

# Monocular Elevation Deficiency/Double Elevator Palsy

## WHAT IS MONOCULAR ELEVATION DEFICIENCY (DOUBLE ELEVATOR PALSY)?

Monocular Elevation Deficiency, also known as Double Elevator Palsy, is an inability to elevate one eye above the horizontal plane [See figure 1]. There are varying degrees of severity from misalignment only in upgaze to the deficit effecting straight ahead gaze, resulting in a hypotropia, one eye that is pointed downward relative to the other eye.



**Fig. 1:** PPatient with left-sided monocular elevation deficiency/double elevator palsy attempting to look upward.

## WHAT IS THE CAUSE OF MONOCULAR ELEVATION DEFICIENCY (DOUBLE ELEVATOR PALSY)?

The lack of upward movement of the eye can be caused by paralysis/weakness of one or both of the elevator muscles of an eye (superior rectus or inferior oblique), or constraint from the downward pulling muscle (inferior rectus). In some patients, there is a combination of both paralysis and restriction. Magnetic resonance imaging usually reveals that the muscles themselves are normal and the cause is usually from the control centers in the brain (supranuclear).



Monocular elevation deficiency is considered part of a group of conditions caused by loss of innervation to an eye muscle called congenital innervation dysgenesis syndrome (CID).

### **IS MONOCULAR ELEVATION DEFICIENCY HEREDITARY?**

This entity is not usually hereditary (inherited). It is a rare condition, and can be congenital (present at birth) or acquired after birth. There have been some reports of families with monocular elevation deficiency, but no single gene has been identified as causative.

### **IS MONOCULAR ELEVATION DEFICIENCY ASSOCIATED WITH PTOSIS (DROOPY EYELID)?**

Yes. The eyelid on the same side that cannot elevate is droopy ([ptosis](#)) 25-75% of the time, and most of the remaining cases have pseudoptosis. Pseudoptosis is the appearance of droopy eyelid caused by the eyeball being hypotropic (downward displaced). This is because when the eye looks downward, the eyelid also is lower naturally.

### **IS MONOCULAR ELEVATION DEFICIENCY ASSOCIATED WITH JAW WINKING?**

Up to 50% of patients with monocular elevation deficiency and congenital ptosis have a phenomenon called Marcus Gunn jaw-winking. This is a condition in which the cranial nerve that controls eyelid movement is mis-wired with the cranial nerve that controls chewing or sucking thus creating a "wink" when chewing or sucking. Marcus Gunn jaw-winking is also considered a CID.

### **IS MONOCULAR ELEVATION DEFICIENCY ASSOCIATED WITH OTHER DISEASES OR DEVELOPMENTAL PROBLEMS?**

There is no known association between Monocular Elevation Deficiency and systemic or neurological diseases. Other disorders can occur with Monocular Elevation Deficiency, but they are not directly related. Amblyopia, a lack of visual development of one eye, may result if a child is unable to use both eyes simultaneously due to the elevation deficit.

### **WHAT ARE THE TREATMENT OPTIONS FOR MONOCULAR ELEVATION DEFICIENCY?**



The treatment of Monocular Elevation Deficiency is surgery or prism in glasses, which are indicated if the eyes can only align with an abnormal head posture, such as large chin up posture due to double vision in straight ahead gaze. The type of surgery depends on the cause of the elevation deficit. Surgery is usually performed on the eye that cannot look up but may sometimes be necessary in the opposite eye to improve the deficit.

### **DO EXERCISES OR VISION THERAPY HELP TO FIX MONOCULAR ELEVATION DEFICIENCY?**

No.

### **SHOULD EYE PATCHING BE PERFORMED FOR MONOCULAR ELEVATION DEFICIENCY?**

Patching is sometimes necessary to treat [amblyopia](#) (weak vision) that can result from misalignment of the eyes. Patching will not realign the eyes; it is done to help strengthen the vision in the eye that has amblyopia.

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