Addressing Psychosocial Factors in MND: A Conceptual Framework for Intervention

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I. INTRODUCTION

Neuromuscular Disorders constitute a major category of non-communicable disorders contributing to significant disability & family burden and pose a threat to quality of life of the affected individual and family. The efforts for a cure for these disorders are still progressing. Among the neuromuscular disorders, Motor Neuron Disease (MND), a degenerative disorder of unknown cause affecting motor neurons, is characterised by a combination of upper and lower motor neuron signs with progression. The disease usually presents in the 5th or 6th decades, and is characterised by a relentless progression to dependence and shortened life span. MND has an incidence of 3/100 000[1] and ranks as the third most common neurological degenerative disorder after Alzheimer's and Parkinson's diseases,[2] giving a prevalence of around 6 per 100 000. Males are usually affected more than females (ratio about 1 · 6:1). The epidemiological transition that is taking place in India [3] has brought the focus to the disease burden of non communicable diseases, of which Motor Neuron Disease (MND) constitutes a major category. Increased age of onset, low forced vital capacity, short time from first symptom to presentation, and bulbar onset are all adverse prognostic indicators.[4] Given the relentless development to paralysis and accompanying incapacity & dependence for activities of Daily living, combined with maintenance of good cognitive function, MND is a unique illness, quite different from other diagnoses with a guarded prognosis, such as cancer .[5]

II. MND AND PSYCHOSOCIAL FACTORS

The physical aspects of MND frequently receive the majority of attention, with psychosocial aspects accorded secondary importance. Helplessness and hopelessness, and the existential issues of purpose in life and value as a person, together with loss and grief, have been reported to be the key parts of the MND experience.[5] Although guidelines exist to direct physicians to attend to the physical care, there is a distinct lack of guidance to address the psychological state of the MND patient. Apart from rehabilitation focused on motor, sensory and cognitive deficits there is a need for psychosocial rehabilitation and effective utilization of community resources.[6,7] As the disease progresses, ALS patients require assistance with mobility, drinking & eating, dressing and nursing care. A family member who assumes the role of primary caregiver often provides this assistance.[8] In this context, the concept of burden may be relevant to MND, as in other chronic diseases.

As the patients require more care and professional support, experiences of services could be an important outcome measure for care.[9] MND being a progressive disorder with fatal outcome, the limited opportunity for active medical intervention might make the patient and the family feel abandoned by the professional team and this adds to the caregiving distress.

III. PSYCHOSOCIAL INTERVENTION IN MND

MND is devastating and it may seem hopeless because there is no cure. At the same time, but there is hope - because there are psychological, attitudinal and coping patterns that can lead to greater psychological well--being and a higher quality of life, within the constraints of the progressive illness.

Studies have emphasised the need for developing interventions that provide direct practical and psychosocial supports for motor neurone disease family caregivers[10] and psychosocial support has been recommended as the part of practice parameters.[11] It is suggested that further research into palliative care and symptom control is of primary importance to improve quality of life of patients and their families, since MND is likely to remain a progressive and fatal disease for several years, if not decades[12]. There is no simple solution or answer to the question of optimizing the quality of life of these persons. The key is to identify areas where increased understanding can lead to greater well-being and quality of life, including addressing and handling the physical challenges associated with the progression of the disease, psychological and social factors, the patient's environment and family relationships, the patient's spirit and the patient's experiences with the health-care system.

Personal factors such as demographic factors (gender, race, age, educational status, and socioeconomic status), emotional states (depression, stress, anxiety, and fear), and coping strategies and styles (problem-based coping and denial), personality, beliefs (includes self-efficacy, religious beliefs, and personal values), attitudes (of the patient) and "other" (perceived social support) have been reported to be important factors in psychosocial adjustment [13,14]and need to be addressed in the intervention.

Psychosocial understanding of MND as a chronic disorder, [15,16] taking into account the relationship between chronic illness and family life cycle provides a conceptual framework for the issues confronted by families with MND, using a family systems perspective. The experience of disease poses a number of psychosocial and emotional demands upon an individual and his/ her family. The very nature of the disease process becomes a determinant giving rise to specific psychosocial demands. These demands can be organized conceptually, within four elements that form meaningful categories. Psychosocial demands for a broad range of chronic illnesses can be categorized, and when applied to Motor neuron Disease, it provides us with a useful framework for understanding the psychosocial needs and stressors that the patients and families go through. This understanding can guide the therapist in assessment, formulation and subsequent intervention. These four elements are:

- *Onset :* Some illnesses has an acute onset, in which the full nature is revealed suddenly, such as Optic Atrophy, which can precipitate rapid blindness, whereas other conditions develop in a gradual way with increasing severity, such as Motor Neuron Disease (MND).

- *Course :* Course of an illness is concerned with how it changes over time. Some conditions are "constant," or non progressive, such as Cerebral Palsy, whereas others are of a progressive nature, such as MND. Some illnesses have a relapsing course that may or may not involve a deteriorating element, such as Multiple Sclerosis, others have an episodic character in which the frequency and intensity of the "attacks" are variable, such as the Epilepsy.

- *Outcome:* Illnesses vary in their outcomes and the morbidity. At one end of the spectrum are illnesses that have little effect on life span, while other disorders are progressive and fatal, such as MND. While the latter condition shortens life span progressively, others include the possibility of sudden death, for example, Cardiopathy.

- *Incapacitation:* Diseases differ in terms of the nature and degree of objective physical problems. These problems can result from and impairment of functioning and structure, with the degree being mild, moderate, or severe.

IV. PSYCHOSOCIAL TASKS

The developmental time phase of the illness is the second dimension of the model, which helps to create a psychosocial schema of motor Neuron Disease. The disease poses different challenges in each phase, and thus the process of coping alters in each of these. Each phase has its own distinctive developmental tasks that require equally distinctive and considerably different strengths, attitudes or modifications from the patient and his/her family. Any illness thus presents a family with different psychosocial tasks at different times and these tasks fall into distinct phases, which Rolland [17] has categorized as (1) the crisis phase, (2) the chronic phase, and (3) the terminal phase.

In MND, the crisis phase might involve handling the tasks involved in dealing with the diagnosis for the patient and the family. While the duration of symptoms and increasing functional loss might differ for individual patients, the communication of the 'bad news' poses a significant crisis in terms of psychosocial demands and will require those skills necessary to meet the tasks of crisis management. During the chronic phase, the reality of the disease and the functional limitations sinks in, and the patient and the family makes the journey towards accepting the disease as irrevocable. As the patient and his/her family makes structural and functional changes to adapt to the unrelenting progression and subsequent effects of the condition, the central task calls for negotiation about family roles. In the terminal phase, the end of life issues come to the forefront, and the emotional tasks related to the imminent death will predominate. Though the life threatening nature of MND is intellectually understood from the time of diagnosis, it is in terminal phase that the grief and bereavement issues become more prominent. Critical transition periods link the three time phases, and would require interventions addressing the specific issues.

The concept of anticipatory loss, as suggested by Rolland [18] does not just refer to the expected loss of the ill family member. It also refers to the losses of anticipated life-cycle stages and tasks that a chronic illness foils. Depending upon the life cycle stage and the family life cycle stage the patient with MND is in, the current derailment posed by the illness is compounded by the likelihood of further losses as the disease progresses. To place the unfolding nature of a chronic condition such as MND into a developmental context, it is important to understand the intertwining of three elements: the illness, and individual, and family life cycles[16]. The life cycle implies a basic succession and evolution of the life course, with its attendant strengths and challenges[19]. Family life cycle perspective implies that family life can be organized into psychologically functional phases that are passed through by individuals, phases in which the development and fulfillment of needs for one individual are meshed with the needs of other family members[20]. This becomes an important focus of understanding the meaning that the patient and family gives to the illness experience, and hence influences their coping (Figure 2).

V. THE CASE OF Ms R

Ms R, 50 year old married lady, home maker, educated up to post graduation, from urban upper middle class family with the diagnosis of Motor Neuron Disease with bulbar symptoms was referred for psychosocial intervention. During the time of referral she was hospitalized for detailed evaluation of complaints of increasing difficulty in speech and swallowing. Initial clinical assessments pointed to a diagnosis of ALS with bulbar onset [21]. During the hospitalization period, she was also undergoing assessment and intervention from different

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supportive disciplines that included Speech Pathology, Neuro Rehabilitation and Neuro Psychology. This multidisciplinary team approach has been recommended to improve the quality of life for patients, and their families, as well as survival rates [22].

Based on the assessment, goals for the intervention were set and daily sessions were planned for during the hospital stay, with the follow up planned on outpatient basis. This short term, intensive therapy was proposed with the goals of facilitating understanding about the condition and meeting the information needs of the family[10], facilitating acceptance and coping and aiding Ms R and her family to put in place a strategy for coping with the progressive nature of the illness[10, 23]. The supportive therapeutic stance by the therapist was based on the assessment that revealed a family with healthy coping strategies and problem solving mechanisms, which have been derailed by a sudden crisis [24].

Her family consisted of husband, elder son who works as software engineer in a leading IT company, one daughter who is a chartered accountant and a younger daughter who had recently completed post graduation in business administration. The husband was working in a private construction firm after retirement from the government service. The son and the younger daughter were the caregivers in the hospital. Ms R was a trained classical vocalist who used to take music lessons for children from her home and used practice every morning. The initial difficulty or change that she and the family members noticed was a change in the quality of her voice while singing devotional hymns which was her practice every day. When they noticed the voice change the family sought the help of the General Physician, the ENT surgeon and then the Dental surgeon, who suggested that they seek the opinion of a Neurologist. During this help seeking process, the voice change had progressed into significant difficulty to speak which became a major cause of frustration to the patient and distress to the family members.

The diagnosis, when it came, was a shock to the family[10]. The son and the younger daughter, who received the news initially, took some time to absorb it. Ms R’s husband continued to be optimistic and kept seeking clarification to rule out any other possible explanation for her difficulties. The crisis posed by the news brought about a significant amount of distress and frustration in the family. The intervention at this phase focused upon handling the emotional response to the diagnosis [23]. By adopting empathic listening and unconditional regard, the therapist relied upon supportive therapeutic techniques [24,25] to enhance the family’s coping and to equip the family with skills required in coping with the relentless progression of the disease.

At this point in the disease process, Ms R continued to function well in her role, except for her difficulty in speech and swallowing liquid food. The individual sessions with her focused on the helplessness that she felt in not being able to articulate and the consequences[5,10]. The family was aware that they were not handling the situation well, and that she has to be informed about the nature of the illness. They were also aware that as a family, they did not have any experience of dealing with chronic illness [26]. Handling the collusion became a focus of the intervention at this point and negotiating with the family to communicate about the illness with Ms R was done. The situation was complicated by the role of Ms R in her family. She was an accomplished homemaker who was the central figure and decision maker in the family. The cultural values and role allocation that gives the women the primary care taker role in the family was very much evident in the family’s functioning prior to the onset of the illness. Her incapacitation resulted in the family’s envisaged loss of emotional anchor and decision making at this point was a difficult process for the family without her involvement.

Ms R was going through significant confusion and personal crisis during this period. She kept asking her son to show the video of her singing to the therapist and kept expressing the hope that she will be able to sing like that again soon. She appeared to be grieving the loss of her voice and what it meant to her, which went beyond the mere use of voice, and started questioning the purpose of living itself[5]. She had also stopped going out for shopping and even for temple, for fear of others noticing her limitation. This further interrupted her everyday functioning and increased the social isolation [27]. Intervention with her at this point focused on addressing her distress and fear about her loss. By addressing the perceived social support and enhancement of internal locus of control,[28] over a number of sessions, she was able to face the likelihood of the changes in her being irreversible. The grief process for her was severe and was addressed in the sessions thoroughly. Ms R was able to derive some comfort from having someone with whom to share her feelings about her loss and her fears about the future.

The family also had to move forward in the process of acceptance about the nature of illness being incapacitating and life threatening. The younger daughter, who was on the verge of taking up a job, postponed the decision so that she can be at home to look after the mother. For Ms R’s husband, the elder daughter’s marriage became an all important goal to that he wanted to accomplish soon. Each member of the family was caught up in their own grief process and resolution that for some time, they lost some emotional connectedness with each other. Intervention at this point focused on normalization of family reactions[24] and facilitating their coping as a unit through ensuring support to each other.

Over the intensive supportive therapeutic sessions that spanned over one and a half months, the family came to better grips with the illness and the resulting incapacitation. Future continued to loom large in front of them as a concern, which they acknowledged that they were not ready to face at present. Given the severe and relentless progression of the disease, the family needed to be supported through the day to day crisis.

VI. CONCLUSION

Adjusting to a chronic illness like MND calls for a myriad of coping strategies from the patient and the family members. Psychosocial interventions has a major role to play in mediating patients' and their families' response to the devastating illness. Rather than being confined to a single point of time during the disease course, the psychosocial support seem to be required throughout the disease process. The framework discussed in the present article becomes a guideline that helps the therapist to facilitate the process of equipping the patient and family members with skills needed to find their way through the maze of everyday living with MND. While we have discussed the framework as applied in a single case, further studies are required in order to find out the efficacy of the model to bring about change and to reach a conclusion about the generalisability of the framework.

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