Creutzfeldt-Jakob Disease (CJD) is a rapidly progressive, inevitably fatal neurodegenerative disorder believed to be caused by an abnormal isoform of a cellular glycoprotein known as the prion protein. In simpler terms, what happens is that the prion protein eat away at the brain and create tiny sponge-like or Swiss cheese like holes in some parts of the brain. These holes cause the brain to slowly deteriorate and eventually this causes symptoms that affect the whole body. Eventually death occurs. Reportedly the occurrence of this disease world wide is about 1 in one million.

Prions can be found in the brain, spinal cord, eye (retinal portion) and other tissues of the nervous system of affected animals or humans. They are highly resistant to heat, ultraviolet light, radiation and disinfectants that normally kill viruses and bacteria. Prions can also infect humans who eat meat from infected cattle. Even thorough cooking of the infected meat does not kill the prion or eliminate the risk of getting the disease. The incubation period can be anywhere from one year to as many as 13 years. When there are finally enough prions in the brain, the symptoms are such that it can be difficult to impossible to diagnose without a biopsy. Some of these symptoms include; depression, anxiety, trouble sleeping, difficulty in walking and dementia and can progress very rapidly.

There are three categories or classifications for how CJD is acquired. The first is sporadic. This means that there is no known infectious source and no family history of the disease. This includes about 85% of the known infective population. The second is inherited, which accounts for 10 to 15 percent of all CJD cases. The inherited cause appears to be a mutation in the gene coding for the prion protein. The third is through infection.

For the Central Sterile professional, the difficulty is two fold. How do we clean without spreading and how can we sufficiently sterilize in order to make it safe to use? The World Health Organization (WHO) has put out guidelines and protocols to insure the safe handling of instruments and other possible infectious items within all areas that may come in contact with CJD prions. It is suggested that any known or suspected case of CJD or vCJD (variant CJD) that is being biopsied for definitive diagnosis, be done only with sterile single use equipment so they can be disposed of after being soaked in Clorox (sodium hypochlorite).

Power equipment should not be used as the splatter from the potentially infective material can be slung from the point of use to contaminate all things in the immediate vicinity including staff. Also, power equipment is difficult to clean and the cleaning and sterilization methods necessary to render the equipment sterile are well known to damage the equipment. Another type of equipment that should not be used for these procedures are flexible endoscopes. Like the power equipment, the flexible endoscopes are damaged by the cleaning and sterilization processes that are necessary to deactivate prions effectively.

If implants have to be used, then the physician needs to tell the OR crew before hand what implants he is going to need so that those are removed from the set before the procedure starts. This action is necessary in order to prevent contamination of implants that are used on other patients. Any implants not used during the case should be discarded and not re-sterilized as this could potentiate the spread of CJD to other patients.

When potentially contaminated instruments are used, it is imperative that all personnel involved wear personal protective equipment (PPE). These precautions should be taken by everyone (OR or CSD) potentially coming in contact with a possible CJD patient or contaminated equipment. This includes and is not limited to the following: repellant gown, gloves, mask, visor or goggles and shoe covers. All protective clothing, covers or liquid should be considered contaminated and should be incinerated. All instrumentation should be kept moist until cleaned.
or decontaminated. This is to insure that the prions do not dry on the instruments which may cause ineffective cleaning processes.

As a Central Sterile professional, what are our responsibilities both to ourselves, our coworkers and to the patient? There currently is no consensus on the best method of managing instrumentation that has been contaminated with prions. It is documented that paracetic acid is not effective and hydrogen peroxide gas plasma is, at best, only partially effective. So just where does that leave the CS team that is trying to decontaminate and sterilize instruments that are reusable for other cases? There is a protocol that has been developed thru extensive research that now at least gives us an opportunity to keep ourselves safe and decontaminates instrumentation for reuse. After use in the OR, an instrument cleaner should be sprayed on or dipped onto the used instruments in order to reduce the amount of contamination. This can be done first in the OR. Again, if the prion contaminated material is allowed to dry on the surface, any methodology used to sterilize the instruments will be ineffective. Alcohol, formalin or glutaraldehyde should not be used as they are drying agents and as such will cause the prions to be dried on the equipment rendering them as fixed. Therefore, the instruments must be cleaned. In order to process these instruments, they must be cleaned using an instrument cleaner.

In the event there is unfamiliarity with the decontamination method for certain instrumentation, the manufacturer must always be contacted as a final authority. Even if the instrument has been sterilized and used since being used on a contaminated patient, these cleaning procedures should be used to attempt to breakdown the prions and clean and sterilize this instrument according to current methodologies.

Once the equipment is thoroughly cleaned, the instruments can be steam sterilized in a prevac cycle of eighteen (18) minutes at a cycle temperature of 272 degrees F or 134 degrees C. If you don’t have a prevac sterilizer, sixty minutes in a gravity displaced sterilizer with a cycle temperature of 132 degrees C or 272 degrees F. The other alternative is to immerse the instruments in 1 N sodium hydroxide (NaOH) for one hour, followed by a water rinse followed by steam sterilization as listed above. An N of sodium hydroxide is 40 grams of Soda Lye in 1 liter of water for those of you that were curious. This is considered the most effective way of cleaning the reusable instrumentation.

After the instruments are cleaned via this process, they are then considered safe enough to reprocess using the current set up of running the instruments thru the washer/decontaminator before handling them to prepare for sterilization. Liquids used for cleaning should be decontaminated in situ by adding NaOH (soda lye) or hydrochlorite (bleach) or even autoclaving at 134 degrees C (272 F) for 18 minutes. In as much as is possible, the cleaning equipment such as brushes, scouring pads and toweling should be considered disposable.

It is well documented that prions can survive for years. Instruments that are found after surgery to have been in contact with prions must be pulled from service and handled as suggested above and then put back into service.

Professionals, whether it is in the OR and/or the CS department must always be aware for the potential of contamination to the patients in order to prevent the spread of CJD by the use of contaminated instrumentation.

References:
WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies
WHO/CDS/CSR/APH/2000

Perioperative Standards and Recommended Practices AORN 2008 Edition
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