

## Management of sixth nerve palsy – different approaches

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### Abstract

Sixth nerve palsy is one of the most common oculomotor nerve palsy, manifested by abolished or limited abduction of the affected eye and diplopia. This condition significantly alters patient's quality of life. The authors wish to emphasize forensic implications of this neuro-muscular dysfunction throughout its management steps, giving some examples of cases, with different etiology and treatment. It's important to highlight the role of a careful imaging and a good interdisciplinary collaboration in management of this paralysis.

**Key words:** sixth nerve, palsy, paralysis, paresis, abduction, diplopia, skull base trauma.

### General features

Sixth nerve palsy / paresis is defined as the abolition or reduction, unilateral or bilateral, of oculomotor sixth nerve function, which innervates lateral rectus muscle, resulting in neuro - muscular

dysfunction, translated by impossible or very limited abduction of the interested eye, manifested by diplopia (1), which significantly reduces patient's quality of life. Sometimes binocular vision is possible in a compensatory position of the head, with face turned toward the affected eye.

Due to its long intracranial course (2), with angulation over the petrous apex of the temporal bone, the sixth nerve is vulnerable to increased intracranial pressure, skull base trauma, meningeal inflammation, and a displacement of the brain stem. Therefore, sixth nerve palsy is most common oculomotor nerve palsies, representing up to 40% of these. (3) It may be also affected, like the other cranial nerves, by toxic substances, acute demyelinating diseases, and some viral illness, even post vaccinal. Expansive intracranial processes and aneurysms are frequent causes of the abducens nerve palsy.

Sixth nerve palsy can occur in many contexts:

- Complications of vasculopathic diseases such as diabetes, hypertension, hyperlipidemia, arthritis that produce dysfunction by affecting vasa nervorum. (4,5)

- Trauma, when the VI-th nerve damage occurs either directly (eg through a skull base fracture) or indirectly by increased intracranial pressure. (2, 4)

- Signs of false localization in cases of lesions leading to increase of intracranial pressure (idiopathic intracranial hypertension, expansive processes etc.) by nerve compression at the petrosus apex, and not necessarily pathological process to be in immediate contact with the nerve. (6)

- Locating signs in various intracranial disorders that may involve sixth nerve from the nucleus to the orbit, and in such cases associated manifestations frequently occur, often falling into syndromes, such as Foville syndrome, Millard-Gubler syndrome, syndrome Gradenigo, acoustic neurinoma, etc. (7)

Therapeutic options are varied and depend on the etiology and degree of neuromuscular dysfunction. Treatment may be conservative (by unilateral occlusion or prescribing prism lenses to correct diplopia) or surgical. It is generally considered a waiting period of at least 6 months after the onset of paralysis, to track the event of spontaneous recovery; if this is not achieved, surgery is usually indicated, depending on the residual function of the neuro-muscular complex. (8, 9)

Generally, if paresis is diagnosed, a

recession / resection technique is the procedure of choice (ipsilateral medial rectus recession, associated with lateral rectus resection of paretic eye). If sixth nerve paralysis is found, transposition techniques is recommended (Hummelsheim, Jensen, etc.), associated or not with medial rectus recession (uni- or bilateral). (8) Assuming that there are chances of spontaneous recovery of deficit and the limits of prismatic correction are exceeded, chemodenervation with botulinum toxin (BTX-A) of the ipsilateral medial rectus can be made, leading to temporary paralysis of adduction of the involved eye, which eliminate diplopia in primary position and prevents ipsilateral medial rectus contracture. (10)

Detailed below are some examples of VI nerve palsy with different etiology and different therapeutic approaches.

## Cases presentation

### Case 1

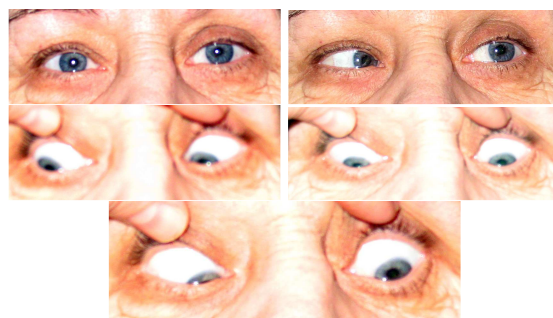
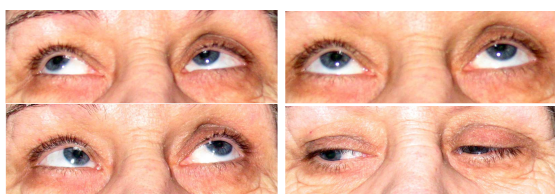
52 year old woman, an economist, is presented with VI-th nerve palsy, gained one year ago from a traffic accident, resulting in multiple trauma and skull base fracture; she sought damages in court, without receiving prevail. The patient was no more able to drive the car and, to avoid marked diplopia, which created confusion and vertigo, she had to cover one eye permanently. Ophthalmologic examination revealed a VI-th nerve palsy in the left eye, with completely abolished abduction, without saccadic velocity, the eye being immobilized in extreme adduction and

ipsilateral medial rectus hyperaction, without contralateral lateral rectus inhibitory paresis. (Figure 1) Surgery was decided, practicing Hummelsheim technique, combined with ipsilateral medial rectus recession. Orthoptic alignment in primary position was achieved postoperative, with minimal esotropia at near (+ 4 prism diopters). (Figure 2) Patient manifested no diplopia in primary position, both at distance and near and recovered useful binocular vision. Left eye abduction was partially recovered (- 3).



**Figure 1**

Preoperative pictures: large esotropia in primary position; abolished abduction on the left eye



**Figure 2**

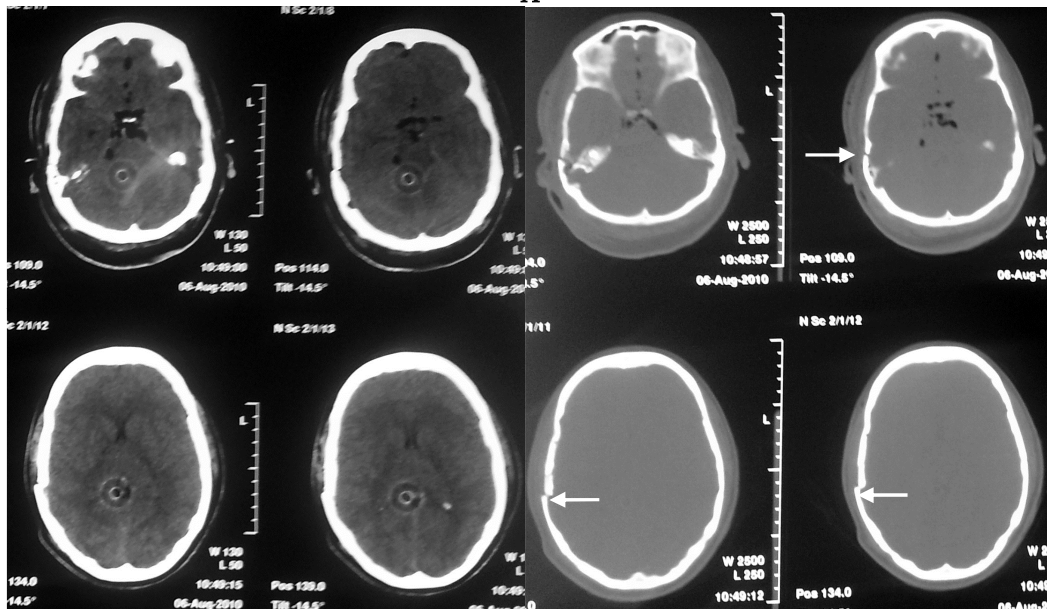
Postoperative pictures: orthoptic alignment in primary position; abduction improved (- 3)

### **Case 2**

36 years old woman, civil servant, was hospitalized in serious condition with multiple trauma and TCC produced in a car crash. Skull radiography and brain CT scan were performed at admission in the hospital. (Figures 3A, 3B) She was evaluated ophthalmologic during hospitalization, and bilateral VI-th nerve palsy was found (Figure 4), considered secondary to increased intracranial pressure. Two weeks after discharge, the patient asks the ophthalmologist to surgically correct her paralytic strabismus. After the examination, the therapeutic options are discussed with the patient, persuading her to be "patient" for few months, to observe if there is a chance for spontaneous improvement. And improvement, indeed, happened. Periodic evaluation at 3 months reveals a significant recovery of both sixth nerves function, and in approximately 5 months, total recovery appears, with useful binocular vision in all gaze positions and no diplopia.



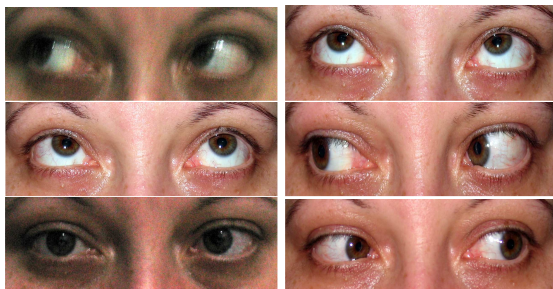
A



B

**Figure 3**

A – Lateral skull radiography shows right temporo-parietal fracture; B – Brain CT scan (native and bone windows) reveals right temporo-parietal fracture and pneumoencephaly

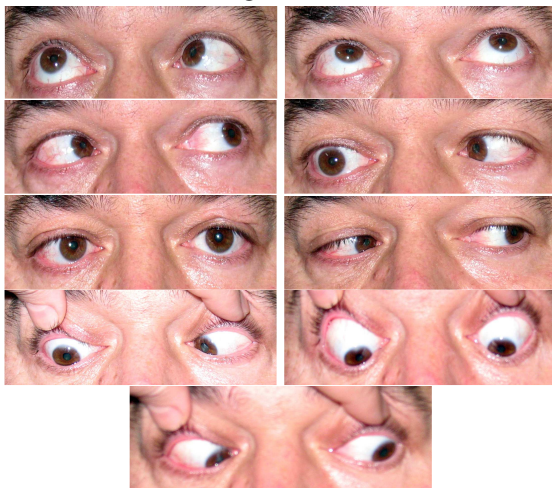


**Figure 4**

Pictures at 3 months after trauma: mild left esotropia, with good enough abduction

**Case 3**

47 years old man, laborer, is presented with horizontal homonymous diplopia, started 18 months ago, with mild worsening of onset within 8 months and relatively steady since then. No etiology was found. Ophthalmological examination reveals isolated right VI-th nerve paresis, with limited abduction (- 3) and esotropia (Figure 5); no history of vascular diseases or diabetes. Cerebral MRI with contrast was again recommended, which revealed a small tumor (probably meningioma) located in the Dorello channel. Prismatic correction possible on eyeglasses did not eliminate diplopia, then surgery was decided; medial rectus recession combined with lateral rectus resection on the right eye was performed. Postoperative the patient shows residual esotropia (10 PD), with improvement of abduction (- 1). At present, the patient has got a small prismatic correction for distance, and no prisms for near (Figure 6).

**Figure 5**

Preoperative pictures: right esotropia: limited abduction on the right eye (-3)

**Figure 6**

Postoperative pictures: mild esotropia in primary position; abduction improved (- 1)

**Case 4**

5 years old boy, released a day before from hospital, where he was admitted in gastro-enterology service for nausea, headache and vomiting, is presenting for ophthalmological exam, accompanied by his mother, who affirms that the boy has double vision and crossed eyes since 3 days. In the examination room the child was turned away, drowsy, with psychomotor retardation. Diplopia was confirmed - horizontal homonymous, given by bilateral sixth nerve paresis. Fundus examination with cycloplegia revealed bilateral papilledema. The patient was sent urgent for brain MRI, which showed a posterior fossa tumor, with hydrocephalus. The child was admitted to the neurosurgery service and underwent external ventricular drainage. Tumor ablation was done few days after.

## Discutions

The nuclei of sixth nerve consist of a paired group of neurons located beneath the floor of the fourth ventricle, near the midline, at the pontomedullary junction. The intrapontine portion of the facial nerve is in close proximity to these neurons. The medial longitudinal fasciculus is located just medial, and the vestibular nuclei are located lateral to the abducens nuclei. Following emerging from the brainstem, the abducens nerve continues through the inferior venous compartment of the petroclival venous confluence in Dorello's canal. It then bends sharply across the apex of petrous portion of the temporal bone, then runs between the petrosphenoidal ligament of Gruber and the dorsum sellae. The abducens nerve then arrive in the cavernous sinus, where walk lateral to the internal carotid artery. The long course of the sixth nerve from the brainstem to the eye make it vulnerable to lesions at different sites, like skull base fractures, especially of the petrous portion of the temporal bone, or aneurysms of the carotid artery in the cavernous sinus. Also, expansive processes that dislocate the brainstem inferiorly can damage the sixth nerve by compressing it between the pons and the tip of the petrosom. The abducens nerve enters the orbit through the superior orbital fissure and goes forward into the the lateral rectus muscle. (11)

In a population-based study, Patel found the incidence of sixth nerve palsy as 11.3 / 10000, with a peak in the seventh

decade of life. In this study, "the most frequent causes was hypertension alone, coexistent hypertension and diabetes, trauma, multiple sclerosis, neoplasms, diabetes alone, stroke, post-neurosurgery, aneurysms, and 34% of cases remain unsolved". (2) Based on these data, the authors suggest that patients with nontraumatic isolated sixth nerve palsy do not need extensive investigations, and may be just periodical followed, while isolated cases which are not neurologically related should be completely evaluated with neuroimaging. On the other hand, in other studies (12) a much higher incidence of more serious pathology was found. These studies have shown that according to causes, patients can be divided into three age groups: children (predominant etiology is either postviral or tumor and trauma), young adults, where the etiology is most difficult to assess, and elderly patients, where microvascular diseases are frequent. In a prospective study that evaluated the role of MRI in 43 cases of isolated sixth nerve palsy, the authors found serious pathology in 63% of cases. (13) Therefore, a cerebral CT and / or MRI scan should be performed as a routine for any sixth nerve palsy with unknown etiology.

Medico legal considerations for VI-th nerve palsy involve several issues that arise at all stages of assessment and treatment of this condition.

1 - The traumatic etiology of VI-th nerve palsy rise the question of forensic issues related to trauma itself: mode of production (traffic accident, accident at

work or home, aggression), many organs and / or systems damaged (polytrauma) and number of days of hospitalization and temporary or permanent work disability.

2 - Although the diagnosis of paralysis / paresis of sixth nerve is not difficult to establish, finding the etiology may encounter, in many cases, difficulties. If paralysis is associated with signs of dysfunction of other cranial nerves, they can guide the doctor to a correct etiologic diagnosis. In cases of isolated VI-th nerve palsy, although it is still considered to be mostly of vascular etiology and does not require costly investigations (economically unprofitable), some studies show that in over 10% of these cases, imaging reveals a significant disease directly related to the pathogenesis of paralysis (tumors, aneurysms, multiple sclerosis). (14) For these reasons, especially since the quality of imaging methods has considerably improved in recent years, is recommended careful investigation of VI-th nerve palsy by means of cerebral imaging, at least by the fact that undetected or late etiologic diagnosis of severe injuries can have medico legal consequences. Vascular etiology should remain a diagnosis of exclusion after other causes have been eliminated and periodic revaluations do not show any new elements; furthermore, spontaneously gradually recovery of the deficit is often seen. (6)

3 - VI-th nerve palsy itself is a condition that can lead to temporary disability, especially for persons working at height, driving vehicles and generally workers in

jobs that require good spatial perception and coordination between visual and musculoskeletal system; the disability occurs through the mechanism of esotropia and diplopia, which creates confusion, reduce field of binocular single vision and lose the stereoscopic vision.

4 - Treatment options should be discussed thoroughly with the patient, so he or she to understand that treating the cause that led to neuro-muscular dysfunction is crucial and conservative or surgical treatment of paralysis itself is not designed to recover lost function (although in some procedures can improve abduction), but to improving symptoms (eliminating diplopia in primary position and recovery of binocular visual field and a useful binocular vision). (8) In order to avoid creating conflict situations, when chemodenervation with BTX-A is recommended, the patient should understand very well that this process will lead to a medial rectus muscle paralysis in the affected eye, but that paralysis is induced deliberately by physician, in order to eliminate diplopia and has a temporary effect, lasting 6-8 months, then gradually returns to normal function. When conservative treatment is exceeded, eye surgery is indicate, to achieve alignment in primary position. Most patients look forward to the time of surgery, in a natural desire to improve their quality of life. A small percentage of patients refusing surgery for two reasons: one is the fear of any invasive medical or surgical maneuver, and the other occurs in patients awaiting a

solution in court related to the circumstances of the event which led to paralysis or seeking employment in a category of disability. In the latter situation, the role of the physician is to state clearly the treatment options for each case.

In a postmortem study, Bulent Sam & col investigated the mechanism of injury of the abducens nerve in severe head trauma, and they found “in microscopic evaluation, remarkable cellular edema and perineural hemorrhage in the all abducens nerve along its course at the petroclival region. It was also noted that, the amount of perineural hemorrhage, particularly at the sites of dural entry zone and petrous apex were in accordance with findings in macroscopic sections” (15). They concluded that “in severe head trauma, the abducens nerve can be injured at the sites of dural entry point, petrous apex and lateral wall of the internal carotid artery, directly proportional with the amount of the trauma. (15) Abducens nerve paralysis is significant as an indicator of the severity and the direction of the trauma applied to the head or neck. Particularly, children exposed to trauma might have sixth nerve paralysis according to the relevant literature. In authors opinion, in autopsy cases suspected of shaken baby syndrome, searching for the hemorrhage and contusions at the abducens nerve entrapping points, together with brain and retina damages will support the diagnose”. (15)

Attention should be paid to children with sudden uni- or bilateral sixth nerve palsy or paresis, especially for those who presents with so called gastro-intestinal symptoms, because the nausea, headache and vomiting may be due to the raised intracranial pressure and the medico-legal implications may be dramatic, since some authors published an article about “Appendectomy – first choice surgery in cerebral tumors at children” mainly to emphasize the tremendous importance of careful evaluation and investigation of brain related signs and symptoms in young patients. (16)

### Conclusion

In conclusion, the sixth nerve palsy should be carefully evaluated clinically and imaging, requiring a good interdisciplinary collaboration and therapeutic options should be weighed according to each case. The forensic implications should be kept in mind throughout the management of this condition.

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