

CASE 5

Investigating Suspected Outbreaks of Rare Infectious Disease Using Surveillance Data: The CJDSS Perspective

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BACKGROUND

Renée Dubois, a public health nurse consultant of fifteen years, sat at her desk at the Public Health Agency of Canada, sipped her tea, and stared out the window. It was a beautiful spring morning in Ottawa, Canada. From the corner of her eye, she noticed a red light flashing on her landline and listened intently to the voicemails left on the Creutzfeldt-Jakob Disease Surveillance System (CJDSS) phone line over the weekend. She listened to the first message, then the second, and then the third. As the train of voicemails ended, her eyes widened. For the first time in a decade, there had been a suspected outbreak of variant Creutzfeldt-Jakob Disease (vCJD) in Ontario.

Dr. Lawson, the referring doctor on the last voicemail, sounded calm despite the situation at hand. “If you would please call me back as soon as possible, I would like to speak further about the next steps.” Dr. Lawson is a neurologist at a well-known neurology center in Toronto, and she called regarding a suspicious cluster of possible variant CJD cases in the Greater Toronto Area. A set of five patients were displaying early symptoms of rapidly progressive dementia. Noting that several of these patients were much younger than the typical age demographic prone to sporadic CJD (Exhibit 1), Dr. Lawson immediately contacted the CJDSS to investigate these cases as possible cases of variant CJD.

Upon receiving this information, Renée immediately sent out a department-wide email notifying the other nurses of the situation. She had previous experience with a possible CJD outbreak a decade prior. Luckily, the cases were not variant CJD (as confirmed by autopsy), and were instead misdiagnosed cases of sporadic CJD. From this experience, Renée knew that the consequences of an outbreak of variant CJD had wide ramifications, which required pre-planning, inter-sectoral collaboration, and effective mobilization of resources to quickly diagnose and isolate the source of a possible outbreak.

CREUTZFELDT-JAKOB DISEASE

Creutzfeldt-Jakob Disease (CJD) is a rapidly progressing, fatal, neurodegenerative disease. It is the human form of bovine spongiform encephalopathy (BSE), popularized as “Mad Cow Disease” in the early 2000s. The worldwide incidence of CJD is approximately one to two cases per 1,000,000 persons (NCJDRSU, 2016). In Canada, this translates to 40-50 new cases per year (Exhibit 2). Since 1994, there have been over 800 cases of confirmed CJD in Canada (Exhibit 3), with a majority of cases in Ontario and Quebec (Exhibit 4).

DISEASE SYMPTOMOLOGY AND PROGRESSION

Creutzfeldt-Jakob Disease is characterized by the accumulation of prion proteins in the brain and neural tissue degeneration, which gives the tissue a spongy appearance (PHAC, 2012).

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At onset, patients often experience psychiatric or sensory symptoms, including anxiety, depression, visual hallucinations, and aphasia (WHO, 2012). As the disease rapidly progresses, neurological defects develop within weeks, resulting in involuntary movements, dementia, and ataxia (UCSF, 2016). The average duration of illness, from initial neurological symptoms to death, can vary from four to 40 months (Kovács et al., 2005). At time of death, patients are completely immobile and mute (WHO, 2012).

TYPES OF CJD

There are three types of CJD: sporadic, genetic, and acquired. In ~85% of patients, the disease occurs sporadically, without any known external source of infection. This is referred to as sporadic CJD. In 10% to 15% of patients, CJD develops in familial clusters, passed down through inherited mutations of the prion protein gene PRNP. This type of CJD is referred to as genetic CJD. Lastly, acquired CJD can develop in several ways and is the rarest form of CJD, occurring in approximately 1% of patients. Acquired CJD is an umbrella term used to describe iatrogenic CJD and variant CJD. As the name suggests, iatrogenic CJD is transmitted iatrogenically, induced accidentally via hospital or diagnostic procedures. Iatrogenic CJD is linked with exposure to contaminated hormones, infected dura mater or corneal grafts, and contaminated neurosurgical instruments. However, *variant* CJD (vCJD) develops from the ingestion of BSE-contaminated meats, or blood transfusions from a vCJD-positive donor (WHO, 2012; Budka & Will, 2015). WHO's case definitions of CJD are presented in Exhibit 5.

PRIONS AS INFECTIOUS AGENTS

The prion protein (PrP) in its normal form is present in all humans and animals (WHO, 2011). However, PrP is in an altered, self-propagating form in diseases like CJD and is referred to as a prion or a proteinaceous infectious particle (Prusiner, 1998). When PrP is in its mutated form, it is able to infect non-mutated forms of the protein, resulting in a cascade effect in the brain. Contact with tiny amounts of prion-contaminated material can initiate the disease process in an otherwise healthy individual. Once they appear, abnormal prion proteins aggregate. The aggregation of proteins contributes to widespread neuron loss. However, the exact molecular mechanism of this process is unknown (Clarke et al., 2001).

The altered form of PrP is responsible for a host of diseases including Kuru, Gerstmann–Sträussler–Scheinker syndrome, and fatal familial insomnia (Will, 2003), with the latter two implicated in genetic routes of transmission (Collins et al., 2001).

Certain surgical procedures can transmit prions. These include dura mater grafts, corneal grafts, and any surgery involving neural tissues (PHAC, 2007). Prions can also be spread via exposure to BSE-containing foods. However, there is no evidence that CJD can spread via social contact, sexual contact, mother-to-child, blood transfusion, or through routine health care (PHAC, 2007). CJD tissue infectivity is outlined in Exhibit 6.

A HISTORY OF VARIANT CJD IN CANADA AND WORLDWIDE

The earliest cases of vCJD were identified in 1994 in unexpectedly young individuals in the United Kingdom (Will et al., 1996). Since then, over 200 cases of vCJD have been identified worldwide, with 175 cases in the UK alone and 49 cases in other countries around the world – including two in Canadian residents (WHO, 2012).

After 1996, the United Kingdom implemented strict food safety measures to prevent further contamination in its food supply. Canada followed suit after the first Canadian case of vCJD was identified in 2002 (Jansen et al., 2003).

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Furthermore, scientists discovered that several patients in the UK were infected with vCJD-positive blood via blood transfusion (Belay & Schonberger, 2005; WHO, 2008). This revelation spurred many countries to take preventive measures to regulate their blood supplies and to minimize the risk of vCJD being spread by blood transfusion. This remains true even today, as Canadian Blood Services and Héma-Québec ensure that blood donations are not accepted from individuals who may be silent carriers of CJD – for example, individuals who may have lived a cumulative of three or more months in the United Kingdom between 1980 and 1996, or five or more years in Western Europe from 1980 to 2007 (Héma-Québec, 2014).

Currently, the Government of Canada regulates the Canadian beef industry to minimize the risk of BSE contamination. It has also minimized the risk of infection through surgical procedures (PHAC, 2007) and through blood transfusion (Héma-Québec, 2014). By implementing various infection-control guidelines, as outlined in the Creutzfeldt-Jakob Disease Quick Reference Guide (PHAC, 2007), the risk of BSE infection in Canada is considered to be extremely low.

CREUTZFELDT-JAKOB DISEASE SURVEILLANCE

The Canadian Creutzfeldt-Jakob Disease Surveillance System (CJDSS) was established by Health Canada in 1998 (PHAC, 2003), in response to the widespread outbreak of vCJD in the United Kingdom. It was created as a national surveillance system for Creutzfeldt-Jakob Disease. Currently, the role of the CJDSS is to assist in the investigation and diagnosis of suspected cases of CJD in Canada, provide support to patients and health care providers, and ultimately protect the health of Canadians by reducing the risk of CJD transmission.

Following the World Health Organization's (WHO) model for comprehensive surveillance of human prion diseases, the CJDSS offers support to collaborating health care professionals. This is completed via initial consultation, referral for laboratory testing, provision of logistic support, education, and awareness. With informed consent from patients, a detailed medical chart review and family interviews are also completed. The resultant diagnostic and epidemiological information gathered by the CJDSS is assembled in a comprehensive online database and is used to support further epidemiological analysis and public health decision-making (PHAC, 2016).

HOW IS CJD DIAGNOSED?

Currently, there is no single definitive diagnostic test for CJD. However, if a patient is exhibiting symptoms of CJD, there are several recommended laboratory tests that can be completed to rule out more common causes of dementia or neurological symptoms, including encephalitis or chronic meningitis (NINDS, 2003). Recommended laboratory testing for CJD is outlined in Exhibit 7.

PUBLIC HEALTH CHALLENGES OF PRION DISEASES

Prion diseases like CJD pose a unique combination of challenges to public health. These include their long incubation periods (upwards of a decade), a lack of host immunity, a strong resistance to decontamination (for example, in neurosurgical instruments), limited diagnostic testing, and potential for zoonotic and health care-based transmission (Coulthart et al., 2015).

Because the early symptoms of CJD are often non-specific, and the disease itself is quite rare, it can be very difficult for physicians to diagnose. The disease also progresses quickly, and it can be difficult for physicians to support families through the quick deterioration of their loved one. Thus, it is important for public health to be aware of CJD and its risks and to be able to mobilize resources to isolate and quarantine the source of a possible outbreak situation.

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BACK TO RENÉE – THE CASES AT HAND

After conducting an initial intake questionnaire with physicians and health care providers, Renée amalgamated essential information regarding the five cases of suspected CJD, taking care to note that all cases occurred within 50 km of Toronto. She recorded the patients' dates of birth, dates of onset, symptoms, and test results. She also noted rapidity of symptom progression.

Some initial test results are recorded in Exhibit 8. However, due to the varied availability of equipment and quality of care experienced by Ontarians, not all recommended diagnostic tests were completed prior to patient death (as in Case #3), while some results are still pending (Case #4 and #5).

MASS MEDIA GETS THE SCOOP

Two weeks after Renée received the first voicemail from Dr. Lawson, Renée received word from Dr. Lawson that the first patient of the five patients passed away. Mr. C. (Case #2) was a young, prolific athlete, who was set to compete in the upcoming Olympics in Brazil. His rapidly declining health was covered extensively in the public eye, with frequent posts to social media outlining his deteriorating physical state.

Local reporters had begun flooding the neurology clinics and hospitals frequented by Mr. C., demanding answers. Several news outlets also released articles sensationalizing the potential return of "Mad Cow Disease," and imploring the federal government to intervene in the investigation. Lobbyists began to spread sensationalized articles demanding a beef ban, and Health Canada was being pressured to release a statement.

DEVELOPING AN INVESTIGATIVE STRATEGY: RENÉE'S DECISION

Renée tasked two nurses to compile the medical records and clinical information for each case. Knowing the causes and methods of transmission of CJD, Renée struggled to come up with the root cause of this outbreak. Without access to each patient's medical history, she was struggling to weave a connecting thread between the individual cases.

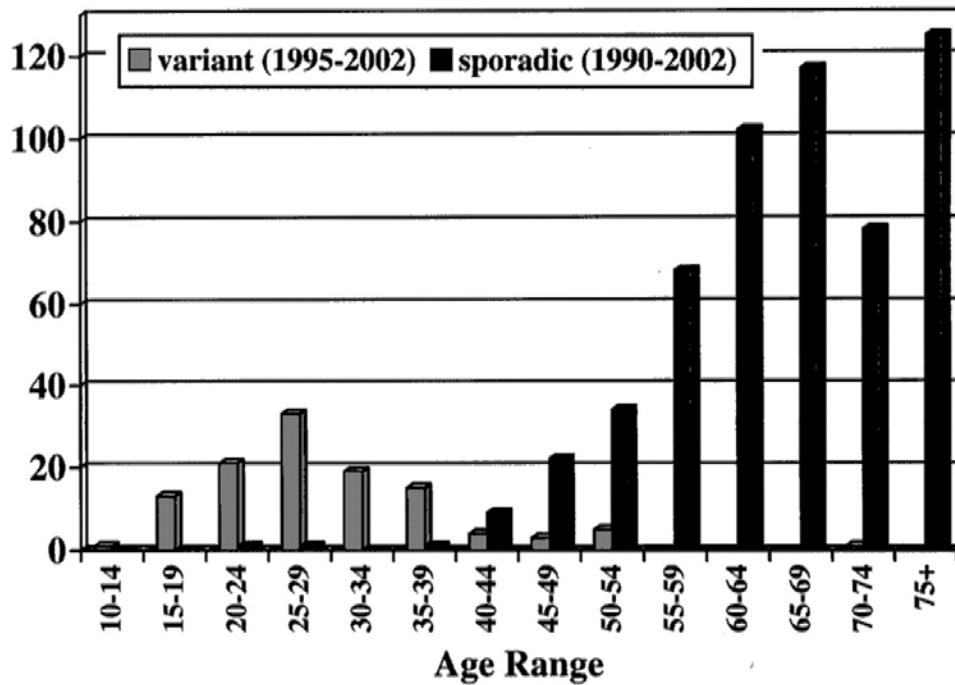
Renée needed to figure out the best course of action to lead the team of nurses to the correct diagnosis. From her previous experiences working in a neurology clinic for 10 years, she was very familiar with CJD diagnostic tests and how to interpret them. What were the best diagnostic tests for her to pursue? What were some potential root causes she could suggest the team to concentrate on to direct their investigation? Were these cases connected? If so, what should she do next?

A confident speaker, Renée often presented about CJD at universities and conferences across Canada. From her experiences as a keynote speaker, Renée knew that referring to CJD as "Mad Cow Disease" would merely fan the flames of public hysteria. Reducing the sensationalized stories in the media was her first goal.

In the case of Mr. C., what essential information should Renée recommend that the hospitals and neurology clinics share to the media? What should Renée's main message be? Who should Renée collaborate with to get her message out to the public? What cautionary information should the public campaign include?

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EXHIBIT 1
Distribution of Variant and Sporadic CJD Cases in the UK, by Age



Source: Will, 2003. Reproduced with permission from Robert G. Will. Acquired Prion Disease: Iatrogenic CJD, Variant CJD, Kuru. British Medical Bulletin (2003) 66 (1): 255-265. Published by Oxford University Press online at: <https://academic.oup.com/bmb/article/66/1/255/284826/Acquired-prion-disease-iatrogenic-CJD-variant-CJD?searchresult=1> For permissions please email: journals.permissions@oup.com.

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**EXHIBIT 2
Incidences of CJD in Canada: 1999-2016
As of June 30, 2016**

Year of Death	Total CJD Cases	Population of Canada	Incidence Rate
1999	32	30,492,106	1.05
2000	35	30,783,969	1.14
2001	30	31,130,030	0.96
2002	36	31,450,443	1.14
2003	29	31,734,851	0.91
2004	44	32,037,434	1.37
2005	44	32,352,233	1.36
2006	44	32,678,986	1.35
2007	39	33,001,076	1.18
2008	49	33,371,810	1.47
2009	53	33,756,714	1.57
2010	38	34,131,451	1.11
2011	51	34,472,304	1.48
2012	63	34,880,248	1.81
2013	51	35,289,003	1.45
2014	55	35,675,834	1.54
2015	51	35,702,707	1.43
2016	11	35,985,751	0.61

Cases with definite & probable diagnosis to date.
[2015 Population Source](#)

Source: PHAC, 2016.

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**EXHIBIT 3
CJD Deaths Reported by the CJD Surveillance System (1994-2016)
As of June 30, 2016**

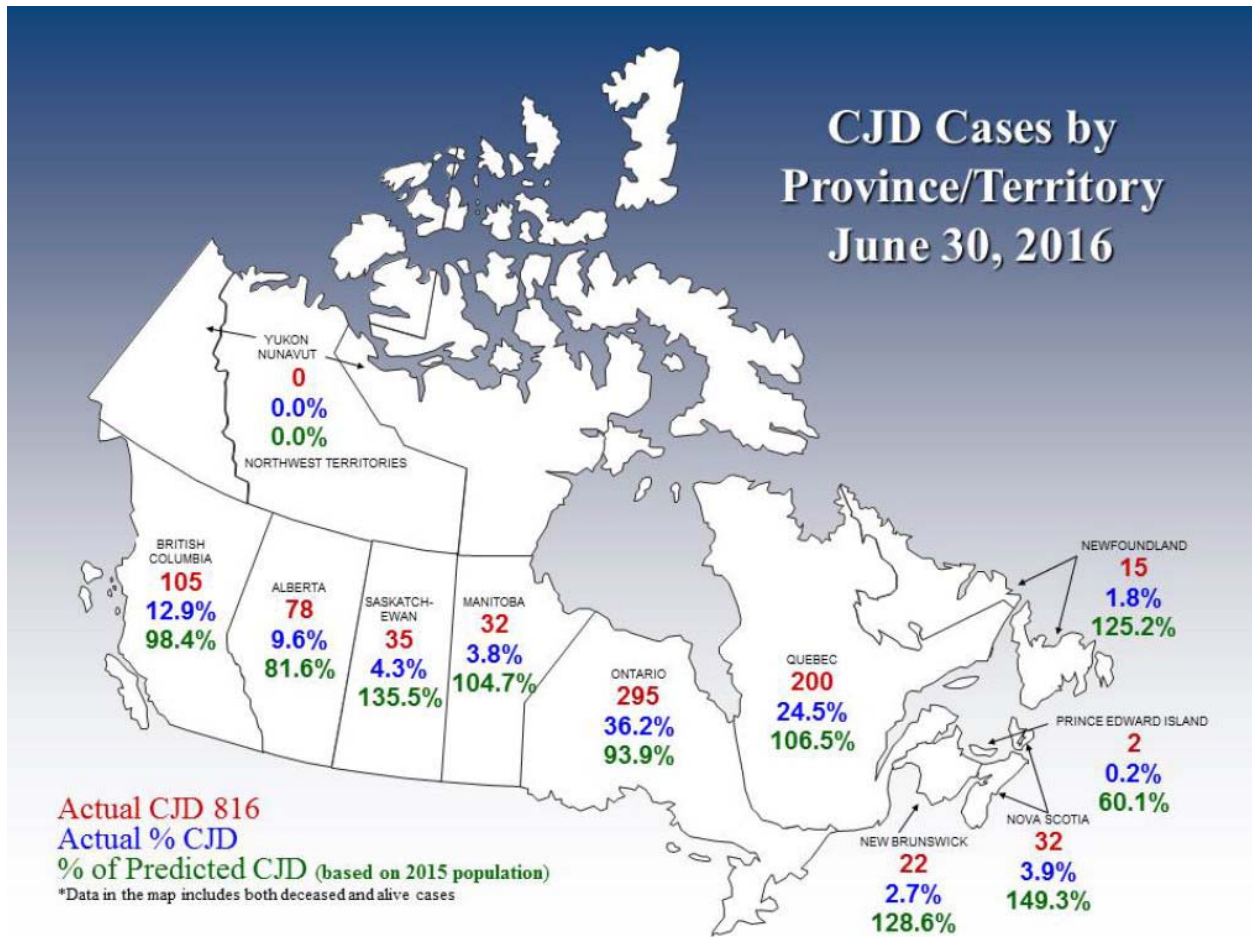
Deaths of Definite and Probable CJD							
Year	Sporadic	Iatrogenic	Familial	GSS	FFI	vCJD	Total
1994	2	0	0	1	0	0	3
1995	3	0	0	0	0	0	3
1996	13	0	0	0	0	0	13
1997	16	0	1	1	0	0	18
1998	22	1	0	1	0	0	24
1999	27	2	2	1	0	0	32
2000	32	0	0	3	0	0	35
2001	27	0	2	1	0	0	30
2002	31	0	2	2	0	1	36
2003	27	1	1	0	0	0	29
2004	42	0	1	1	0	0	44
2005	42	0	1	1	0	0	44
2006	39	0	1	3	1	0	44
2007	35	0	0	4	0	0	39
2008	48	0	1	0	0	0	49
2009	48	0	3	2	0	0	53
2010	35	0	3	0	0	0	38
2011	46	0	3	1	0	1	51
2012	62	0	1	0	0	0	63
2013	50	0	0	0	1	0	51
2014	50	0	4	0	1	0	55
2015	44	0	4	1	2	0	51
2016	10	0	1	0	0	0	11
Total	751	4	31	23	5	2	816

Cases with definite & probable diagnosis to date.

Source: PHAC, 2016.

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**EXHIBIT 4
Map of CJD Cases in Canada
As of June 30, 2016**



Source: PHAC, 2016.

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EXHIBIT 5 Case Definitions of CJD

At time of death, all possible CJD cases in Canada are classified according to the WHO diagnostic criteria for CJD:

Definite CJD

- Requires neuropathologic examination
- Confirmed by cranial autopsy

Probable CJD

- Case is lacking neuropathologic examination
- Based on combination of non-neuropathologic criteria:
 - Clinical profile (ataxia, rapid progressive dementia, etc.)
 - Results of cerebrospinal fluid (CSF) protein immunoassays
 - EEG
 - MRI
 - Genetic analysis
 - Brain/tonsil biopsy

Possible CJD

- Information is even more limited.

Source: WHO, 2008.

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**EXHIBIT 6
CJD Tissue Infectivity**

High Infectivity	Low Infectivity
<ul style="list-style-type: none"> - Brain - Cerebrospinal fluid - Dura mater - Pituitary gland - Posterior eye - Spinal cord and ganglia - Trigeminal ganglia 	<ul style="list-style-type: none"> - Cornea - Kidney - Liver - Lung - Lymph nodes - Placenta - Spleen
No Detected Infectivity	
<ul style="list-style-type: none"> - Adipose tissue - Adrenal gland - Appendix - Blood (including cord blood) - Blood vessels - Bone marrow - Breast milk - Dental pulp - Epididymis - Esophagus - Feces - Gingival tissue - Heart - Ileum - Jejunum - Large intestine - Nasal mucosa - Nasal mucous - Ovary 	<ul style="list-style-type: none"> - Pancreas - Pericardium - Peripheral nerves - Placental fluids - Prostate - Saliva - Semen - Seminal vesicle - Skeletal muscle - Skin - Sweat - Tears - Testis - Thymus - Thyroid gland - Tongue - Tonsil - Trachea - Urine

Source: PHAC, 2007.

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EXHIBIT 7 Recommended Laboratory Testing for CJD

Upon initial intake of a patient, it is recommended that the following tests be conducted in order to confirm or rule-out the possibility of CJD:

1. CEREBROSPINAL FLUID (CSF) TESTING

In all cases of CJD, CSF testing is highly recommended. CSF is collected via lumbar puncture and is tested for levels of various proteins. Some protein tests that are conducted using CSF include 14-3-3, S100B, and tau protein tests (Coulthart et al., 2011). Elevated levels of these proteins have been reported to support a CJD diagnosis. However, each test has varying levels of sensitivity and specificity, and the threshold limits of each protein can differ from patient to patient. Currently, CSF tau shows the best overall diagnostic accuracy, compared to 14-3-3 or S100B protein markers (Coulthart et al., 2011).

2. COMPUTERIZED TOMOGRAPHY (CT) SCANS

Computerized tomography (CT) scans of the brain can sometimes rule out the possibility that exhibited symptoms may have resulted from another neurological issue including strokes or tumours in the brain (WHO, 2008).

3. ELECTROENCEPHALOGRAPHY (EEG)

Electroencephalography (EEG) measures waves of electrical activity in the brain. This test can be valuable as certain sharp wave complexes are characteristic in sporadic CJD. However, EEG is not useful in the diagnosis of vCJD (WHO, 2008).

4. MAGNETIC RESONANCE IMAGING (MRI)

Magnetic resonance imaging (MRI) brain scans also can reveal characteristic patterns of brain degeneration that can help diagnose CJD (WHO, 2008).

5. GENETIC TESTING

Blood samples can be used to prepare DNA for genetic analysis. Specific mutations in the PRNP gene, including P102L, E200K and D178, are CJD-causing variations. However, some variations are silent, and do not cause disease – for example, M129V (Swietnicki et al., 1998).

6. TONSIL BIOPSY

Prion proteins can often be found in tonsil tissues in CJD-positive patients. However, this test is not useful in diagnosing sporadic or genetic CJD (WHO, 2008).

7. BRAIN AUTOPSY

While the results of the aforementioned six diagnostic tests may lead to a strong diagnosis of CJD, the only definite method of knowing whether the patient has CJD is through brain autopsy at time of death. However, autopsy is a lengthy process, requiring several months to complete. Some characteristic changes in a CJD-positive brain include:

- Spongiform change
- Loss of neurons
- High astrocyte density
- Presence of abnormal prion proteins

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**EXHIBIT 8
Case Characteristics and Test Results**

Case #	Age at Onset	CSF Results	EEG	MRI	Status
1	42	Positive	Negative	Negative	Alive
2	25	Positive	Negative	Positive	Dead
3	33	Pending	Unknown	Unknown	Dead
4	44	Positive	Negative	Pending	Dead
5	36	Pending	Negative	Pending	Alive

Source: Created by author.

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INSTRUCTOR GUIDANCE

Investigating Suspected Outbreaks of Rare Infectious Disease Using Surveillance Data: The CJDSS Perspective

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BACKGROUND

A suspected cluster of variant Creutzfeldt-Jakob Disease (CJD) cases (a fatal neurodegenerative disease) has been identified in Toronto. Renée Dubois, a public health nurse, must utilize epidemiological data to recognize the outbreak, and mobilize resources to support patients and health care providers. The assembled epidemiological information gathered by the CJD surveillance system must be used to further support data analysis, public health decision-making, and public messaging. This case may be used to supplement lectures covering relevant public health events or epidemic analysis.

OBJECTIVES

1. Understand the importance of case reporting and laboratory data for surveillance of infectious diseases and outbreak detection.
2. Identify limitations of diagnostic tests and laboratory data, as well as the importance of ethical guidelines and informed consent in patient sampling.
3. Devise an emergency preparedness management strategy in the event of a communicable disease outbreak.
4. Interpret epidemiological data to understand the spread, origin, and effects of communicable disease.
5. Interpret laboratory data in context of epidemiological data in order to make recommendations for timely public health action.

DISCUSSION QUESTIONS

1. What is an outbreak?
 - a. What are some modern examples of outbreaks?
2. How can you recognize an outbreak?
3. What sources of information would you consult to confirm that it is an outbreak?
 - a. How would you access this information?
 - b. What are the limitations of diagnostic tests and laboratory data?
4. What information is most relevant for public health decision-making?
5. What is the role of media in outbreak investigation and communication?
 - a. How do you ensure confidentiality in a high profile outbreak?
 - b. What is the public message you would communicate, and what would that look like?
 - Relate this to current public health events.

KEYWORDS

Epidemiology; Creutzfeldt-Jakob Disease (CJD); outbreak management; surveillance data; PHAC.