Right Brain: The fragile sense of self in neurodegenerative disease

A brief philosophical examination

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Progressive neurodegenerative diseases such as Alzheimer disease, Parkinson disease, and Huntington disease are all associated with cognitive decline that may eventually result in advanced dementia and loss of the many elements that contribute to consciousness and self-awareness. This is a confronting prospect for many people living with these diseases, their families, and society as a whole.

The concepts of self-awareness and consciousness are notoriously elusive brain functions to define. They are not only practical neurologic descriptors but also terms that are widely used in the literature, philosophy, and psychology. Consciousness may be practically defined by the physician as "the state of awareness of self and environment, and responsiveness to external stimulation and inner need."¹ This requires the normal functioning of several facets of the nervous system for the integration and association of multiple sensory, motor, and emotional inputs into a single unified percept.² Philosophical perspectives can inform the discussion of self and self-awareness in neurodegenerative disease and may contribute to the foundation of a more positive approach.

Neurodegenerative diseases are characterized by the insidious, rather than sudden, loss of mental faculties. This is a challenge for patients, families, and professionals as there is no definable moment in which a person loses their self-awareness, agency, and purpose, but this is, rather, a gradual process. Alzheimer disease is characterized by the early emergence of a significant and progressive episodic memory deficit. This may be associated with other cognitive deficits including neuropsychiatric changes, executive and language dysfunctions, and visual processing impairments.³ Dementia is also common in Parkinson disease and may be termed Parkinson disease dementia (PDD) when it is preceded by the clinical motor features of Parkinson disease by at least 1 year or dementia with Lewy bodies (DLB) when dementia occurs simultaneously or within 1 year of motor parkinsonism.³ Other core features of DLB include well-formed visual hallucinations, cognitive fluctuations,

and sleep disturbance. Finally, Huntington disease typically presents with the combination of a movement disorder and a frontal dementia characterized by a range of psychiatric and cognitive symptoms. Individuals with Huntington disease develop prominent cognitive deficits on tests of attention, semantic verbal fluency, processing speed, and executive function.³ All of these diseases are associated with varying degrees of anosagnosia. This further compromises self-awareness by creating a disconnect between the internal and external projections of self.

While dementia represents the gradual erosion of cognitive capacity and self-awareness, the state of delirium illustrates that this can also be a dynamic construct. Those with neurodegenerative disease are at higher risk of developing delirium, which can simply be defined as an acute confusional state typically affecting older people with an acute medical illness.³ One criterion for a diagnosis of delirium is a fluctuating course, and therefore, individuals' degree of self-awareness and cognitive capacity may vary greatly within the course of a single day. Notably, cognitive fluctuations are also typical of those with DLB.

Much of Western philosophy is premised on the intact mind. For example, Descartes famously concluded, "I think, therefore I am" and conceived a sharp separation between the mind and body, termed Cartesian dualism.⁴ By extension, those individuals with advanced dementia who are no longer capable of rational thought could be considered not to exist. Cartesian dualism implies that the mind can outlive the body, but in dementia, many would say that the converse is true. Certainly, caregivers of those with dementia report that the loss of "personhood" and "social death" predate biological death.⁵

John Locke, the philosopher, conceived a thought experiment in his discussion of personal identity:

For should the soul of a prince, carrying with it the consciousness of the prince's past life, enter and inform the body of a cobbler, as soon as deserted by his own soul, every one sees he would be the same person with the prince, accountable only for the prince's actions: but who would say it was the same man?⁶

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In this scenario, it would not be fair to hold the man with the body of the prince responsible for the prince's former actions as he would have no recollection of performing them.⁴ This is an interesting notion in relation to dementia. It is perhaps fortunate that long-term autobiographical memory is usually lost late in the course of the disease as this appears integral to individual identity. However, in advanced dementia, when, for instance, people are no longer able to recognize even their siblings, it may be fair to say that they are only faint facsimiles of the individuals they used to be.

Kant conceptualized individuals as rational beings who are capable of leading their own lives and therefore we should accept their autonomy.⁴ This relates to one of the commonest ethical dilemmas facing doctors treating those with cognitive decline. It can be difficult to define the point at which it becomes morally acceptable to deny individual autonomy to people who have lost the capacity for rational thought. For Kant, persons are rational beings, and therefore, those who have lost the capacity for rational thought are no longer persons. However, this has been challenged by Kitwood and others. Kitwood's notion of person-centered care for dementia, emphasizing the essential humanity of the person rather than the disease and its deficits, is crucial to the maintenance of dignity and "personhood."⁷

While there have been developments in our understanding of dementia and resources are directed toward the development of potential treatments, most types of dementia remain incurable. The prospect of losing one's mental faculties and the inevitable progressive decline culminating in death is an understandably challenging one for most people. With advances in other areas of medicine and increasing life expectancies, particularly in the Western world, these issues will become more prominent.

Aldous Huxley provides a particularly graphic solution for those confronted by the idea of aging and dying in his dystopian novel *Brave New World*. At the "Park Lane Hospital for the Dying,"⁷ middle-aged people accelerate through their later years and into a state of second infancy soothed by the ubiquitous psychotropic drug *soma*.⁸ While this is an extreme fictional account of a possible future, there are nevertheless uncomfortable comparisons to be drawn with some modern nursing homes in which those with neurodegenerative diseases are kept safely away from the majority of society and pacified with multiple psychotropic medications. It is perhaps no surprise that people kept in these artificial environments and denied their autonomy can become depressed, angry, or even violent and be transferred back to acute hospital environments that are, in some ways, even more dehumanizing.

Two ideas that may be comforting in the face of such devastating diseases and the social isolation

they may portend are derived from Buddhism and the work of the Austrian psychiatrist Victor Frankl. Contrary to much of Western philosophical thought, the Buddha taught that "you" are not an integral autonomous entity.⁹ If we can see through the delusion of attachment to an individual self, we experience that which is not subject to birth and death. A logical extension of this idea is that we are part of a broader society. Working to integrate those with neurodegenerative disease into the society rather than ostracising and isolating them in institutions may have practical advantages for these individuals. This relates both to those still living in the community with varying degrees of independence and to those who have been institutionalized. In addition, for the broader society, exposure to people with such conditions may go some way to ameliorating the fear of the unknown.

Victor Frankl's experiences as a Jew incarcerated in a concentration camp during the second world war inspired him to write "Man's Search for Meaning" and develop logotherapy. Frankl determined that in order to survive, one must have optimism in the face of tragedy and in view of human potential.¹⁰ He conceived that the search for meaning should be the primary motivation in life and that there is an inherent but largely constructive tension between "...what one has already achieved and what one still ought to accomplish...."¹⁰ This philosophy can also be applied to human suffering, with Frankl writing:

...[W]e may also find meaning in life even when confronted with a hopeless situation, when facing a fate that cannot be changed. For what then matters is to bear witness to the uniquely human potential at its best, which is to transform a personal tragedy into a triumph, to turn one's predicament into a human achievement.¹⁰

This concept is readily translatable to those living with neurodegenerative disease. Finding meaning in life and ensuring that people still engage in meaningful activities even at a stage at which they are profoundly disabled can only be a positive endeavor.

The prospect of losing oneself with the inevitable progression of a neurodegenerative disease associated with cognitive decline is a daunting one, and there are a wealth of philosophical ideas that relate to this predicament. While some reinforce the notion of "social death" and loss of "personhood," hope can also be derived from the Buddhist notion of letting go of the "self" and enabling these individuals to be better integrated into society. Lastly, finding meaning in life allows those with neurodegenerative disease to remain positive and correspondingly bolsters their resilience.

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