Ocular Motor Apraxia
Revisited

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Key Signs

Failure to initiate horizontal saccades.
Head thrusts trigger vestibular-induced saccades to change gaze.
Head may overshoot to use the vestibulo-ocular reflex to drag the eyes around.
Defect in generating quick phases of nystagmus
Tonic lateral deviation of the eyes present during sustained (vestibular) rotation of the body
Saccadic Subtypes

Intentional – volitional, purposeful, on command

Visually guided – saccade to fixate a target

Memory guided – saccade to a previously presented target i.e. visual memory

Anti-saccade – after instruction to look in the opposite direction of a suddenly appearing target.

Quick phases of vestibular and optokinetic nystagmus.
A Type of Congenital Ocular Motor Apraxia - Presenting Jerky Head Movements

- Jackson Memorial Lecture
A saccade is a rapid eye movement to redirect the line of sight from one target to another.
Observations

Watch the child’s random eye movements when sitting
Check the co-ordination of the head movement with movement of the eyes
Observations

Time the latency period for the initiation of a voluntary saccade after the child is shown a target to look at. A delay >200 msec is significant.

Rotate the infant at arm’s length and look if the eyes show tonic lateral deviation in the opposite direction to rotation of the body.
History

This 5 month old baby boy was born at term after an uneventful pregnancy and normal delivery.

In the first few months of life he was thought to be blind because he did not look at objects with his eyes.
Presentation

The baby fails to fix normally and may be thought to be blind

At age 4 to 6 months, characteristic horizontal head thrusts develop but if the infant has poor head control head thrusts are delayed or absent

Vertical eye movements and pursuit intact
History

The mother of this 16 month old infant boy noted from birth that he turned his head instead of his eyes and frequently tried to fix an object on either side by making a quick turn of the head past the object of regard.

Random conjugate vertical eye movements were normal.
Question

Is the delay at cortical level?

In support of a delay in maturation of the frontal eye field is the observation of impaired initiation of horizontal saccadic eye movements in patients with fronto-temporal dementia.
Saccades Initiated

By using the vestibulo-ocular reflex - the horizontal canal stimulates the vestibular nuclei and produces the VOR.

By simultaneously elicited eye and head movements which *use central head commands* while suppressing the VOR.

In normal subjects head commands usually precede the saccadic command.
Tonic lateral deviation of the eyes in the opposite direction to rotation of the body
Central Gaze –
Head Movement Commands

The analysis illustrates the hierarchical organization of both the saccadic eye movement system and the mechanisms by which changes of gaze are facilitated by head movements.
Eye Lid Blinks

OMA

Eye lid blinks are usually absent in OMA. Here, a partial blink fails to initiate a saccade to the left.

The child makes a rapid head thrust to the left to stimulate the VOR and intact slow phases of vestibular nystagmus to move the eyes to the left.
Eye Lid Blinks

Eye lid blinks turn off omnipause neurons in the pons to allow the excitatory burst neurons to fire and generate a saccadic eye movement.
Natural History

Slow motor development typically clumsy awkward and prone to stumbling
Difficulty reading
Occasional alternating esotropia
Gradual lessening of the head thrust
Pathogenesis

The pathogenesis of this congenital selective horizontal saccadic palsy remains unknown.

Cogan concluded that it may reflect a delay in the normal development of the mechanisms that control voluntary saccades.
Selective Saccadic Palsy

Congenital Ocular Motor Apraxia is a selective saccadic palsy affecting only voluntary horizontal saccades.
Voluntary Horizontal Saccade to the left

1. R Frontal Eye Field
2. R saccade center-the superior colliculus
3. L horiz. gaze premotor burst neurons in the paramedian pontine reticular formation
4. L 6\textsuperscript{th} nucleus –abduction L eye
5. R medial longitudinal fasciculus
6. R 3\textsuperscript{rd} medial rectus subnucleus-adduction R eye
Testing Voluntary Saccades

• Instruct the patient to look left, right, up and down
• Determine
  – Are saccades promptly initiated?
  – Are they of normal velocity?
  – Are they accurate?
  – If dysmetric do they undershoot -hypometric or overshoot- hypermetric on return gaze to the primary position
  – Using an OKN drum look if the eyes show normal quick phases of optokinetic nystagmus - reflexive saccades
Dysmetria

A : accurate, right on target.
B : short of the target, hypometric
C : overshot the target, hypermetric
Acknowledgments

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