CASE REPORT

# Anesthetic considerations in patients with Dyke-Davidoff-Masson syndrome: a case report and review of literature

Aparna Bagle[1], Tripti Nagdev[2], Guneet Chadha[3], Jateen Amonkar[2]

*[1]Associate Professor; [2]Resident; [3]Lecturer*
*Department Of Anesthesiology, Dr. D. Y. Patil Medical College & Hospital, Pune, Maharashtra, (India)*

**Correspondence:** Dr Tripti Nagdev, Department of Anesthesiology, Dr. D. Y. Patil Medical College & Hospital, Pimpri, Sant Tukaram Nagar, Pune, Maharashtra 411018, (India); E-mail: triptinagdev@ymail.com

## ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disorder of cerebral hemiatrophy. The clinical presentation may consist of facial asymmetry, contralateral atrophy (including the trunk and extremities), hemiparesis, speech difficulties, mental retardation and epilepsy.

It involves multiple systems, especially problems of the airway, occult myopathy and seizure disorder. Anesthesia for such patients is a challenge to the anesthesiologist. We report the anesthetic care of 9 year old female child of DDMS for fractional lengthening of tendons of the forearm. Airway management, induction technique, pathophysiology of the disease, drug selection and other concerns of anesthesia have been discussed reviewing the sparse literature.

**Key words**: Dyke-Davidoff-Masson syndrome; Seizure; Cerebral Hemiatrophy; Hemiparesis

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## INTRODUCTION

The Dyke-Davidoff-Masson syndrome (DDMS) is defined as atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) which is secondary to brain insult in fetal or early childhood period.[1]

The clinical features are variable and depend on the extent of brain injury. More commonly they present with recurrent seizures, facial asymmetry, contralateral hemiplegia, mental retardation or learning disability and speech and language disorders. Seizures can be generalized or focal.[2] Typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of skull and sinuses. Syndrome had been documented mainly in adolescent and adults.[3-5] However, it can also be seen in children.[6]

## CASE REPORT

A nine year old female child presented for a pre-anesthesia check up with complaints of shortening of right lower limb, and stiffening of and inability to extend right elbow joint. She was posted for fractional lengthening of tendons. Parents gave history of a febrile seizure at the age of three years. She developed weakness of right upper and lower limb after seizure.

She had been delivered by emergency cesarean section at full term in view of fetal distress, after which she had to be nursed in NICU for one week in view of birth asphyxia due to meconium aspiration. Birth weight was 3 kg. No developmental delay or delayed milestones. Not properly vaccinated till date. There was no history of similar illness in any other sibling or family member.

On examination she was conscious and oriented. Her facial features showed mild facial asymmetry with deviation of mouth to left side (Figure 1).

Body weight was 30 kg. There was mild mental

**Figure 1: Showing deviation of mouth to left side**

retardation (delayed schooling). She was afebrile, alert and had a scissoring gait. Cardiovascular and respiratory systems were normal. Neurological examination revealed power in right upper limb and right lower limb 4/5 and 3/5 respectively. Power at wrist joint was 0/5 (Figure 2).

**Figure 2: Showing Right upper limb**

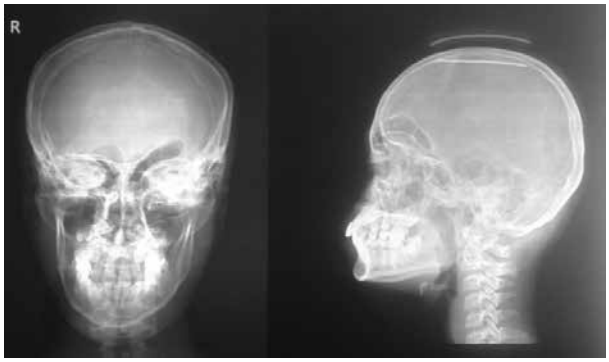


**Figure 3: X-ray skull showing thickening of skull vault and enlargement of left frontal sinus also ethmoidal air cells.**

Right plantar was extensor. There was no sensory deficit, cranial nerve, or bowel bladder involvement. Thoracic spine scoliosis to left was present. Mallampati classification was I, thyromental distance and inter incisor distance were normal. Complete blood count and all routine biochemical investigations (renal and liver function tests, random blood sugar and serum electrolytes levels) were within normal range.

Chest x-ray was normal.

Her x-ray skull AP and lateral view showed thickening of skull vault (compensatory) on left side and enlargement of left frontal sinus and enlargement of the ethmoidal air cells (Figure 3).

MRI Brain revealed atrophy of left cerebral hemisphere (consistent with Dyke-Davidoff-Masson syndrome). Chronic ischemic changes in left periventricular white mater, left corona radiata and centrum semiovale (Figure 4).

The patient was posted for fractional lengthening of tendons of forearm. The procedure demanded general anesthesia. The procedure was explained to the patient's parent and the patient was accepted under ASA grade III. Written informed high risk consent was taken and possibility of Intensive Care Unit stay was explained. Patient was kept NBM for 6 hours and taken in the operation theatre.

In the operation theatre, after attaching the pulse oximeter, ECG, and non-invasive blood pressure cuff the baseline vitals were recorded. Intravenous line was secured with 22 G cannula.

Premedication with inj glycopyrrolate 0.12 mg, inj midazolam 0.6 mg, inj ondansetron 3

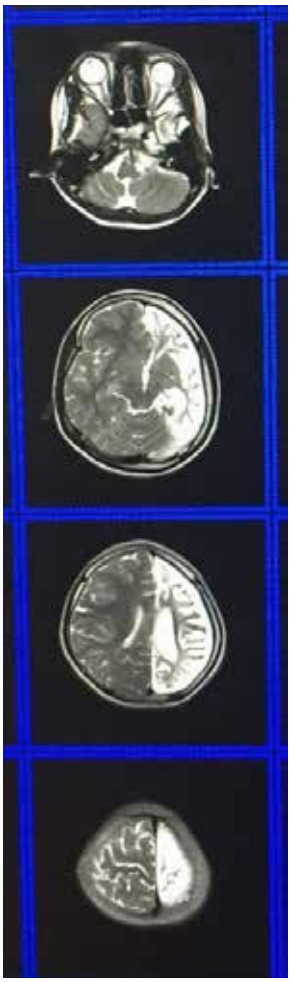

**Figure 4: MRI brain showing atrophic hemicerebrum**

mg and inj fentanyl 60 $\mu$g was done. Patient was preoxygenated for 3 min. Patient was anesthetized using inj thiopentone 150 mg and inj vecuronium 3 mg and sevoflurane 2-3%. Oral Intubation was done with endotracheal tube size 6 (cuffed). Tracheal placement was confirmed with $EtCO_2$. Anesthesia was maintained with nitrous oxide-oxygen (50:50) and sevoflurane and vecuronium. Surgery lasted for two and a half hours and was uneventful. Intra operative blood loss was about 100 ml. After attaining spontaneous respiratory efforts, patient was reversed with neostigmine 1.5 mg and glycopyrrolate 0.3 mg. She was extubated uneventfully and shifted to recovery room for observation postoperatively.

## DISCUSSION

In 1933, CG Dyke, LM Davidoff and CB Masson first described the syndrome in plain radiographic and pneumoencephalographic changes in a series of nine patients.[7] The syndrome causes cortical and subcortical atrophy, porencephalic cysts, contralateral hemiparesis, infarction and gliosis of middle cerebral artery with compensatory hypertrophy of skull and sinuses.

The syndrome is characterized by asymmetrical growth of cerebral hemisphere with atrophy or hypoplasia of one side and midline shift, ipsilateral osseous hypertrophy with hyperpneumatisation of sinuses mainly frontal and mastoid air cells with contralateral paresis. Other features are enlargement of ipsilateral sulci, dilatation of ipsilateral sulci, dilatation of ipsilateral ventricle and cisternal space, decrease in size of ipsilateral cranial fossa and unilateral thickening of skull. [8]

Cerebral hemiatrophy can be of two types; infantile (congenital ) and acquired.[9] The infantile variety results from various etiologies such as infection, neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies and coarctation of the mid aortic arch.[9,10] Patient becomes symptomatic in perinatal period or infancy. The main causes of acquired type are trauma, tumor, infection, ischemia, hemorrhage and prolonged febrile seizures. Hageman et al. proposed the term cerebral hemi-hypoplasia or unilateral cerebral hypoplasia for the congenital cerebral atrophy because there is a lack of cerebral development rather than atrophy.[11] When the cerebral hemiatrophy develops in utero or during first 2 years of life, it is associated with certain cranial changes like ipsilateral hypertrophy of the skull and sinuses as a compensatory change to take up the relative vacuum created by the hypoplastic cerebrum. Age of presentation depends on time of insult and character of insult. Changes maybe be seen only in adolescence or adult.[12]

The condition needs to be differentiated from basal ganglia germinoma, Sturge Weber syndrome, Fishman syndrome, Silver – Russell syndrome and Rasmussen encephalitis.[12]

Challenges to the anesthetist in cases of DDMS are seizure disorder and cerebral insult, difficult airway/ intubation, pediatric age group, involvement of multiple systems and occult myopathies.[13]

Avoid anesthetic drugs and techniques which decreased cerebral perfusion and oxygenation, and trigger seizure activity. Attenuating the pressor response during intubation and a smooth extubation is also of prime importance in these patients.

We need to keep in mind the occult myopathy. So, succinylcholine to be avoided because of rhabdomyolysis and hyperkalemia, but usually there is normal response to depolarizing muscle relaxant.

Patients with DDMS can be successfully managed by avoiding further insult to the brain by maintaining adequate blood pressure, avoiding hypoxia, avoiding the presser response during intubation and rise in intracranial pressure due to anesthetic drugs and techniques.

## CONCLUSION

Successful management of such patients includes anticipating problems in the peri-operative period, vigilant monitoring and prompt management.

**Conflicts of interest:** None declared by the authors

**Author contribution:** All authors took part in the preparation of this case report.

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