

Congenital Mirror Movements in a Child with Chiari Type 1 Malformation

Mirror movements refer to involuntary movements, which occur in a muscle group or limb on one side of the body in response to an intentionally performed movement in the corresponding contralateral muscle group or limb(1). Mirror movements may be congenital or acquired. We report a case of congenital mirror movements.

A 9-year-old boy presented to the Child Guidance Clinic at the Department of Pediatrics, Medical College, Calicut with abnormal hand movements. He gave history of simultaneous, involuntary movements of the hand associated with voluntary movements of the opposite hand. The involuntary movements were noticed from the first year onwards. There was history of dropping objects in one hand whenever the other hand was moved. He was made fun of by his classmates because writing with the right hand caused similar movements of the left fingers. There was no history of any other neurological symptoms.

The child was born of non-consanguineous parents. There was no family history of similar illness, epilepsy or mental illness. His developmental milestones were normal. He was studying in the fourth standard and had average academic performance. Examination showed involuntary, simultaneous movements of the left hand whenever the right hand was moved. The movements were confined to the joints of the fingers and shoulder, elbow and wrist joints were not involved. The mirror movements were not present in the legs. He was a right-handed individual with no neurological deficits. Deep tendon reflexes were normal and plantar bilaterally flexor. Psychological evaluation showed a normally intelligent child with no psychotic or depressive features.

MRI scan showed inferior migration of the cerebellar tonsils below the McRae's line into the cervical posterior subarachnoid space at C1 level suggestive of tonsillar herniation and Chiari type 1 malformation. Vertebral bodies and appendages

were normal and there was no syringohydromyelia or hydrocephalus.

Three categories of congenital mirror movements are described. A physiologic form in which the movements occur during early infancy and disappear by the age of 8-10 years along with maturation of the nervous system, a hereditary form with predominantly autosomal dominant mode of inheritance and a pathologic form associated with nervous system disorders(2). Klippel-Feil Syndrome is the commonest disease associated with mirror movements. Other causes include Kallmann's syndrome, agenesis of the corpus callosum, basilar invagination of the skull, spina bifida occulta, Friedrich's ataxia, Usher's syndrome and hemiplegic cerebral palsy(2,3). Causes of acquired mirror movements include cerebrovascular accidents, subarachnoid hemorrhage, trauma and tumors(2). Mirror movements, following surgery for intractable epilepsy is reported(4).

The exact pathophysiology of mirror movements is not known. The causes postulated include abnormal development of pyramidal decussation and transcallosal inhibitory pathways(2). In the present case the child had herniation of the cerebellar tonsils into the cervical posterior subarachnoid space, which may be the cause of the mirror movements. It is postulated that the effects of hindbrain herniation on the pyramidal decussation lead to aberrant development of the neural pathways and mirror movements(5).

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**P. Krishnakumar,
P. Jisha,**

*Department of Pediatrics,
Institute of Maternal and Child Health
Medical College, Calicut,
Kerala, India.*

E-mail: krshnakumar@sancharnet.in

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Ketamine

The article, "Safe and efficacious use of procedural sedation and analgesia (PSA) by non-anesthesiologists in a pediatric hematology-oncology unit" in the April 2006 issue was indeed very interesting. Regarding the use of ketamine for PSA in children outside the operation theater, certain important points need to be emphasized.

Firstly, though midazolam and ketamine is probably the best combination available for PSA, the incidence of oxygen desaturation is much higher with this as compared to use of a single drug alone. This combination is also reported to produce prolonged sedation and psychedelic effects in children which may adversely alter the child's comfort and parental satisfaction(1).

Secondly, as mentioned in the article, vigilant monitoring for occurrence of known complications like apnea, hypoxia, laryngospasm, seizures, arrhythmias, vomiting, etc; is essential. As far as possible, adequate preventive measures such as addition of an antiemetic and anticholinergic is recommended. Another exceptional feature of ketamine is that as opposed to other sedative drugs with cardio-depressant properties, it causes a rise in heart rate as well as both systolic and diastolic blood pressure(6).

Besides ketamine, PSA has been administered successfully using alternate drugs like propofol, etomidate and methohexital. In comparison to ketamine, propofol has shown more rapid recovery, smoother emergence, and shorter stay in the pediatric critical care department(3).

Sona Chaturvedi,

*Department of Anesthesiology,
Mahatma Gandhi Institute of Medical Sciences,
Sewagram 442 102,
Maharashtra, India.*

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