--Acquired: Associated with...
Arnold-Chiari malformation

Monocular Nystagmus of Childhood
Acquired
Congenital Motor
Congenital Sensory
Periodic Alternating

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

Latent nystagmus
--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

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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**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

**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

**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 the fixating eye
- Only nystagmus to change direction with fixation

**Sensory nystagmus**

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

**Bielschowsky phenomenon**

- 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 muscle responsible for a...
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
--Acquired: Associated with Arnold-Chiari malformation

Monocular nystagmus of childhood
Acquired nystagmus
Congenital motor nystagmus
Congenital sensory nystagmus

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

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

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

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

Alternative nystagmus

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

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

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

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**Periodic alternating nystagmus**
--Jerk in one direction → slows → stops → jerks in the other
--Changing null point may lead to alternating...
--Plane of action always...
--Acquired: Associated with...
--Total cycle repeats every...
4 minutes or so
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**Latent nystagmus**
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--Jerk nystagmus toward...fixating eye
--Only nystagmus to change direction with fixation

**Spasmus nutans**
--Triad of nystagmus + head nodding + torticollis
--Nystagmus amplitude very...small (‘shimmer’) / Usually bilateral
--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

**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'

**Congenital motor nystagmus**
--Usually...
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--Vision usually...good
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**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 (note that this step is called the Bielschowsky head-tilt test)

**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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### Periodic alternating nystagmus

--Jerk in one direction
→ slows → stops → jerks in the other
--Changing null point may lead to alternating...

### Changing null point may lead to alternating...

--Plane of action always...

### Acquired: Associated with Arnold-Chiari malformation

### Latent nystagmus

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

---

### 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'*

---

### 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... via cover testing
**Step 2:**
**Step 3:**

---

### Can...? Yes

What...good

It is referred to as ‘the Heimann-Bielschowsky phenomenon’
**Spasmus nutans**
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**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

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--Secondary to early bilateral poor vision
--Waveform depends on visual acuity:
  --20/60 – 20/100: Jerk
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  --<20/200: ‘Searching’

**Periodic alternating nystagmus**
--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

**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:**
**Step 3:**

**Sparing**
--Involved eye never changes
--Neuroimaging if signs of anterior pathway disease present

**Heimann-Bielschowsky phenomenon**

**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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Periodic alternating nystagmus

- Jerk in one direction → slows → stops → jerks in the other
- Changing null point may lead to alternating...
- Plane of action always horizontal
- Acquired: Associated with Arnold-Chiari malformation

Latent nystagmus

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

Monocular Nystagmus of Childhood

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

Congenital Sensory Nystagmus

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

Sensory nystagmus

Spasmus nutans

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Latent nystagmus

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Step 1: Determine which eye is higher via cover testing
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Step 3:

What a good reminder you are. It is referred to as 'the Heimann-Bielschowsky phenomenon'
Periodic alternating nystagmus
--Jerk in one direction → slows → stops → jerks in the other → Changing null point may lead to alternating...

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Monocular Nystagmus of Childhood

Acquired
Congenital Motor
Congenital Sensory

Periodic Alternating Nystagmus

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

Childhood

Conagenital Motor

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

Can you name the disease that caused her condition?
Yes

What is the name of the disease?
Arnold-Chiari malformation
--Involved eye... never changes
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**Periodic alternating nystagmus**
--Jerk in one direction → slows → stops → jerks in the other
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**Latent nystagmus**
--Occurs when one eye is… occluded
--Jerk nystagmus toward… fixating eye
--Only nystagmus to change direction with fixation

**Sensory nystagmus**
--2ndry to early bilateral poor vision
--Waveform depends on visual acuity:
  --<20/60: jerk
  --20/60 – 20/100: pendular
  --<20/200: ‘Searching’

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**Step 2:** Identify the gaze (ie, right vs left) in which the deviation gets worse
**Step 3:** Tilt the head toward each bodypart, and see which direction worsens v improves

---

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**The Heimann-Bielschowsky Phenomenon**

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**Monocular nystagmus of childhood**

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