

Exploring Dyke-Davidoff-Masson Syndrome: Key Insights From A Comprehensive Case Series

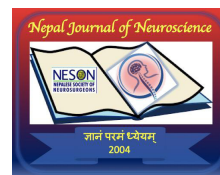
Asit Parmanand Patil¹, Sushma Uttam Save²

¹ MD (Pediatrics), Fellowship in Pediatric Neurology and Epilepsy, Consultant Pediatric Neurologist,

Early Intervention and Rehabilitation Centre for Children, Mumbai (Branch of TNMC and BYL Nair Hospital, Mumbai)

².Professor (Additional), Incharge of PICU and Pediatric Neurology and Epilepsy clinic, TNMC and BYL Nair Hospital,

Mumbai



Date of Submission: 25th June 2026

Date of Acceptance: 12th July 2026

Date of Publication: 15th September 2026

Abstract

Dyke Davidoff Masson syndrome (DDMS) also called cerebral hemiatrophy is a rare syndrome presents with cerebral injury leading to hypoplasia in one of the cerebral hemispheres. It is classified into congenital and acquired. DDMS have variable clinical features with almost consistent neuroimaging findings. We aim to highlight clinical features, neuroimaging and electroencephalographic reports to help elucidate the readers with the wide spectrum of features in this disease.

Key words: Dyke Davidoff Masson syndrome (DDMS), weakness or hemiplegia, seizures, atrophied cerebral hemisphere (unilateral), Multidisciplinary approach.

Introduction

Dyke-Davidoff-Masson (DDMS) first described the Dyke, Davidoff, and Masson syndrome in 1933¹. Since its identification, fewer than 100 cases of DDMS have been documented. This rare condition is characterized by cerebral hemiatrophy, compensatory ipsilateral skull hypertrophy, and hyperpneumatization of the paranasal sinuses, as seen in radiographic imaging. Clinically, DDMS presents with contralateral hemiparesis, epilepsy resistant to treatment, facial asymmetry, and intellectual disability. The exact causes of

DDMS remain debated². The management involves controlling seizures with suitable anti-seizure medications as most individuals with this condition experience refractory seizures. In addition, physiotherapy along with occupational and speech therapy, play an essential role in treatment³.

We report the variable clinical presentations and radiological signs of this syndrome through case series of 3 patients as represented in Table 1 below.

Discussion

DDMS is a rare syndrome with only a limited number of cases documented in medical literature⁴. DDMS encompasses a spectrum of symptoms with varying degree of clinical presentation. It is linked to epileptic seizures, which are frequently resistant to treatment. These seizures can be focal or generalized, and the condition may also involve hemiplegia, intellectual impairment, facial asymmetry, cleft palate, cavus foot and extremity atrophy. The severity of the disorder is influenced by the onset of its occurrence during brain development⁵. Our case series present with variable clinical features including recurrent seizures, low IQ, developmental delay with microcephaly. Power and tone was variable and dependent on intervention and rehabilitation. Seizures semiology varied though electroencephalography showed focal epileptiform discharges. Two of the cases had hemiparesis and one had dyslexia with behavioural abnormalities in the form of aggression. The primary cause of DDMS is cerebral insult, which can occur either in utero or during early childhood. Prenatal factors include congenital anomalies, vascular malformations, cerebral infarction, and infections. Perinatal factors involve hypoxia, birth

Access this article online

Website: <https://www.nepjol.info/index.php/NJN>

DOI: <https://doi.org/10.3126/njn.v23i3.98025>

HOW TO CITE

Patil, A. P., & Save, S. U. (n.d.). Exploring Dyke-Davidoff-Masson Syndrome: Key Insights from a Comprehensive Case Series. *NJNS*. 2026;23(3):67-69



Address for correspondence:

Dr. Asit Parmanand Patil

MD (Pediatrics), Fellowship in Pediatric Neurology and Epilepsy,

Consultant Pediatric Neurologist,

Early Intervention and Rehabilitation Centre for Children,

Mumbai (Branch of TNMC and BYL Nair Hospital, Mumbai)

E-mail: asitpatil030@gmail.com

Copyright © 2023 Nepalese Society of Neurosurgeons (NESON)

ISSN: 1813-1948 (Print), 1813-1956 (Online)



This work is licensed under a Creative Commons Attribution-Non Commercial 4.0 International License.

	Case1	Case2	Case3
Age	16 years	14 years	10 years
Sex	Female	Male	Female
Anthropometry	Head circumference 42cm (-3SD), height 160cm (0to-1SD), weight 26.4kg (-3SD), BMI 10.3 (-3sd)	head circumference 46cm (-3SD), height 158cm (0to-1SD), weight 44kg(0to-1SD), BMI 17.6 (0to-1SD)	head circumference 47cm (-3SD), height 138cm(0to+1SD), weight 27kg(0to-1SD), BMI 14.2 (-1to-2SD)
Clinical features	Weakness of right upper and lower limbs, Speaks sentences with few words. Walks with support, Right sided circumduction gait.	No global developmental delay, Mild cognitive decline Increased tone on left side with power of 3/5 at all joints in upper & lower limbs Hemiparetic gait.	Mild weakness of right upper and lower limb noted since 1 year of age with epilepsy on anti-seizure medication and an anticoagulant. Learning disability in the form of difficulty in reading predominantly. Behavioural abnormalities in the form of increased aggression (hitting her school companions).
Seizure semiology	Only myoclonic seizures multiple episodes.	The patient had febrile seizures with changing semiology: 1st at 1 year (generalized tonic-clonic with fever), 2nd at 2 years (left limb tightening, fever, and lateral rectus palsy), 3rd at 3 years (left limb deficit), and 4th at 4 years (left tonic-clonic, incontinence, no fever or postictal drowsiness). Diagnosed with focal impaired awareness seizures.	At 6 months of age febrile status epilepticus and required intensive care, later recognised to have right sided weakness. Repeat episode of febrile status epilepticus at 3 years of age Later duration of febrile seizures reduced and acquired generalised tonic clonic semiology. Last seizure was noted in January 2024.
Etiology	Eventful perinatal period. Meconium aspiration syndrome requiring Neonatal intensive care unit stay and ventilator for 12 days	Antenatal and Birth history were uneventful. Post natal ischemic insult as confirmed on MRI Brain. Febrile seizures.	Antenatal and Birth history were uneventful. Post natal chronic infarcts. Febrile seizures
EEG (Interictal)	focal right sided frontocentral epileptiform activity.	focal right temporo-parieto-occipital epileptiform activity	left focal fronto-temporal epileptiform activity.
MRI Brain	Severe atrophy of left cerebral hemisphere with mild thickening of overlying calvarial bones.	Moderate atrophy of right cerebral hemisphere with areas of gliosis in right parietal and occipital region causing ex vacuo dilation body and occipital horns of right ventricle sequalae to old ischemic insult, encephalomalacia in right posterior parietal region, right cerebral hemiatrophy.	Cystic encephalomalacia (suggestive of evolving chronic infarcts) involving both frontal and left temporo-occipital lobes and multiple areas of blooming in both cerebral hemispheres. Repeat MRI Brain: Hemi-cerebral atrophy of left half of cerebral hemisphere with adjacent calvarial thickening of left frontal and occipital bone and ipsilateral falcine displacement to left in its posterior aspect.
Management	Anti-seizure medication and symptomatic management. Physiotherapy and occupation therapy.		

trauma and intracranial haemorrhage. Postnatal hemiatrophy may result from cerebral tumours, infections, trauma and febrile seizures. The infantile (congenital) form of DDMS, unlike the acquired form, is characterized by an enlarged calvarium, diploic space, and paranasal sinuses. These cranial changes occur as a compensatory response to the vacuum created by the atrophied cerebral hemisphere⁶. The congenital form suggests that the cerebral insult likely occurs in utero, possibly due to cerebral infarction, vascular malformation, gestational vascular occlusion, coarctation of the mid-aortic arch, or infections. In contrast, the acquired form indicates that the cerebral insult likely occurred during the perinatal or postnatal period, resulting from factors such as birth trauma, intracranial haemorrhage, hypoxia, tumours, infections or prolonged febrile seizures⁷. Our patients had acquired form of DDMS with distinct etiologies namely meconium aspiration syndrome requiring ventilator, recurrent febrile seizures and febrile status epilepticus with neuroimaging suspecting stroke. The differential diagnoses are sturge weber syndrome and chronic Rasmussen encephalitis. However, sturge weber syndrome shows enhancing pial angiomas and cortical calcifications, Rasmussen encephalitis doesn't present with calvarial changes³. As there is no established protocol for treating DDMS, management focuses mainly on symptom relief, including the use of anti-seizure medications to control seizures. For cases with severe, intractable seizures, hemispherectomy is a possible neurosurgical treatment, with an 85% reported success rate. Long-term care involves physiotherapy for hemiparesis, occupational therapy, speech therapy for communication difficulties, psychiatric support, and medication as needed⁴.

Conclusion

Multidisciplinary approach including neurologist, occupation therapist, physiotherapist, speech therapist, radiologist have important role aiding the child's mental and physical development. Neuroimaging is crucial for diagnosis with accurate neurological history to identify the etiology. Many conditions have similar presentation thus awareness of DDMS needs to be improved to consider it as an important differential diagnosis and avoid it getting misdiagnosed. Early diagnosis and intervention with appropriate rehabilitation is the mainstay for management. This case series provides valuable information regarding variable clinical presentation and etiology in DDMS.

Financial Support

No funding was obtained for this study.

Conflict Of Interest

The author(s) declare that they do not have any conflict of interest regarding the publication of this article.

References

1. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy and homolateral hypertrophy 17 of the skull and sinuses. *Surg Gynecol Obstet.* 1933;57:588–600.
2. Wang B, Jiang W, Yan W, Tian J, Xu J, Li Y, Zhao Y, Dai Y, Cheng G, Hou G. Clinical 19 characteristics and neuroimaging findings of seven patients with Dyke Davidoff Masson 20 syndrome. *BMC neurol.* 2021 Dec;21:1-9.
3. Arora R, Rani JY. Dyke-Davidoff-Masson syndrome: imaging features with illustration of 22 two cases. *Quant Imaging Med Surg.* 2015 Jun;5(3):469.
4. Anirudh CV, Nishitha K, Kumar PS, Prajwal MM, Nalamalapu RR. Dyke-Davidoff-24 Masson Syndrome: A Case Report. *Cureus.* 2023;15(2).
5. Bahlouli N, Chait F, Laasri K, El Harras Y, Allali N, Chat L, El Haddad S. Dyke-Davidoff-26 Masson syndrome in an 8-year-old child: Report of a case. *Radiol Case Rep.* 2024 Jan 27 1;19(1):239-41.
6. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: classical imaging 29 findings. *J Pediatr Neurosci.* 2010 Jul 1;5(2):124-5.
7. Kalaskar R, Kalaskar AR. Classical oral manifestations of Dyke-Davidoff-Masson 31 syndrome: a case report with review of the literature. *J Korean Assoc Oral Maxillofac Surg.* 32 2018 Aug 1;44(4):198-203.