Motor neuron disease – a challenge to medical ethics: discussion paper

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Introduction

Motor neuron disease (MND) designates a progressive neurological disorder, appearing at the average age of 60 years, which culminates in death, usually within three years from diagnosis. With a creeping paralysis the body systematically deteriorates, including the legs, arms, and the ability to eat, swallow and breathe, until pneumonia or a choking seizure induces death. Throughout the course of the disease, mentation and all forms of sensation remain intact, even when all physical movement, including speech, has disappeared.

Once the diagnosis has been made, based on clinical history, examination and electromyography, and once the grave prognosis is realized by the attending physician – namely that the patient will observe, in one form or another, his or her own incremental demise – difficult questions arise concerning treatment. For example, how should the diagnosis and prognosis be presented to the patient, if at all, and what therefore should be done in the way of symptomatic management for the patient experiencing a 'demeaning illness?'[1] And can certain ethical principles be developed to apply in all cases? Or, must the doctor rely strictly on intuition and clinical judgments: 'For only by exposure to this poignant problem can one acquire the experience and skill to handle it, with medical professional skills on the one hand, and kindliness and human dignity on the other'[2].

This paper will argue that neither clinical judgment alone nor intuition can be considered as the sole criterion concerning treatment of the MND patient: explicit ethical judgments are equally and fundamentally important. Although the disease is of unknown aetiology, and is remorseless and inexorable in its progress unto eventual death, the doctor must still respond to the patient with a course of treatment, which involves both a clinical expertise and well developed ethical position. Such an ethical position must have considered both theoretical questions and their concrete applications.

This paper will consider such substantive questions as truth-telling – as it concerns whether and how the reality of the terminal condition should be conveyed to the patient, and also how such truth-telling relates to the common practice of emphasizing hope that research might discover a cure for the patient. In a related sense, too, it will consider whether the common practice of deception is ethically justified, or the use of placebos. Finally, it will explore what is meant by offering a 'rational treatment plan' in the way of symptomatic management of this difficult neurological disease[3]. Or, as two neurologists asked recently, 'Can we do better?'[4] If physicians ought to do better in management, then when does it go beyond the ordinary means of preserving life and into extraordinary applications, which may not be ethically, as well as medically, justified? All of these questions indicate how difficult clinical treatment is and why MND presents such a complex challenge to both medicine and medical ethics.

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In considering MND as a challenge to medical ethics, it should initially be stated that the attitude here is one of prudence. For prudence refers to the correct knowledge about things to be done. It is the intellectual virtue whereby a human being recognizes in any matter at hand what is good and what is evil. With neurological diseases, the need for prudence by the attending physician is particularly evident. Pellegrino and Thomasma[5] are correct when they write that the modern neurological situation in medicine has become such that new literature is so abundant on technical advances that greater need is evident for the clearer understanding of what correct knowledge is involved in making both a clinical and ethical judgment. Thus, it is difficult to insist upon a standard policy, except to reinforce the meaning of a prudent attitude as one concerned with that case which is 'at hand'; how the physician perceives the unmitigated truth of the MND diagnosis, the truth of the patient's condition, and relevant materials, given the particular circumstance; will be important. A prudent approach considers all pertinent materials.

Truth-telling

The physician's knowledge of the truth of the patient's MND diagnosis becomes information initially unknown to the patient. Whether it is to be told depends on how the physician understands the issue of respect for autonomy, and its applications to a particular patient. Three basic attitudes exist to truth-telling and the autonomy of the dying patient: (1) the physician has the obligation not to do harm: informing a patient as to a terminal condition could inflict harm; (2) physicians cannot be sure of the diagnosis and prognosis and to attempt clinical truth presentations might be misleading or misunderstood; and (3) the patient does not wish to be told[6].

It will be recalled that a prudential attitude forms the basis for ethical judgments. This means that none of the positions could be successfully argued as an ethical absolute, applicable in all cases, as shall be demonstrated. In the first case, informing a patient as to the diagnosis of MND may not be prudent. For example, if the patient should then enquire for the rest of the truth concerning the dying process, and were to learn of such expectations as probably choking to death or becoming completely paralysed, then the patient might live in mortal terror of any choking spell, however minor, or slowness of gait as signalling the onset of paralysis. If the patient should also live longer than the statistical mean of one to three years, it would also be the case that frank statements initially about the probable course of MND may be untrue in a particular case, and more harmful or even evil.

Some patients, however, may believe the truth to be important. Arrangements can be made. A will can be prepared or updated; certain devices installed in the home to assist the patient as his or her condition deteriorates. In a different way, psychological or spiritual preparations can be made. In some religious traditions, in fact, there is the obligation to inform a patient of impending death so that certain sacramental rites might be performed. If such a patient should be deceived, and should discover the diagnosis by some other means, then there is the understandable questioning of the physician's credibility in the eyes of the patient. For the patient and family adhering to a particular religious tradition, there is the serious matter of concern for body and soul – in life and death. To withhold the truth of finality is to violate the patient's faith.

Doctors must be careful, as the second point indicates, with being sure of their diagnosis and prognosis. As studies reveal, 20% of those diagnosed with MND will not die within three years. Indeed, it is the case that there might have been a misdiagnosis, or that the course of the disease does not heed established norms[7]. Such a possibility, that one-fifth of patients will survive longer than expected, should remind the physician that in truth-telling the clinical judgment may not be altogether final. In another way, informing the patient as to a considered truth involves the problems with what is to be conveyed, and how. Medical facts may not be comprehensible, whether for reasons of intellect or, for that matter, psychological denial in operation: despite the reality of the patient being considered autonomous and competent. To tell the truth may be quite different from discovering what is actually understood. So what responsibility does the doctor have?

Saunders[8] states that 'Every patient needs an explanation of his illness that will be understandable and convincing to him if he is to cooperate in his treatment or be relieved of the burden of unknown fears'. Her sense of explanation, however, suggests that 'There are many different truths'. She believes the physician must produce the type of truth needed at a given stage. Saunders' approach, however, might well produce but janus-faced veracity, for lack of a better concept, unless prudence is exerted. In another way, if the particular type of truth told is not grasped by the patient, then it might be suggested that what the physician has offered may not be as 'truthful' as the physician might think it is.

Gert and Culver[9] would no doubt also criticize this use of truth and its paternalistic problems. Certainly they accept the fact that paternalism can be used ethically, but only when the patient is not in a position to respond competently, such as when unconscious. Thus it is important to understand that when truth is at stake, it is one thing to promulgate a clinical judgment and run the risk of being misunderstood with the facts by a competent patient; and quite another matter to try to camouflage the truth or fail to respect the patient as a moral agent[10].

Saunders' 'different truths' might be said to present prudent conveyance of information. Yet the problem with such prudence is that technique becomes operative over truth. Conveyed information may be misunderstood; also, the presentation may not strike the patient as credible or authentic. If such credibility is lost, then the physician may find more explanations demanded of other matters pertaining to treatment, and the burden of fears centring on the doctor's credibility instead of the disease.

The third point concerns the fact that some patients do not wish to be told of their terminal disease. One writer has actually suggested from his review of some of the literature that 'Although these surveys are now distinctly elderly they at least cast substantial *doubt* upon the claim that most patients do not wish to know the truth'[6]. Such a conclusion is unjustified. For example, two of the quoted sources indicate the opposite. In the 1976 *Lancet* paper[11], only a small minority of patients with undisclosed diagnosis of cancer desired direct information. Their decisions were intentional; concomitant anxiety chosen. In another one of the five papers cited, it is true that two-thirds of the patients were glad to know the diagnosis; but, as the authors indicated[12], these were people with curable cancer. It would be reasonable to assume that people would welcome good news or happiness. Learning that a state of good health might be restored would certainly be the cause of encouraging such disclosure. In the case of an untreatable disease, such as MND, refraining from truth-telling might be justified in order to spare the patient undue harm and to prevent despair – even though studies indicate that most MND patients possess 'an extremely high internal locus of control'[13].

Conspiracy of silence

If truth-telling is not directly sanctioned for prudential reasons, then what course of action should be taken and for what ethical reasons? The most obvious would be a conspiracy of silence[14]. To be effective, it must be complete. From the day of diagnosis to the moment of death, the patient is never made aware of the terminal eventuality. This approach probably comes the closest to fulfilling the means of avoiding truth-telling: a conspiracy of silence causes no harm by disturbing the patient with the awareness of death; if the diagnosis and prognosis should be in error, then the patient is not made to endure the possible emotional hardship and other setbacks occasioned by faulty evaluation or change in health for the good; and in a conspiracy, the physician and patient are not involved in the dilemmas about whether the truth should be told at all because the patient may not wish to be informed.

Conspiracy of speech

It would seem more likely that the patient will be aware to a certain extent of a problem, and the conspiracy of silence may not be as pure or simple in reality. If the conspiracy of silence should not be considered appropriate, then what ought to be said? Let us consider in detail the following plan of treatment suggested by Walton[15]:

> There is no doubt that a responsible relative should be told the truth, even if one stresses the variability of the clinical course of the condition, emphasizing that some cases are more benign. It has been my custom to tell the affected individual first that the condition is one which is well-recognized, if of unknown cause, and to explain something of research now in progress. In order not to destroy all hope, I believe that it is best to say also that the condition progresses slowly up to a point but then usually becomes arrested, and may even subsequently improve spontaneously, while making it clear that no-one can predict when and if arrest will occur. Comparatively few patients seem to be aware of the deception, even to the end.

This clinical stance indicates a basic deception all around: a responsible relative is to be informed of 'the truth', but not so much as to stress the dark reality of the probable outcome. The patient is then made aware of a 'condition' - not a disease - and this is explained in the context of research being conducted so as 'not to destroy all hope', but in reality only as a deceptive means to conceal the real truth. The neurologist also speaks of a usual arrest, but one that never occurs quite as the patient might be led to believe - an admitted deception.

The neurologist would, no doubt, defend these deceptions as intended to be benevolent. But where physicians demand veracity from their patients in the name of helping them, then how can such sustained deceptions be justified?[16] The neurologist might also argue that it is his duty to treat both patient and relative as he sees fit - a paternalistic argument. He might also say that such clinical decisions were judged to be in their best interests. The stark truth of MND must remain within his clinical domain.

Hope

The pivotal idea expressed, and one that is ultimately meant as deception, concerns hope: to the relative, that MND is not necessarily fatal; to the patient, that research may produce a cure. Other than the percentage of patients who will not die within the first three years, the statistical evidence is such that death will result. What would obviously prevent death from MND would be a cure stemming from medical research. But it is clear that nothing even remotely possible has been forthcoming and, even if this were the case, an incipient solution would not be perfected to save the lives, given the reality of the disease, of those presently afflicted. The type of hope intended then might be regarded more as a lie than truthful divulgence, a conspiracy of speech.

What should be made of hope where MND is involved? Walton[15] believes it is important not to destroy all hope; in doing so, however, the result is a conspiracy of speech. The patient is indeed told something about the disease, and the need for hope, as Saunders would no doubt confirm as being a 'different truth'[8], but what is directly intended is a deception: the patient is led to believe recovery will occur. Through this deliberate deception both patient and physician are spared from confronting reality in the name of hope.

The French phenomenologist Gabriel Marcel is quite correct when he states that 'Any physical theory of hope is absurd'[17]. In other words, to speak of hope is to address the transcendent dimensions of humanity. It is not to dictate the flesh and other forms of physical matter. The physician must especially refrain from engaging in transcendental meditations at the expense of medical reality.

Campbell[18], another neurologist, has written concerning MND that:

> It is our policy to encourage optimism and, later, simple resignation. We do not stress the inevitably fatal outcome but quote the marked variability in the time course of progressive disability and encourage the patient to remain active for as long as possible with our help.

Like Walton, Campbell stresses the variability of the disease. He also speaks of hope in the form of encouraging optimism but then, unlike Walton, 'encourages' simple resignation; in this case, assisting the MND patient to come to terms with the disease and the fact of finality. Such stoic resignation is still not enough. For resignation speaks of acquiescence; and never is despair admitted. Despair marks the emotional element so important in both hope and resignation.

Marcel identifies this despair when he writes of the need for 'inward consolation' when confronting struggles. As he also says about the transcendent dimension within the person confronting a struggle: 'I shall rise infinitely above this *fatum* to which I have never allowed myself to shut my eyes'[17]. With the assistance of the physician, the patient is able to face MND with both ears and eyes open - and still have hope.

Rabin, an American endocrinologist diagnosed in the 1980s as having MND, and who progressed over the course of three years to an almost totally paralysed state, found that hope played upon the intellect. As an endocrinologist, fully aware of the likelihood of death as well as the usual devastating problems along the way, Rabin[19] believed that even the informed mind could live with the disease. Another patient found hope in God important[20]. Still another expressed hope that doctors would become more conversant in the MND literature - and willing to discuss the material with patients without embarrassment and a need for deceptive practices[21].

For all of these reasons, one must challenge the statement by Matthews and Miller[22] that 'It is usual to try to maintain morale by the use of a placebo'. Morale, in this reading, would be directly linked to hope: that by dutifully swallowing certain pills, assumed to be of benefit for a diseased state, some good would result. The patient would hope for health. As we have seen, hope stems from something quite different.

It is a documented reality that the suicidal rate for those aware of their MND, including a general idea of what might be expected, is remarkably low. Considering the distinctly foreboding expected progressive symptoms, this lowness might seem surprising. But studies indicate that the patient's will to live, combined with the support of family, exerts a decisive influence[7]. In other words, even in the knowledge of what might be expected, the MND patient endures - despite the usual outcome of a restricted environment as leading to depression and contemplated suicide[23].

The point is that deception need not be used in order to foster hope. Hope assumes different forms in MND, depending on the concerns of the patient. Deception seems to serve no purpose, for those aware of MND function as best they can in mind and body, just as those unaware will inevitably express some hope for something in their lives. Offering a placebo does not accomplish anything more than serving as a fallacious 'ought': that something ought to be done, if just for the sake of doing something.

Prudent judgment, then, must take account of ethical principles and also empirical studies, including statistics and the thoughts of patients who seek to engage in their own truth-telling about the disease. By and large, what they express is the hope that more truth-telling would be forthcoming; for what might be considered worse than a deception is the failure to be honest when confronted with explicit questions, as one patient expressed in his frustrating experience with the physician who shied away from his wondering if an episode of impotence could be associated with his MND[21].

Rational treatment plan

In no other aspect do all the discussed ethical problems become more highlighted than in the question of what type of 'rational treatment plan' should be advocated in response to an increasing concern for what ought or could be done to assist MND patients[3]. What prudent type of symptomatic management emerges when such themes as autonomy, truth-telling, and the general desire to be non-maleficent are recognized? With no definitive treatment acknowledged, and no certainties as to the particular course of each case, the resultant problems are both theoretical and concrete.

In 1975 Smith and Norris[3] published an important paper arguing that symptomatic therapy could be based on rational principles. Most therapy could be limited to simple and low-cost procedures, which the authors felt was correct. Where weakness was found, an appropriate aid, such as a brace or splint, would suffice. Cramps could be controlled with appropriate medications. Dysphagia, impairment of the ability to swallow, could be controlled at first by a careful monitoring of diet, perhaps followed by medication or the necessity of a nasogastric tube, and maybe surgery. Sialorrhea, drooling, normally responds to medication or to surgery. Aspiration may require surgery. Aphonia, the loss of the power of speech, presents an overwhelming emotional burden. Yet surgery cannot really correct the problem. Instead, an amplifier of some sort might assist. All of these treatments Smith and Norris would consider as ordinary means of preserving life. For they will show a reasonable hope of benefit and do not involve excessive expense, pain, or inconvenience.

Attending to such problems will relieve the patient of manageable problems. In another way, respiratory management would be considered as prudent if the following principles[24] were followed:

(1) Respiratory obstruction (e.g. obstruction secondary to the tongue falling back) should be treated.
(2) A patient should not be allowed to 'drown' in his own secretions, but should receive adequate suctioning.
(3) Hypoxia (e.g. that secondary to pneumonia) should be treated if there is adequate accessory muscle strength.

In each case, ordinary means are used to keep the patient alive. The prudent physician is not causing harm in responding to these conditions, nor is there any real doubt about the benefits to the patient. For the patient aware of MND, informed consent is really not an issue at this point: prolonging the life produces benefits and not undue burdens.

In most cases, however, there will come the time when death seems imminent. Given the nature of MND, the patient may be horribly incapacitated and suffering terrible mental anguish. In such situations, prudence would dictate that it would be fruitless to prolong such a life. Pneumonia being the usual cause of death, such last moments can be eased by the introduction of a generous narcotic medication rather than antibiotics, so that the patient is freed from pain and harm. Such introductions may still require the permission of the competent patient, if the autonomy is to be respected, or at least an earlier approval of this course of treatment. If this should not be the case, and explicit approval cannot be given due to incompetence or unconsciousness, then the doctor can still justify such a paternal act in terms of its being perceived as the good medical practice in the imminent terminal case.

The concern for symptomatic management is an increasing consideration for medical ethics. Medicine and surgery can respond to certain presenting complaints associated with MND. Medically, certain drugs may be given but, quite frankly, they do not contribute to the overall problem. Surgically, certain procedures can be performed, but are of limited value. The financial cost may not justify their use. In a much more serious line of thought, the ordinary resources for private hospital care will probably not cover accumulated expenses; for that matter, few private nursing facilities can provide for advanced MND patients without formidable costs, including equipment and the need for available staff. For these reasons, treatment must be limited to ordinary care.

Conclusion

When the neurologists Newrick and Langton-Hewer ask 'Can we do better?', as they did in the *British Medical Journal* in 1984[4], the answer should indeed be yes, in terms of their suggestions that the neurological community should attend closer to the ordinary means of treatment. The prudent physician would do no less than to provide adequately for a patient in reasonable need. Assessing those needs involves both clinical and ethical skills.

To do any less would be to warrant confirmation of the statement once made by an MND patient after diagnosis when he felt his physician provided neither good clinical nor ethical judgment in his treatment: 'He is no more use to me than the milkman'[1]. The physician who fails to consider the theoretical and concrete issues involved in the prudent treatment of MND, or who neglects to consider an honest type of hope, may find him or herself existing in a professional paralysis, promoting a deception or conspiracy in medical practice where neither the patient nor the physician benefits and medical and ethical truth go out the window. In this regard, at least the milkman delivers promised goods upon the doorstep faithfully.

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