



## Update On Mad Cow Disease

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Cow infected by Bovine Spongiform Encephalopathy (BSE) that destroys brain tissue, on right, with a myriad of holes that resemble a sponge. Photographs courtesy [www.mad-cow.org](http://www.mad-cow.org).

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**February 11, 2001 Atlanta, Georgia** - *The London Times* reported this week that animal feed protein contaminated with bovine spongiform encephalopathy, also known as BSE or mad cow disease, is estimated to have reached 70 countries through exports by a British company between 1988 and 1996. The company, Prosper de Mulder based in Doncaster, northern England, admitted to the *Times* that its animal feed was exported as pig and poultry food which were not banned until 1996, but could still have been mixed up with cattle feed which was illegal. The BSE-contaminated pig and poultry food was exported to Indonesia, Israel, Japan, Kenya, Lebanon, Malta, Saudi Arabia, Singapore, South Korea, Sri Lanka, Taiwan and Thailand. The United Nations is now warning all countries that have imported cattle or animal feed from western Europe, especially Britain, to be concerned about the risk of BSE and variant CJD.

In Britain, the number of new variant Creutzfeldt-Jakob victims has now reached 94. Also this week, *The Guardian* newspaper reported that thirteen of those victims donated blood which had been used in transfusions for 23 people and other blood products such as immunoglobulin. The newspaper said, "Thousands of patients worldwide, and an unknown number of hemophiliacs in Britain, might have received treatments with the blood products between 1996 and 2000. Malcolm Tibbert, Chairman of the Human BSE Foundation which represents families of the victims, told reporters, "Lessons of the past have not been taken on board. It is bad enough we have vCHD in this country, and it is clear BSE was exported. But this blood contamination problem would take it to another level."

Since 1998, England switched to purchasing most of its blood plasma from the United States which has been strictly monitoring for BSE disease in cattle since the middle of the 1980s. But another unknown factor in all prion disease is the lag time between ingestion of prions and their destruction of the brain and spinal cord which causes death in animals and people.

This week I talked with a medical epidemiologist at the Center for Disease Control in Atlanta, Georgia about the time lag and some other questions that listeners have e-mailed to me since my other *Dreamland* reports about Mad Cow Disease and New Variant CJD. I began by reading a question e-mailed from a medical doctor: "Are prions killed by irradiation that would be used to

## **Interview:**

**Ermias Belay, M. D., Medical Epidemiologist, CJD Surveillance Program, Center for Disease Control, Atlanta, Georgia:**

### **"WHAT CAN DESTROY PRIONS?"**

They are not destroyed by irradiation. Prions have been shown to be extremely resistant to conventional methods of sterilization or disinfecting, including radiation. There are some autoclaving methods and chemical methods that have been shown to be effective against prions, but not radiation.

### **CAN YOU EXPLAIN FOR A LAY PERSON ABOUT WHAT COULD KILL A PRION?**

There are two chemical agents including sodium hydroxide and also sodium hypochloride. Sodium hypochloride would be undiluted 100% household bleach. In high concentrations, it has been shown to be effective against the prions if the material that's contaminated is exposed for one to two hours. In autoclaving which is the normal way of inactivating instruments potentially contaminated with infective agents in a hospital set up not the routine autoclaving, but autoclaving with high temperature and increased duration has been shown to be effective also.

### **LONG HIGH HEAT AND A BLEACH-LIKE CHEMICAL.**

Hypochloride and a second chemical, sodium hydroxide in very high concentrations.

### **THE TRANSMISSION OF CJD, MAD COW AND ANY OF THESE PRION-RELATED DISEASES MUST COME THROUGH CONSUMING SOMETHING INFECTED WITH THE PRIONS?**

Classic CJD has not been transmitted through the consumption of anything known yet. We have to make a distinction between the classic form of CJD and BSE which people normally call Mad Cow Disease. Mad Cow Disease affects cattle and it is a new disease relatively speaking and was first identified in the U.K. and subsequently in other European countries in the cattle population. In contrast, the regular or classic form of CJD is not a new disease. It has been around since 1920 at least and occurs worldwide and has not been in most patients been associated with consumption of anything. It's reported to have occurred spontaneously in the vast majority of patients and has nothing to do with beef consumption. It is true that the BSE problem in the cattle population, the Mad Cow Disease, may have been transmitted to humans in the U. K. and several other European countries and that is a new disease in the human population is different from the classic CJD that occurs in the U. S.

### **THAT IS THE ONE THAT THE HYPOTHESIS IS THAT PEOPLE HAVE CONTRACTED IT BY EATING THE BSE-CONTAMINATED BEEF...**

That's correct...

### **THAT INGESTION HAS SOMEHOW AFFECTED THE PRION PROTEIN IN THE HUMAN CAUSING THIS DISEASE?**

That's correct. The new form of the disease in humans is called New Variant CJD, or human BSE because it originated from BSE in cattle.

### Do BSE Prions Spread Through Dairy Products?

### **WOULD THAT APPLY ALSO TO DAIRY PRODUCTS FROM CHEESE TO MILK?**

Dairy products have not been identified to be the vehicle of transmission for BSE even in the U. K. Where they've had a lot of problems with BSE, they have done experimental studies and none of the studies indicated that dairy products from infected animals would transmit the disease. So, as far as we can tell, and

as far as experimental studies are concerned, even in the U. K. where the BSE problem is a big problem, dairy products have not been identified to transmit the BSE agent.

**BUT IF PEOPLE HAVE CONTRACTED THE NEW VARIANT CJD FROM EATING BEEF PRODUCTS IN BRITAIN, IRELAND, FRANCE AND SO FORTH, THAT MEANS THAT THE PRIONS ARE IN THE MEAT THEY ARE EATING, RIGHT?**

The way the prions made it into the beef is generally believed because from the slaughterhouse down to the meat products being served to humans, there is a lot of opportunity for that product to be contaminated with the brain tissue and the spinal cord. It's not that the solid meat or muscle tissue that has been implicated in transmitting the disease, rather beef products have had the opportunity to get contaminated with the highly infected tissues such as the brain and spinal cord.

**YOU'RE MORE LIKELY TO GET THE NEW VARIANT CJD FROM EATING SAUSAGE, FOR EXAMPLE, AS OPPOSED TO A FRESH CUT OF STEAK?**

That's possible, correct.

**BUT IT'S NOT KNOWN FOR CERTAIN?**

They have been trying to pin down what the new variant CJD patients consumed in order to get the agent. And as far as I know, they have not been able to come up with a specific product. The general consensus in understanding is that these patients may have consumed materials that may have been contaminated with the nervous tissue coming from BSE-infected animals.

Where Do Prions Concentrate in the Body?

**WHAT IS YOUR CURRENT UNDERSTANDING ABOUT WHY THE MISSHAPEN PRIONS THAT CAUSE THE DISEASE SHOULD CLUSTER ONLY IN THE BRAIN AND THE SPINAL CORD AND NOT OUT INTO THE REST OF THE BODY?**

We don't have all the answers to that. For some reason, they lodge in the brain and the disease process starts in the brain and brain cells causing destruction of the neurons or brain cells. And as cells are destroyed, neurologic signs and symptoms start to appear. Now what actually triggers this transmission from one to another in the brain is poorly understood.

**WHEN YOU THINK OF NERVES, THE BRAIN COMMUNICATES TO THE REST OF A BODY BY ALL THE NERVE GANGLIONS THAT GO ALL THE WAY THROUGH EVERY PART OF A BODY. HAVE THEY EXAMINED NERVES THROUGH ARMS, LEGS AND TORSO TO SEE IF THEY HAVE THE SAME KINDS OF HOLES AS FOUND IN SPINAL TISSUE AND THE BRAIN?**

The pathology, the structure of the brain is different in the brain than in the areas you just mentioned. The disease seems to be very localized, as I said, in the brain. It's one thing you need to understand that the disease in different species may have different characteristics. The classic form of CJD we have in the U. S. and which occurs worldwide and has nothing to do with beef consumption - the agents seems to be very localized to the brain only and probably also the spinal cord. In some cases, the cornea tissue.

Cornea Tissue of Eyes?

**WHICH?**

The cornea or eye tissue.

**CORNEAS IN EYES CAN BE AFFECTED WITH THE SAME SPONGIFORM HOLES?**

Not the spongiform lesions, but the presence of the agent.

**PRESENCE OF THE PRIONS.**

Correct.

### **WHY WOULD THE PRIONS BE IN THE CORNEA OF THE EYE?**

We like to think of the cornea as an extension of the brain. If you think about your eyes, it's the back of the eye right next to the brain, almost attached to the brain. And if you look at how the tissues develop from conception, the fetus grows up or develops, the retina and parts of the eye originate from the same place that the brain and spinal cord develop from. So technically speaking, it's an extension of the brain. Right behind your eyes is the brain tissue.

### **THEN EYES ARE AN EXTENSION OF THE BRAIN?**

That's correct. In many ways, it is. There are a lot of nervous tissue that extend right from the brain into the eyes and even by proximity they are extremely close right next to the brain. What I'm telling you applies to the classic form of CJD.

When you go to other prion diseases that affect different species, they behave differently. So we cannot talk about all prion diseases in the same terminology, where the agents are found and which parts of the tissue harbors the infection agent. It's different in different species.

### **WHAT HAPPENS WITH NEW VARIANT CJD IN HUMANS OR BSE IN COWS OR EVEN SCRAPIE IN SHEEP WHICH IS A DIFFERENT KIND OF PRION DISEASE OR CHRONIC WASTING DISEASE IN ELK AND DEER WHICH IS YET ANOTHER PRION DISEASE - ALL OF THEM ARE STILL NOT FULLY UNDERSTOOD AND EACH ONE IS IT'S OWN MEDICAL PROBLEM.**

It's got its own different distribution of the agent in the body, let me put it that way. In that regard, the classic form of CJD is very different because the agent seems to be very highly localized in the brain and nervous tissue.

### **WHAT WE DON'T KNOW NOW ABOUT THE NEW VARIANT CJD IS WHETHER OR NOT THE PRIONS ARE INGESTED FROM THE MEAT AND MUSCLE TISSUE OF THE INFECTED COWS IN EUROPE, OR IF IT IS COMING ONLY THROUGH PRODUCTS IN WHICH BRAIN AND SPINAL CORD HAVE BEEN GROUND UP?**

There is evidence in the U.K. that the muscle tissue from cattle infected with BSE is not infectious. One of the studies they did in the U.K. was to take different parts of the animal and put it into experimental animal models. So they took tissues from the brain and all other parts of the infected animal and wanted to see if that tissue would be infectious in other animal models. Afterward, they determined that the muscle tissue was not infectious, even from infected animals.

### **IF YOU ARE IN EUROPE NOW AND YOU WANT MEAT, AT LEAST THE SAFEST THING TO DO IS TO GET A FRESH STEAK, BUT NOT EAT ANY GROUND UP MEAT PRODUCTS?**

That's correct. If you are dying to get meat and beef, then it would be more advisable to eat a solid cut of muscle tissue.

### What Is The Lag Time Between Destructive Prion Ingestion and Disease?

### **IT'S KNOWN THERE IS A LAG TIME BETWEEN INGESTION AND DISEASE IN BSE CASES IN CATTLE AND NEW VARIANT CJD IN EUROPE SUGGESTING THAT YOU COULD INGEST THE PRIONS SAY 10 TO 20 YEARS AGO AND THEN THE NEUROLOGIC SYMPTOMS ARE NOW JUST EMERGING?**

Right, the lag time can be measured in years as you correctly suggested. Now, what that exact time period is, we don't know.

### **IF THERE WAS THIS EMERGENCE OF THIS NEW VARIANT IN 1996 IN ENGLAND, THIS COULD BE THE TIP OF THE ICEBERG OF CASES YET TO EMERGE?**

That's correct.

### **THAT'S EVERYBODY'S CONCERN?**

That's correct. In fact, if you look at the 94 cases that have been reported as of last month, there have been 94 in the U. K., if you plot those cases by year, you'll see an increase, an increasing trend in the number of cases year after year. And how much it would increase and what the eventual size of that outbreak will be, nobody knows."

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### **Websites:**

**<http://www.mad-cow.org/>**

### **Credits**

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