



Q

## Miscellaneous Neuro

- type of *nystagmus* usually indicates a cerebellar or cervical-medullary problem



# A

## Miscellaneous Neuro

- *Downbeat* nystagmus usually indicates a cerebellar or cervical-medullary problem



# A

## Miscellaneous Neuro

- *Downbeat* nystagmus usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - 
  -



# Q

## Miscellaneous Neuro

- **Downbeat** nystagmus usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - eponym-eponym malformation
  - one word atrophy



# A

## Miscellaneous Neuro

- **Downbeat** nystagmus usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy



# Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari malformation**
  - Cerebellar atrophy

*In brief, what constitutes the Arnold-Chiari malformation?*



# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari malformation**
  - Cerebellar atrophy

*In brief, what constitutes the Arnold-Chiari malformation?*

The herniation of the cerebellar tonsils into the foramen magnum, with resulting compression of the cervico-medullary junction. There are several variants classified according to the degree of herniation and severity of the sequelae.



# Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a anatomical site 1 or anatomical site 2 problem





# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem



# Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **symptom** is highly steroid-responsive; however, the **sign** problem can take **amount of time** to resolve.



# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve.



# Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

What is the Tolosa-Hunt syndrome?



# Q/A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*

two words--a symptom, and a sign

secondary to

a two-word cause

of the

a two-word anatomic location



# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** parasellar or 3<sup>rd</sup> ventricle
- Re **Tolosa-Hunt**. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*  
Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus



Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*  
Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*



# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*

Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*

Very rare (literally, 1 in a million rare)





Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** parasellar or 3<sup>rd</sup> ventricle
- Re **Tolosa-Hunt**. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*

Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*

Very rare (literally, 1 in a million rare)

*Is there an age predilection?*



# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*

Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*

Very rare (literally, 1 in a million rare)

*Is there an age predilection?*

No



Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*  
Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*

Very rare (literally, 1 in a million rare)

*Is there an age predilection?*

No

*Is there a gender predilection?*



# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* parasellar or 3<sup>rd</sup> ventricle
- Re ***Tolosa-Hunt***. T responsive; however can take weeks to r

*What is the Tolosa-Hunt syndrome?*  
Painful ophthalmoplegia secondary to noninfectious inflammation of the cavernous sinus

*Is it common, or rare?*

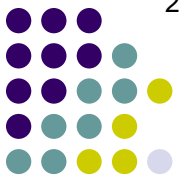
Very rare (literally, 1 in a million rare)

*Is there an age predilection?*

No

*Is there a gender predilection?*

No

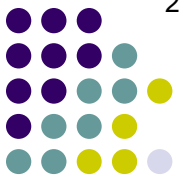


## Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re **Tolosa-Hunt**. The **pain** is highly steroid-responsive; however, the **motility** problem

*The (well-deserved) reputation for being steroid-responsive enjoyed by Tolosa-Hunt can be highly misleading--why?*



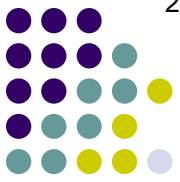
# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re **Tolosa-Hunt**. The **pain** is highly steroid-responsive; however, the motility problem

*The (well-deserved) reputation for being steroid-responsive enjoyed by Tolosa-Hunt can be highly misleading--why?*

Because other, far more common causes of painful ophthalmoplegia are steroid-responsive too. So **steroid-responsiveness should not be interpreted as confirming the diagnosis of Tolosa-Hunt.**



Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re **Tolosa-Hunt**. The **pain** is highly steroid-responsive; however, the **motility** problem

The (well-deserved) reputation for being steroid-responsive enjoyed by Tolosa-Hunt can be highly misleading--why?

Because other, far more common causes of painful ophthalmoplegia are steroid-responsive too. So steroid-responsiveness should not be interpreted as confirming the diagnosis of Tolosa-Hunt.

What are some of these causes?

--  
--  
--



# Q/A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- **Re Tolosa-Hunt.** The **pain** is highly steroid-responsive; however, the **motility** problem

The (well-deserved) reputation for being steroid-responsive enjoyed by Tolosa-Hunt can be highly misleading--why?

Because other, far more common causes of painful ophthalmoplegia are steroid-responsive too. So steroid-responsiveness should not be interpreted as confirming the diagnosis of Tolosa-Hunt.

What are some of these causes?

--Neoplasms, especially

one word

--Vascular lesions, especially

abb.

--Infections, especially

class of  
bug

(eg,

member  
of class

)





# A

## Miscellaneous Neuro

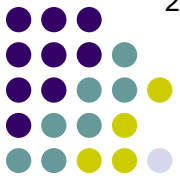
- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- **Re Tolosa-Hunt**. The pain is highly steroid-responsive; however, the motility problem

The (well-deserved) reputation for being steroid-responsive enjoyed by Tolosa-Hunt can be highly misleading--why?

Because other, far more common causes of painful ophthalmoplegia are steroid-responsive too. So steroid-responsiveness should not be interpreted as confirming the diagnosis of Tolosa-Hunt.

What are some of these causes?

- Neoplasms, especially lymphoma
- Vascular lesions, especially CCF
- Infections, especially fungal (eg, *Mucor*)



## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- **See-saw nystagmus** parasellar or 3<sup>rd</sup> ventricle

**The takeaway point:** Tolosa-Hunt, being so much rarer than its imitators, should not be diagnosed by anyone other than a trained neuro-oph. Refer all suspected cases! (BTW, that's not me talking--it's straight from Dr Andrew Lee.)

What is the Tolosa-Hunt syndrome?  
Painful ophthalmoplegia secondary to noninfectious inflammation of the

sinus

n, or rare?

terally, **1 in a million rare**

age predilection?

Is there a gender predilection?

No

*What are some of these causes?*

--Neoplasms, especially lymphoma

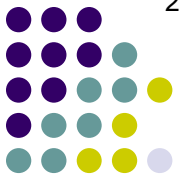
--Vascular lesions, especially CCF

--Infections, especially fungal (eg, *Mucor*)



## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve
- In *CPEO*:

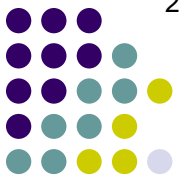


# Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to resolve
- In **CPEO**:

*In this context, what does CPEO stand for?*



# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to resolve
- In **CPEO**:

*In this context, what does CPEO stand for?*

Chronic progressive external ophthalmoplegia



# Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to resolve
- In **CPEO**:
  - In this context, what does CPEO stand for?*  
Chronic progressive external ophthalmoplegia
  - Briefly, what is it?*



# A

## Miscellaneous Neuro

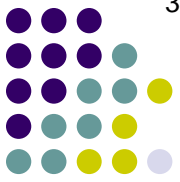
- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to resolve
- In **CPEO**:

*In this context, what does CPEO stand for?*

Chronic progressive external ophthalmoplegia

*Briefly, what is it?*

A mitochondrial disorder characterized by relentlessly progressive, symmetric paralysis of the extraocular muscles



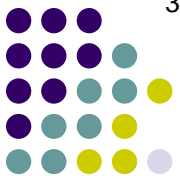
## Q

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve
- In *CPEO*:
  - Bell's phenomenon...is 

present v absent
---------------------





# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve
- In *CPEO*:
  - Bell's phenomenon...is **absent**



Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*: The *pain* is highly steroid-responsive; however, the motility problem

*In this context, what is Bell's phenomenon?*

- In *CPEO*:
  - **Bell's phenomenon**... is absent



# A

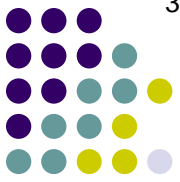
## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the motility problem

*In this context, what is Bell's phenomenon?*

Involuntary elevation of the eye when attempted lid closure is thwarted (ie, the eye rolls up if you hold the lids open during attempted closure)

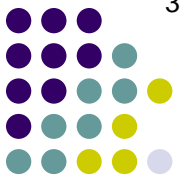
- In *CPEO*:
  - **Bell's phenomenon**... is absent



# Q

## Miscellaneous Neuro

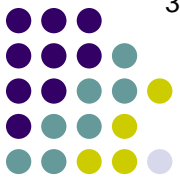
- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve
- In *CPEO*:
  - Bell's phenomenon...is **absent**
  - Patients  complain of diplopia



# A

## Miscellaneous Neuro

- **Downbeat nystagmus** usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - **Arnold-Chiari** malformation
  - **Cerebellar** atrophy
- **See-saw nystagmus** usually indicates a **parasellar** or **3<sup>rd</sup> ventricle** problem
- Re *Tolosa-Hunt*. The **pain** is highly steroid-responsive; however, the **motility** problem can take **weeks to months** to resolve
- In *CPEO*:
  - Bell's phenomenon...is **absent**
  - Patients **do not** complain of diplopia



# Q

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to months to resolve
- In *CF*
  - Bell's phenomenon... is absent
  - Patients **do not** complain of diplopia

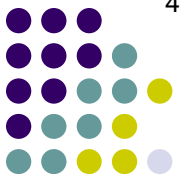
Why don't CPEO pts c/o diplopia?



# A

## Miscellaneous Neuro

- *Downbeat nystagmus* usually indicates a cerebellar or cervical-medullary problem; e.g.:
  - Arnold-Chiari malformation
  - Cerebellar atrophy
- *See-saw nystagmus* usually indicates a parasellar or 3<sup>rd</sup> ventricle problem
- Re *Tolosa-Hunt*. The *pain* is highly steroid-responsive; however, the *motility* problem can take weeks to months to resolve
- In *CF*
  - Why don't CPEO pts c/o diplopia? Because as mentioned, the ophthalmoplegia is symmetric, and thus both globes are similarly limited in their ability to move
  - Bell's phenomenon... is absent
  - Patients **do not complain of diplopia**

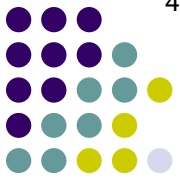


# Q

## *Miscellaneous Neuro*

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?



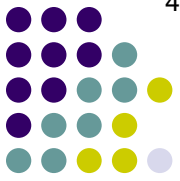


# A

## *Miscellaneous Neuro*

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?

**Infantile botulism**



# Q

## *Miscellaneous Neuro*

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?

**Infantile botulism**

- What is the classic history?



# A/Q

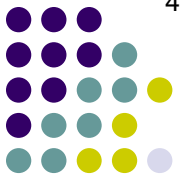
## Miscellaneous Neuro

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?

**Infantile botulism**

- What is the classic history? **Ingestion of...**

two words



# A

## Miscellaneous Neuro

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?

**Infantile botulism**

- What is the classic history? **Ingestion of...**  
***raw honey***



# Q

## Miscellaneous Neuro

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?

**Infantile botulism**

- What is the classic history? **Ingestion of...**  
***raw honey***

- What is the treatment?



# A

## Miscellaneous Neuro

- What tops the differential for an infant with rapid onset of bilateral ophthalmoplegia + dilated fixed pupils + ptosis + bulbar weakness?  
**Infantile botulism**
- What is the classic history? **Ingestion of...**  
***raw honey***
- What is the treatment? **Supportive**