Chicago Department of Public Health

CD Info

City of Chicago
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Immunization and Communicable Disease Programs

Chicago Department of Public Health
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CD Info is a surveillance newsletter intended to promote prevention of morbidity and mortality by providing useful data and practical recommendations for clinicians, laboratorians, and infection control personnel who diagnose, treat or report infectious diseases in Chicago. (Written by Saadeh Ewaidah, MD, MPH) September 7th, 2016.

Creutzfeldt - Jakob Disease

Background

Creutzfeldt-Jakob disease (CJD) belongs to the family of diseases known as transmissible spongiform encephalopathies (TSEs). TSEs are also known as prion-related diseases because they are thought to be caused by abnormal forms of a host protein, termed prion protein. There are three types of CJD in humans: sporadic or classic (85%), familial (15%), and acquired [iatrogenic and variant] (1%).[1]

Surveillance

CJD is the most common form of TSE in humans, with an estimated annual incidence of 1 case/1 million population worldwide. Persons ≥ 50 years old have a higher risk of CJD with an average annual rate of approximately 3.4 cases per million.[1]

CJD became reportable in Illinois effective March 3, 2008. From 2008 to 2015 the Chicago Department of Public Health (CDPH) received 75 CJD suspected case reports. Of these 75 reported cases, eight were confirmed and four were probable (Figure). As of 2014, only confirmed CJD cases are reportable to Illinois Department of Public Health. Confirmed cases have pathologic evidence of disease and probable cases have clinical, laboratory and radiographic features consistent with CJD. (http://www.cdc.gov/prions/cjd/diagnostic-criteria.html)

Of the 12 confirmed and probable cases, 7 (58%) cases were female. The age range was 51 -72 years, with a median age of 62 years. Regarding race/ethnicity, seven cases were white (58%), three cases (25%) were Hispanic, and one case each (8%) was black, and Asian/Pacific Islander. Based on pathology results reported from the National Prion Disease Pathology Surveillance Center of the eight confirmed cases, three were sporadic, two were familial, and three had prion protein present consistent with CJD, but type was undetermined**.**

Clinical Course

The incubation period for CJD ranges from years to decades.[3] Patients with sporadic CJD present with rapidly progressive dementia, visual or speech abnormalities or cerebellar dysfunction, including loss of muscle coordination and gait abnormalities. During the course of the disease most patients develop abnormal reflexes, spasticity, tremors, and rigidity. Patients may show behavioral changes including agitation, depression, or confusion.[4] CJD is invariably fatal with a median illness duration of four months (mean of 7.6 months). Most CJD cases (85-90%) expire within 12 months of illness onset. There is no treatment for CJD.[1]

Infection-Control Guidelines

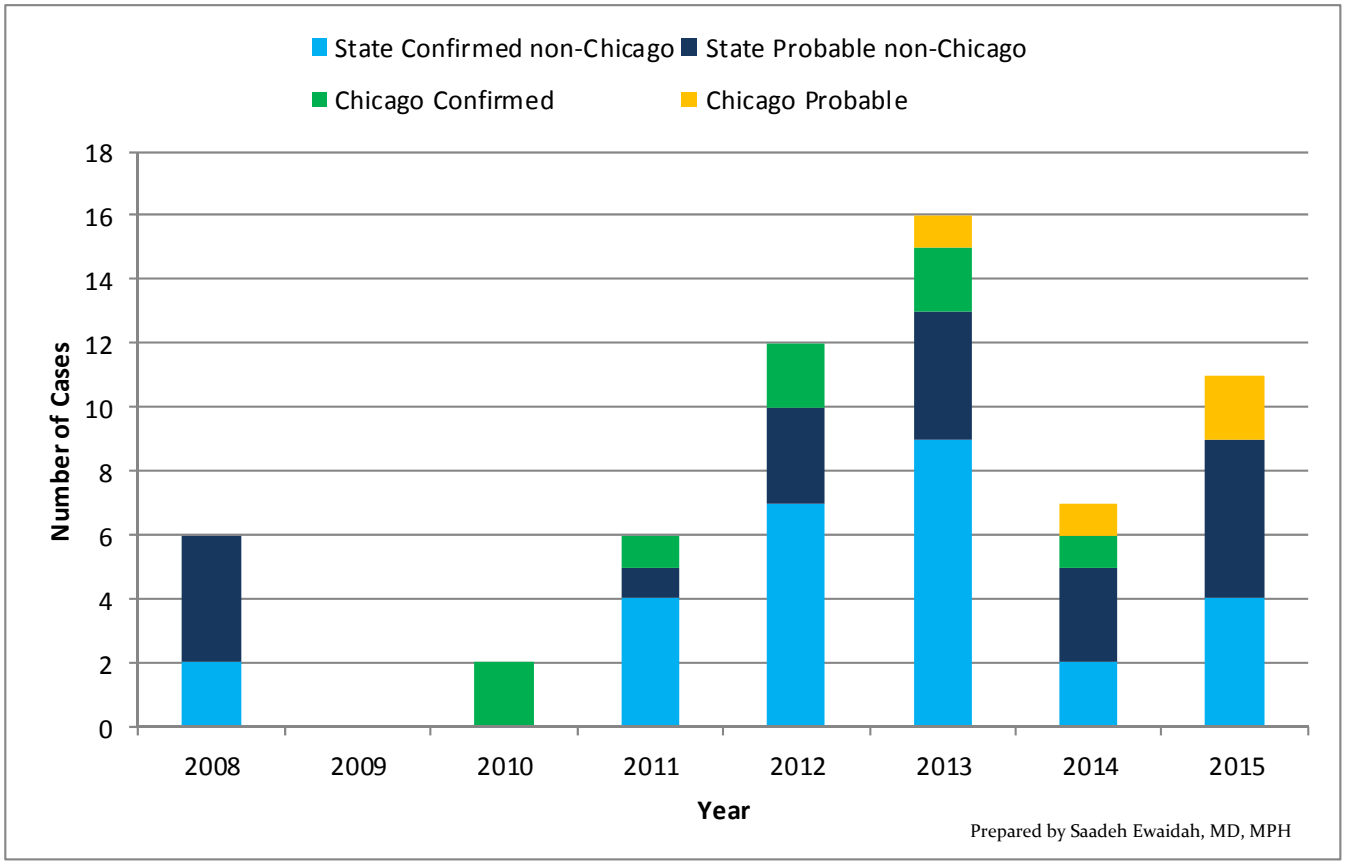
Investigations of potential CJD cases by CDPH have demonstrated a need for facilities to review policies and procedures pertaining to TSE. Published literature describes iatrogenic CJD exposures linked to use of cadaveric pituitary hormones, dura mater, corneal grafts and neurosurgical instruments.[5] Four of six cases linked to neurosurgical procedures from 1950s-1976 were related to contaminated neurosurgical instruments and two were related to implantable electrodes.[5] Conventional chemical and physical methods of decontamination are insufficient for reprocessing instruments potentially contaminated with TSEs. The World Health Organization (WHO) has developed TSE infection control guidelines that provide a framework to determine infectivity risk for patient tissues, secretions and excretions. Specific recommendations address patient care, occupational exposures, laboratory investigations, decontamination procedures, waste disposal, and precautions after death. No TSE patient should be denied admission to a health facility, kept in isolation, or deprived of an indicated procedure.[7]

In a review of management of neurosurgical instruments and patients exposed to CJD, over half of the procedures were likely performed as diagnostic work-up for the patients.[3] Infection preventionists should review infection control policies for intracranial diagnostic procedures. Providers evaluating patients with acute neurodegenerative illness of undetermined etiology should consider CJD in the differential diagnosis and plan ahead to use appropriate reprocessing measures for any instruments used during diagnostic procedures. Instruments should be kept moist and promptly cleaned and sterilized according to WHO guidance[6] or quarantined until an alternative diagnosis is identified. Timely communication between physicians, the operating room staff, central sterilization and infection preventionists is critical to ensure appropriate instrument handling.[3] In addition, facilities should consider use of an instrument tracking procedure in order to trace potentially contaminated neurosurgical instruments linked to an identified case, should the need arise. Instruments from neurosurgical sets should not be mixed with those from general surgical sets.[3]

Reporting

Creutzfeldt-Jakob Disease (CJD) is reportable within 7 days of identification. Clinicians and infection control practitioners should report suspect cases in Chicago by calling the CDPH Communicable Disease Program at (312) 746-5916. Laboratories should report patients who have a positive result on any laboratory test indicative of and specific for detecting CJD. Clinical specimens should be forwarded to the National Prion Disease Pathology Surveillance Center for disease confirmation.[7,8]

Figure. Confirmed and probable cases of Creutzfeldt - Jakob Disease (CJD) among Chicago and Illinois residents by year, 2008-2015.



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