CASE STUDY

An Integrated Multimodal Intervention of Remedial and Adaptive Approaches in Secondary Dystonia of Hand: A Case Report

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ABSTRACT

Purpose: *To evaluate an integrated approach of remedial and adaptive interventions that were employed to improve functional independence of a young woman with childhood-onset secondary dystonia of the right hand.*

Methods: *The intervention involved inhibiting dystonic movements, functional training with correct movement patterns, and activity modifications. The techniques combined multiple modalities (static and dynamic orthoses, electrical stimulation, purposeful activities, and adaptations), therapeutic exercises (stretching, inhibitory strategies and strengthening), sensorimotor training and motor learning strategies.*

Results: *After a year-long intervention of supervised therapy sessions and home programmes, there were improvements in dystonia severity (25%), hand functions (38%) and disability scores (30.3%).*

Conclusion: *The intervention outlined is a positive therapeutic approach to enhance function in secondary dystonia of the hand.*

Key words: *women with disabilities, movement disorders, rehabilitation, orthoses, electrical stimulation, activity adaptations*

INTRODUCTION

Women with disabilities are a marginalised section in India (Rao, 2005) and there is limited research pertaining to their functional rehabilitation. Dystonia is a neurological syndrome characterised by involuntary sustained contractions of the agonist and antagonist muscles, leading to twisting and repetitive movements

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or abnormal postures (Fahn, 1988). Based on etiology, dystonias are classified as primary or secondary. In the primary type, dystonia is the only feature and is usually of genetic etiology, e.g. focal hand dystonia seen in writer's cramp and musician's dystonia. Secondary or symptomatic dystonia arises as a consequence of an underlying neurological disease and has an identifiable acquired cause such as trauma, drugs, infections and ischaemia (Fahn, 2011). Dystonias are associated with significant disability, and treatment involves medications, surgical interventions and rehabilitation (Torres-Russotto and Perlmutter, 2008; Messina et al, 2012).

Recent research in rehabilitation of primary dystonia supports the use of remedial techniques involving sensorimotor training to restore normal brain organisation (Byl et al, 2009; Rosenkranz et al, 2009; Bleton et al, 2011). Therefore, it is possible that retraining the brain to restore sensorimotor organisation could also have therapeutic benefits in secondary dystonia. Rehabilitation of secondary dystonia has primarily involved the use of Lycra splints and orthoses (Mirlicourtois et al, 2009; Elliott et al, 2011). This case report aims to evaluate the effectiveness of a multimodal intervention of remedial and adaptive approaches to improve function in a young woman with secondary dystonia of the hand.

METHOD

DP was a 19-year-old Indian woman presenting to a tertiary care hospital with involuntary movements of the right hand ever since post-encephalitic right hemiplegia at 6 years of age. Up to the age of 12, she had seizures which were treated with oral medications. There were no other significant medical antecedents.

Following medical management for encephalitis, there was improvement in the right lower limb and independent ambulation was achieved. Recovery in the right upper limb was relatively slow. Gradually, abnormal involuntary movements of the right hand were observed which were diagnosed as post-encephalitic secondary dystonia. Pharmacological management was started with Benzhexol and Tetrabenazine, and she was referred for Occupational Therapy.

DP had begun to use her left hand for writing and feeding. However, she reported activity limitations and required assistance for tasks such as washing hair, dressing, grooming, menstrual hygiene, cooking, household chores, money management, and college and academic activities (carrying and manipulating books, etc). She was particularly concerned about her inability to accept *prasadam* (sacred food) in temples with her right hand, which has cultural and religious

significance in India. She reported low self-esteem and avoided social interactions. Her parents were concerned about her dependence in matters of self-care, as well as her ability to perform the roles of wife, mother and home-maker if she were to marry in the future.

Informed consent was obtained for publication of data relating to her assessment and intervention.

Assessment: It was observed that the right upper limb was atrophied. Dystonia was present at rest and while attempting any movement with the right upper limb. The dystonic movements primarily involved repetitive finger movements with wrist flexion and ulnar deviation, thumb adduction, elbow flexion and forearm pronation. The Flexor Carpi Ulnaris (FCU) was the strongest component of the dystonia. DP was unable to retain her grasp on any object and had incorrect movement patterns of the right upper limb. She did not use the affected limb spontaneously in movement transitions and daily activities.

On examination, reduced soft tissue extensibility of the long finger flexors, thumb adductors, FCU, Pronator Teres, elbow flexors and shoulder internal rotators was found. Manual muscle testing showed weakness of the right upper limb, particularly in elbow extensors and supinators (grade 3), wrist muscles and finger flexors (grade 2), and thumb and finger extensors (grade 1). There were no sensory and cognitive impairments.

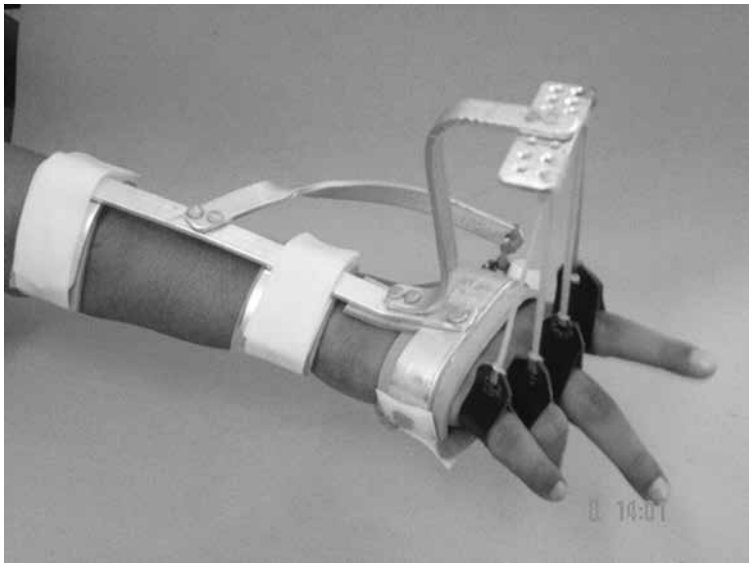
The primary outcome measures included the Burke-Fahn-Marsden (BFM) scale for dystonia and the Dystonia Disability Scale (DDS) (Burke et al, 1985), Jebson's Hand Function Test (JHFT) (Jebson et al, 1969) and the 30-item Disabilities of the Arm, Shoulder and Hand (DASH) symptom scale (Hudak et al, 1996). The writing subtest of JHFT was not administered as DP could not hold a pen. The maximum time for the JHFT was fixed as 120 seconds (Duncan et al, 1998). The time taken for the 6 subtests was summed up and the mean time for JHFT was computed. All assessments were repeated after one year.

Intervention: The intervention involved remedial and adaptive approaches with multiple modalities. The remedial approach involving inhibitory techniques and teaching correct patterns of movement in functional activities, aimed to decrease abnormal tone (dystonia) at neural and non-neural levels (Mayston, 2002; Lennon, 2003; Raine, 2007), to promote a balance between the agonist and antagonists, and facilitate normal muscle contractions. The inhibitory techniques are summarised in Table 1.

Table 1: Inhibitory techniques

Regain normal muscle length	• **Stretching and mobilisation techniques**: Active and passive stretch, massage. • **Orthotic devices:** A static thermoplastic orthosis with wrist in extension, neutral deviation and thumb in opposition to elongate the shortened FCU and promote functional position of wrist.
Inhibiting dystonic movements	• **Positioning** (including bed) antagonistic to the dystonic posturing, emphasising shoulder abduction with external rotation, elbow extension, supination and wrist extension. • **Movement combinations** deviating from the dystonic posturing to weaken the strong association of involved muscles, e.g. - carrying a tray bilaterally with arms straightened out in front (combine shoulder flexion of dystonia with elbow extension and supination) - carrying books or a tray bilaterally (elbow flexion of dystonia with supination) - playing on inclined chess board (pronation of dystonia with wrist extension using wrist orthosis). • **Weight bearing** on the right upper limb with active weight shifts in developmental positions (supine to side-lying/sitting, prone-on-elbows, extended elbows, and quadruped) that progressed to functional activities, e.g. weight bearing on right upper limb while wiping the floor in quadruped position, stabilising book/paper with right hand while writing with left hand. This also encouraged increased use of the affected limb. • **Relaxation exercises** • **Avoiding excessive voluntary effort** • **Incorporate these inhibitory techniques in daily activities**
Strengthening	• **Activities with graded resistance** with emphasis on the muscles antagonistic to those involved in the dystonia, i.e. shoulder extensors, abductors, external rotators, elbow extensors, supinators, wrist and finger extensors. • **Functional Electrical Stimulation** for weak or trace muscles, i.e. finger and thumb extensors followed by isometric contractions, active assisted movements and then progressed to active movements. A **dynamic finger extension outrigger orthosis** (Figure 1) was also provided to facilitate the finger extensors and encourage release.

Figure 1: Dynamic finger extension outrigger orthosis



A task-oriented approach was used in relearning correct patterns of muscle activation and movement (Poole, 1991). Initially, movements were passively guided to 'give the sensation' of the correct movements. Visual, proprioceptive, tactile and verbal feedback was provided to facilitate learning. Treatment then progressed to the use of purposeful activities with manual guidance and lastly, to active independent movements (Dutton, 1995). DP was also encouraged to use the right arm and hand in bilateral activities such as making the bed, folding clothes, light household chores, self-care, etc.

Adaptive approach: This involved the suggestion of techniques and simple cost-effective modifications (adaptations) to promote functional independence and encourage maximal use of the affected right hand, e.g., front opening brassiere, stick-on sanitary pads with modified donning techniques, simple dress-wear modifications (elastic waist-band for the skirts, pajamas, and salwar - loose pajama-like trousers; avoiding zippers at the back of upper body garments like the kurta - a long shirt or tunic), using a shoulder carry-bag for convenient handling of contents, using a brush with handles and soap-holder for washing clothes, use of shower for washing hair, towel with loops at the ends, etc.

DP resided in a remote place and could not attend regular therapy sessions. Hence the year-long intervention consisted of 45 supervised sessions, accompanied by home programmes. She would attend 8 to 10 successive supervised sessions (one

session per day of approximately 60 to 90 minutes duration) and then be provided with a structured home programme in writing, supplemented with diagrams, to facilitate adherence. The next round of supervised sessions would be scheduled 10 to 12 weeks later. Techniques of stretching, massage, manual guidance, and active assistive movements were demonstrated to the mother with hands-on practice. Table 2 provides an outline of the structured home programme which was required to be done at least twice a day.

Table 2: Outline of structured home programme

Treatment components	Examples of home programme activities
Regain normal muscle length	Stretching and massage done by mother, use of static orthosis for gradually increased durations. Later, progressed to self-stretching procedures. Use of static orthosis continued though duration decreased to maintain muscle length and encourage use of hand in function.
Inhibiting dystonic movements	Activities described in Table 1, done with assistance of mother. Later, progressed to incorporating inhibitory techniques in performance of functional tasks such as stabilising garment with right hand while ironing, stabilising objects (jars, bottles, vessels, books) with right hand, using right hand to bear weight in common movement transitions such as rising from a bed or chair.
Strengthening	Strengthening exercises of supervised sessions were initially done with mother's assistance. Later weighted sand bags tied distally to upper limb were used. Resistance was gradually increased. With improved gross grasp, strengthening done using functional activities such as lifting bottles with water /sand, bouncing ball against wall, sweeping with a broom, hitting ball with a bat for shoulder and elbow muscles, cooling tea, turning cards and pages for supination-pronation. For wrist and hand, activities included rolling chappattis, moulding clay/dough, striking marbles/coins initially with dynamic orthosis, and later without orthosis.
Correct movement patterns	Initially mother provided manual guidance and feedback in performance of selected functional activities such as eating biscuits/bananas, drinking water from a glass, picking/sorting onions, combing hair, etc. Later, progressed to performing in front of a mirror for visual feedback. Manual guidance was gradually decreased.

DP and her mother were encouraged to gradually generalise exercises and activities of the supervised therapy sessions and home programme to other activities of daily living. The compliance with the home programme was considered "good" as she reported for follow-up appointments, showed keen interest and participated in the intervention. Motivation and family support facilitated compliance.

RESULTS

The Pre-assessment and Post-assessment scores are summarised in Table 3 (BFM, DDS and DASH) and Table 4 (JHFT).

Table 3: Dystonia and Disability assessments

Assessments	Pre-assessment	Post-assessment	% change
Burke-Fahn-Marsden scale	16	12	25%
Dystonia Disability Scale	14	11	21.4%
Disabilities of the Arm, Shoulder and Hand scale	66	46	30.3%

Table 4: Jebson's Hand Function Test

Subtests	Pre-assessment (seconds)		Post-assessment (seconds)	
	Right	Left	Right	Left
Page turning	21	12	21	10
Lifting common small objects	59	12	56	13
Simulated feeding	120	15	55	14
Stacking checkers	120	4	57	6
Lifting large light objects	30	9	21	6
Lifting large heavy objects	22	9	18	7

At Pre-assessment, on the BFM scale for dystonia, she had dystonia of right hand at rest (provoking factor = 4) and no useful grasp (severity factor = 4). On the DDS, she had difficulty in writing (4 = unable to grasp and maintain hold on pen with right hand), feeding (4 = completely dependent), and hygiene and dressing

(3 = needs help with most activities). According to the DASH symptom scale, her disability was rated as considerable. She was unable to perform the JFFT subtests of simulated feeding and stacking checkers. The right hand was slow as compared to the left hand in all subtests. The mean time for JHFT subtests was 62 seconds.

At Post-assessment, the severity factor of the BFM scale had decreased (score = 3).On the DDS, she was able to eat food with the fingers of her right hand (score = 3) and required help in some activities of hygiene and dressing (score = 2 in each). As seen in Table 3, the overall scores of BFM, DDS and DASH had improved. On the JHFT, the mean time for subtests had decreased to 38 seconds, suggesting an improvement of 38%.

DISCUSSION

The Pre-assessment scores suggest dystonia of considerable severity, associated with significantly impaired hand functions and overall disability. Following the year-long rehabilitation, the subject's dystonic movements had not decreased, as she continued to have abnormal movements at rest. The severity of dystonia had decreased as seen in BFM scores. The decreased scores on the DDS and DASH appeared to indicate improved functional abilities. The 20-points decrease in the overall DASH scores exceeds the values for the minimal clinically important difference (change exceeding 15 points) and the minimal detectable change at 95% confidence interval (8 to 17 points) reported for the DASH scale (Beaton et al, 2001; Institute for Work & Health, n.d.). The JHFT scores, especially of stacking checkers and simulated feeding subtests, seemed to indicate improved hand functions. DP reported an overall improvement in her ability to perform daily activities of self-care (especially bathing, dressing and menstrual hygiene), household chores, and improved participation in college-related tasks. She was especially happy with the culturally acceptable ability to eat food using her fingers and accept *prasadam* (sacred food) in temples with her right hand. Thus, the intervention decreased her activity limitations and improved participation in meaningful tasks.

In dystonia, single-focus interventions have not been found to be effective (Zeuner et al, 2002, 2003, 2005) and a multifactorial approach is recommended (Byl et al, 2003).The intervention outlined in this case report is a comprehensive programme of remedial (therapeutic exercises, sensorimotor training, motor learning, task-oriented approaches) and adaptive approaches. Multiple modalities such as

activities, orthotic devices and functional electrical stimulation were also used. In India, access and affordability of long-term supervised therapy is difficult. The case report suggests that structured home programmes could have some potential to improve treatment outcomes in rehabilitation.

Limitations

Limitations of the case report include the lack of intermediate assessments and follow-up assessment after termination of therapy. Also, adherence to the home programme, frequency and intensity were not monitored objectively. The integrated multi-modal approach outlined in this case report could be a useful adjunct to pharmacological management of dystonia. However, since this is a single case report further research is required, involving larger study samples and experimental study designs, to determine the effectiveness of this approach.

Dystonia cases in this tertiary hospital are relatively uncommon. There were few more cases but due to socioeconomic and accessibility problems, most do not report for follow-up. DP's was one case with adequate compliance and follow-up that could be studied. Efforts are on to obtain more cases to enable statistical analysis and improve research evidence.

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