

## Case Report

# Dyke-Davidoff- Masson syndrome presenting with mirror movement

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### Abstract:

Mirror movement is a rare neurologic soft sign associated with many conditions. Dyke-Davidoff- Masson syndrome (DDMS) is a rare condition characterized by unilateral cerebral atrophy and facial asymmetry, hemiparesis, seizures and mental retardation. Association of mirror movements with DDMS which was previously suggested in a few studies, will be presented and discussed. We report a case of 10 year old male with infantile right hemiparesis now presented with 5 year history of mirror movements of both upper limbs. There is right hemiatrophy with spastic right hemiparesis. MRI brain revealed hemiatrophy of left hemisphere, left half of midbrain with thickening of left skull vault with normal cervical spine, features consistent with DDMS. Although treatment of both conditions is conservative, goal of treatment is improving the quality of life of the patient so that with existing limitation, patient can lead more meaningful life.

**Introduction:** Mirror movement is a rare and interesting but often overlooked neurologic soft sign characterized by simultaneous contralateral, involuntary, identical movement that accompany voluntary movements.<sup>1</sup> Dyke-Davidoff-Masson Syndrome (DDMS) is characterized by convulsions, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. The etiology is not clear, but prenatal infections, vascular pathologies, genetic mutations, hypoxia, intracranial hemorrhage and trauma, febrile convulsions and infections occurring during birth which may cause in utero brain damage or during the early periods of life are thought to be the main causes<sup>2</sup>.

We report a case of rare association of DDMS with mirror movement.

**Key words :** Mirror movement, Dyke-Davidoff-Masson Syndrome, Hemiatrophy

### Case Report:

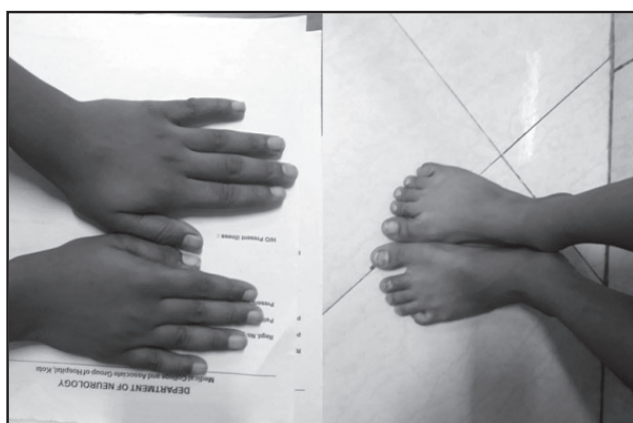
Our patient is a 10 year male born by normal vaginal delivery with uneventful antenatal and perinatal history brought with complaints of insidious onset, progressive involuntary movement of one upper limb mimicking the voluntary movement of other limb noticed since last 4 to 5 years. The severity of these movements is relatively static since long time. There is also history of weakness of right half of body which was noticed by his parents at 6 month of age, static since then. No history of seizures, cognitive decline was present. Other past and family history was negative with normal social and language milestone history. On examination, he has mild atrophy of right half of body, mild facial asymmetry (Figure 1), short right upper and lower limbs compared to left side (Figure 2) with spasticity, mild weakness (4/5 power) and brisk reflexes on right side. EEG (Electroencephalography) and IQ testing were normal (94). MRI brain revealed hemiatrophy of left hemisphere, left half of midbrain with thickening of left skull vault (Figure 3) with normal cervical spine, features consistent with DDMS. Patient was treated conservatively with physiotherapy with partial relief of symptoms and is under regular follow up.

### Discussion:

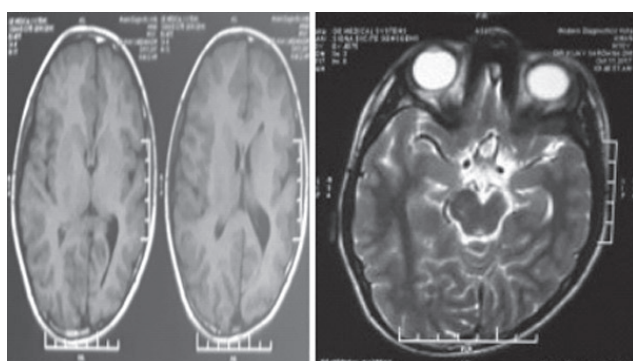
Erlenmeyer first applied the term mirror



**Figure 1 :** Image of the patient showing facial asymmetry



**Figure 2 :**Image of the patient showing short right upper and lower limbs compared to left side



**Figure 3 :** T1 & T2 MRI images showing Left hemispheric and Left half of midbrain atrophy with dilatation of Occipital horn of ipsilateral Lateral ventricle and thickening of the overlying skull bone

movement in 1879<sup>3</sup>, the first description of these movements as “involuntary, synkinetic mirror reversals of an intended movement of opposite side”, was given by Cohen et al, in 1991<sup>4</sup>. Usually, do not persist beyond age of 10 years. Mirror movements are more common in the upper limb than the lower limb. Several rare diseases known to be associated with mirror movements include cerebral palsy, Klippel-Feil Syndrome, phenylketonuria, Friedreich’s ataxia, schizophrenia, parkinsonism and Kallman syndromes<sup>5</sup>. Patients with DDMS usually present with refractory seizures but this case never had history of seizures.

Pathophysiology of Mirror movements is poorly understood. Possible mechanisms defined are abnormal decussation of the corticospinal tract, abnormal interhemispheric inhibition and bilateral cortical activation of primary motor areas during intended unimanual movements and an abnormal involvement of the supplementary motor area during both unimanual and bimanual movements<sup>6</sup>.

DDMS is a congenital or acquired in early infantile period, characterized by facial asymmetry, contralateral hemiparesis, mental retardation and seizures due to ipsilateral hemiatrophy, usually due to an insult in fetal or early childhood period<sup>7</sup>. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses<sup>8</sup>.

In 2002, Bhattacharya A and Lahiri A, reported twenty three patients with mirror movements, studied over a period of six years among which 4 patients had symptomatic epilepsy, only one had DDMS<sup>1</sup>.

In a study done by Bhargava A et al. in 32 patients of DDMS, only one patient had associated mirror movement<sup>9</sup>.

There is no proven drug treatment available for mirror movements. Adaptation of the school environment, limitation of the amount of handwriting helps parent to consider a future profession that does not require complex bimanual

movements<sup>10</sup>. Few case reports demonstrates that Botulinum toxin may be useful to treat Mirror movements. Physiotherapy, occupational and speech therapy play a significant role in long-term management. Appropriate anticonvulsants if the child develop seizures should be used. Hemispherectomy is the treatment of choice for children with intractable disabling seizures and hemiplegia with a success rate of 85%<sup>11</sup>.

**Conclusion :**

Both DDMS and Mirror movements are rare neurologic manifestations. Their association in a patient was reported previously in few reports. Treatment of both conditions is mainly conservative with target of controlling seizures whenever present and improving the quality of life of the patient. Identification of this entity may help in advising long term prognosis and occupational therapy so that with existing limitation, patient can lead more meaningful life.

**Declarations:**

**Contribution of authors:** Vijay Sardana-concept, strategy, guidance. Rahi Kiran Bhattiprolu-revision of manuscript, data collection.

**Conflict of interest:** No

**Funding source and its role in the study:** No

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