



## Dyke-Davidoff-Masson Syndrome- A Case Report

Authors

**Dr Tabassum Saher<sup>1</sup>, Dr Priya Chauhan<sup>2</sup>, Dr Joginder Yadav<sup>3</sup>**

<sup>1</sup>Assistant Professor, Faculty of Physiotherapy, SGT University Gurugram

<sup>2</sup>Assistant Professor, Faculty of Physiotherapy, SGT University Gurugram

<sup>3</sup>Dean, Faculty of Physiotherapy, SGT University Gurugram

### Abstract

*Dyke-Davidoff-Masson Syndrome (DDMS) is a syndrome in which the diagnosis is mainly done by various clinical presentations along with positive radiological findings. Generally, patients complain of facial asymmetry, seizures, learning difficulties, and contra lateral hemiparesis. The case was on 2.5 years old male child presented with left side spastic hemiparesis and was having a history of delayed cry, delayed milestones. He was diagnosed clinically with DDMS (Dyke Davidoff Masson Syndrome) based on EEG and MRI report's findings.*

**Keywords:** *Dyke Davidoff Masson Syndrome, Spastic hemiparesis, Facial asymmetry.*

### Introduction

Dyke Davidoff Masson Syndrome is an uncommon condition which refers to atrophy or hypoplasia of one cerebral hemisphere resulting due to an insult during the developing brain in fetal or in early childhood period. The clinical feature varies and depends upon the extent to which the brain is injured. The prominent features include facial asymmetry, recurrent seizures, contralateral hemiplegia, mental retardation or learning disability and various speech and language disorders<sup>(1-3)</sup>. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults<sup>(3-6)</sup>. However, it can be seen in children<sup>(3,7)</sup>. We present here a 2.5 year old male child with typical clinical and imaging features and treatment options of DDMS.

### Case report

A 2.5 year old male child presented with complaints of focal seizures, characterized by brief episodic movements of left side of body after which child regains consciousness. He had a delayed milestone of development in the form of not able to communicate or understand language. He had microcephali without any focal convulsions or facial asymmetry. The bilateral carotid pulsations were normal. Vision and hearing were normal and cranial nerves were intact. There was no history of head trauma. Neurological examination revealed left sided spastic hemiparesis with brisk tendon reflexes and extensor plantar response. Other systemic examination came out to be normal. A multiplaner and multisectional MRI acquired in T1W, T2W, DWI and flair sequences revealed gliencephalomalacic area in right cerebral hemisphere with

hemiatrophy of right mid brain and pons and compensatory hypertrophy of left cerebral hemisphere and ganglionic thalamic complex. There was no midline shift. Tectal plate and the cerebelloquadrate were normal sellar and suprasellar structure were normal. Posterior fossa structures were normal with normal shape of fourth ventricle. From the above MRI finding a diagnosis of DDMS was made. Treatment history revealed consulting local medical practitioner and patient was on carbamazepine. However, patient had stopped responding recently. Other antiepileptic drugs were added in his medication and he almost became seizure free. He was advised speech therapy too and was kept on regular follow-ups.

### Discussion

Dyke-Davidoff-Masson syndrome refers to variable degrees of hypoplasia or atrophy of one cerebral hemisphere with compensatory changes of the calvarium. The etiopathogenesis could be either vascular insult during intrauterine life resulting in hypoplasia of a cerebral hemisphere or acquired causes like trauma, infection, vascular abnormalities and intracranial hemorrhage in the perinatal period or shortly thereafter causing hemispheric atrophy. Cerebral atrophy in turn results due to reduction in the formation of brain derived neurotrophic factors by these causes<sup>(8,9)</sup>. The syndrome was first described by Dyke, Davidoff and Masson in 1933 on plain skull radiographs and pneumo encephalograms in a series of nine patients. The clinical features depend on extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation or learning disability, speech or language disorders and facial asymmetry. Rarely patients can also present with sensory symptoms and psychiatric disorders like schizophrenia. Nevertheless, seizures may appear months to years after the onset of hemiparesis and mental retardation is not seen in all cases<sup>(9-11)</sup>. Management consists of control of seizures with appropriate anticonvulsants, as most patients with this

disorder present with refractory seizures. Physiotherapy is found to be a very important and crucial option for these patients.

Hemispherectomy is indicated in patients with hemiplegia and intractable disabling seizures and is successful in 85% of the cases. Prognosis is poor in cases of prolonged or recurrent seizures and if hemiparesis occurs before two years of age. Hence, it is indeed very important for neurologists, paediatricians and radiologists to be familiar with this condition for its early diagnosis and treatment<sup>(9-12)</sup>.

### References

1. Dyke C G, Davidoff L M and Masson C B. Cerebral Hemiatrophy and homolateral hypertrophy of the skull and sinuses. *Surg Gynecol Obstet* 1933; 57: 588-600.
2. Sharma S, Goyal I, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff-Masson Syndrome. *Ind J Radiol Imag*. 2006;16:165-6.
3. Manas R Behera, Sibabrata Patnaik, Aswini K Mohanty. Dyke-Davidoff-Masson Syndrome. *J Neurosci Rural Pract*. 2012 Sep-Dec; 3(3): 411-3.
4. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson Syndrome: Classical imaging findings. *J Pediatr Neurosci*. 2010;5:124-5.
5. Koshy B, Surendrababu NR. Dyke-Davidoff-Masson Syndrome. *Ann Acad Med Singapore*. 2010;39:501-2.
6. Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson Syndrome. *Neurol India*. 2003;51:136.
7. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson Syndrome. *Indian Pediatr*. 2008;45:927-8.
8. Lee JH, Lee ZI, Kim HK, et al. A case of Dyke-Davidoff-Masson syndrome in Korea. *Korean J Pediatr* 2006;49:208-11.
9. Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff-Masson syndrome. *Indian J Radiol Imaging*

2006;16:165-6.

10. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: Classical imaging findings. *J Pediatr Neurosci* 2010;5:124-5.
11. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. *J Neurosci Rural Pract* 2012;3:411-3.
12. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. *Indian Pediatr* 2008;45:927-8.