Blood recipients and CJD: to notify or not to notify, that is the question

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In brief

SHOULD RECIPIENTS WHO HAVE RECEIVED BLOOD from a person who developed Creutzfeldt–Jakob disease be notified, even though there is no proof the disease will be transmitted? Does notification simply cause unnecessary anxiety? Two recipients explained their polar-opposite positions during a recent Health Canada conference.

En bref

FAUDRAIT-IL PRÉVENIR LES PERSONNES AYANT REÇU DU SANG de donneurs chez qui la maladie de Creutzfeld-Jakob s'est déclarée par la suite, même si l'on ne peut prouver que la maladie sera transmise? Risque-t-on de provoquer seulement une anxiété inutile? Dans le cadre d'une récente conférence de Santé Canada, deux transfusés ont exposé à ce sujet des points de vue diamétralement opposés.

Is Creutzfeldt–Jakob disease (CJD) another HIV-blood scandal in the making? Another hepatitis C disaster? Jacques Pelletier thinks so. "This could be strike 3," says Pelletier, who received a transfusion during a heart operation and was later notified that 1 of his blood-product donors has subsequently developed CJD.

"Can you afford not to notify the victims?" Pelletier asked doctors and others attending a recent Ottawa conference on CJD, which was sponsored by Health Canada's Laboratory Centre for Disease Control. "I don't think so. I received [blood from a person with] CJD and I have to live with it. I'm not afraid of it, I'm afraid of you trying to make my decisions, trying to think for me."

Paula Grau disagrees completely. In 1996 she was notified that her son had received a blood product while a patient at Toronto's Hospital for Sick Children, and one of the donors may have had CJD. "Initially I was glad I was told," she recalls, "and now I wish I hadn't been. Notification, I think, is an injustice, because the risk is only theoretical."

She says she has worried for 2 years but had nowhere to turn for answers to her many questions: "How do I tell my son? Do I? Should I stop him from donating blood?"

Grau also spoke at the conference, where delegates reviewed the science and ethical issues surrounding the transmissibility of CJD through blood. Is notification justified given that there is only a theoretical risk of transmission? And if it is justified, why are those who have been notified still allowed to donate blood?

There is no conclusive evidence that CJD can be transmitted in blood or blood products, but blood obtained from donors who develop CJD or have a significant risk factor for the disease has been withdrawn since 1995 because of what Health Canada describes as a "theoretical risk." The responsibility of Health Canada and the blood-collecting agency — formerly the Red Cross, now Canadian Blood Services — ends with the withdrawal of the blood and blood products. It is then up to individual hospital blood banks and transfusion

centres to decide whether to notify those who have received the "theoretically" infectious products. At the moment, few do.

The 300-member Canadian Creutzfeldt–Jakob Disease Society, which is based in Calgary, says about 300 000 Canadians have received blood from withdrawn lots and 2500 of them have been notified. After a multimillion dollar withdrawal in July 1995, only a few governments and hospitals decided to notify recipients.

In Calgary, for example, the city's health authority notified all hospitals, which in turn informed patients. In Toronto, the Hospital for Sick Children sent letters notifying parents, and St. Michaels Hospital contacted patients' attending physicians; it was then up to the doctor to decide whether to notify the patient, and 28.7% decided against it. British Columbia created a hot line that people could call if they wanted to know whether the blood they had received was from a withdrawn lot.

Dr. Andrew Kaegi, the director of the Red Cross Blood Centre in Calgary, says it's not fair to notify people when there's no proof CJD is even transmitted by blood, and there's no way of diagnosing the disease, much less treating it. CJD is a neurodegenerative disorder — 1 of 4 human prion-related dementias — that typically affects people between age 60 and 65. It is always fatal. Symptoms include presenile dementia, myoclonus and progressive motor dysfunction. Its incubation period ranges from 18 months to 30 years, but survival time is short once symptoms appear and patients generally live less than a year. The incidence of CJD is relatively rare globally, with between 0.5 and 1 case per million people annually.

"Notification has done a lot of harm"

Kaegi attributes the decision to notify blood recipients to a "fit of enthusiasm" that followed the discovery of the variant form of CJD in the UK — there have been no documented cases of this variant form in Canada — and to fear of another blood scandal. "I think people went off a little half-cocked," says Kaegi, who became director of the Calgary blood centre after that city's massive CJD notification effort in 1995. "We were trying to be proactive here, and I see the damage."

Kaegi recently attended a meeting of the CJD society and "was surrounded by a dozen people who were scared silly every time they forgot a number or a key. Many went to see a physician or psychiatrist about their fears. This notification has done a lot of harm. I think we have to take some of the blame here for notifying people about a disease that's exceedingly rare and, to our knowledge, not transmitted by blood."

But do blood recipients want to be notified? Dr. Lynn Boshkov, an assistant professor at the University of Alberta, asked 1216 Alberta residents this question, and 67.8% of respondents said they did want to know. However, 54.3% agreed that contacting recipients would do more harm than good and 62.4% said the money would be better spent on educating the public about the risks associated with blood products.

During the conference, Boshkov said results indicated that people should have more individual choice. In a survey of patients and families who were notified at the Hospital for Sick Children, 84% said they wanted to be notified if there was another recall (see p. 771).

In 1996, delegates at another national conference sponsored by Health Canada decided that the public did have a right to be informed but didn't determine how this should be done. At this year's conference they were still debating the same question. And even though it wasn't geared to creating guidelines, organizer Graham Sher ex-

Physicians' help needed in CJD study

If you are treating someone with Creutzfeldt–Jakob disease, Health Canada would like to know. The Laboratory Centre for Disease Control is conducting 2 studies: a general surveillance for new variant-CJD, for which there have been no reported cases in Canada, and a 5-year case-control study to assess the risk the transfusion of blood products holds for the development of CJD.

Dr. Neil Cashman, coprincipal investigator at the Centre for Research in Neurodegenerative Diseases at the University of Toronto, says 4 similar studies haven't had a big enough sample, although all of them revealed no evidence of risk. "Our study is 5 times bigger than the next biggest and in that sense it's unique. We have a better chance of seeing a link." The study will look at an estimated 100 patients between 1996 and 2000, and results are expected late in 2000.

To protect confidentiality, physicians are being requested to ask patients or their families if they'd like to take part in the study. It involves giving a blood sample for genetic testing, a 2 to 3 hour interview and a chart review to obtain a complete history, including record of transfusions. To speak to a study member, phone 888 489-2999.

pects the conference could eventually result in a series of "loose recommendations." Minutes from the June CJD Conference will be on-line later this fall at **www.hc-sc.gc.ca/hpb/lcdc/hp_eng.html**.

Progress is slow and Pelletier, a retired railway engineer, says he's immensely frustrated. The vice-president and founding member of the CJD society, which was formed in January, says 6 members already have symptoms: memory, eyesight and balance problems. [CJD has not been diagnosed in these patients. — Ed.]

"The cat is out of the bag in Calgary," says Pelletier, "and soon it will be out all across Canada." If all the blood recipients involved align, he says, the government will be facing pressure from a major group. For the moment, Pelletier would like to see monitoring of everyone who has received blood from donors who developed CJD.

Grau says she too is frustrated. "I assume I was told so that my son wouldn't donate blood," she says, but now she finds there's nothing to prevent him from doing just that when he gets older. "It's not hazardous enough to prevent donors from donating, yet it's hazardous enough to notify us."

The FP's role

Both Pelletier and Grau also think family doctors should be better informed about CJD. Pelletier thinks a well-informed FP could go a long way toward soothing patients fears about the risks. "Not advising everyone involved is a breach of trust," he says.

[A support and discussion group has been formed for those who have been notified that they received blood from someone who developed CJD: **members.aol.com/debbieoney/blood.htm**.] ❓