Commentary

Advance Care Planning in Huntington’s Disease

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Pre-press 23 March 2023

Abstract. Advance care planning (ACP) is a useful tool that benefits adult patients, care providers, and surrogate decision makers, through providing opportunities for patients to consider, express, and formalize their beliefs, preferences, and wishes pertaining to decisions regarding future medical care at a time when they retain decision-making capacity. Early and timely consideration of ACP discussions is paramount in Huntington’s disease (HD) given the potential challenges in ascertaining decision-making capacity in the advanced stages of the disease. ACP helps to empower and extend patient autonomy, providing clinicians and surrogate decision makers with reassurance that management is consistent with a patient’s expressed wishes. Regular follow up is vital to establish consistency of decisions and wishes. We outline the framework of the dedicated ACP clinic integrated within our HD service to highlight the importance of a patient-centred and tailored care plan that fulfils the patient’s expressed goals, preferences, and values.

Keywords: Advance care planning, Huntington’s disease

BACKGROUND

Huntington’s disease (HD) is an autosomal dominant neurodegenerative condition with a chronic progressive course characterized by the combination of cognitive, motor, and neuropsychiatric disturbance. Although there are several potential disease-modifying treatments currently in development, to date there is no approved disease-modifying treatment for patients with this disease [1]. HD has an ultimately fatal trajectory, and the condition is devastating to patients and their families. Supportive and symptomatic management remains the mainstay of treatment.

The focus of this commentary is to emphasize the guiding principles and importance of advance care planning (ACP) for a patient with HD, their family, and the multidisciplinary team (MDT) involved in provision of their care. We will also outline the model and structure of the dedicated ACP clinic integrated within our HD service.

A multidisciplinary Delphi panel of international ACP experts established a consensus definition of ACP as “...a process that supports adults at any age or stage of health in understanding and sharing their personal values, life goals, and preferences regarding future medical care [2].” The consensus definition also incorporates that the goal of ACP is “...to help ensure that people receive medical care that is consis-
tent with their values, goals and preferences during serious and chronic illness. [2]” This definition provides a uniform framework in which to deliver ACP clinical interventions [2]. Although this definition of ACP is specific for adults [2], ACP nonetheless needs to be considered, individualized, and tailored to all patients, irrespective of age. ACP is voluntary and depicts an ongoing process of discussions on a continuum over time, fostering opportunities for an adult, who retains decision-making capacity, to contemplate and express their individual concerns, goals, preferences, values and wishes regarding future medical care [3].

ADVANCE CARE PLANNING IN HUNTINGTON’S DISEASE

Patients with HD usually have many years to engage in ACP given that the course of the disease may often span up to two decades from diagnosis to death. Early and timely consideration of ACP discussions is paramount in HD given the potential challenges in ascertaining decision-making capacity in the advanced stages of the disease. ACP helps to empower and extend patient autonomy and provides clinicians and surrogate decision makers with reassurance that management is consistent with the individual’s expressed values and wishes. Documentation of ACP discussions and completion of relevant legal documents, for example, advance decisions and lasting power of attorney (LPA), is essential in order to ensure care and treatment plans align with the patient’s expressed preferences.

A three-stage qualitative study produced and piloted a care pathway for advance decisions and power of attorney using HD as a model [4]. Five major themes emerged during the modelling phase of this study, including information deliberated and method of delivery, individuals involved and location (e.g., clinical setting, home), duration of process and timing, assessment of capacity and form of documentation [4]. Rapport with an expert in HD to facilitate the overall process was highlighted as a dominant theme [4]. Although professionals were reportedly reluctant to approach asymptomatic service users too early due to concern of causing distress, services users expressed positivity regarding early introduction of advance decisions to promote individual autonomy; and thus earlier routine introduction was implemented in development of the second care pathway following the first pilot [4]. The first stage of the care pathway incorporates the introduction of end-of-life issues to patients in the clinic; the second stage outlines the process of education and decision-specific capacity assessment; and the third stage depicts the process after completion of an advance decision and the follow up review of the advance decision [4]. Resource implications need to be considered in service integration, particularly relating to administration and time, given that the duration of sessions in education, capacity assessment and advance decision completion took an hour each on average [4].

Initiating ACP discussions with individuals with HD can be facilitated using the Huntington Disease Quality of Life End of Life (HDQLIFE EOL) Planning measure [5]. This is a 16-item patient-reported assessment tool that has been developed to evaluate end-of-life (EOL) preferences for individuals with HD [5]. This HD-specific EOL measure explores domains and preferences relevant to ACP discussions, including: advance directive, health care power of attorney, nursing home care, location of death preference, conversations about death and dying, living will, life insurance, palliative care, child care planning, finances, estate planning, support to make decisions, hospice care, resuscitation preference, funeral arrangements and preference about death [5]. Whilst reliability and validity data are preliminary [5], the HDQLIFE EOL Planning measure offers potential for meaningful and productive conversations relating to ACP.

There are no standardized guidelines about how or when to broach the topic of ACP with patients with HD. Ultimately readiness to engage in ACP discussions is context-dependent and patient-specific and needs to be tailored according to whether the patient is ready and willing to discuss future decision making. A qualitative research study aimed to investigate the presence of thoughts or wishes surrounding EOL in patients with HD or identified gene carriers visiting the outpatient clinic via a questionnaire [6]. There was a response rate of 55.4% out of 242 questionnaires sent, with non-responders younger in age and of lower education compared to the demographics of responders [6]. There was no significant differences between sex. Unified Huntington’s Disease Rating Scale (UHDRS) Total Functional Capacity (UHDRS-TFC) or motor (UHDRS-M) scores between responders and non-responders [6]. Of note, 77.2% of responders discussed their wishes with family members yet only 42.6% of responders discussed their wishes with a healthcare professional, with not being ready for such discussions contributing to part
of the reasoning [6]. In addition, familiarity with HD in the family was significantly correlated to the presence of any thoughts about EOL in respondents in this study [6]. This influence may be reflective of specific characteristics in HD patients who are familiar with the disease and trajectory through first-hand experience of witnessing the disease course in family members across generations.

The issue of decision-making capacity is substantial in ACP discussions as cognition declines during the late stages of HD. Thus, we should encourage patients to discuss their wishes regarding future care and medical interventions during a point in time when decision-making capacity is retained. An advance statement and an advance decision to refuse treatment (ADRT) can be completed during the ACP process. This underpins the fundamental ethical principle of respect for autonomy. A retrospective chart review of advance directive documentation in a HD clinic demonstrated a completion rate of only 24.2%, with patients with moderate and late-stage HD more likely to have documented advance directives [7]. Similarly, a larger cross-sectional study of patients with prodromal or manifest HD demonstrated that 15.3% of patients across all stages had not thought about getting an advance directive whereas only 38.2% of participants across all stages had advance directives [8]. Although these findings should be generalized with caution given the majority of participants in both studies were non-Hispanic [7, 8], the findings nonetheless highlight that ACP in this cohort is underutilized in clinical practice. Potential barriers to ACP discussions include lack of awareness and knowledge about ACP, uncertainty regarding the role of initiating ACP discussion and perceived concerns about jeopardizing rapport with the patient [9]. Addressing these potential barriers, we outline the model and structure of the dedicated ACP clinic that has been integrated within our HD service.

FRAMEWORK OF ACP CLINIC INTEGRATED WITHIN THE HD SERVICE

A dedicated ACP clinic has been integrated within our multidisciplinary HD service since 2015 with development of an ACP booklet for HD titled ‘Preparing for the Future’ [10]. The clinic is led by a Clinical Nurse Specialist (CNS) with input sought from physicians within the specialties of neurology and palliative are as needed. The framework of the dedicated ACP clinic integrated within our HD service is illustrated in Fig. 1.

Readiness to engage in ACP discussions is context-dependent and patient-specific. A patient is referred to the dedicated ACP clinic following review in the departmental HD MDT clinic when a patient with decision-making capacity feels ready and is willing to discuss future decision making. The initial appointment is given a one-hour slot, which offers protected time to introduce the concept of ACP and to promote self-reflection and initial communication of goals and values. Understanding goals and values at this stage can translate into more specific discussions relating to the patient’s care and treatment preferences at a later stage. The initial appointment may also include a discussion of prognosis, if appropriate, which again is tailored to the patient’s readiness to hear prognostic information.

Relatives and surrogate decision makers are encouraged to be present as this can help to prepare and support decision making, particularly at a potential time in the event of a patient’s incapacity. Care needs become complex in the advanced stages of HD with deterioration in cognition, subsequent loss of decision-making capacity, inability to communicate needs, swallowing difficulties and impaired mobility. The foundation of the clinic is to support patients and families to plan and prepare for the future. The clinic supports families in managing expectations and helps to minimize the burden of care and experience of guilt often prevalent in relatives caring for an individual with HD.

Formalized outcomes of ACP within the National Health Service (NHS) England framework include advance statement(s), ADRT and LPA [11].

An advance statement is an expression of the individual’s preferences and wishes towards the EOL or when the individual becomes unable to make decisions. An advance statement is not legally binding; however, this helps to inform best interest decisions in the future. The discussions are documented and regularly reviewed as the individual’s views may change over time. Advance statements may include: religious or spiritual beliefs, preferred place of care, thoughts about treatments or types of care the individual may be offered (e.g., hospital admission, major surgery), a person the individual wishes to be consulted on their behalf at a later time, personal preferences (e.g., likes and dislikes), organ donation and funeral preferences.

An ADRT only covers the refusal of a specific future treatment. If the treatment to be refused could be considered life-sustaining, the ADRT document
must be written, signed, and witnessed. ADRT will only come into effect if the individual loses the capacity to make decisions. This is legally binding if valid and applicable. Examples of specific issues that individuals with HD may choose to refuse through an ADRT include: clinically assisted nutrition and hydration (e.g., percutaneous endoscopic gastrostomy feeding, radiologically inserted gastrostomy feeding, nasogastric tube feeding, intravenous feeding, intravenous or subcutaneous infusion), antibiotic

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**Fig. 1.** The framework of the Advance Care Planning (ACP) clinic integrated within our Huntington’s disease (HD) service. ACP, advance care planning; ADRT, advance decision to refuse treatment; CADs, court appointed deputies; CPR, cardiopulmonary resuscitation; IMCAs, independent mental capacity advocates; IV, intravenous; LPA, lasting power of attorney; MDT, multidisciplinary team; NGT, nasogastric tube; PEG, percutaneous endoscopic gastrostomy; RIG, radiologically inserted gastrostomy; SC, subcutaneous.
treatment, artificial ventilation and cardiopulmonary resuscitation.

LPA is the appointment of an individual to make decisions on the patient’s behalf when the time comes that they lack the mental capacity to make decisions for themselves. In the United Kingdom, this must be in a prescribed form and must be registered with the Office of the Public Guardian whilst the individual still retains decision-making capacity. There are two types of LPA, for Health and Welfare and Property and Affairs. Other advocacy services include Court Appointed Deputies and Independent Mental Capacity Advocates.

We recognize that patients with HD may also wish to discuss euthanasia within ACP discussions. Although euthanasia is currently illegal throughout the United Kingdom (UK), euthanasia is legal in selective countries when strict conditions and criteria are met. For example, euthanasia has been legal in the Netherlands since The Dutch Euthanasia Act was approved by parliament in 2002 [12, 13]. In addition, Canada legalized medical assistance in dying (MAID) in 2016 and MAID has been utilized within a multidisciplinary HD clinic [14]. Euthanasia may come up in ACP discussions with individuals affected by HD and thus knowledge about the legislation and requirements of law is emphasized.

It is important to emphasize that ACP is voluntary and depicts an ongoing process of discussions on a continuum over time. The discussions are documented and regularly reviewed as the individual’s views may change over time.

**CONCLUSION**

Integrating ACP discussions within the provision of care and management of patients with HD is essential. Early and timely consideration of ACP discussions is paramount in HD given the potential challenges in ascertaining decision-making capacity in the advanced stages of the disease. Ultimately readiness to engage in ACP discussions is context-dependent and patient-specific and needs to be tailored according to whether the patient is ready and willing to discuss future decision making. ACP helps to empower and extend patient autonomy and provides clinicians and surrogate decision makers with reassurance that management is consistent with the individual’s expressed values and wishes. ACP is voluntary and depicts an ongoing process of discussions on a continuum over time. Regular follow up and review of ACP discussions is vital to establish consistency of decisions and preferences. We outline the framework of the dedicated ACP clinic integrated within our HD service to highlight the importance of a patient-centered and tailored care plan that fulfils the patient’s expressed goals, preferences, and values.

**CONFLICT OF INTEREST**

The authors have no conflict of interest to report.

**REFERENCES**


