Prion disease is both feared and misunderstood by healthcare professionals. As the leading source of ophthalmic allografts, it is our responsibility to keep our customers informed. Cristoph Schopf, Senior Scientific Officer at Tutogen Medical recently forwarded us the paper *The Sclera, the Prion and the Ophthalmologist*. This paper published in the British Journal of Ophthalmology, is an overview of prion disease and the growing concern in the ophthalmic community.

Creutzfeldt-Jakob Disease, CJD is the primary transmissible spongiform encephalopathy (TSE) disease in humans. Mad Cow Disease is the most newsworthy and is often the limit of exposure to the general public. Several human TSE cases in Europe have been linked to the consumption of beef. This new variant CJD is believed to have crossed the cow/human barrier and is the source of the growing concern with potentially pandemic prion disease. To put this into perspective, in 2002 FDA implemented policy restricting blood donation from anyone living in at-risk European countries from 1980 through the mid ’90s.

Prions are abnormal proteins that self-replicate in cell nuclei. TSEs disrupt cell function causing incremental cell death or apoptosis leaving a sponge-like necrosis. The disease affects innervated tissues and therefore concentrates in the brain and nerve tissues of the spinal chord. The sclera is a direct extension of the dura matter the outer layer of the brain where TSEs show the first signs physical evidence. The ocular surface also contains many nerves harboring infectious concentrations of prions. Therefore TSE disease is a growing concern to the ophthalmic community.

TSE disease is 100% fatal. The process of infection takes several years but is usually characterized by the progressive onset of memory loss, dementia, and decreased motor control. The long incubation period imposes challenges to healthcare professionals regarding the source of infection. Since the incidence of spontaneous CJD is 1:1,000,000, health officials will investigate every case. Was it the Beef Wellington consumed at Heathrow back in 1988 or that sclera patch with the glaucoma surgery in 1995? The meal will be difficult to trace but the graft is listed on the patient’s chart.

Most of the reported cases of CJD transmission in the ophthalmic community have been from cornea transplants. This risk may be acceptable in that there is no disinfection option with organ transplants nor validated donor-screening process. The use of sclera patch grafts carries the same risk unless it is treated to inactivate prions. Healthcare regulators suggest all neurosurgical instruments be soaked in sodium hypochlorite or sodium hydroxide and steam autoclaved to inactivate prions. The Tutoplast process utilizes NaOH at the recommended levels.

Surgeons need to ask themselves, "What does my tissue supplier do to inactivate prions?" Our objective is to make sure the ophthalmic community never needs to publish the paper, *CJD Transmission from Donor Sclera*.

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1. Screening
   - FDA neurological testing protocols are followed. No donor pooling is allowed. Tissues are inspected if the donor has a medical or surgical history that indicates risk of infectious disease or malignancy.

2. Tissue Processing - Phase 1
   - Tissues are cleaned with a saline solution of various concentrations. This process results in the osmotic cleansing of the collagen matrix.

   - Osmotic Treatment
     - Cellular components are removed.
     - Potential viruses are exposed for inactivation.
     - Antigens are reduced.

3. Tissue Processing - Phase 2
   - Inorganic agents are employed in specific concentrations to inactivate HIV and hepatitis. The denaturation is achieved without significant alterations in the extracellular matrix and collagen structure.

   - NaOH Treatment
     - Virus inactivation
     - Collagen preservation

4. Preservation
   - Non-sterile, sterilized tissues with disinfectant properties
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5. Final Packaging
   - The tissue is cut to specified size and packaged in trays for transplant.
   - The grafts are then terminally sterilized using gamma-irradiation to medical device standards.