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Considerations in Medical Decision-Making for an Unrepresented Patient With Late-Stage Creutzfeldt-Jakob Disease: Who Speaks for the Dying?

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Abstract

Creutzfeldt-Jakob disease (CJD) is a rare, rapidly progressive, and universally fatal neurodegenerative disease. Beyond its diagnostic and prognostic challenges, CJD presents profound ethical dilemmas when patients lack the capacity to participate in medical decisions. This case presents a middle-aged patient with paranoid schizophrenia, recent incarceration, and complex psychosocial determinants of health who was admitted for subacute progressive encephalopathy after being found down during a wellness check and had no contact with his family for decades, lacked an advance directive or identifiable surrogate, and was judged to lack decision-making capacity. The hospital course was complicated by incidental findings of acute pulmonary embolism and a rapid neurological decline which was later revealed to have a high likelihood of prion disease. We present the interdisciplinary process in diagnosing and developing a treatment plan that best reflects the patient's values and goals of care. This case also highlights essential considerations for clinical practice when caring for an unrepresented patient with irreversible loss of decision-making capacity at the end of life, including the application of ethical principles, adherence to institutional policies and legal frameworks, and the implementation of a patient-centered approach that preserves dignity, promotes comfort, and protects vulnerable individuals.

Categories: Neurology, Palliative Care, Geriatrics
Keywords: advance care planning, clinical ethics, end-of-life care, patient autonomy, surrogate decision-making, unrepresented patients

Introduction

Unrepresented patients are those who lack an advance directive or identifiable surrogate and are judged to lack decision-making capacity. An increasing number of patients admitted to critical care settings are unrepresented [1]. This can result in delays in care and unnecessary or prolonged non-beneficial treatments, among other consequences for this vulnerable population [2]. In a landmark study reviewing data from the Health and Retirement Study, the majority of patients who needed to make a treatment decision were incapable of doing so [3]. Protocols within some hospitals do not typically address comfort care; thus, unrepresented patients may unintentionally receive aggressive care [4].

We report the case of a patient in his late 50s who was admitted to the hospital with an unclear presentation of subacute progressive encephalopathy and a fragmented psychiatric medical history. Additionally, the patient lacked an advance directive and any identifiable next of kin who could make medical decisions on his behalf. In our case, we present an interdisciplinary approach to the management of an unrepresented patient with psychiatric comorbidity and limited social support who was diagnosed with late-stage Creutzfeldt-Jakob disease (CJD), a rapidly progressive disease with a poor prognosis.

Although the clinical manifestations and prognosis of CJD are well established, the ethical complexities of end-of-life decision-making for unrepresented patients remain less frequently addressed in the literature. This case offers practical insights into the role of interdisciplinary collaboration, institutional safeguards, and ethical oversight in ensuring that end-of-life care respects patient dignity and prioritizes comfort.

Case Presentation

A man in his late 50s with schizoaffective disorder, bipolar type, and past suicidal ideation who had experienced recent incarceration and housing instability was admitted to the Jennifer Moreno Department of Veterans Affairs Medical Center, San Diego, California, USA, in mid-2024 for acute subsegmental pulmonary embolism, urinary tract infection, and subacute progressive encephalopathy after being found unresponsive in a motel room during a wellness check. Collateral information obtained from his case manager indicated that his presenting mental status and behavior represented a marked departure from his

reported baseline four months earlier. The case manager described fluctuating levels of consciousness, behaviors suggestive of responding to internal stimuli, episodes of laughter without an apparent external stimulus, speaking aloud in the absence of an identifiable conversational partner, and inability to recognize her. Neurological examination revealed decreased concentration, impaired delayed recall despite preserved verbal repetition, mild to moderate dysarthria, left upper extremity weakness, and bilateral dysmetria on finger-to-nose testing, more pronounced in the left upper extremity.

The patient exhibited deficits in multiple cognitive domains, including attention, word recall, and following simple commands. The differential diagnosis included infectious, autoimmune, and paraneoplastic encephalitis, toxic-metabolic encephalopathy, hypoxic-ischemic brain injury, seizures, and other rapidly progressing neurodegenerative disorders. Multidisciplinary evaluations involving neurology, psychiatry, infectious disease, and other relevant specialties were conducted. Additional diagnostic testing was deemed necessary to distinguish prion disease from these clinical mimics. As shown in Figure *1*, the patient's brain magnetic resonance imaging (MRI) revealed right greater than left curvilinear/gyriform restricted diffusion, predominantly involving the right cerebral hemisphere and basal ganglia, concerning for CJD, an ultimately fatal neurodegenerative disease attributed to the misfolding of prions in the brain [5].

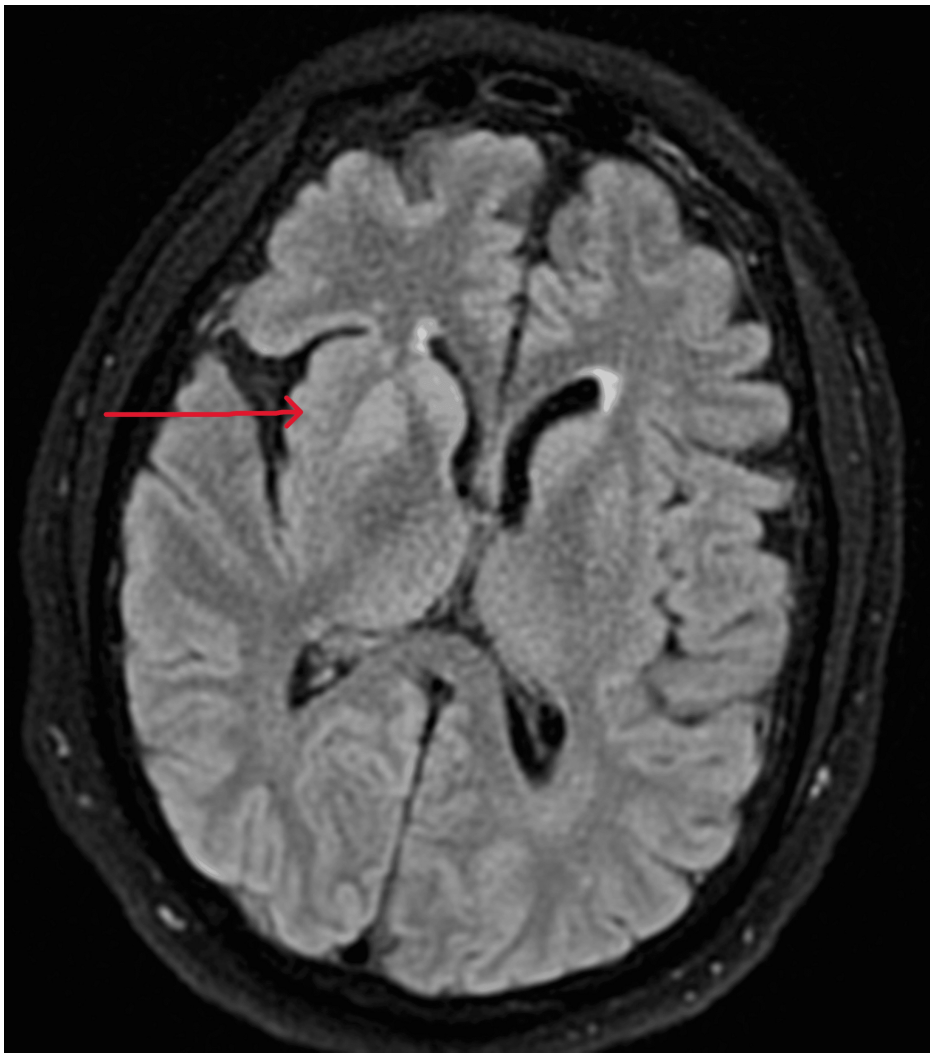


FIGURE 1: T2-weighted magnetic resonance image of the brain.

Cortical/gyriform restricted diffusion involving the right greater than left cerebral hemispheres and basal ganglia.

Multiple healthcare team members attempted to contact the patient's next of kin but were unable to reach anyone listed in his medical record. An ethics consultation was placed for review and adherence to the VA policy. Accordingly, procedures requiring informed consent, such as the lumbar puncture, were performed after joint approval by the attending physician and the chief of service [6]. Cerebrospinal fluid analysis revealed positive real-time quaking-induced conversion (RT-QuIC) and abnormally elevated T-tau >20,000 pg/ml (reference range: 0-1149 pg/ml) and 14-3-3 protein >16,000 AU/ml (reference range: 173-1999 AU/ml), with a prion disease likelihood ratio of >98%. A probable sporadic CJD (sCJD) diagnosis can be established by

the presence of a neuropsychiatric disorder combined with positive RT-QuIC resulting in the cerebrospinal fluid [7]. Given the results from the comprehensive workup and the patient's rapid progressive decline, including severe oropharyngeal dysphagia, the patient was estimated to have been symptomatic with CJD for approximately four months, with a prognosis of weeks.

An exhaustive search for potential surrogates was conducted, including an extensive review of the patient's electronic medical record and contact with collateral agencies involved with the patient to inquire about possible next of kin. Social work contacted remote relatives and close friends who either could not be reached or were unable to participate. From those contacts, three weeks after admission, the highest-ranking surrogate was identified and agreed to become the voice of the patient. After discussing the benefits and burdens of potential treatments and exploring the patient's core values, the next of kin provided surrogate consent to transfer the patient to the inpatient hospice unit for comfort care. The patient passed away comfortably, roughly three weeks later following rapid decline. The tissue was sent to the National Prion Disease Pathology Surveillance Center, confirming sCJD.

Discussion

While treating patients with CJD is not unique in the literature, our case study describes the challenges and questions posed when treating unrepresented patients admitted with an unclear presentation, particularly those from underserved populations (e.g., homelessness, incarceration history, and low socioeconomic status) and those with psychiatric comorbidities. Although CJD is incurable and inevitably fatal, the requirement for informed consent to transition a patient from life-sustaining treatments to comfort care can be a logistical and ethical barrier [1,6-9]. Refusal of further testing before a thorough diagnostic workup can pose particular challenges to treatment decisions when the patient lacks decision-making capacity and a representative is unavailable [1]. Even for family members who are present, surrogate decision-making can be fraught with stress and anxiety [9,10].

The literature has indicated that providers may at times avoid end-of-life discussions in anticipation that such discussions will provoke uncomfortable thoughts of dying, including suicide [11]. The stigma that mental illness may impair complex decision-making is also prevalent [11,12]. Perhaps more importantly, the state of being unrepresented can create a dilemma between autonomy and beneficence. Clinicians must balance respect for the patient's previously expressed values, religious or spiritual beliefs, and cultural preferences, including preferences for pursuing or declining aggressive interventions, with their obligation to act in the patient's best interests by appropriately diagnosing and treating the underlying disease. What may be clear is the adherence to the principle of proportionality: the intended outcome of treatments and testing should outweigh the burden of those interventions [13]. However, where and when to draw the line for further testing and treatment is less clear-cut in practice.

Variability and differences between state and federal laws further complicate surrogate selection, especially for clinicians who work at multiple sites [6,14,15]. For instance, VA follows a rigid hierarchy (spouse, child, parent, sibling, grandparent, grandchild, followed by a close friend), while California law, for example, permits healthcare providers to select a familiar and reasonable surrogate without strict hierarchical constraints, theoretically allowing a close friend to serve over a spouse.

Recognition of nontraditional surrogates warrants further discussion. For example, Georgia explicitly permits incarcerated individuals to serve as surrogates for other incarcerated individuals, while other states explicitly ban it. Most states lack clear guidance for incarcerated individuals near the end of life as surrogates or as patients [15]. In this case, an individual who was incarcerated alongside the patient could have had some familiarity with the patient's personal values. This may raise additional ethical questions about whether such a person should be considered a close friend for the purposes of surrogate decision-making. However, even where legally permissible, logistical barriers can be challenging.

A novel clinical project at one VA site facilitated a health professional-led group discussion on advance healthcare planning for veterans presenting to the hospital for other reasons. It revealed that a substantial number of veterans had made at least some progress or had given forethought to advance healthcare planning, even if they did not yet have an advance directive in their medical record. Furthermore, a large majority of patients initiated at least one step of advance care planning after the intervention [16]. Clinicians should prioritize advance care planning discussions with patients at risk of becoming unrepresented, particularly those with multiple social risk factors. Some institutions mandate discussions surrounding advance directives annually [16].

Given that CJD has no curative treatments available, early and accurate diagnosis could rule out aggressive treatments and allow for earlier engagement in goals-of-care discussion. This can prevent the need to make complex ethical decisions and allow patients time to cope with and prepare for the end of life. Clinician decision-making in patients with irreversible neurological diseases is no different from other shared medical decision-making, which includes an obligation to honor the ethical principles of autonomy, beneficence, nonmaleficence, and justice [17]. Ethics committees can provide guidance and oversight to support ethically sound, patient-centered decisions, particularly when considering the benefits and burdens of potentially non-beneficial interventions in patients with incurable diseases and poor prognoses. Finally, the legal

procedure at the healthcare institution caring for unrepresented patients is paramount when choosing a surrogate and whether to continue life-sustaining treatment.

Conclusions

This case underscores the practical and ethical challenges of managing rapidly progressive neurocognitive decline in patients without an advance directive or identifiable surrogate and judged to lack decision-making capacity.

Given the absence of curative treatment for CJD, early and accurate diagnosis is essential to guide individualized care. Treatment decisions should consider prognosis, potential burdens of intervention, and the patient's known values, including religious and spiritual beliefs, while avoiding non-beneficial interventions.

We believe that proactive advance care planning, including early multidisciplinary goals-of-care discussions, is essential for patients at risk of becoming unrepresented. Timely documentation of advance directives and periodic reassessment of treatment preferences may facilitate appropriate palliative care interventions, preserve patient autonomy, and minimize delays in medical decision-making.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Critical review of the manuscript for important intellectual content: Zaida Albarracin

Supervision: Zaida Albarracin

Acquisition, analysis, or interpretation of data: Esther Emenalom

Disclosures

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References

1. Dixon JD, Josyula AV, Javier NM, et al.: American Geriatrics Society position statement: making medical treatment decisions for unrepresented older adults. J Am Geriatr Soc. 2025, 73:1353-64. 10.1111/jgs.19288
2. Catlin CC, Connors HL, Teaster PB, Wood E, Sager ZS, Moye J: Unrepresented adults face adverse healthcare consequences: the role of guardians, public guardianship reform, and alternative policy solutions. J Aging Soc Policy. 2022, 34:418-37. 10.1080/08959420.2020.1851433
3. Silveira MJ, Kim SY, Langa KM: Advance directives and outcomes of surrogate decision making before death . N Engl J Med. 2010, 362:1211-8. 10.1056/NEJMsa0907901
4. Pope TM, Bennett J, Carson SS, et al.: Making medical treatment decisions for unrepresented patients in the ICU. An official American Thoracic Society/American Geriatrics Society Policy Statement. Am J Respir Crit Care Med. 2020, 201:1182-92. 10.1164/rccm.202003-0512ST
5. Mead S, Rudge P: CJD mimics and chameleons. Pract Neurol. 2017, 17:113-21. 10.1136/practneurol-2016-001571
6. Life-Sustaining Treatment Decisions: Eliciting, Documenting and Honoring Patients' Values, Goals and Preferences. Department of Veterans Affairs, Washington, D.C.; 2017. https://www.ethics.va.gov/docs/policy/VHA_Handbook_1004_03_LST.pdf.
7. Clinical overview of Creutzfeldt-Jakob Disease (CJD). (2024). Accessed: November 18, 2025: https://www.cdc.gov/creutzfeldt-jakob/hcp/clinical-overview/diagnosis.html.
8. Hall WA, Masood W: Creutzfeldt-Jakob disease. StatPearls [Internet]. 2025 (ed): StatPearls Publishing, Treasure Island (FL);
9. Torke AM, Sachs GA, Helft PR, Montz K, Hui SL, Slaven JE, Callahan CM: Scope and outcomes of surrogate decision making among hospitalized older adults. JAMA Intern Med. 2014, 174:370-7.

10.1001/jamainternmed.2013.13315

10. Sulmasy DP, Hughes MT, Yenokyan G, et al.: The Trial of Ascertaining Individual Preferences for Loved Ones' Role in End-of-Life Decisions (TAILORED) study: a randomized controlled trial to improve surrogate decision making. J Pain Symptom Manage. 2017, 54:455-65. 10.1016/j.jpainsymman.2017.07.004
11. Elie D, Marino A, Torres-Platas SG, et al.: End-of-life care preferences in patients with severe and persistent mental illness and chronic medical conditions: a comparative cross-sectional study. Am J Geriatr Psychiatry. 2018, 26:89-97. 10.1016/j.jagp.2017.09.018
12. Foti ME, Bartels SJ, Van Citters AD, Merriman MP, Fletcher KE: End-of-life treatment preferences of persons with serious mental illness. Psychiatr Serv. 2005, 56:585-91. 10.1176/appi.ps.56.5.585
13. Hermerén G: The principle of proportionality revisited: interpretations and applications . Med Health Care Philos. 2012, 15:373-82. 10.1007/s11019-011-9360-x
14. Health care decisions: decisionmakers and surrogates. (2022). Accessed: May 16, 2025: https://legiscan.com/CA/text/AB2338/id/2609209.
15. Helmly V, Garica M, Williams B, Howell BA: A review and content analysis of U.S. Department of Corrections end-of-life decision making policies. Int J Prison Health. 2022, 18:165-75. 10.1108/IJPH-06-2021-0060
16. Garner KK, Dubbert P, Lensing S, Sullivan DH: Concordance between veterans' self-report and documentation of surrogate decision makers: implications for quality measurement. J Pain Symptom Manage. 2017, 53:1-4. 10.1016/j.jpainsymman.2016.10.356
17. Taylor LP, Besbris JM, Graf WD, Rubin MA, Cruz-Flores S, Epstein LG: Clinical guidance in neuropalliative care: an AAN Position Statement. Neurology. 2022, 98:409-16. 10.1212/WNL.0000000000200063