# *Serious Neurologic Disease Presenting as Comitant Esotropia*

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# *Comitant and Incomitant*

- *The onset of comitant strabismus in most patients occurs during early infancy or childhood. The two major categories of early-onset comitant esotropia are congenital (infantile) and accommodative.*

- *there is general agreement that they are not causally related to any serious underlying central nervous system pathologic process.*

- *In contrast, an older patient who presents with acute esotropia and diplopia should prompt careful consideration of whether the strabismus might be a sign of central nervous system disease.*

- *First, one needs to establish whether the deviation is comitant or incomitant.*

- *There is no doubt that the vast majority of neuropathic and/or myopathic cases of strabismus present as an incomitant deviation.*

# *Comitant and Incomitant*

- *Until recently, most authorities would probably have agreed that* ***Acute-onset esotropia that is comitant without divergence insufficiency*** *is benign in nature.*

- *it is now apparent that this simple algorithmic way of viewing acute-onset esotropia, although valid in the vast majority of cases, is not inevitably appropriate, because many exceptions exist.*

- *Comitance in acute onset esotropia does not rule out the possibility of a serious underlying neurologic condition.*

## *Types of Acute Comitant Esotropia that lack of any serious underlying neurologic disease.*

- *Common to all three groups are an acute onset, comitance, a relatively large angle of deviation, good binocular potential, and the lack of any serious underlying neurologic disease.*

- ***Type 1: Acute-Onset Comitant Strabismus After Occlusion (Swan Type) :***

  *This form of strabismus may occur after therapeutic patching or as the result of monocular or asymmetric visual loss. The resulting strabismus in children and young adults is usually an esotropia, whereas exotropia predominates in adults.*

# *Types of Acute Comitant Esotropia that lack of any serious underlying neurologic disease.*

- ***Type 2: Comitant Convergence Strabismus (Franceschetti Type) :***

  *These patients develop an acute-onset esotropia that at first may be intermittent but quickly becomes constant. The refractive error is usually minimally hyperopic, with a normal AC/A ratio.*

  *There are reports that this type of esotropia may occur in multiple siblings as well as in monozygotic twins.*

- ***Type 3 : Comitant Convergent Strabismus Associated with Myopia (Bielschowsky Type) :***

  *The consistent features of this group were myopia of 5 D or less, esotropia at distance but maintained fusion at near, and no evidence of lateral rectus paralysis.*

  *However, these patients often present with a reasonably small angle of esotropia (10 PD or less) but gradually develop an increasingly large angle that may preclude the use of prisms.*

  *direct damage to the lateral rectus muscle may be the primary pathologic problem.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Hydrocephalus*

- *It is useful to divide patients with comitant esotropia and hydrocephalus into two distinct categories :*

- ***First**, a significant number of patients with hydrocephalus are seen with an early-onset large-angle comitant esotropia, resembling infantile esotropia.*

  *They present in early childhood with a large-angle esotropia, usually an A pattern, and obvious neurologic problems associated with hydrocephalus or other congenital central nervous system anomalies.*

  *The A pattern often seen in these patients clearly speaks against the notion that sixth nerve paresis is the underlying pathologic process responsible for the esodeviation, because a small V pattern is expected in sixth nerve palsies.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Hydrocephalus*

- *In contrast, a **second** and smaller group of patients with hydrocephalus may present with the acute onset of comitant esotropia as a sign of dramatic elevation in intracranial pressure and/or ventricular shunt failure.*

- *The comitant esotropia seen in these patients is usually a large angle one, An A pattern is distinctly unusual in this group.*

- *Most of these patients will experience realignment of the ocular axes after restoration of normal intracranial pressure.*

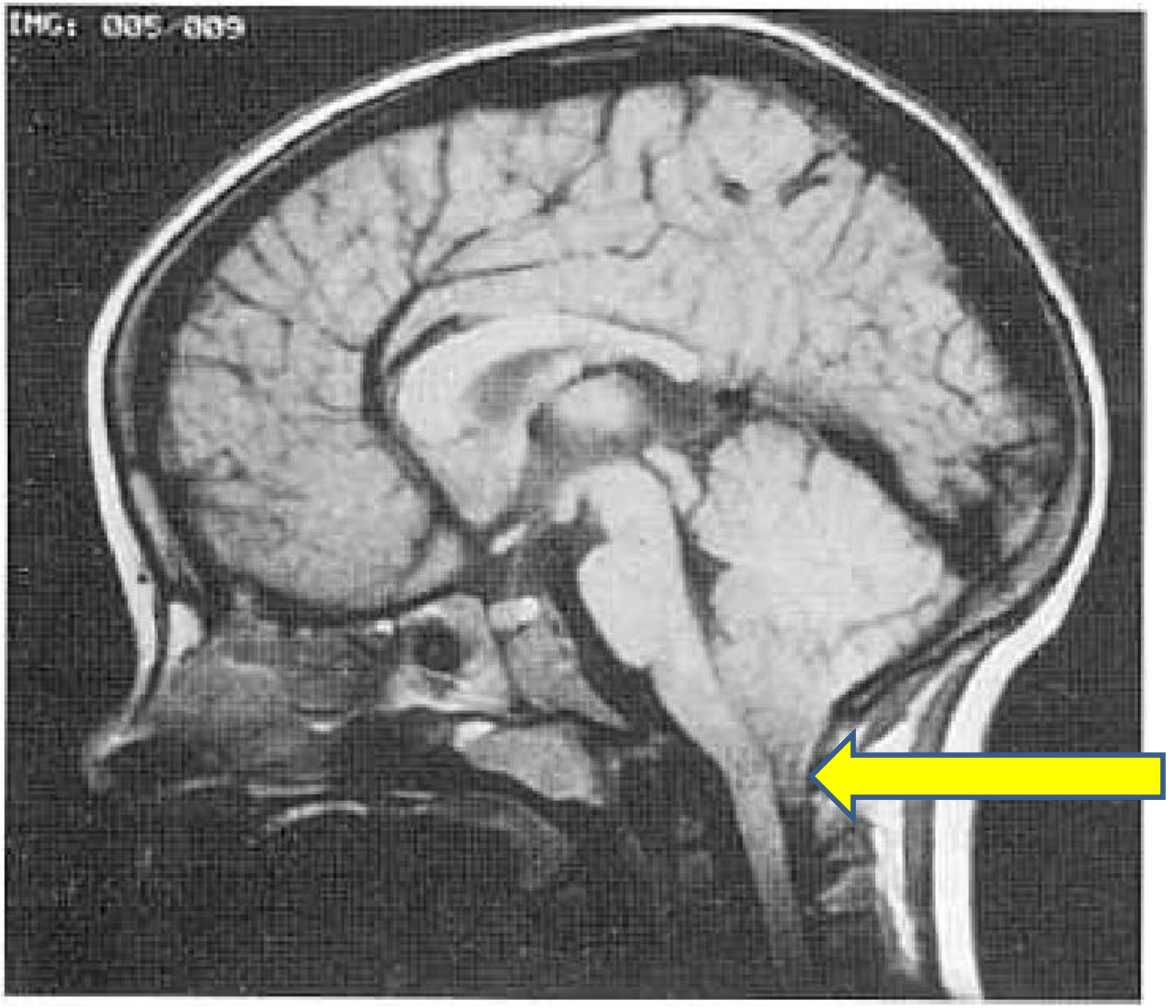
- *Indeed, the acute onset of comitant esotropia with diplopia is now well recognized by neurologists and neurosurgeons as a sign of shunt failure.*

- *However, our experience even in these cases suggests that esotropia is rarely the presenting sign in an otherwise healthy child. In the vast majority of patients, hydrocephalus has been previously diagnosed, usually in infancy, and the esotropia is a sign of shunt failure.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Chiari Type 1 Malformation*

- *The Chiari type I malformation involves displacement of the cerebellar tonsils into the upper cervical canal.*
- *Unlike many congenital malformations of the central nervous system, patients with this condition usually remain asymptomatic until late childhood or the early adult years. The most frequent symptoms are neck pain and headach.*
- *Ophthalmic abnormalities may be the sole manifestation of Chiari type I malformation. Symptoms may include diplopia and oscillopsia secondary to various forms of nystagmus.*
- *as in long-standing comitant esotropia associated with hydrocephalus, an A pattern is frequent.*
- *Comitant esotropia may be the presenting sign of a Chiari type I malformation, even in the absence of nystagmus or other obvious brain stem signs.*




***Chiari type 1***
***displacement of the cerebellar tonsils into the upper cervical canal***

# *Comitant Esotropia Associated with Neurologic Disorders*

## ***Brain Tumors***

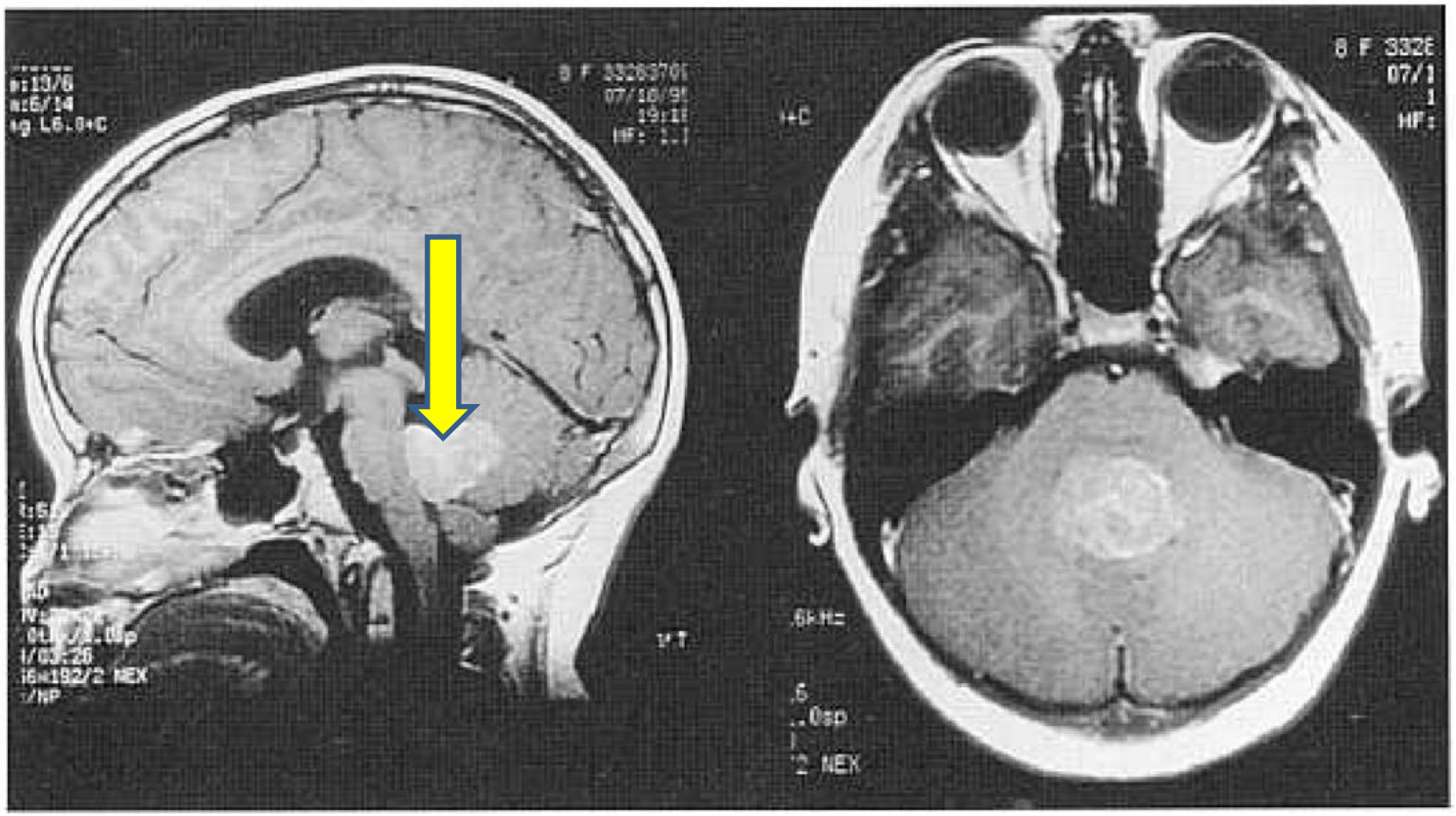
- *acute-onset comitant esotropia may be associated with a brain tumor .*
- *No single type or site of brain tumor can account for all cases described thus far.*
- *It is essential to note that an A pattern is distinctly unusual in this group of patients. This is in sharp contrast to patients having long-standing hydrocephalus and a Chiari type I malformation.*
- *A small V pattern is common in patients having comitant esotropia and brain tumor and should suggest the possibility that the primary problem is subclinical paresis of the abducens nerve.*
- *It seems clear that paresis of the sixth nerve cannot explain all cases of comitant esotropia associated with brain tumors and more central part of the* ***ocular motor control system*** *is involved in development of the strabismus.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Brain Tumors*

- *Most unnerving about these patients is that the comitant esotropia is often the initial and only sign of intracranial pathology .*
- *A significant proportion of patients with brain tumors and comitant esotropia will exhibit nystagmus, particularly nystagmus in abduction .*
- *Tumors of the cerebellum, brain stem, sellar region , and corpus callosum all have been associated with acute-onset comitant esotropia. However, the cerebellum is by far the most common tumor site .*

# *Brain Tumors*



***A 7-year-old girl with a medulloblastoma and no hydrocephalus presented with an acute onset comitant esotropia of 25 PD.***

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Thalamic Disease*

- *An acute acquired comitant esotropia may be a prominent sign of pathology involving the thalamus.*
- *This is particularly true for adults with hypertensive bleeding within the thalamus, it is a direct result of interference with vergence neurons in the midbrain.*
- *The comitant esotropia may be associated with downward deviation of the eyes and miosis.*
- *Both computed tomographic analysis and autopsy cases suggest that only patients whose thalamic hemorrhage extends into the dorsal midbrain are likely to develop these signs.*
- *These adult patients rarely present a diagnostic dilemma, because associated neurologic signs invariably point to this life-threatening condition.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Myasthenia Gravis*

- *Whereas most patients with myasthenia gravis and diplopia will have evidence of incomitant strabismus with restricted ocular movements, occasionally in children this is not necessarily the case.*

- *Comitant esotropia and exotropia have been described as the initial presenting sign of infantile myasthenia gravis.*

- *These comitant deviations may continue to be the only sign of myasthenia gravis for several months before the variable nature of the disorder and associated ptosis become apparent.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Seizures*

- *In some case reports a constant comitant esotropia has been associated with epileptic activity.*
- *Of interest is the fact that, with medical control of the seizure disorder, esotropia resolved in these cases.*
- *The mechanism whereby esotropia is produced by cortical dysfunction in epilepsy is not known. It is a rare occurrence in a common disorder and may represent nothing more than decompensation of a previously unrecognized esophoria.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Diagnosis*

- *The vast majority of cases of acquired comitant esotropia will not be associated with serious underlying neurologic disease. When there is a history of previous strabismus, occlusion therapy, monocular visual loss, or myopia, acquired comitant esotropia need cause little worry.*

- *However, if a patient has no apparent cause for acute-onset comitant esotropia, the possibility of an underlying neurologic disorder should at least be considered.*

- *An A pattern in an acquired comitant esotropia almost invariably indicates that the patient has an underlying neurologic problem, either hydrocephalus or the Chiari type I malformation (or both).*

- *V pattern may be present in patients with or without neurologic disease, and careful reassessment is essential to be certain that it is not an early sign of sixth nerve palsy.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Diagnosis*

- *Forms of manifest and latent nystagmus are frequently seen in patients with infantile esotropia.*
- *Nystagmus is absent in all benign forms of acquired comitant esotropia.*
- *Nystagmus therefore is an important sign suggesting underlying neurologic disease.*
- *Downbeating nystagmus localizes the pathologic process to the area of the craniocervical junction, but any form of nystagmus seen in a patient with acquired comitant esotropia should suggest an underlying neurologic process.*
- *Nystagmus may be seen in patients with hydrocephalus, the Chiari type I malformation, or a brain tumor.*

# *Comitant Esotropia Associated with Neurologic Disorders*

## *Diagnosis*

- *Although further studies are needed to evaluate fusion potential in patients with comitant esotropia and a brain tumor, it appears at present that a lack of significant motor and sensory fusion, even after appropriate optical and/or surgical treatment, is found in a considerable number of patients.*

- *We believe that a patient who presents with an acquired comitant esotropia and whose eyes cannot be made to fuse with hand-held prisms or on synoptophore examination is very likely to have an underlying neurologic problem, especially a brain tumor.*

# *Comitant Esotropia Associated with Neurologic Disorders*

- *The patient with acquired comitant esotropia needs to be carefully examined to detect other neuro-ophthalmic signs of neurologic disease*

- *It should be emphasized once again, however, that a number of patients with posterior fossa tumor have presented with comitant esotropia and no other signs of cerebellar dysfunction, even when examined by a competent neurologist*