# PHYSIOTHERAPY MANAGEMENT IMPROVES FUNCTIONAL **RECOVERY IN ACQUIRED DYKE-DAVIDOFF MASSON SYNDROME**

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## ABSTRACT

Background: The Acquired Dyke-Davidoff Masson Syndrome (ADDMS) or cerebral hemiatrophy is rarely encountered case in common rehabilitation practice. Also there is lack of study which explores the physical therapy intervention and rehabilitation protocol. The aim of present study is to propose and explore the physiotherapy intervention based on literature available aimed to enhance functional recovery and cortical reorganization.

Case Description: A 27 year old female presented with episodes of seizures and difficulty in performing ADLs with right upper extremity in physiotherapy OPD. At the age of 3 year, her parents reported that there was a developmental difference between right side upper and lower extremities with lack of movements. The MRI revealed evidence of Encephelomalacia with surrounding gliosis. There was also loss of volume of left cerebral hemisphere with mild ipsilateral calvarian thickening.

Outcome: After the physiotherapy session the patient showed recovery in functional activity. However there was no any significant improvement in strength of muscles. Gripping and other fine motor skills was also improved in Upper extremity.

Discussion: Plastic changes can occur at the cortical level in a number of ways. Evidences suggest that enriched environment and skill learning in adults are associated with growth of dendrites. Possible strategies to enhance the human brains response after injury can be a somatosensory stimulation, motor training, cortical stimulation and combination of sensory stimulation and motor training which is task specific.

Conclusion: Physical therapy intervention improves the functional recovery in ADDMS.

KEY WORDS: Acquired Dyke-Davidoff Masson Syndrome, Cerebral Hemiatrophy, Physical therapy, Rehabilitation.

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## INTRODUCTION

The Dyke-Davidoff Masson Syndrome (DDMS) is trauma in foetal or early childhood period. The defined as the atrophy of one cerebral hemisp-

here or hemiatrophy which is secondary to brain DDMS was first described by Dyke, Davidoff and

Masson in 1933. The study conducted on 5000 CT studies of the head over 2-year period found 10 subjects with hemiatrophy of cerebral cortex [1]. This type of cases are rarely encountered in Clinical practice. The DDMS is characterised by variable degree of facial asymmetry, seizers, contralateral hemiplegia, and mental retardation. Radiological characteristic shows unilateral loss of cerebral volume and compensatory bone alteration in the calvarium, such as thickening, hyperpeuma-tization of the paranasal sinuses and mastoid cells as well as elevated petrous ridge and greater wing of the sphenoid bone [2].

The Cerebral injury that may occur early in life or in utero are the main cause of these findings. The etiology can be categorised as congenital and acquired. In congenital type, the symptoms are present at birth or immediate after birth and there is no etiological factor. Intrauterine vascular occlusion might be the cause for cerebral damage during intrauterine life. In acquired type, the symptoms are related to cerebral damage that occurs in the perinatal period or later stage. The gold standard for diagnosis of DDMS is CTscan and MRI [2]. As this is a rare case there is lack of evidence for diagnosis and management. Most of the available studies is in the form of case report and case series [3].

Management consist of control of seizures with appropriate anticonvulsant, as most patient with this disorder present with seizures. Additionally physiotherapy, occupational therapy have crucial role in functional training [4]. However, upto our knowledge no literature is available which explores the physiotherapy management of acquired DDMS, i.e. the aim of present case report is to explore the management strategies.

# **CASE DESCRIPTION**

A 27 year old female presented with episodes of seizures and difficulty in performing ADLs with right upper extremity in physiotherapy OPD. Subjective examination reveals that she was born with normal delivery and was apparently normal at birth to 3 years of age. At the age of 3 year, her parents reported that there was a developmental difference between right side upper and lower extremities with lack of movements. Her behaviour and communication was normal. At the age of 22 years she got married and after 1 year she had delivered a normal child. She had difficulty in performing ADL with right upper limb, as there was a significant developmental change in skin and muscles (Fig.1).

She was able to walk without assistance but lack of bilateral arm swinging was reported. On examination the elbow & wrist flexors were spastic grade-2 in Modified Ashworth Scale (MAS). Her sensory function was normal. She also reported low back pain since 2 years. Functionally she was independent. The MRI revealed evidence of encephelomalacia with surrounding gliosis. Overlying sulcal spaces and left sylvian fissure were found to be prominent. There was dilatation of left lateral ventricle. Left cerebral hemisphere was relatively smaller in size with signs of wallerian degeneration. There was also loss of volume of left cerebral hemisphere with mild ipsilateral calvarian thickening. Left thalamus and basal ganglia too showed the loss of volume (Fig.2).

However upto our knowledge there is lack of study which explores the physiotherapy management of acquired DDMS, therefore the aim of present case report is to propose or explore the treatment approaches based on available literature in cortical reorganization and cortical plasticity.

**Fig. 1 (a,b):** Showing the attitude of limb and shortening of right upper extremity.







MMT
5
4
3
2
1
0
week1 week2 week3 week4 week5 week6
Shoulder Muscles Arm Muscles Wrist muscles

## INTERVENTION

As the patient had complaint of Seizers she was asked to take prescribed medication during treatment session. Intervention includes Transcutaneous electrical stimulation (TENS) in affected upper limb for 30 minutes following the task related training. Treatment session was for 6 weeks and about 90 minutes of session per day, 6 days a week. Stimulation was applied with 0.2 ms pulse at 100 Hz frequency in constant mode within patient's tolerance level. She was asked to perform reaching activities, with various difficulty levels by altering the sitting surface, adding the weights, changing the objects and gripping exercises. Conventional intervention was also applied to manage the spasticity and to improve the strength. Home based exercise was also advised to the patient which includes practicing the task in home environment.

## **OUTCOMES AND RESULT**

Functional abilities was measured by Upper Extremity Functional Index (UEFI). Modified Ashworth Scale was used to measure spasticity in Upper extremity. Muscle strength was

Week1 week2 week3 week4 week5 week6

UEFI

0

measured for shoulder and arm musculature by MRC grading of Manual Muscle Testing. After the 6 weeks of intervention upper limb function was improved including the gripping and fine motor skills. However there was no significant improvement in muscle strength. Spasticity was also reduced in forearm and wrist flexors from 5<sup>th</sup> week. (Fig. 3 a,b,c)

# DISCUSSION

Acquired Davidoff, Masson Syndrome as diagnosis of indexed case is first described by radiological investigation. The clinical features of acquired DDMS depend upon extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation, learning disability, and behavioural problems [4]. In DDMS the compensatory skull changes reflect an adaptation to unilateral reduction in brain volume. The development and growth of brain proceeds in newborn child and reaches three fourth of full size by 3 years of age [5]. The differential diagnosis can be considered on the basis of clinical features like Ramussen encephalitis, Sturge-Weber Syndrome, Silver-Russel Syndrome, linear nevus syndrome, progressive multifocal Leukoencephalopathy, Fisherman Syndrome and basal ganglia germinoma [3]. The physiotherapy intervention included Transcutaneous electrical stimulation, Task specific exercise and conventional physiotherapy. Treatment protocol for indexed case was given to enhance functional recovery which is based on literature available in reorganization of cortex areas.

Plastic changes can occur at the cortical level in a number of ways. Evidences suggest that enriched environment and skill learning in adult animals are associated with growth of dendrites. Also a work in animals has demonstrated that focal damage in adult brain affects cortical areas are more able to change structure and function in response to afferent signals as a developing brain [6].

The study done by Nudo demonstrated that the hand representation region expanded into region occupied by elbow and shoulder representation areas by exercise in an experimentally induced motor cortex lesion in monkeys, which presents an evidence that the brain plasticity is induced by physical activity [7]. Possible strategies to enhance the human brains response after injury can be a somatosensory stimulation, motor training, cortical stimulation and combination of sensory stimulation and motor training which is task specific [6,8].

It has been reported that repetitive activity alone is not enough to produce representational plasticity. Functional reorganization is found to be greater for task that are meaningful [9]. So it can be hypothesised that in acquired DDMS task specific exercises or purposeful task, Constraint induced movement therapy, Dual task training [10], along with repetition will useful in regaining function and enhanceing cortical reorganization. Additionally the somatosensory stimulation in the form of transcutaneous electrical stimulation, Cortical stimulation can be combined with above mentioned rehabilitation protocol to enhance functional recovery and cortical representation plasticity in DDMS. The study on 12 healthy subjects who performed hand, finger, wrist and elbow movements under controlled condition revealed a clear large scale somatot-ophy of the M1 with distinct subregion controlling the foot, arm, and tongue [11].

Statistically significant differences were found in geometric centres of gravity between elbow, wrist and fingers. So the above mentioned rehabilitation protocol can be incorporated in management of DDMS aimed to enhance functional recovery and brain plasticity. However the present report is focused to explore the rehabilitation strategies and not on treatment effects, the further experimental study is required to elaborate the effective rehabilitation protocol and its results to improve the functional activity and cortical reorganization in case of acquired DDMS.

# CONCLUSION

ADDMS is rarely encountered case in physiotherapy practices. The present case study states functional recovery after the physical therapy intervention. However present report cannot be generalised for this population because it is a single case report. Further study is required with this population to form and elaborate an effective treatment protocol.

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#### **Conflicts of interest: None**

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