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PRION DISEASE AND ORAL CAVITY – THE UNKOWN FACTS

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ABSTRACT:

Prions are misfolded proteins with the ability to transmit their misfolded shape onto normal variants of the same protein. They characterize several fatal and transmissible **neurodegenerative diseases in humans** and many other animals. The word prion derives from **"proteinaceous infectious particle".**

Human prion disorders are classified into **Creutzfeldt Jakob Disease (CJD), Gerstmann-Straussler-Scheinker (GSS) syndrome, and Kuru**. It also further subclassified into 2 main etiologic categories:

1. Inherited Prion Disease
2. Acquired Prion Diseases

There is also evidence suggesting prions may play a part in the process of Alzheimer's disease, Parkinson's disease, and amyotrophic lateral sclerosis (ALS), these have been termed **prion-like diseases.**

Prions cause neurodegenerative disease by aggregating extracellularly within the central nervous system to form plaques known as amyloids, which disrupt the normal tissue structure. This disruption is characterized by "holes" in the tissue with resultant spongy architecture due to the vacuole formation in the neurons. Other histological changes include astrogliosis and the absence of an inflammatory reaction. While the incubation period for prion diseases is relatively long (5 to 20 years), once symptoms appear the disease progresses rapidly, leading to brain damage and death. Neurodegenerative symptoms can include convulsions, dementia, ataxia (balance and coordination dysfunction), and behavioural or personality changes**.**

Oral symptoms occur rarely in patients with prion disease. But oral manifestations are commonly seen in prion diseases as dysphagia (difficulty in swallowing) and dysarthria (speech disorder as characterized by poor articulation). In vCJD, orofacial dysesthesia (abnormal sensations experienced in the absence of stimulation), paraesthesia (tingling, pricking or numbness of skin) or loss of taste and smell (only one case has been reported so far).

There is a theoretical, yet real risk of prion disease transmission via dental instruments and dental treatment, although the magnitude of the risk has yet to be determined. Health care

workers should understand these emerging diseases so that practical and reasonable changes to dental public health and infection control policies can be implemented. A proper and

precise case history should be taken before any dental treatment is given to the patient. And also due to the difficulty to define the risk of CJD at present because it is unrelated to family history, therefore, to reduce the risk of prion disease transmission, the best practice is to treat every person as potentially infectious. By improving universal infection-control precautions for decontamination of instruments and waste in dental practice is now the best way to prevent current prion disease trends.

Keywords: Prions, prion-like diseases, Alzheimer's disease, Parkinson's disease, Creutzfeldt Jakob Disease (CJD), Oral symptoms.