Creutzfeldt-Jakob Disease, Classic (CJD)

What is Classic CJD?
Classic Creutzfeldt-Jakob disease (CJD) is a rare, fatal, degenerative brain disease caused by abnormal, transmissible proteins called prions.

There are three types of classic CJD: (1) Sporadic CJD, (2) Familial CJD, and (3) Iatrogenic CJD. Sporadic CJD occurs occasionally with no known cause. It accounts for approximately 85-90% of diagnosed classic CJD cases. Familial CJD is an inherited form of CJD that occurs in families. Familial CJD accounts for approximately 10-15% of diagnosed classic CJD cases. Iatrogenic CJD occurs in a patient who was infected during a medical or surgical procedure. Iatrogenic CJD accounts for less than 1% of diagnosed classic CJD cases.

Classic CJD occurs at a rate of approximately one case per 1 million per population per year. The risk of CJD increases with age, and in persons over 50 years of age, the annual rate is approximately 3.4 cases per million.

How is classic CJD transmitted?
The risk of CJD is low. The disease can't be transmitted through coughing or sneezing, touching or sexual contact. The three ways it develops are: (1) spontaneously, (2) by genetic mutation, and (3) by contamination.

A small number of people have developed CJD after being exposed to infected human tissue during certain medical procedures, such as cornea or skin transplants and dura mater grafts from infected donors. There have also been documented cases of CJD for persons receiving injections of pituitary hormones from infected cadavers. Also, because standard sterilization methods do not destroy abnormal prions, a few people have developed CJD after undergoing brain surgery with contaminated instruments and after being exposed to contaminated electrodes used during electroencephalographic (EEG) procedures.

All equipment-related cases of CJD occurred before the routine implementation of sterilization procedures currently used in healthcare facilities. There have been no such cases reported since 1976, and no iatrogenic CJD cases associated with exposure to the CJD agent from surfaces such as floors, walls, or countertops have been identified.

What are the symptoms of classic CJD?
Symptoms of CJD are marked by rapid mental deterioration, usually within a few months. Initial signs and symptoms of CJD include:

- personality changes
- anxiety
- depression
- memory loss
- impaired thinking
- blurred vision
- insomnia
- difficulty speaking
- difficulty swallowing
- sudden jerky movements

As the disease progresses, mental symptoms worsen. Most people eventually lapse into a coma. Heart failure, respiratory failure, pneumonia or other infections are generally the cause of death.

The disease usually runs its course in about seven months, although a few people may live up to one or two years after diagnosis. There is no cure for CJD; the disease is ultimately fatal.

Are certain people at risk for getting classic CJD?
Most cases of CJD disease occur for unknown reasons, and no risk factors can be identified. However, a few factors seem to be associated with different kinds of CJD and these include age (associated with Sporadic CJD), genetics (associated with Familial CJD), and exposure to contaminated tissues (associated with Iatrogenic CJD).

What is the treatment for classic CJD?
Symptoms of the disease are treated, but there is no treatment available that slows or stops the disease.

How can classic CJD be prevented in the healthcare setting?
The World Health Organization (WHO) has developed CJD infection control guidelines (http://www.who.int/csr/resources/publications/bse/WHO_CDS_CSR_APH_2000_3/en/) that can be a valuable guide for healthcare workers involved in the care of CJD patients. The WHO guidelines provide guidance upon which infection control practitioners, healthcare workers, medical officers, and all those involved with the care of persons infected with CJD can base their care and infection control practices.

For more information regarding CJD, visit the following resources:

- http://www.cdc.gov/ncidod/dvrd/cjd/index.htm
- http://www.cdc.gov/ncidod/dvrd/cjd/qa_cjd_infection_control.htm
- CJD Infection Control Guidelines (WHO) - http://www.who.int/csr/resources/publications/bse/WHO_CDS_CSR_APH_2000_3/en/