

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-

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47 *tent with their values, goals and preferences during*
48 *serious and chronic illness. [2]” This definition pro-*
49 *vides a uniform framework in which to deliver ACP*
50 *clinical interventions [2]. Although this definition*
51 *of ACP is specific for adults [2], ACP nonetheless*
52 *needs to be considered, individualized, and tailored*
53 *to all patients, irrespective of age. ACP is voluntary*
54 *and depicts an ongoing process of discussions on a*
55 *continuum over time, fostering opportunities for an*
56 *adult, who retains decision-making capacity, to con-*
57 *template and express their individual concerns, goals,*
58 *preferences, values and wishes regarding future medi-*
59 *cal care [3].*

60 **ADVANCE CARE PLANNING IN** 61 **HUNTINGTON'S DISEASE**

62 Patients with HD usually have many years to
63 engage in ACP given that the course of the disease
64 may often span up to two decades from diagno-
65 sis to death. Early and timely consideration of ACP
66 discussions is paramount in HD given the potential
67 challenges in ascertaining decision-making capacity
68 in the advanced stages of the disease. ACP helps
69 to empower and extend patient autonomy and pro-
70 vides clinicians and surrogate decision makers with
71 reassurance that management is consistent with the
72 individual's expressed values and wishes. Document-
73 ation of ACP discussions and completion of relevant
74 legal documents, for example, advance decisions and
75 lasting power of attorney (LPA), is essential in order
76 to ensure care and treatment plans align with the
77 patient's expressed preferences.

78 A three-stage qualitative study produced and
79 piloted a care pathway for advance decisions and
80 power of attorney using HD as a model [4]. Five
81 major themes emerged during the modelling phase
82 of this study, including information deliberated and
83 method of delivery, individuals involved and loca-
84 tion (e.g., clinical setting, home), duration of process
85 and timing, assessment of capacity and form of doc-
86 umentation [4]. Rapport with an expert in HD to
87 facilitate the overall process was highlighted as a
88 dominant theme [4]. Although professionals were
89 reportedly reluctant to approach asymptomatic ser-
90 vices users too early due to concern of causing distress,
91 services users expressed positivity regarding early
92 introduction of advance decisions to promote individ-
93 ual autonomy; and thus earlier routine introduction
94 was implemented in development of the second care
95 pathway following the first pilot [4]. The first stage

96 of the care pathway incorporates the introduction of
97 end-of-life issues to patients in the clinic; the second
98 stage outlines the process of education and decision-
99 specific capacity assessment; and the third stage
100 depicts the process after completion of an advance
101 decision and the follow up review of the advance deci-
102 sion [4]. Resource implications need to be considered
103 in service integration, particularly relating to admin-
104 istration and time, given that the duration of sessions
105 in education, capacity assessment and advance deci-
106 sion completion took an hour each on average [4].

107 Initiating ACP discussions with individuals with
108 HD can be facilitated using the Huntington Disease
109 Quality of Life End of Life (HDQLIFE EOL) Plan-
110 ning measure [5]. This is a 16-item patient-reported
111 assessment tool that has been developed to evaluate
112 end-of-life (EOL) preferences for individuals with
113 HD [5]. This HD-specific EOL measure explores
114 domains and preferences relevant to ACP discus-
115 sions, including: advance directive, health care power
116 of attorney, nursing home care, location of death
117 preference, conversations about death and dying, liv-
118 ing will, life insurance, palliative care, child care
119 planning, finances, estate planning, support to make
120 decisions, hospice care, resuscitation preference,
121 funeral arrangements and preference about death [5].
122 Whilst reliability and validity data are preliminary
123 [5], the HDQLIFE EOL Planning measure offers
124 potential for meaningful and productive conversa-
125 tions relating to ACP.

126 There are no standardized guidelines about how or
127 when to broach the topic of ACP with patients with
128 HD. Ultimately readiness to engage in ACP discus-
129 sions is context-dependent and patient-specific and
130 needs to be tailored according to whether the patient
131 is ready and willing to discuss future decision mak-
132 ing. A qualitative research study aimed to investigate
133 the presence of thoughts or wishes surrounding EOL
134 in patients with HD or identified gene carriers visit-
135 ing the outpatient clinic via a questionnaire [6]. There
136 was a response rate of 55.4% out of 242 question-
137 naires sent, with non-responders younger in age and
138 of lower education compared to the demographics
139 of responders [6]. There was no significant differ-
140 ences between sex, Unified Huntington's Disease
141 Rating Scale (UHDRS) Total Functional Capac-
142 ity (UHDRS-TFC) or motor (UHDRS-M) scores
143 between responders and non-responders [6]. Of note,
144 77.2% of responders discussed their wishes with fam-
145 ily members yet only 42.6% of responders discussed
146 their wishes with a healthcare professional, with not
147 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 care 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

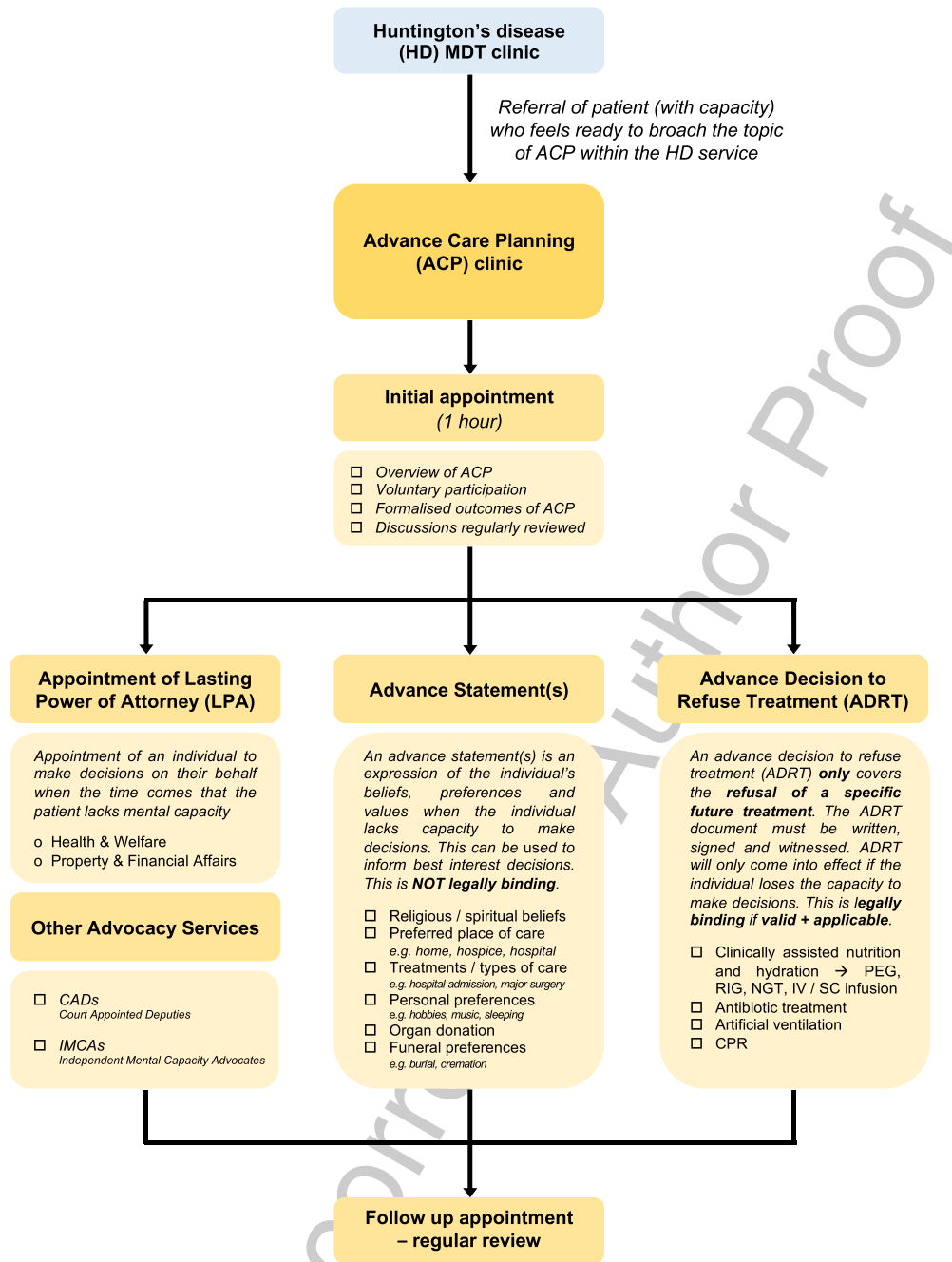


Fig. 1. The framework of the Advance Care Planning (ACP) clinic integrated within our Huntington's disease (HD) service. ACP, advance care planning; ADRT, advance decision to refuse treatment; CADs, court appointed deputies; CPR, cardiopulmonary resuscitation; IMCAs, independent mental capacity advocates;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

248 must be written, signed, and witnessed. ADRT will
 249 only come into effect if the individual loses the
 250 capacity to make decisions. This is legally binding if
 251 valid and applicable. Examples of specific issues that
 252 individuals with HD may choose to refuse through

an ADRT include: clinically assisted nutrition and
 hydration (e.g., percutaneous endoscopic gastro-
 stomy feeding, radiologically inserted gastrostomy
 feeding, nasogastric tube feeding, intravenous feed-
 ing, intravenous or subcutaneous infusion), antibiotic

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 254
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258 treatment, artificial ventilation and cardiopulmonary
259 resuscitation.

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

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

285 It is important to emphasize that ACP is volun-
286 tary and depicts an ongoing process of discussions
287 on a continuum over time. The discussions are doc-
288 umented and regularly reviewed as the individual's
289 views may change over time.

290 CONCLUSION

291 Integrating ACP discussions within the provision
292 of care and management of patients with HD is
293 essential. Early and timely consideration of ACP
294 discussions is paramount in HD given the potential
295 challenges in ascertaining decision-making capac-
296 ity in the advanced stages of the disease. Ultimately
297 readiness to engage in ACP discussions is context-
298 dependent and patient-specific and needs to be
299 tailored according to whether the patient is ready and
300 willing to discuss future decision making. ACP helps
301 to empower and extend patient autonomy and pro-
302 vides clinicians and surrogate decision makers with
303 reassurance that management is consistent with the
304 individual's expressed values and wishes. ACP is vol-
305 untary and depicts an ongoing process of discussions
306 on a continuum over time. Regular follow up and

review of ACP discussions is vital to establish con- 307
sistency of decisions and preferences. We outline the 308
framework of the dedicated ACP clinic integrated 309
within our HD service to highlight the importance of 310
a patient-centered and tailored care plan that fulfils 311
the patient's expressed goals, preferences, and values. 312

313 CONFLICT OF INTEREST

The authors have no conflict of interest to report. 314

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