EPIDEMIOLOGY AND IMMUNIZATION SERVICES BRANCH

MONTHLY COMMUNICABLE DISEASE REPORT

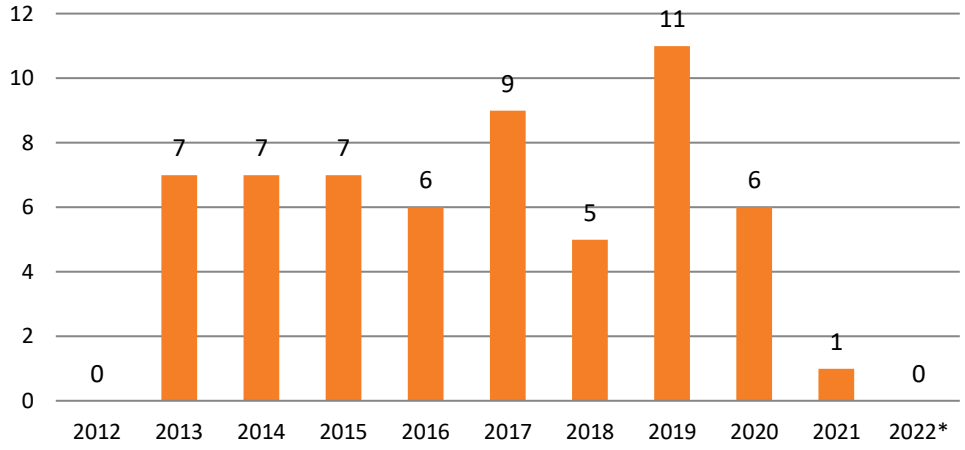
JANUARY 2022

Volume 6, Issue 1: February 16, 2022

Creutzfeldt-Jakob Disease

Creutzfeldt-Jackob Disease (CJD) is a rare, but rapidly progressive and always fatal neurodegenerative disorder belonging to a family of human and animal diseases known as transmissible spongiform encephalopathies (TSEs) or prion disease. It is believed that the etiologic agent of TSEs are abnormal conformers of a host-encoded cellular protein known as the prion protein. The normal prion protein is naturally found throughout the body and is abundant in the nervous system.

Figure 1. Creutzfeldt-Jakob Disease Cases, San Diego County, 2012-2022*



*Year to date. 2022 includes data through 1/18/2022. Includes suspect, probable, and confirmed cases following CDPH case criteria. Death defined as person reported dead at time of case closure. Data are provisional and subject to change as additional information becomes available. Grouped by CDC disease years.

CJD is the most common form of TSEs. The disease is characterized by long incubation periods with spontaneous onset of rapidly progressing dementia, visual disturbance, inability to speak (akinetic mutism), tremors, sudden and involuntary jerking of a muscle or group of muscles (myoclonus), spasticity, rigidity, and difficulties with balance and coordination. Death typically occurs within one year after disease onset.

There are three major categories of CJD: sporadic (identified in approximately 85% of patients), hereditary or familial (identified in approximately 10-15% of patients), and acquired or iatrogenic (identified in less than 5% of patients). Mode of transmission for CJD differs by category. Sporadic CJD (sCJD) has no recognizable pattern of transmission, and it is defined as spontaneous transformation of normal proteins to infectious prion proteins. Familial CJD (fCJD) is believed to be developed through inherited mutations of the prion protein gene. Iatrogenic CJD (iCJD) has been linked to exposure to infectious material through use of human cadaveric-derived pituitary hormones, dural and cornea homografts, and contaminated neurosurgical equipment during invasive medical interventions. The last reported case of iCJD acquired through contaminated equipment was reported in 1976.

There is also a very rare form of prion disease, variant CJD (vCJD), first recognized in the United Kingdom in 1996, that is associated with bovine spongiform encephalopathy (BSE) or “mad cow” disease. There is strong scientific evidence that vCJD transmission is through consumption of BSE infected cattle products. To date in the United States (U.S.), there have only been four cases of vCJD and all four cases were exposed abroad.

Continued on next page

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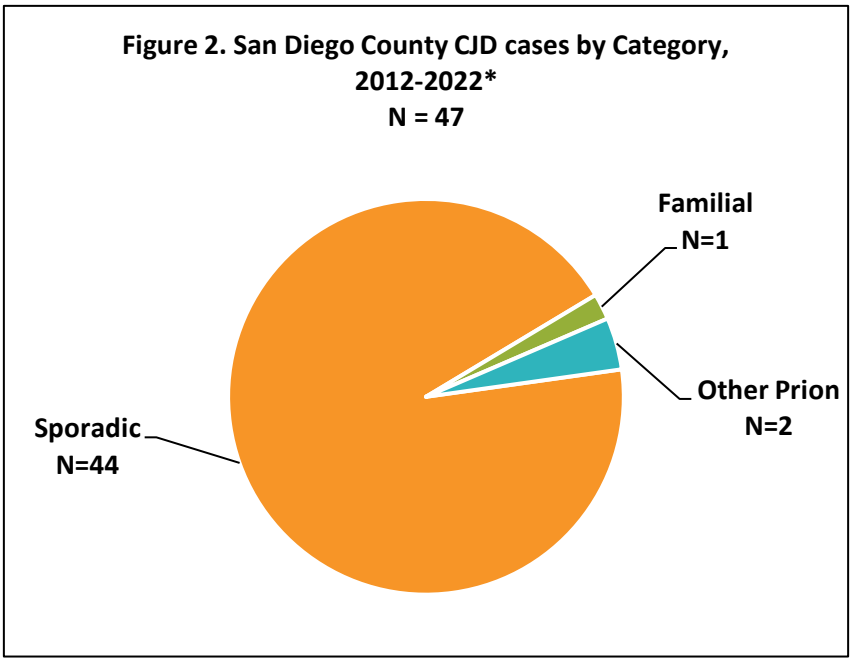
Creutzfeldt-Jakob Disease, continued

Enhanced surveillance of TSEs, including CJD, in California was initiated in 1999 by the Centers for Disease Control and Prevention (CDC) and the California Department of Public Health (CDPH); it became a notifiable condition at the state level in 2007. Currently, CJD is not a nationally notifiable disease. sCJD occurs in the United States at a rate of roughly 1 to 1.5 cases per 1 million population per year, with risk increasing with age. The CDC estimated the 1970-2019 U.S. sCJD average annual rate to be 3.6 cases per million in persons 50 years of age or older.



*Year to date. 2022 includes data through 1/18/2022. Cases that did not have category information were excluded in this graph (N=12). Data are provisional and subject to change as additional information becomes available. Grouped by CDC disease years.

Annual case counts for CJD are relatively low in San Diego County (Figure 1) and nationally. In 2019, an estimated total of 561 CJD deaths were reported in the United States. In San Diego County, preliminary numbers for 2021 indicate only one reported case. Among the San Diego County cases where a CJD category could be identified, 92% were sCJD (Figure 2). Over 85% of San Diego County CJD cases had died due to the illness at time the investigation had been completed. Approximately 50% of San Diego County CJD cases were male; the median age was 66 years.

There are several methods to help diagnose CJD: electroencephalograms (EEG), cerebrospinal fluid-based tests (i.e., Real Time-Quaking-Induced Conversion (RT-QuIC)), and magnetic resonance images (MRI). However, there is only one way to confirm diagnosis of CJD: a brain biopsy or autopsy. Due to the dangers involved with brain biopsies this procedure is discouraged unless it is needed to rule out a treatable disorder.

Currently, treatment of prion diseases remains supportive, and no specific therapy has been shown to stop the progression of these diseases. There are infection control guidelines for iCJD that have been developed by the CDC and the World Health Organization (WHO) for infection preventionists and health care workers involved in caring for CJD patients.

Federal Resources

- Centers for Disease Control and Prevention (CDC) Creutzfeldt-Jakob Disease website
- National Institute of Neurological Disorders and Stroke (NIH) Creutzfeldt-Jakob Disease website

State Resources

- California Department of Public Health Creutzfeldt-Jakob Disease website

Suggested citation: Dionicio M, Maroufi A. Creutzfeldt-Jakob Disease. County of San Diego Monthly Communicable Disease Report 2022; 6(1):1-2.

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Table 2. Select Reportable Diseases		2022			Prior Years		
Disease and Case Inclusion Criteria (C,P,S)		Current Month	Prior Month	Year-to-Date (YTD)	2021 YTD	Avg YTD, Prior 3 Years	2021 Total
Botulism (Foodborne, Infant, Wound, Other)	C,P	0	0	0	0	0.0	3
Brucellosis	C,P	1	0	1	0	0.0	3
Campylobacteriosis	C,P	45	58	45	33	60.7	905
Chickenpox, Hospitalization or Death	C,P	0	0	0	0	0.3	3
Chikungunya	C,P	0	1	0	0	0.0	2
Coccidioidomycosis	C	0	22	0	55	50.7	452
Cryptosporidiosis	C,P	2	4	2	0	3.3	53
Dengue Virus Infection	C,P	0	0	0	0	0.7	2
Encephalitis, All	C	0	3	0	6	5.0	34
Giardiasis	C,P	15	12	15	13	16.0	160
Hepatitis A, Acute	C	0	0	0	0	1.3	10
Hepatitis B, Acute	C	2	0	2	0	1.0	16
Hepatitis B, Chronic	C,P	82	75	82	64	69.0	814
Hepatitis C, Acute	C,P	0	0	0	10	8.0	61
Hepatitis C, Chronic	C,P	160	158	160	373	382.0	3,497
Legionellosis	C	10	8	10	8	5.3	62
Listeriosis	C	0	2	0	0	0.3	8
Lyme Disease	C,P	0	0	0	2	1.0	3
Malaria	C	0	0	0	0	1.0	8
Measles (Rubeola)	C	0	0	0	0	0.0	0
Meningitis, Aseptic/Viral	C,P,S	3	2	3	3	6.7	46
Meningitis, Bacterial	C,P,S	3	2	3	4	4.3	19
Meningitis, Other/Unknown	C	0	5	0	3	2.7	24
Meningococcal Disease	C,P	0	0	0	0	1.0	1
Mumps	C,P	1	1	1	0	1.7	2
Pertussis	C,P,S	2	5	2	5	46.3	68
Rabies, Animal	C	0	0	0	1	0.7	4
Rocky Mountain Spotted Fever	C,P	0	0	0	0	0.0	2
Salmonellosis (Non-Typhoid/Non-Paratyphoid)	C,P	22	30	22	20	30.3	581
Shiga toxin-Producing *E. coli* (including O157)	C,P	6	14	6	8	10.0	133
Shigellosis	C,P	26	35	26	8	29.7	430
Typhoid Fever	C,P	3	0	3	0	1.7	9
Vibriosis	C,P	2	2	2	0	1.7	51
West Nile Virus Infection	C,P	0	0	0	0	0.0	3
Yersiniosis	C,P	6	2	6	2	2.0	21
Zika Virus	C,P	0	0	0	0	0.0	0

Case counts are provisional and subject to change as additional information becomes available. Cases are grouped into calendar months and calendar years on the basis of the earliest of the following dates: onset, lab specimen collection, diagnosis, death, and report received. Counts may differ from previously or subsequently reported counts due to differences in inclusion or grouping criteria, late reporting, or updated case information. Inclusion criteria (C,P,S = Confirmed, Probable, Suspect) based on Council of State and Territorial Epidemiologists/Centers for Disease Control and Prevention (CSTE/CDC) surveillance case criteria.

Figure 3. Select Enteric Infections by Month
February 2021 – January 2022

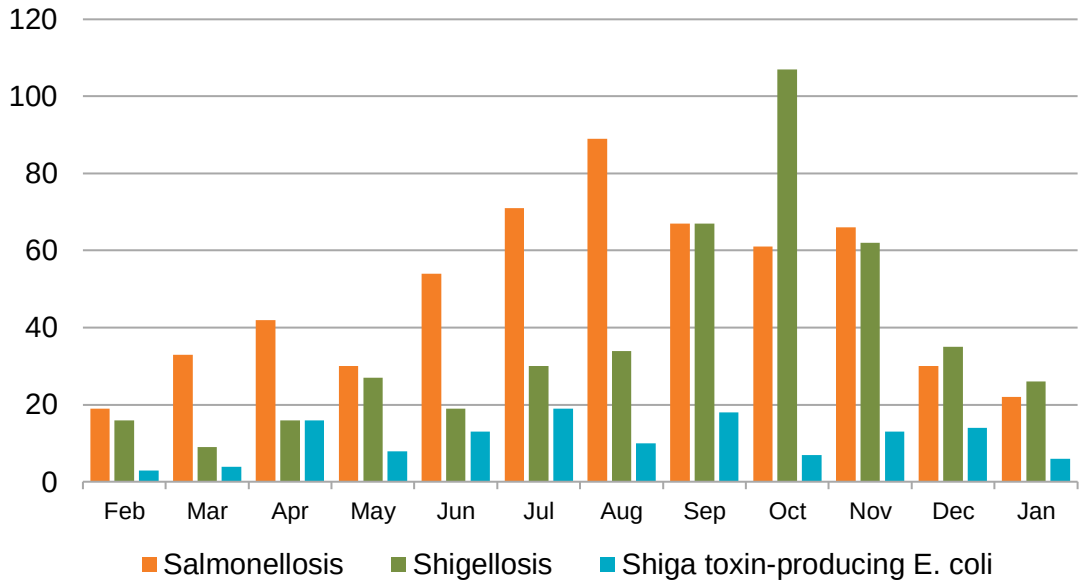

Figure 4. Select Vaccine-Preventable Infections by Month
February 2021 – January 2022

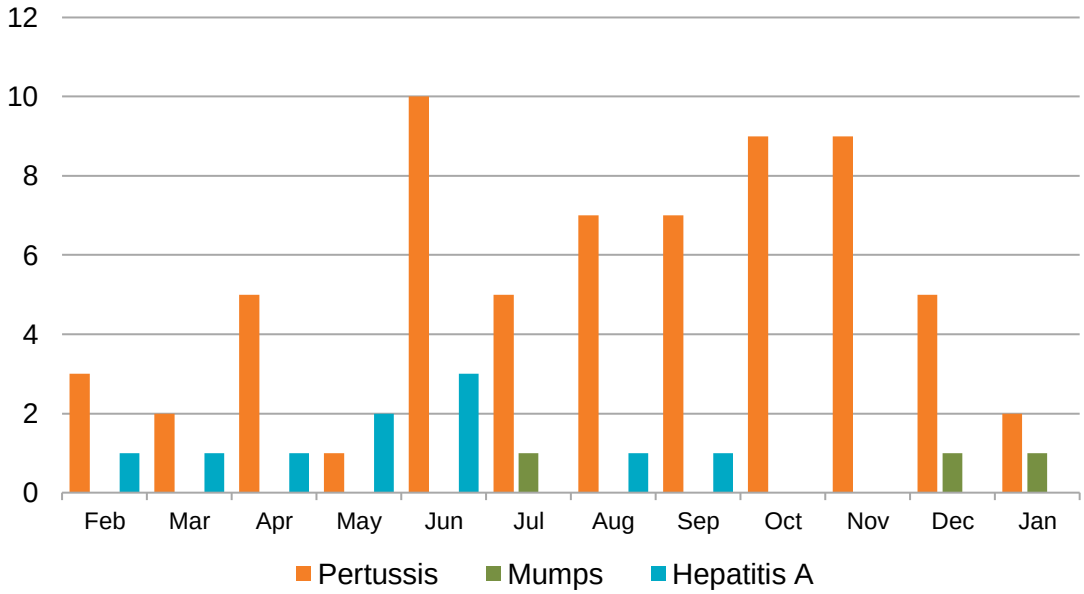

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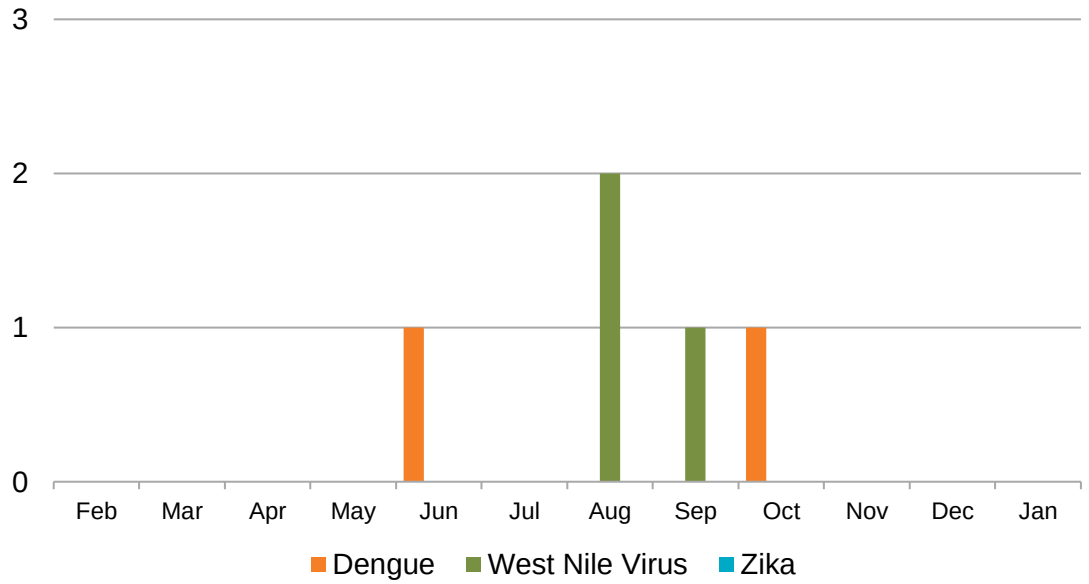
Figure 5. Select Vector-Borne Infections by Month
February 2021 – January 2022



All of the dengue and Zika virus cases are travel-associated. For additional information on Zika cases, see the HHSA Zika Virus webpage. For more information on West Nile virus, see the County West Nile virus webpage. ***Case counts are provisional and subject to change as additional information becomes available.*** Cases are grouped into calendar months and calendar years on the basis of the earliest of the following dates: onset, lab specimen collection, diagnosis, death, and report received. Counts may differ from previously or subsequently reported counts due to differences in inclusion or grouping criteria, late reporting, or updated case information. Inclusion criteria (C,P,S = Confirmed, Probable, Suspect) based on Council of State and Territorial Epidemiologists/Centers for Disease Control and Prevention (CSTE/CDC) surveillance case criteria.

Disease Reporting in San Diego County

San Diego County communicable disease surveillance is a collaborative effort among Public Health Services, hospitals, medical providers, laboratories, and the San Diego Health Connect Health Information Exchange (HIE). The data presented in this report are the result of this effort.

Reporting is crucial for disease surveillance and detection of disease outbreaks. Under the California Code of Regulations, Title 17 (Sections 2500, 2505, and 2508), public health professionals, medical providers, laboratories, schools, and others are mandated to report more than 80 diseases or conditions to San Diego County Health and Human Services Agency.

To report a communicable disease, contact the Epidemiology Program by phone at (619) 692-8499 or download and print a Confidential Morbidity Report form and fax it to (858) 715-6458. For urgent matters on evenings, weekends or holidays, dial (858) 565-5255 and ask for the Epidemiology Program duty officer. For more information, including a complete list of reportable diseases and conditions in California, visit the Epidemiology Program website, www.sdepi.org.

Tuberculosis, sexually transmitted infections, and HIV disease are covered by other programs within Public Health Services. For information about reporting and data related to these conditions, search for the relevant program on the Public Health Services website, http://www.sandiegocounty.gov/content/sdc/hhsa/programs/phs.html.