

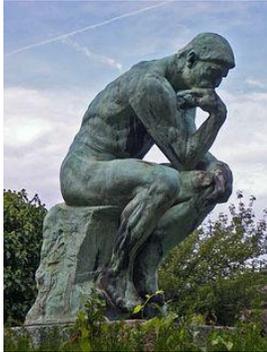
EYE MOVEMENTS IN DEGENERATIVE DISEASES OF THE CEREBELLUM : A VIDEO TUTORIAL



Cerebellar atrophy: SCA6

BEFORE WE START:

- Is CEREBELLAR ANATOMY complex?
 - YES, but it is orderly and manageable.
- Is CEREBELLAR PHYSIOLOGY complex?
 - YES, highly complex, and it can be OVERWHELMING
- But can we extract from the anatomy and physiology easy to use, easy to remember, practical principles for our clinics?
 - YES, WITHOUT A DOUBT!
- Do we need fancy equipment and detailed quantitative analysis to evaluate eye movement disorders?
 - ABSOLUTELY NOT! The focused history and careful, ordered bedside exam usually points us to the diagnosis, and always directs us down the correct path for management. (Testing of course helps to confirm our diagnoses, and provides data for research BUT)
- **HANG IN THERE!** Rather than thinking that eye movements take you to the “**Gates of Hell**”, you will receive “**The Kiss**” from your patients at the end of the day.



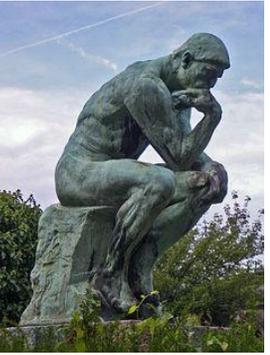
Auguste
Rodin



“Gates of Hell”



“The Kiss”



**Auguste
Rodin**

A Road Map for Today

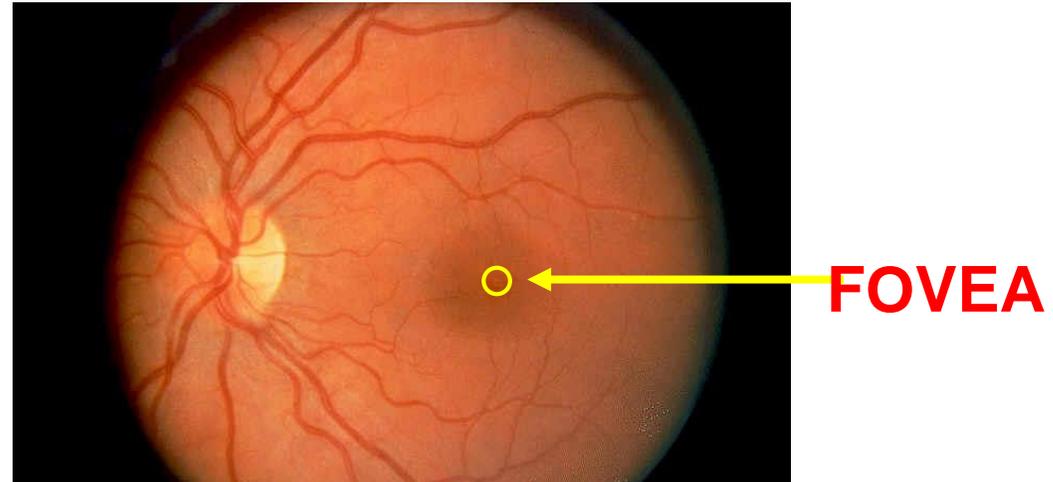


A Road Map for Today

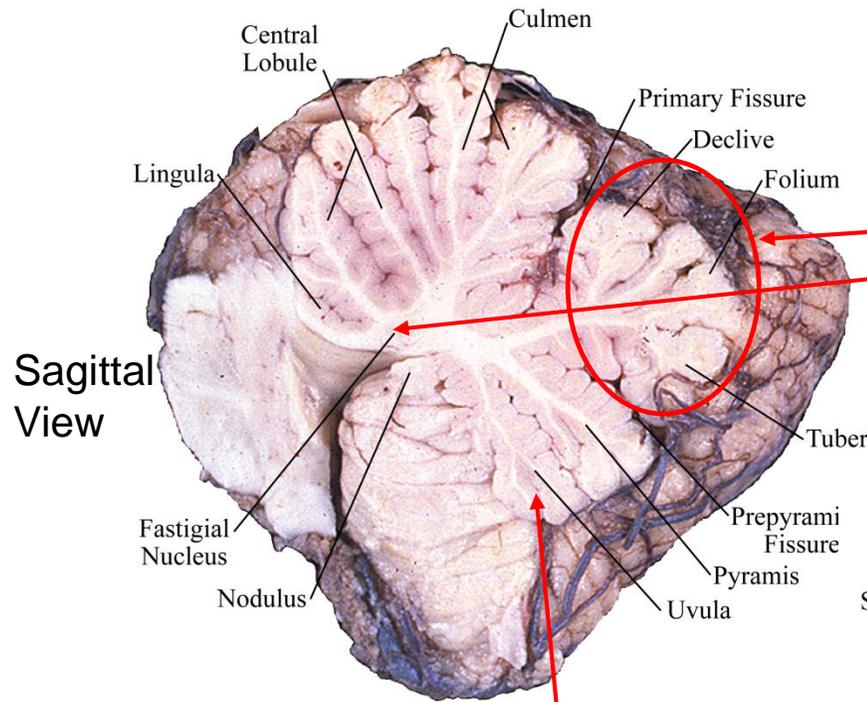
- **Key cerebellar anatomy**
- **Flocculus and paraflocculus (tonsil): downbeat, gaze-evoked, rebound nystagmus**
- **Bedside vestibular testing in cerebellar patients**
 - **Head impulse**
 - **Head-shaking**
 - **Hyperventilation**
 - **Valsalva**
- **Nodulus: periodic alternating nystagmus**
- **Dorsal vermis and fastigial nucleus: Saccade dysmetria**
- **Strabismus and the cerebellum (ESO and HYPER deviations)**

Why do we need eye movements?

Sharp, detailed vision is possible only at the **fovea**, the small area at the center of the retina, and when images are **held steady** there.

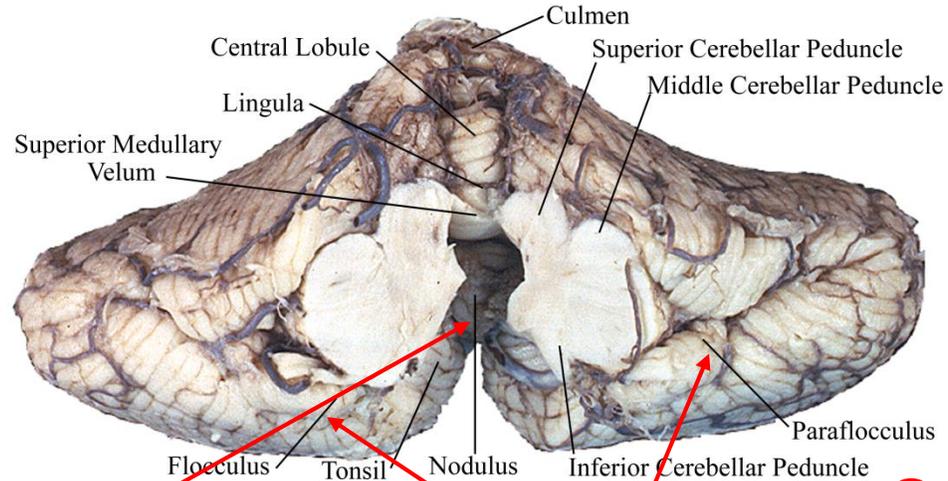


KEY ANATOMY: Three basic functional-anatomical cerebellar syndromes



Syndrome of the dorsal vermis (OMV) & posterior fastigial nucleus (FOR)

Saccade accuracy



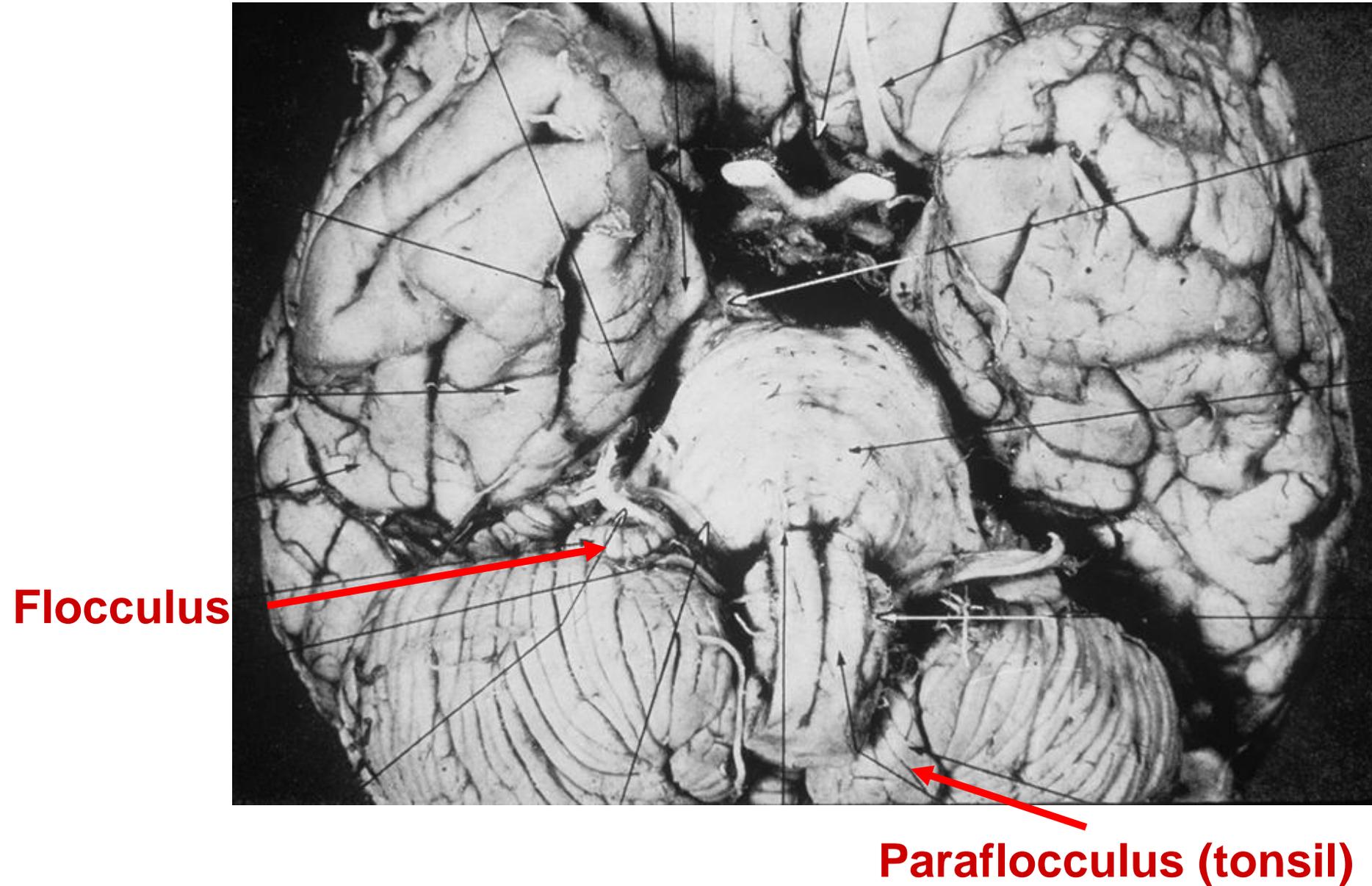
Vestibular (low-freq, slowly changing)

Syndrome of the nodulus & ventral uvula

Syndrome of the flocculus and paraflocculus (tonsil)

Gaze-holding, pursuit, vestibular (high-freq, rapidly changing)

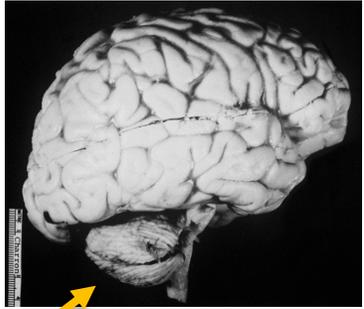
Cerebellar flocculus and paraflocculus (tonsils)



Flocculus

Paraflocculus (tonsil)

Flocculus/Paraflocculus syndrome: Downbeat, gaze-evoked and rebound nystagmus in cerebellar atrophy



Cerebellar atrophy: SCA6



Flocculus/Paraflocculus syndrome

Impaired pursuit and vestibuloocular reflex cancellation (fixation suppression)

Pursuit and VOR
cancellation

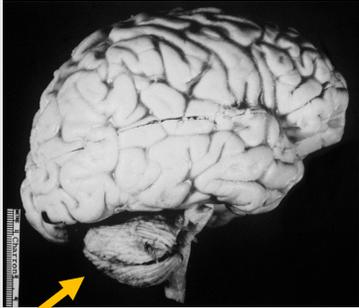


Testing of the VOR: Head impulse sign in a unilateral peripheral labyrinthine lesions

**Catch-up saccade
during brief, high-
acceleration, head
rotation (left-sided
loss)**

**Head-impulse sign in
unilateral labyrinthine
loss**

Abnormal VOR in cerebellar disease: Abnormal direction



Head-shaking nystagmus (HSN) in cerebellar disease



PEARL: Think central if HSN is

- Directed **DIFFERENTLY** than head motion (cross-coupled), e.g, vertical nystagmus with horizontal head-shaking.
- Directed **OPPOSITE** to spontaneous nystagmus
- If there is an **EARLY** and **STRONG REVERSAL** of the direction of HSN

Hyperventilation-induced (HVN) downbeat nystagmus

PEARL: HVN

- **Cranial-cervical junction anomalies**
- **Cerebellar degenerations**

- **Compressive lesions on VIII CN
(microvascular compression, tumors)**

- **Demyelinating diseases (e.g., MS)**

- **Labyrinthine fistula and SCC dehiscence**

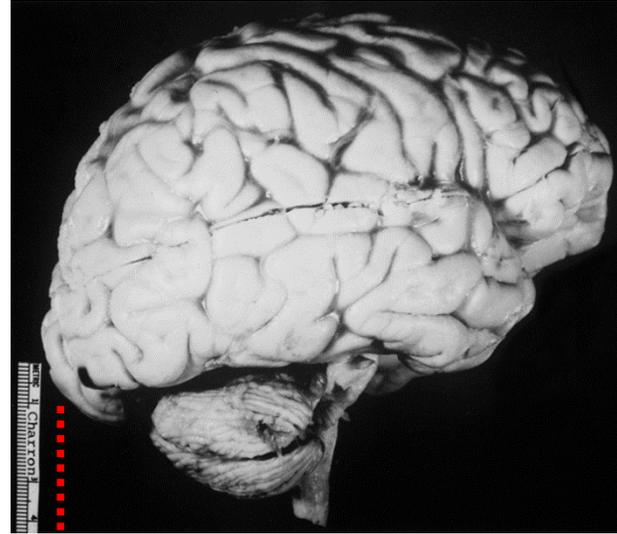
Valsalva-induced vertigo



Valsalva-induced nystagmus

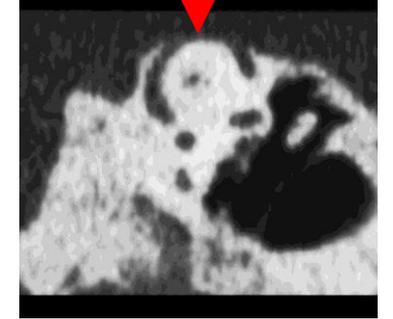
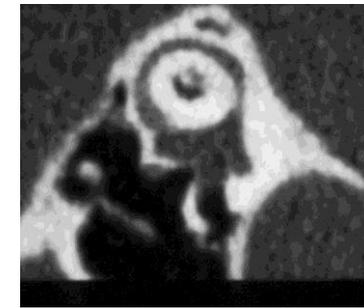


**Cranial-cervical junction:
Chiari**



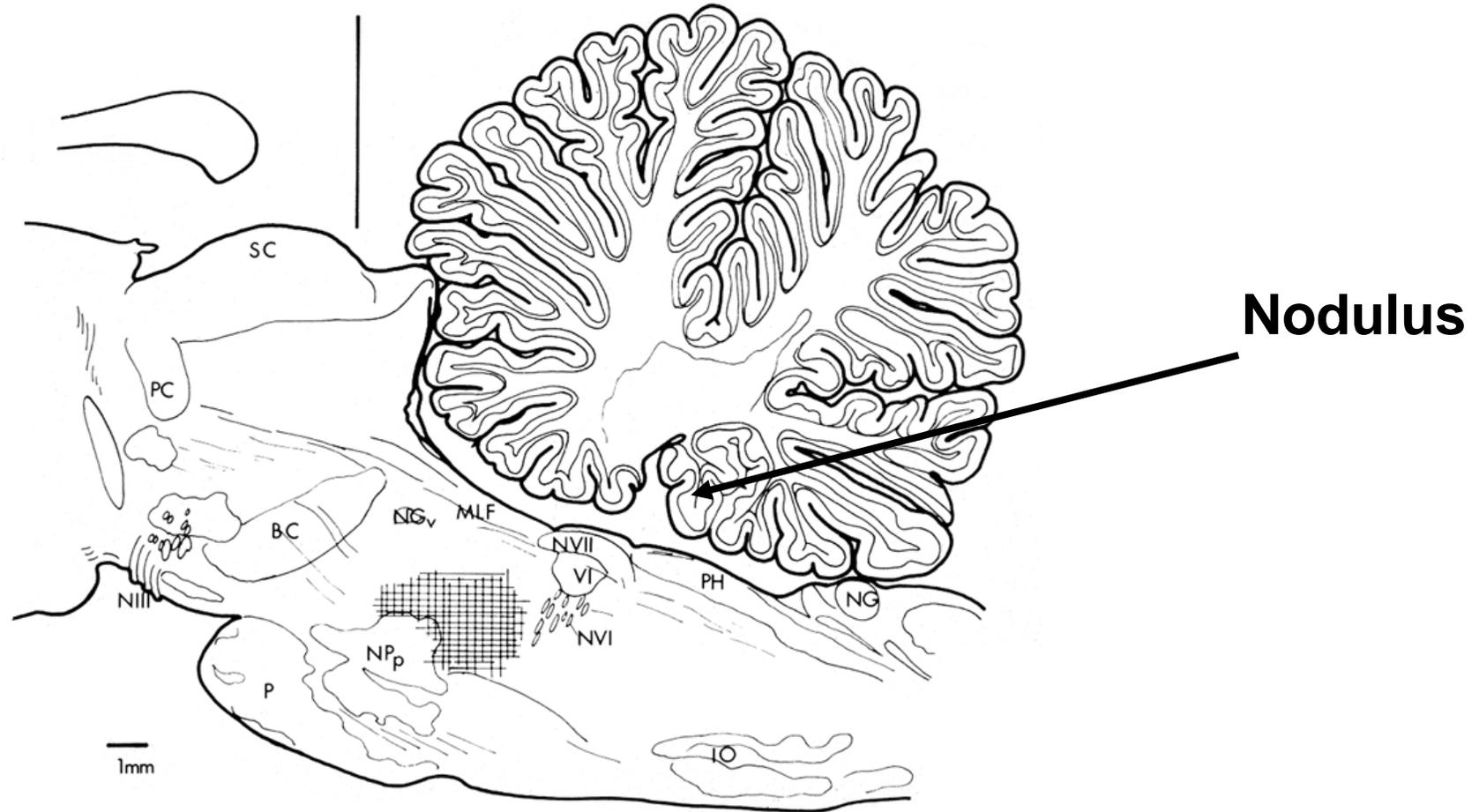
**Cerebellar atrophy:
SCA6**

Superior Semicircular Canal dehiscence



PEARL: Remember Valsalva-induced vertigo with cranial-cervical junction anomalies and with labyrinthine fistula and SCC dehiscence

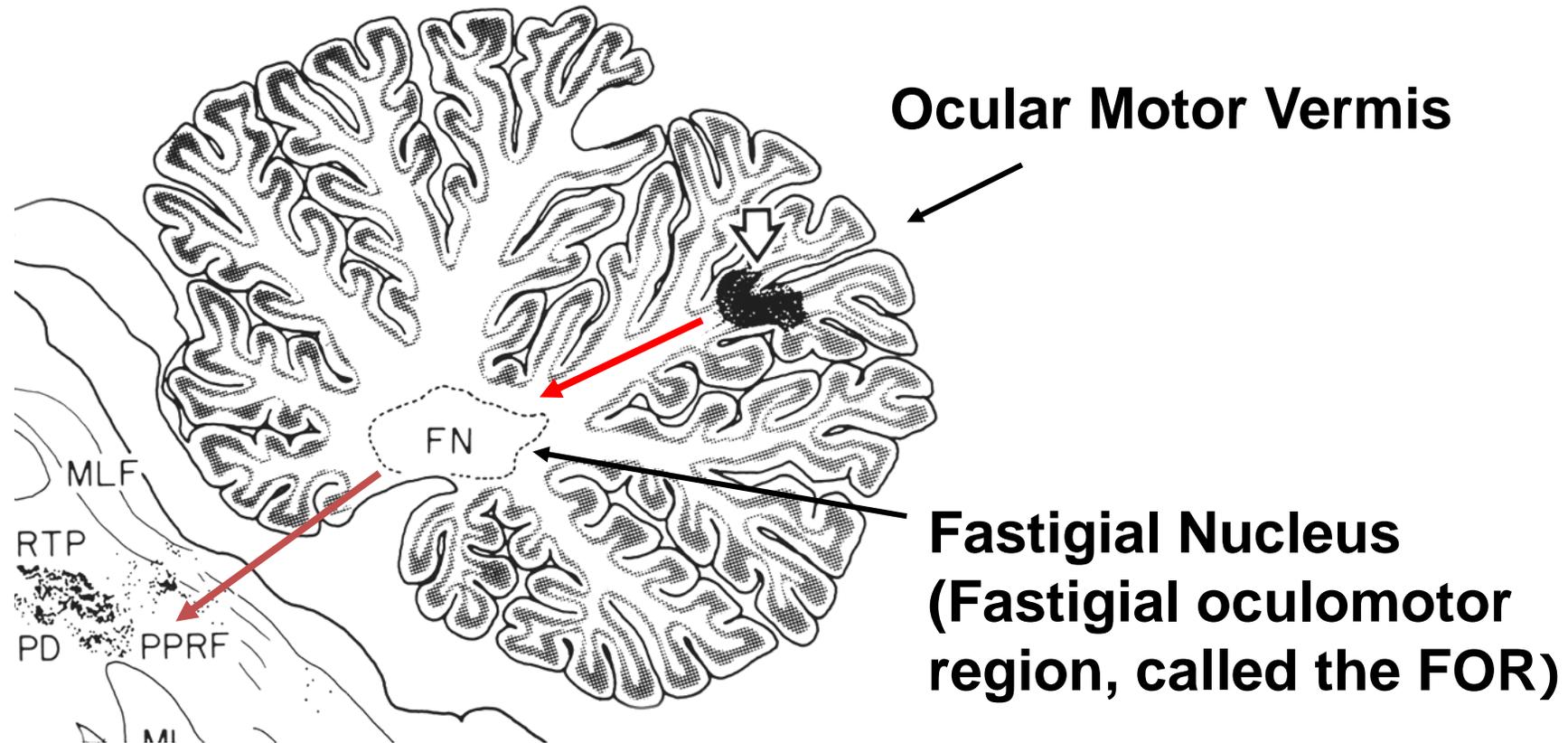
Ocular motor disorders with nodulus lesions: Periodic Alternating Nystagmus and Central Positional Nystagmus



PAN: Pathogenesis and Treatment

- In PAN, instability in velocity storage is produced by **loss of (gaba-mediated) inhibition** from the Purkinje cells of the nodulus onto the vestibular nuclei.
- Short-term adaptation (which is working normally) causes reversals of nystagmus leading to sustained oscillation.
- **Baclofen (GABA-b)*** provides the missing inhibition and stops the nystagmus.
 - Usually need only 10 mg PO TID.
 - Avoid precipitous discontinuation.
 - Does not work as well in congenital PAN.
 - **Memantine*** may be of help.

Cerebellum and saccades

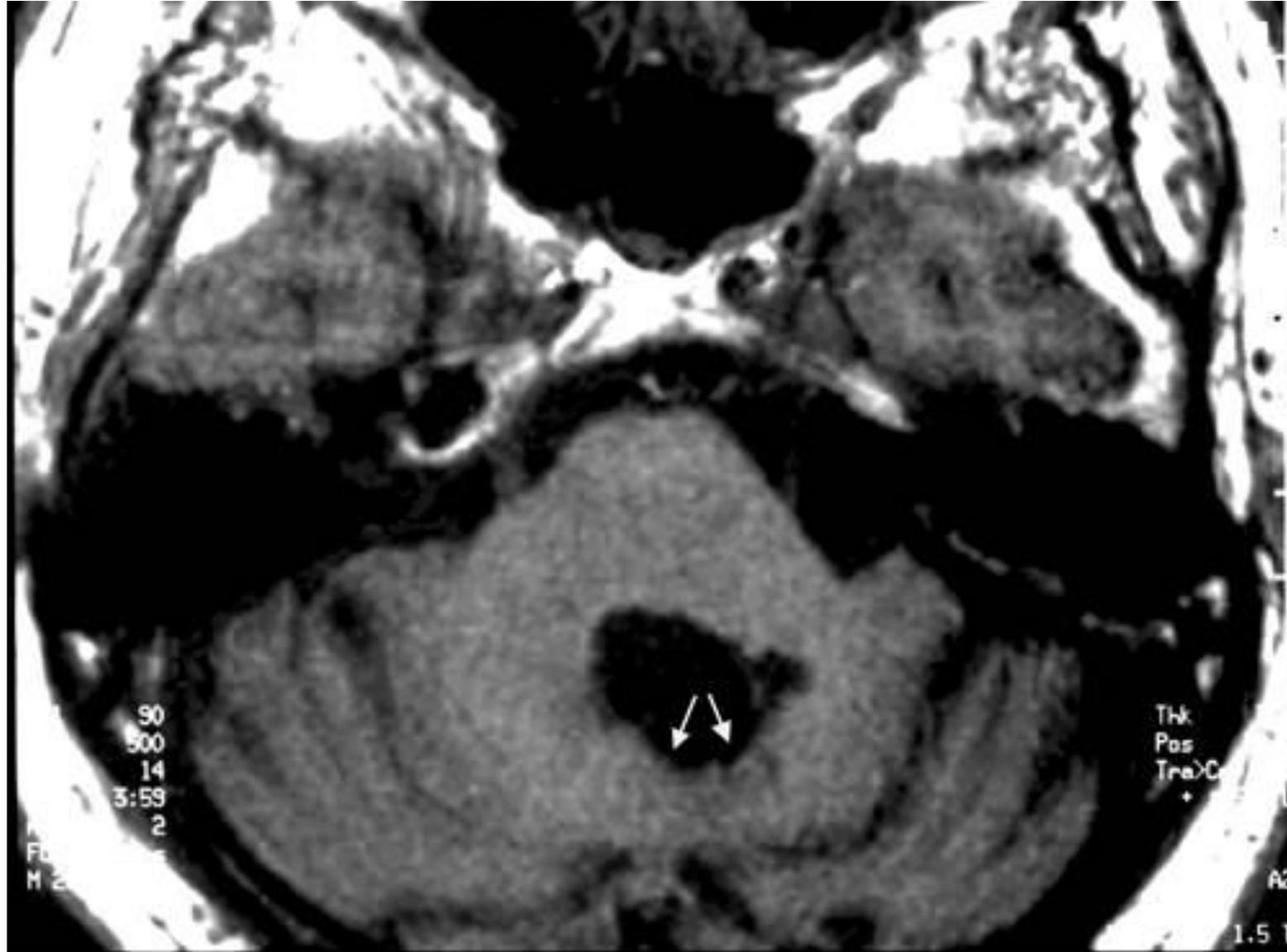


REMEMBER:

1) The vermis contains Purkinje cells and they INHIBIT their target neurons in the deep nuclei (FOR)

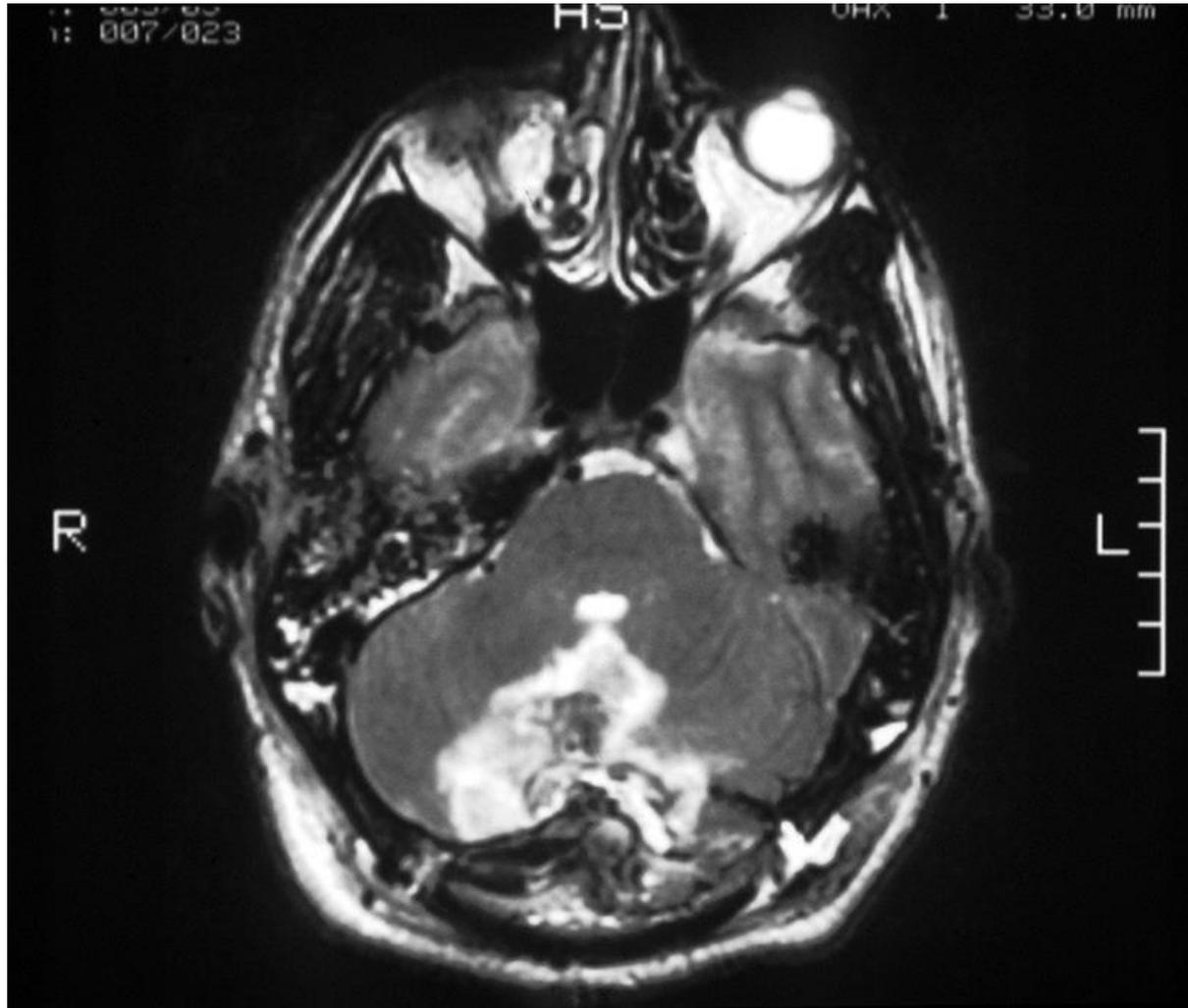
2) Each FOR normally stops ipsilateral saccades

Cerebellar fastigial nucleus lesions produce saccade *hypermetria*

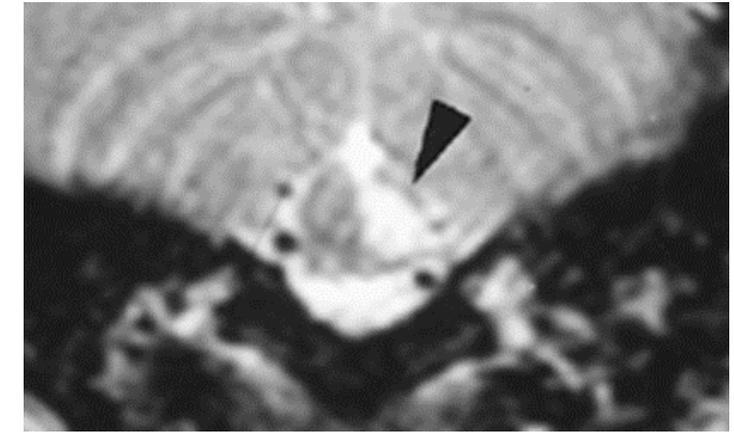


**Cerebellar dorsal vermis lesions
produce saccade *hypometria***

**Hemangiopericytoma
Involving dorsal
vermis**



Dysmetria of saccades: *Overshoot* to one side, *undershoot* toward the other, called *lateropulsion* of saccades

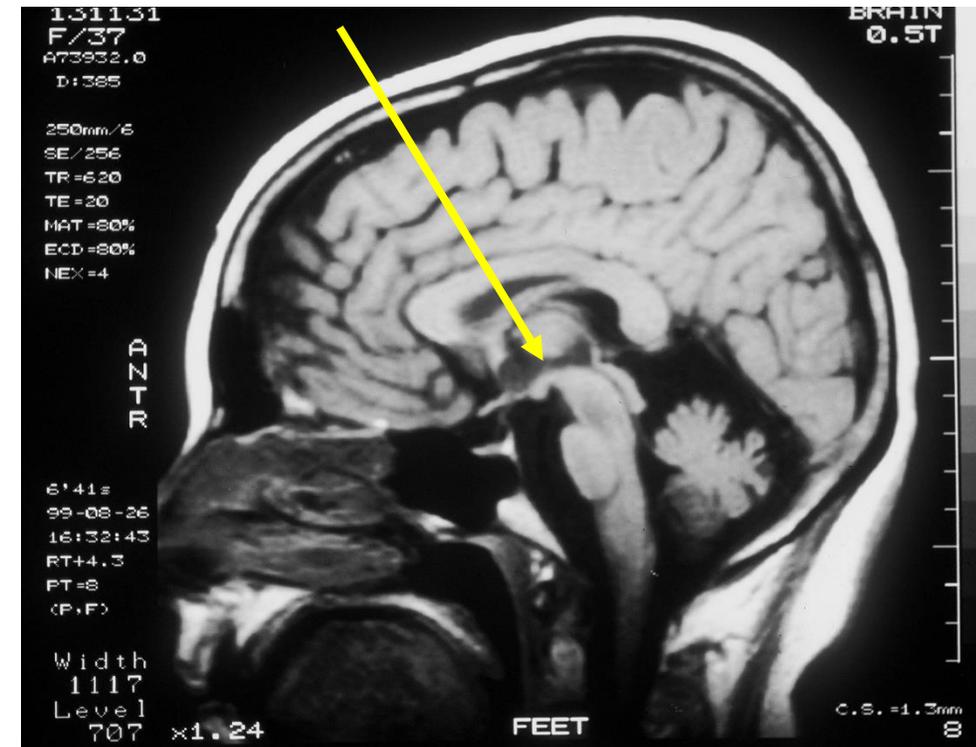


**Wallenberg's
Syndrome – Posterior
Inferior Cerebellar
Artery distribution
infarct
involving the
dorsolateral medulla**

MRI in Progressive Supranuclear Palsy (PSP) Steele-Richardson-Olszewski syndrome



PSP small midbrain, 'hummingbird' or 'emperor penguin' sign



SCA2 small pons

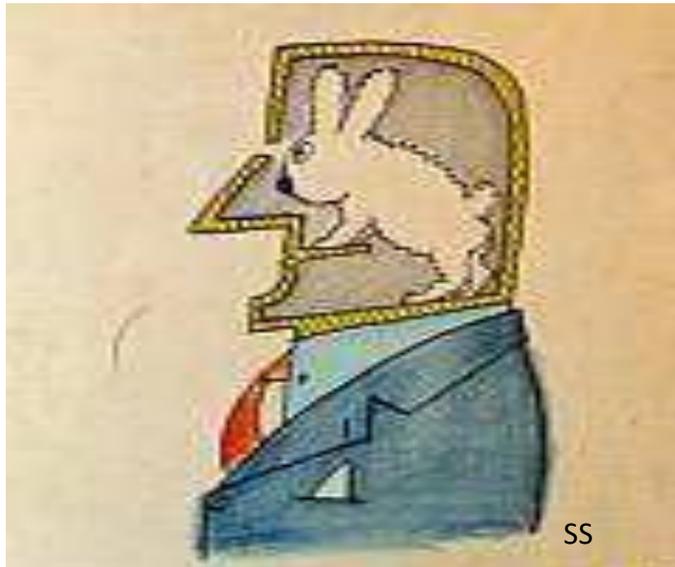
THE ALIGNMENT CHANGES IN PATIENTS WITH CEREBELLAR DISEASE



- Esodeviation (eyes turn in with distance viewing, mimics a divergence paralysis)
- 'Skew' (vertical misalignment (alternating hyperdeviation, usually abducting eye is higher))



WHY this pattern? We ALL have a lateral-eyed rabbit inside our 'human' brains.



This is reflected in the Ocular Tilt Reaction (OTR) – in which (the rabbit) emerges when there is imbalance in otolith (utricle) responses

A FEW TAKE HOME POINTS:

- LOOK FOR DOWNBEAT NYSTAGMUS: WORSE ON LATERAL GAZE AND CONVERGENCE
- LOOK FOR HORIZONTAL GAZE-EVOKED NYSTAGMUS
- IF YOU SEE PENDULAR NYSTAGMUS, LOOK AT THE PALATE
- TEST SACCADE ACCURACY (CEREBELLUM) AND SPEED (BRAINSTEM)
- CEREBELLAR SACCADE DYSMETRIA (HYPERMETRIA (DEEP NUCLEI) OR HYPOMETRIA (VERMIS))
- CEREBELLAR STRABISMUS

ESO AT DISTANCE

ALTERNATING HYPERDEVIATION ON CHANGING LATERAL GAZE DIRECTION,
(ABDUCTING EYE USUALLY HIGHER)

TREAT WITH BACLOFEN, MEMANTINE, 4 AMINO PYRIDINE