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End of life with Huntington's Disease:
how can it be characterized?

Introduction

Huntington's disease (HD) is a rare, hereditary neurodegenerative disorder. Patients may have physical, cognitive and psychiatric symptoms for about twenty years before they die. There is still no cure for the disease, which adds to the importance of improving the quality of life and dying. It is unclear to what extent palliative care is provided in practice and what an effective palliative intervention entails[2]. Therefore, we first need to know what the last phase looks like. We aim to describe the clinical characteristics in the last month of living with HD, change in ACP and describe the reported causes of death.

Methods

A review of charts of patients (n=82) who died between 2017 and 2021 in a Dutch specialized nursing home for HD. No exclusion of charts. The data included is during the last month of life. The charts were reviewed by two researchers, who compared their data entry to achieve uniformity.
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Huntington's Disease[1,2]

- Inheritance: autosomal dominant
- Huntingtin gene (*Htt*) on chromosome 4: ≥36 repeats CAG → mutant Huntingtin protein → neurodegeneration
- Prevalence of 5-10 per 100.000
- Onset of symptoms: ± 30-50 years
- Length of disease duration: ± 15-20 years
- Trias of symptoms:
  - Movement disorders
  - Psychiatric symptoms
  - Cognitive impairment
- Other symptoms: weight loss, swallowing disorders, sleeping problems

Watch this short **video**, using the QR-code, to get an idea of patients in the advanced stage

Results (N=82)

Demographics	**Male**	**Female**
Male/female (N(%))	37 (45)	45 (55)
Age at time of death (mean(SD))	61.4 (11.4)	60.5 (11.2)
Length of stay in years (median(IQR))	2.3 (1.3-5.5)	5.2 (2.0-10.2)

Place of death (N(%))	
Nursing home	76 (93)
Hospital	
Physician assisted dying with organ donation	3 (4)
Admitted to hospital because of pneumonia	2 (2)
Other	1 (1)

Physical symptoms, during last month (N(%))	
Dysphagia	73 (89)
Constipation	66 (81)
Pain	64 (78)
Chorea	56 (68)
Pressure ulcers (mainly stage 1 or 2)	48 (59)
Nausea/vomiting	47 (57)
Body Mass Index (N=65), (median(IQR))	22.4 (19.5-25.6) 20.9 (17.6-23.5)

Psychiatric symptoms, during last month (N(%))	
Agitation	51 (62)
Resistiveness to care	47 (57)
Anxiety	41 (50)
Screaming	34 (42)
Sleeping problems	33 (40)
Obsessive-compulsive behavior	30 (37)
Aggression	23 (28)

Use of psychiatric medication (N(%))	**78 (95)**
Antipsychotics	72 (92)
Benzodiazepines	65 (83)
Antidepressants	49 (63)
Anticonvulsants	28 (36)

Mobility problems, during last month (N(%))	
Minor abnormalities	11 (13)
Immobility	19 (23)
Bedridden	28 (34)
Falls	36 (44)

Change in ACP, during last month (N(%))	**50 (61)**
Patient involved in change of ACP (N(%))	16 (21)

Reported causes of death (N(%))	
Aspiration pneumonia	25 (31)
End-stage HD	16 (20)
Unable to swallow	11 (13)
Palliative sedation due to refractory agitation	8 (10)
Other (malignancy, cardiac failure, COVID-19, ruptured aneurysm, CVA)	7 (9)
Physician assisted dying	6 (7)
Sudden death	5 (6)
Choking	2 (2)
Suicide	1 (1)
Fracture	1 (1)

Conclusions

The end of life of HD is a complex stage because of the variety and high frequency of physical and psychiatric manifestations. Therefore, no typical terminal stage can be described in HD. The physical symptoms become more severe and lead to increased dependency[1]. Psychiatric symptoms warrant high use of psychiatric medication. Change in ACP was needed in 61%, 21% of the patients were able to participate in the ACP discussion. Most patients die from complications of HD, such as severe dysphagia and end-stage HD, which can be seen as a combination of, among others, cachexia, autonomic disfunction and exhaustion.

Discussion

Despite this wide range of symptoms, 'specific' HD symptoms as chorea and obsessive-compulsive behavior may be underreported, when these were seen as 'normal' for the patient and not leading to problems. Severe disruptive behavioral symptoms may need palliative sedation. This, or, for example, withdrawal of food and drinks due to severe dysphagia, indicates that difficult choices are pertinent in the last month of life.
Physician assisted dying is slightly higher in HD than the general population in The Netherlands (5,1% in 2022[3]).
ACP discussions were initiated from the moment of admission to the nursing home and were evaluated at least every year. A change in ACP in 61% may show the difficulty of predicting the terminal stage in HD, despite patients are in the late stage for several years. Nevertheless, a change of ACP short before death was also found in nursing home patients with dementia[4]. ACP discussions are important for providing appropriate care; a multidisciplinary approach is warranted to relief symptoms and to improve comfort.
Only two HD patients died in the hospital because of an acute situation, which is less than the average of hospitalized nursing home residents in The Netherlands[5], indicating good nursing home care for Huntington's disease and in the importance of timely ACP conversations.
Whether or not the symptoms in HD are persistent during the disease progression, is worth further research.

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ZORG ONDER STEUNINGS FONDS

Topaz Overduin

Huntington Center of Expertise Topaz Overduin is the world's largest center specialized in HD. We will soon be moving into a brand-new facility in the Dutch dunes in Katwijk. We provide in-house care for over 80 residents and see approximately 110 patients in our outpatient clinic and 25 in our day-care program. Our vision is that all aspects of life matter and should be considered in caring for people with HD and their families.

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Topaz Het hele leven telt

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