ORIGINAL ARTICLE

# Cerebral Hemiatrophy (Dyke-Davidoff-Masson Syndrome): Prospective Study of 36 Cases

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## Abstract

*The purpose of this study was to assess predominantly the cerebral parenchymal and CSF space changes in patients suffering from hemiparetic cerebral palsy (Dyke-Davidoff-Masson Syndrome) using CT and MRI Imaging. Over a period of 48 months, about 15000 CT scans and 7000 MRI studies of brain were reviewed in a diagnostic center of Dhaka city. Thirty six patients were diagnosed radiologically as Dyke-Davidoff-Masson Syndrome - 27 patients by CT scan and nine by MRI. Incidence was found to be about one in 600 patients. Twenty were male and sixteen were female patients. Age ranged from about one year to twenty five years. Fifty three percent was diagnosed in the first decade of life and thirty seven percent in the second decade. The main CT and MRI findings of cerebral parenchyma was unilateral cerebral volume loss in fifteen patients and regional (MCA territory) in twenty patients with low density on CT images and T1 hypo- and T2 hyperintensity on MRI images. Brainstem and thalamic atrophy were also noted in six patients. Ipsilateral midline shift (six patients), gliosis (six patients), encephalomalacia (eight patients), porencephaly (four patients) were found the associated findings. The commonest cranial skeletal finding was Ipsilateral calvarial thickening followed by overpneumotization of the paranasal sinuses.*

*Keywords: Dyke-Davidoff-Masson Syndrome, Hemiatrophy, CP, Calvarial, Porencephaly, Encephalomalacia*

## Introduction

C. G. Dyke, L. M. Davidoff and C. B. Masson first described Dyke-Davidoff-Masson syndrome of cerebral hemiatrophy in 1933. As per original description, the syndrome consists of cerebral hemiatrophy with hemicranial hypertrophy (homolateral hypertrophy of skull and sinuses causing facial asymmetry and elevation of sphenoid wing and petrous ridge) in association with contralateral hemiplegia, seizure disorders, mental retardation and difficulty and impairment in speech.

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The cranio-cerebral morphological and pathological changes of this syndrome are sequelae of some form of injury to the brain. Any form of insult to the immature brain either in-utero or in early life results in neuronal loss and impaired growth of the brain. Causes of this syndrome may be congenital or acquired.

Intrauterine causes are not well understood, but thought to be due to vascular occlusion usually involving middle cerebral artery territory (Sener et al[5]), resulting from infection, congenital malformation etc. Internal carotid artery may also be involved affecting the ipsilateral cerebral hemisphere.

Acquired causes are usually the sequelae of ischaemic insult to the brain parenchyma and

occurring within first two years of life before the brain and the overlying skull completely develops. The commonest causes are neonatal asphyxia, meningitis, intracranial haemorrhage, trauma etc.

The patient clinically presents with combination of the following features, depending on the time of insult and extent of brain injury: unilateral hemiparesis / hemiplegia, seizure disorders, mental retardation, behavioral changes, learning difficulties and cranio-facial asymmetry.

Demonstration of the imaging features depends on whether the disease occurs in-utero or early life and the extent of the brain damage. CT and MRI are the imaging modalities of choice to assess the radiological features. However, ultrasonogram in neonatal period is helpful to assess the brain parenchymal changes and CSF spaces. X-ray of skull shows some bone changes such as thickened calvarium, hyperpneumotization of the paranasal sinuses etc.

CT and MR Imaging features can be divided in two distinct groups:

a) Features due to brain damage: parenchymal and CSF space changes. Cerebrospinal fluid changes are compensatory enlargement of the homolateral ventricle and extra-ventricular CSF spaces.

b) Features due to secondary skull bone changes: the secondary calvarial changes are compensatory and occur to compensate the vacuum created by unilateral loss of volume of brain. Thickening of the overlying calvarium with widening of diploic space, hyperpneumotization of the ipsilateral paranasal sinuses, overgrowth of ipsilateral mastoid air cells with elevation of the petrous pyramid, high sphenoid ridge and orbital roof and decreased size of the anterior and/or middle cranial fossa are features usually observed.

Though CT and MRI are complementary, MRI is superior for assessment of the etiology and extent of the cerebral parenchymal involvement. Computerized Tomography is better to evaluate the secondary skull bone changes.

## Materials and Method

This study was conducted in a multi-modality diagnostic center in Dhaka city over the period of four years from July 2000 to June 2004 (48 months) using Hitachi Spiral CT Scanner and 0.3T Hitachi Permanent Magnet MRI system. The CT and MRI films were read by two radiologists initially individually and again jointly to avoid or minimize perceptive errors.

During this 48 months period, about 15000 CT Scans and 7000 MRI of brain were reviewed. Thirty six patients of different ages were diagnosed as Dyke-Davidoff-Masson syndrome. More emphasis was given on the patterns of cerebral parenchymal and CSF space changes.

## Results

Among 36 patients, 27 were diagnosed by CT scan and 9 by MRI.

Incidence of Dyke-Davidoff-Masson syndrome in this study was164 per 100,000 patients. There were 20 male and 16 female patients, indicating slight male predilection, though not remarkable. (M:F =5:4).

Age ranges from eight months to 25 years. About 50% of the patients presented in first decade (between 0 – 10 years). The remaining 50% presented during the second and the third decades.

**Table I:** Age Distribution of the patients

| Age | No. of patient | Percentage |
|---|---|---|
| 0 – 05 Years | 07 | 19.45% |
| 05 – 10 Years | 11 | 30.55% |
| 11 – 20 Years | 12 | 33.33% |
| 20+ Years | 06 | 16.67% |

Left hemispheric involvement was noted in 21 (59%) patients and right hemispheric involvement was noted in 15 (41%) patients. There were total unilateral hemispheric involvement in 14 patients (44 %) and predominantly MCA territory involvement in 18 patients (56 %). Ipsilateral midline shift was seen in 05 patients. Ipsilateral thalamic and brainstem atrophy was noted in five patients. Hypoplasia of ipsilateral internal capsule was also found.

Other findings included gliotic changes (six patients), encephalomalacia (seven patients), porencephaly (four patients) – communicating and con-communicating, brainstem and thalamic atrophy (five patients), periventricular leukoencephalopathy (one patient) and minimal to mild similar changes in the contralateral side (three patients).

## Discussion

The common parenchymal findings on CT and MRI were unilateral loss of brain volume - focal, regional (limited within MCA territory) or whole of the cerebral hemisphere. Left hemispheric involvement was seen in about 59% cases. Aetiology of this left sided predominance is not well understood.

**Table II:** Cerebral involvement pattern among the patients

| Pattern | No. | % |
|---|---|---|
| Unilateral hemispheric | 15 | 41.67 |
| Left hemispheric | 23 | 63.89 |
| Right hemispheric | 13 | 36.11 |
| MCA territory | 20 | 55.55 |
| Thalamic and midbrain atrophy | 05 | 13.89 |
| Ipsilateral midline shift | 06 | 16.67 |

MRI demonstrated a good degree of concordance with CT in evaluating cerebral parenchymal changes. On CT images, the involved region(s) shows atrophy with parenchymal hypodensity. A few patients showed no appreciable parenchymal density changes.

On MRI, hypointensity is seen on T1 weighted images and hyperintensity on T2 weighted images. Ipsilateral thalamic and brainstem atrophy were noted in five patients. Hypoplasia of ipsilateral internal capsule was also found. These findings were commonly seen in cases of total hemiatrophy, thought to be occurring due to in-utero insult.

Encephalomalacic changes were manifested by low density/intensity area on CT/MR images. Porencephalic changes were seen as CSF density/intensity area on CT/MRI images. Both communicating and non-communicating forms were noted.

Gliotic changes, a common finding on MRI, were manifested by hyperintensity in the involved regions on T2 weighted images and FLAIR images. Ipsilateral midline shift was found in five patients. This finding is usually seen in patients, when the insult occurs in-utero. Gliotic changes are absent in these patients.

CSF changes are usually compensatory/ex-vacuuo enlargement of the ipsilateral ventricle and adjacent superficial CSF spaces (sulci, fissures and subarachnoid spaces), depending on the extent and location of the parenchymal involvement and volume loss. Among the cranial changes, thickening of the overlying calvarium with widening of diploic space was the commonest feature. The next common feature was hyperpneumatised ipsilateral paranasal sinuses.

Dyke-Davidoff-Masson syndrome should be differentiated from the following entities-

1. **Hemimegalencephaly**: the lateral ventricle of the ipsilateral side is smaller.
2. **Sturge-Weber-Syndrome**: gyral calcification, portwine naevus etc.
3. **Focal Cortical Atrophy**: no associated calvarial thickening is present.

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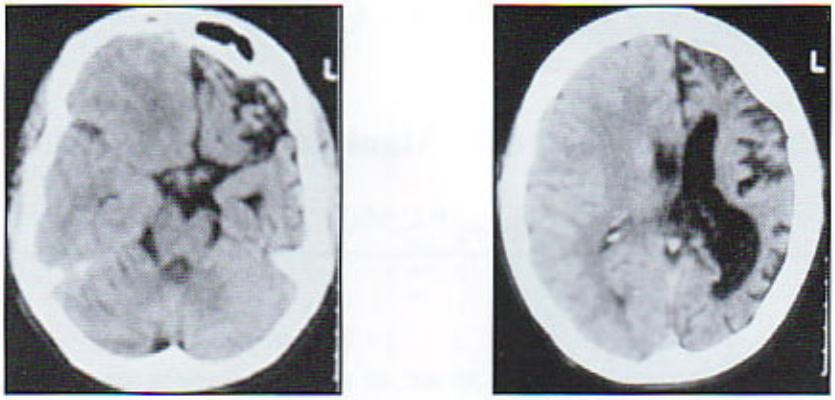
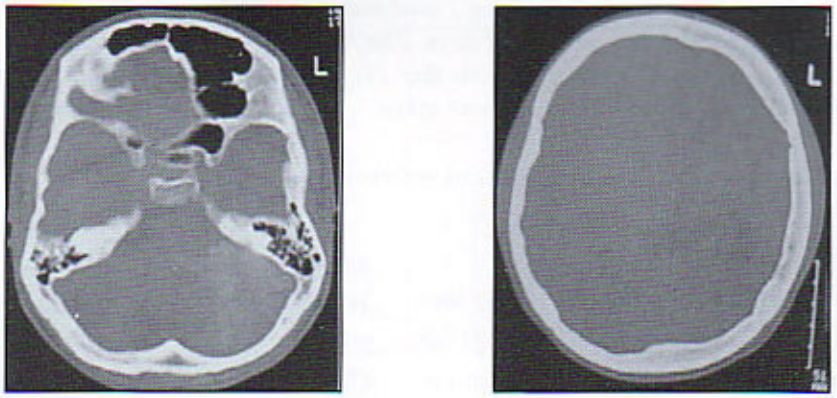
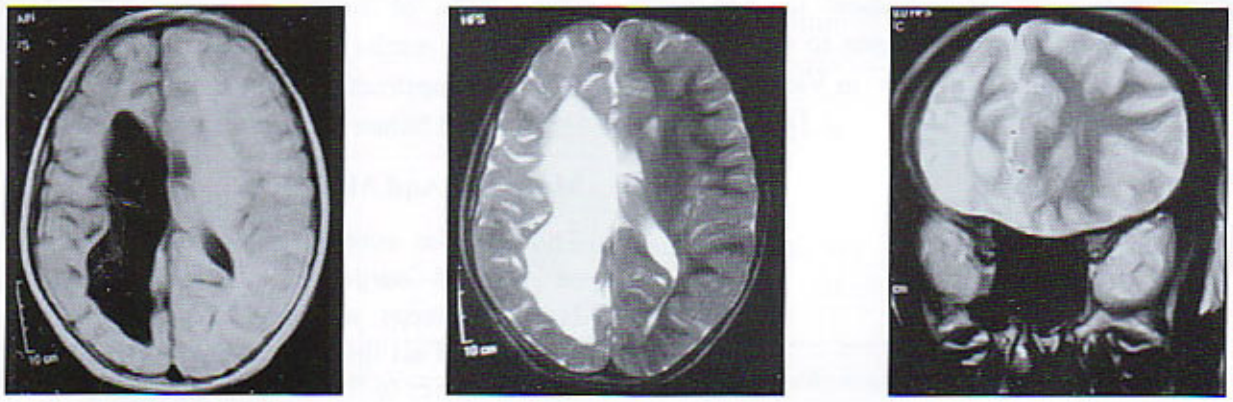


1(a) 1(b)



1(c) 1(d)

Fig-1 (a-d): CT images: Left cerebral hemiatrophy with enlarged CSF spaces (a & b). Thickening of ipsilateral calvarium with hyperpneumatization of left paranasal sinuses and left mastoid with elevation of left petrous ridge (c & d).



a) Axial T1 Weighted b) Axial T2 Weighted c) Coronal T2 Weighted mage

Fig-2 (a-c): MRI of brain. T1 weighted and T2 weighted axial and coronal images showing hemiatrophy of right cerebral hemisphere, dilatation of the ipsilateral lateral ventricle, slightly enlarged superficial CSF spaces and mild thickening of right calvarium.