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REVIEW ON CREUTZFELDT-JAKOB DISEASE

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ABSTRACT

Creutzfeldt jakob disease is an extremely rare degenerative brain disorder it affects about one in every million people for AIR Worldwide people with cjd typically develop symptoms later in life and may show changes in behaviour memory troubles lack of coordination and vision problems as the disease progresses there are many rapidly progressive deterioration of mental functioning memory and muscle control cjd is a fatal disease in most affected individuals life-threatening complications develop less than a year after the start showing symptoms .[14,15]

KEYWORDS: Definition, introduction, epidemiology, etiology, signs and symptoms, Therapy.

INTRODUCTION[1,2,3]

Cruetzfeldt jakob disease is classified as familial sporadic or acquired regardless of the type the disease has a rapid clinical course that is uniformly fattal there are a some consistencies on physical examination radiographic studies and electroencephalogram but the most common form is sporadic cjd and it follows the theme of heterogenous ATI the infectious agent is normally scrapy form of the host encoded CellularTree on protein that causes a post translational modification in to the disease from accumulating in the brain and causing neurodegeneration.

Cjd is a red neurodegenerative condition with the Rapid disease course and a mortality rate of hundred percent several forms of disease have been described and the most common is the sporadic type the most challenging aspect of this disease in is it diagnosis the gold standard of Definite diagnosis is considered to be histopathological confirmation with New alternative test are providing means for an alternate diagnosis invasive less invasive than brain biopsy.[16,19]

The human Prion disorders are heterogeneous with different phenotypes epidemiology and pathogenesis sporadic cjd is a common is human Prion disease accounting for around 85% of cases 10 to 15% are associated with mutations of p r and p and 1% are Iatrogenic most frequently associated with their treatment with human pituitary derived hormones or human Dura matter grafts. Variant cjd is a normal human Prion disease which occurs predominantly in the UK and has been linked to the consumption of beef beef products contaminated with the agent of cattle disease bovine spongiform encephalopathy.[2,16]

There have been recent developments in Diagnostic investigations in cjd and although the variant cjd outbreak is in decline there are continuing concerns for public health in relation to the prevalence of infection in the normal population these issues and the potential relevance of Prion disease to other neurodegenerative disorders are the main topics of this article.[12,17]

DEFINITION[3,4,5]

Cjd is a fatal disease presenting with rapidly progressive dementia and most patients die within a year of clinical onset cjd process a potential risk of iatrogenic Transmission as it can incubate asymptomatically in humans for decades before becoming clinically Apparent.

EPIDEMIOLOGY - globally cjd incident data are gathered by cjd international surveillance network Euro cjd however this online source was last updated in May 2015 data obtained through personal communication with Euro cjd what dated to 2018 UK referrals are suspected definite or probable cjd related deaths are covered by the national cjd research and surveillance unit this source estimates that since 1990 There are have been 3873 UK referrals for investigation and 2541 debts of Definite or probable cjd as of Jan 31 2019 the Global incidence of cjd is reported to be around 122 cases per million per year on the basis of surveillance studies published from 2005 onwards reports are increased incidence might be more probable in areas with access to establish a surveillance units for referring suspected cases of the Prion disease.[1,2,3,4,5]

ETIOLOGY

Cjd is rapidly progressive invariably fattal neuro degenerative disorder believed to be caused by an

abnormal isoform of cellular protein glycoprotein known as the Prion protein.[2,13]

CLINICAL CHARACTERISTICS

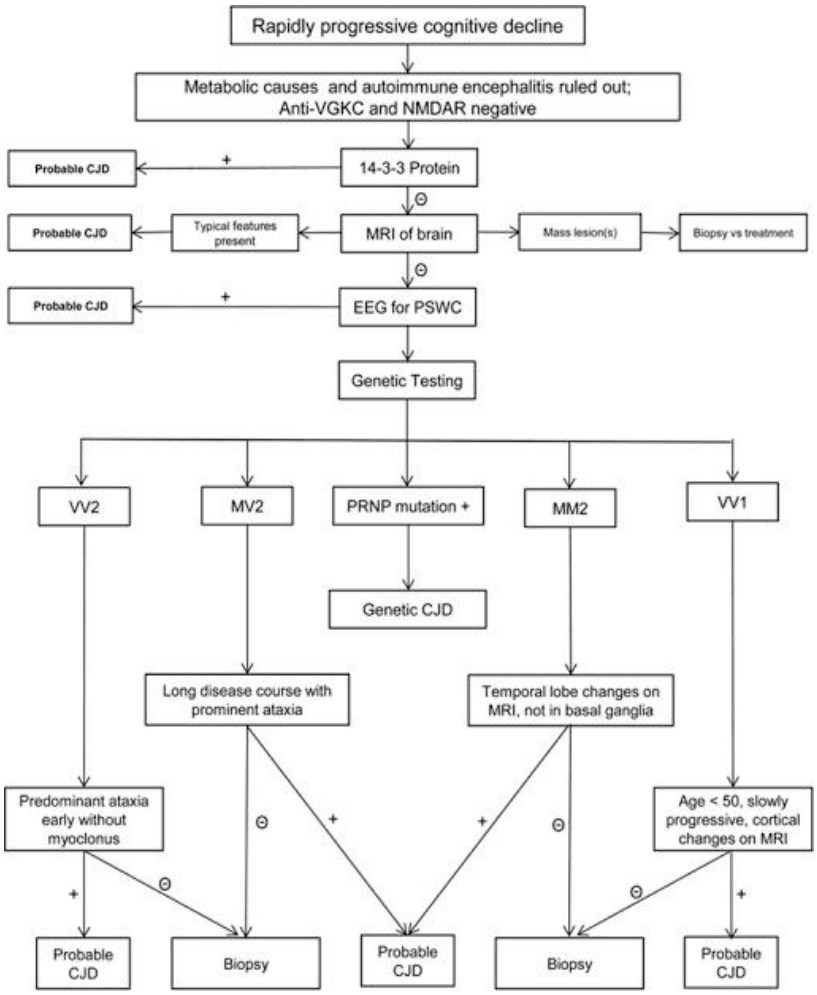
Median age of death – 68yrs
Median duration of illness – 4-5 months.[1]

CLINICAL SIGNS AND SYMPTOMS

- Dementia
- Early neurological sign
- Prominent psychiatric behavioral symptoms
- Painful diastasis.
- Periodic sharp waves on electroencephalogram are often present.[5,6]

PATHOPHYSIOLOGY[7,8,9]

- To understand some of the clinical findings of this disease one must first TV reporter officially of how healthy protein becomes abnormal and destructive to the brain prpc is found in lipid raft of the cell surface on normal brains the function of the protein is unknown on protein knockout mice completely lacking the protein do not show any obvious abnormality and have normal brain development.
- The central pathological event is a formation of the abnormal P RPSC from the wild type cellular form of prpc this is hypothesized to occur in a pathway where PRP SSC serves as the template for prpc to fold abnormally into the pathogenic confirmation this is auto catalytic process that is poor understood but the change in the protein shape is the Hallmark of the Pathology.[20]
- Both forms have an identical Amino acid sequence primary structure but the post translational changes caused the CrPC 40% Alpha helix to refill into a form with 45% beta sheet composition this makes the protein not only highly insoluble but also resistant to protein is digestion throughout the brain parenchyma and include the Classic fungiform change collation of Grey matter by micro logical activation and neuronal loss during to the progressive neurodegeneration and Astroglios over time.[10,11]



DIAGNOSIS - diagnosis related on clinical examination electroencephalogram and cerebrospinal findings.[5,6,7,8]

1. **ELECTROENCEPHALOGRAM FINDINGS-** periodic sharp waves complexes are found in the easy recordings of approximately two-thirds of patients with classic cjd and have therefore been

incorporated into probable classic cjd Diagnostic criteria.[15]

2. **MRI FINDINGS -** initial studies investigating the usefulness of the number in the diagnosis of cjd used to imaging pattern consistently the technological advancements in MRI enabled physicians to use flat diffusion-weighted imaging and Apparent diffusion coefficient improving both the negative and positive predictive value that term pulvinus sign or bilateral flaire hyper intensity of the pulmonary area was coined after review in 86 patients with variant cjd.
3. **BIOMARKERS IN CEREBROSPINAL FLUID -** several CSF biomarkers were proposed to play a significant role in the diagnosis of the Classic CID the most commonly studied is 14-3-3 protein which is a surrogate Marker for cjd in a after neuronal destruction although there is still some debate in the literature several studies conclude that the sensitivity of positive 14-3-3 protein in CSF for Classic cjd is 92 to 96%.[18]
4. **NEW DIAGNOSTIC CRITERIA -** criteria for diagnosis of classic cjd that includes New York informative test and incorporates phenotype is trained provide a less invasive means of Definite diagnosis and will include the atypical patient and updated Diagnostic criteria and algorithm should provide confidence in the diagnosis while avoiding unnecessary cost waste of resources and the potential morbidity as associated with the biopsy.

Topography	Characteristic Appearance	Most Usual Sequences
Cerebral cortex	Focal or diffuse, symmetric or asymmetric involvement Perirolandic area usually spared	FLAIR and mainly DWI/ADC Signal intensity abnormality may fluctuate
Basal ganglia	Symmetric or asymmetric involvement, particularly of caudate and putamen Anterior-posterior gradient	FLAIR and mainly DWI/ADC Increase in both extent and degree of signal intensity abnormality as disease progresses
Cerebellum	Atrophy	Typically negative at imaging Only a few reports show clear DWI hyperintensity

TISSUE CONFIRMATION TEST - the reason for the low Diagnostic yield of brain biopsy is the methodology used in the pathological sections histopathologically investigation must show why collation which may be missing on specimens their needs to be ample amyloid deposits and present for a positive immunohistochemical test for PRP.

TYPE[4]

Groups	Subtypes
Acquired	Kuru Variant CJD Iatrogenic CJD
Familial (10%–15%)	Genetic CJD Fatal familial insomnia Gerstmann-Sträus-sler-Scheinker disease
Idiopathic (85%)	Sporadic CJD (sCJD) Sporadic fatal insomnia Variably protease-sensitive prionopathy

TREATMENT[2,3]

Treatment of Prion diseases remains supportive for example using clonazepam for the first treatment of myoclonus are being used. no specific therapy has been shown to stop the progression of the disease.

CONCLUSION

Creutzfeldt jakob disease started out a description of unusual neuropathological and clinical findings Association with other diseases in a series of 6 patients over the last 90 years has been lead to well described clinical and pathological entity with typical imaging. It is a fatal neurological disease that often perplexus that threatening physician giving its rare publisher Diagnostic criteria 8 in the approach to diagnosis but these are unfortunately outdated and do not incorporate modern research laboratory methods the proposed updated Diagnostic criteria and treatment algorithm are based on the latest research and help guide the work up in a disease that can be difficult to diagnose knowledge different strains phenotypes is crucial to making and accurate diagnosis and brain biopsy need to be performed only rarely in a typical patient because QULC appears to be the best way to provide a confirmed diagnosis.

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