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Prions to Pathways: Safeguarding Against Creutzfeldt-Jakob Disease in the Operating Room

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Introduction

As defined by the Center for Disease Control (CDC, 2018), Creutzfeldt-Jakob Disease (CJD) is a rapidly progressive, rare, transmissible, and fatal illness. Miscoiling of healthy proteins caused by an abnormal isoform of cellular glycoprotein resulting in protein folding has been found to be the molecular mechanism during the pathological process of this malady (Figure 1), (Castle, Gill, 2017).

The current problem is that few recommendations are available for organizations to consider how to manage potential CJD patient cases, and institutions are left to compile and develop their own guidelines and/or protocols for clinical practice. The standards of care for this rare disease in healthcare are evolving quickly and are often forged from experiences (Thomas *et. al.* , 2013).

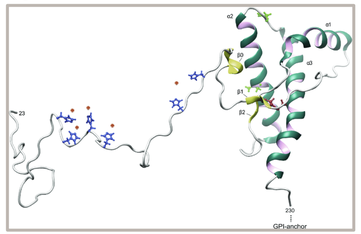

Figure 1. Schematic representation of cellular prion protein (Legname, 2017).

This poster displays a quality improvement project and synthetizes the process and experiences of how one organization transformed and optimized CJD prevention strategies. A pathway was developed that enhances patient identification, promotes multidirectional coordination, utilizes communication tools, and supports interprofessional debriefings, resulting in harmonized CJD patient care.

Approach

The potential presence of CJD in the perioperative patient requires interprofessional care and team collaboration because the signs and symptoms of this disease can be misleading. This includes not just perioperative staff, but also experts representing Neurology, Neurosurgery, Anesthesia, Pathology, Infection Prevention, and Safety Management, amongst others. Inpatient care teams, the patient, and families are also essential partners for collaboration.

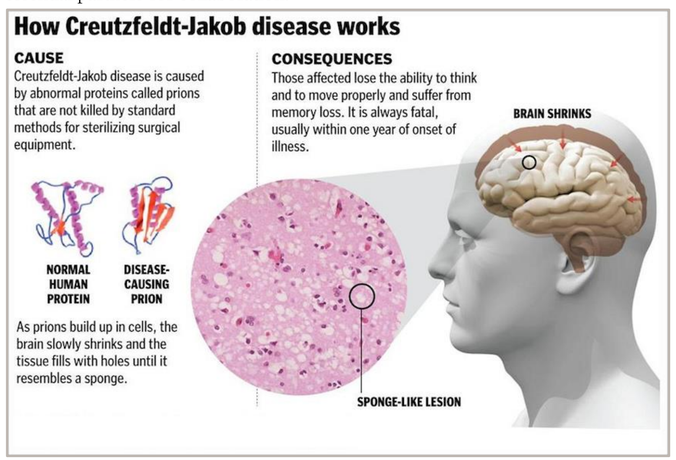

Figure 2. Causes and consequences of CJD (World Health Organization, Centers for Disease Control and Prevention, National Institute of Neurological Disorders and Stroke, AP).

For a patient, the symptoms of CJD are demonstrated by increasing memory impairment that progresses to dementia and the disease is always fatal usually within one year of onset of illness (Figure 2).

Pathway Development and Implementation

The process started in November, 2016 to create prevention strategies and a policy for the interprofessional stakeholders in a large tertiary care organization. Pathway development was a multilayered shared process requiring extensive exploration and review of current evidence to achieve consensus amongst stakeholders. The sequential efforts included practice gap identification, decision making, and role clarification on how to standardize care for the potential CJD population.

Literature Review and Expert Consultations

- The CDC and World Health Organization were consulted. They provided general guidelines and recommendations for the management of the patient with CJD in the hospital organization.
- A comprehensive literature search and review was completed. The content of the current literature focused primarily on the microbiological aspect of the disease, the transmission, the incidence, and the fatality of CJD.
- Fifteen round table discussions were conducted with experts from Perioperative Leadership, Neurology, Neurosurgery, Anesthesia, Pathology, Infection Prevention, Safety Management and Risk Management. These meetings focused on risk screening and symptom identification.

Patient Identification

- **Agreed screening criteria:** any patient with symptoms of rapidly progressive dementia, ataxia, encephalopathy or neuropsychological symptoms without clear etiology will be identified as a risk for CJD.
- **Testing:** at risk patients will undergo further testing and have a lumbar puncture screening for 14-3-3 protein. If positive, a non-directed brain biopsy will follow. If negative, additional testing needs to be considered.
- **Operating Room:** any patient with rapidly progressing neurological symptoms and undergoing non-directed brain biopsy will be treated under the high infectivity prion protocol.

Multidirectional Coordination

- **Roles, responsibilities, and patient flow coordination:** the Surgeon in collaboration with the Neurologist and provider caring for the patient, who have identified a patient at risk for prion disease, will determine and coordinate the most appropriate location for testing.
- Once the proceduralist and the appropriate location have been determined, Infection Prevention (IP) is notified of the proposed surgery/procedure. IP then deploys communication to affected departments in preparation for procedure.
- **Timeline:** no less than 24 hours is given to allow for involved departments and teams to coordinate preparations to conduct procedure.

Communication Tools

- **Policy:** is readily and electronically available for all departments and stakeholders.
- **Huddle:** pre-procedure huddle in the OR is crucial and called upon by the sponsor of the policy. This huddle includes: Neurosurgeon, Anesthesiologist, Circulating RN, Surgical Technologist, and support staff. The policy is reviewed and roles and responsibilities are clearly assigned.
- **Additional communication tool**: red **Protocol Stop Signs** are placed on all OR entrances and exits where the procedure will be performed.

Interprofessional Debriefings

- **Debriefing:** is organized by the policy sponsor after the procedure. This is designed as a structured process and the goal is to analyze the implementation of the procedure whilst adhering to the policy. The focused discussion provides the interprofessional team an opportunity to enhance performance, optimize future procedures, identify communication breakdowns, and also highlight implementation success.
- **Documentation:** content of the discussion is electronically recorded and available for the leadership team of the organization.

Outcome

This topic magnified the institutional vulnerability and safety risks inherent to a disease that is high risk, low volume, and with limited availability of guidelines. The outcome of the process was an institutional policy describing the identification and management of patients with concerns for CJD. This policy extends to, but is not limited to, the perioperative area, interventional radiology, and the patient bedside. The provider is enabled with an interprofessional team to safely deliver evidence-based care for a patient whose neurological diagnosis is unclear. This work has introduced and discussed an approach that is replicable in other organizations that may consider developing a pathway and policy, in anticipation and preparation for managing patients who may undergo the undirected brain biopsy.

Implications for Perioperative Care

The estimated occurrence of CJD worldwide and in the United States has been reported to affect approximately 1:1,000,000 people per year (CDC, 2018). The prevalence of CJD can be anticipated to increase (Figure 3). The signs and symptoms of the disease are deceptive and brain biopsy is used to support conclusive diagnosis. Increased awareness and proactivity, as indicated also by AORN, will safeguard the OR environment, staff, patients and families, and the institution against this insidious disease (Anderson, 2018). Our recommendation is that every institution considers a policy and a pathway that provides guidance to the interprofessional team clarifying expectations, roles and responsibilities, and offering tools for managing patients with concerns for CJD. Together, this work serves to prepare for a one in one million opportunity.

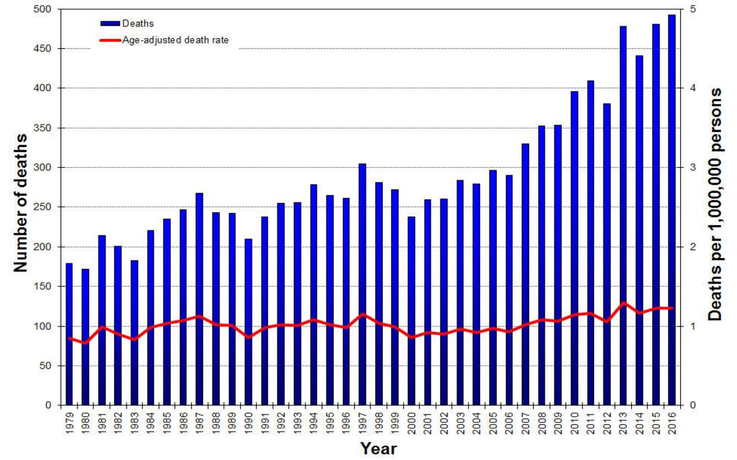

Figure 3. Creutzfeldt-Jakob Disease Deaths and Age-Adjusted Death Rate, United States, 1979-2016 (Centers for Disease Control and Prevention, 2018).

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