Tonsillectomy and variant Creutzfeldt–Jakob disease

Tonsillectomy and adenoidectomy remain common operations of proven benefit. The risk of developing variant Creutzfeldt–Jakob disease (vCJD) and its transmission on surgical instruments have led central government to pilot the use of disposable instruments for this operation. This editorial discusses the issues related to this decision and the risks associated with implementing it.

INTRODUCTION

In 1999, 60 000 tonsillectomies (with or without adenoidectomy) and 9500 adenoidectomies were carried out in England. The mean age at the time of tonsillectomy is 15 years and of adenoidectomy 6 years (www.doh.gov.uk/hes/standard_data/index.html). The main indication for tonsillectomy is recurrent sore throat, and for adenoidectomy alone it is otitis media with effusion. The effectiveness of these operations has been challenged, but evidence supports their value (Blair et al, 1996; Maw, 1997).

There have been 90 confirmed deaths in the UK caused by vCJD since 1996 (www.doh.gov.uk/cjd/cjd_stat.htm). Despite this small number, it is important to exercise caution and take steps to minimize the risk of transferring vCJD during surgery, particularly on the CNS, the retina and lymphoid tissue.

1. If eating bovine spongiform encephalopathy (BSE) affected beef is the cause of vCJD, a large proportion of the British population has been exposed to the causative prion
2. Prion diseases have very long incubation periods. Large numbers of people could be incubating vCJD
3. Tonsil tissue is known to accumulate pathological prion protein in vCJD
4. Prion protein (PrPc) resists destruction by conventional sterilization techniques
5. PrPc has a strong affinity for metal surfaces and resists removal by standard decontamination procedures.

NORMAL PRION PROTEIN AND PRION DISEASES

PrPc is a normal constituent of some cells. Neurones have the highest concentration, with the next highest concentration in cells in the germinal centres in peripheral lymphatic tissue. PrPc has three-dimensional α helices in its structure. In prion diseases, these undergo an isomeric change into a structure which is rich in β sheets (PrPSc = scrapie prion protein or pathological prion protein).

PrPc is metabolized within cells by proteases, but PrPSc is resistant to these enzymes and accumulates in the cells, leading to cell death, with vacuolation of brain tissue, giving rise to the pathological description spongiform encephalopathy. The process explains the predominant clinical feature of these diseases – progressive, lethal neurodegeneration. Despite this, prion diseases are not a single clinical entity and the clinical features differ between them.

TRANSMISSION OF PRION DISEASES

Transmission can be inherited, as in familial CJD, or by ingestion or inoculation of infected material. Kuru affects the Fore people of New Guinea and is transmitted to susceptible individuals when they eat the brains of other infected humans. The incubation period is long – years or even decades. Transmission appears to be achieved by the infectious agent crossing the gut wall intact, entering the relevant cells and slowly converting PrPc to PrPSc.

In iatrogenic CJD, dural grafts and growth hormone from affected humans have been implanted or injected into others who then develop the disease. Such material has been subject to conventional decontamination measures, but the infectious agent survives this.

GENETIC SUSCEPTIBILITY TO PRION DISEASES

A genetic susceptibility is well recognized. In sheep, the prion disease scrapie can be bred out of a flock by the selection of resistant genotypes. Humans whose genes code for methionine/methionine (M/M) at position 129 on PrPc are most susceptible to Kuru and vCJD, whereas those who code for valine/valine (V/V) are most resistant. Those with M/V are of intermediate susceptibility. It might be concluded that the presence of M/M at position 129 allows the PrPc to change shape to β sheets, whereas the presence of V/M or V/V at this position stops this. However, the resistance may not be absolute. Recent evidence regarding Kuru indicates that the V/V genotype can develop the disease, but the incubation period is extended (Lee et al, 2001).

INTER- AND INTRA-SPECIES TRANSMISSIBILITY

There appears to be a barrier for the transfer of prion diseases between species. Scrapie has been known in sheep in Britain for well over a century. Despite the consumption of sheep (brains included), no prion disease has been identified in humans which can be linked to scrapie. The use of sheep offal in cattle feed has been blamed for the emergence of the BSE epidemic in British cattle, but this remains open to question. BSE is an epidemic prion disease which arose in the 1970s and 1980s. Given that humans have been exposed to scrapie for over a century, it is difficult to understand why BSE should be the cause of vCJD.

To date, 90 deaths from vCJD have been confirmed in the UK since 1996. The annual incidence has crept up to a maximum of 27 in 2000, but this does not reflect the steep rise seen in BSE cases in the 1980s. Epidemiology does not support the hypothesis that we are on the edge of an epidemic (Venters, 2001). However, in the experimental setting, BSE and vCJD have been transferred into the same species of experimental animal and an apparently identical spongiform encephalopathy develops.

Given the long incubation period of these diseases, an impending epidemic remains a possibility. If there is a barrier between species, there may be increased susceptibility to infection once the disease is established within a species, which could lead to increased transmission on vectors such as surgical instruments.

vCJD AND SURGERY

The relevance of this discussion to tonsillectomy in particular and surgery in general only becomes apparent when two other characteristics of PrPSc are considered. First, PrPSc is remarkably stable. Containing no nucleic acid, it is resistant to sterilization by radiation. It is also more resistant to high temperatures and chemical destruction than other infectious agents. This has an obvious bearing on the current methods used to sterilize surgical instruments.

The other cornerstone for the safe use of surgical instruments is the removal of all organic material from the instruments. Surgical instruments are increasingly complex, expensive and difficult to clean. Perhaps cleaning instruments is considered to be a menial task in many hospitals and it has not been given the priority it deserves. This is obviously a sensitive issue as indicated in *Panorama* (BBC1, 12 November 2001), where the ineffectiveness of the current systems for decontamination of surgical instruments was exposed. PrPSc has a particular affinity for metal surfaces, making effective cleaning even more important.

GOVERNMENT ACTION

On 4 January 2001, the Minister of Health issued a press statement in which he announced the allocation of £200 million to improve standards of sterilization of surgical instruments. This was triggered by concerns about the quality of cleaning of surgical instruments and about vCJD. In September 2000, the Spongiform Encephalopathy Advisory Committee (SEAC) suggested the use of disposable instruments to reduce the risk of spreading vCJD, provided it did not increase the risk of surgery. SEAC suggested tonsillectomy as a pilot operation because PrPSc is known to accumulate in the tonsil in vCJD (Wadsworth et al 2001), the operation is performed on an otherwise young, healthy population and the instrumentation is relatively simple. The minister allocated £20 million for this change.

CONSEQUENCES OF THE INTRODUCTION OF DISPOSABLE INSTRUMENTS

This announcement had major repercussions for ear, nose and throat surgeons in the UK. They were informed that they could continue to use existing surgical instruments if they discussed the risks with patients. The information concerning these risks was not available, and the immediate response from around the country, supported by the medical defence organizations, was that this surgery should not be performed except in urgent cases until disposable instruments became available. Over the ensuing months, instruments had to be sourced, assessed, selected and supplied. Despite efforts to balance quality and cost, there have been major problems with supply and quality. There has also been a significant rise in the rate of postoperative haemorrhage in many departments as reported to the British Association of Otorhinolaryngologists.

There are two lessons to learn from this experience. First, the development of disposable instruments requires close cooperation between surgeons, suppliers and manufacturers. This may seem obvious, but it is difficult to achieve. Second, such change may increase the risk to patients. For tonsillectomy, this is hopefully transient, but those who recommend such changes need to recognize the risk inherent in changing practice.

RISK ASSESSMENT

The Department of Health carried out a detailed assessment to look at the risks of transferring vCJD on instruments (www.doh.gov.uk). Of necessity, it set a wide range of parameters, but concluded that the size of any epidemic would increase by up to 10% if this route of infection was not addressed.

When the press refers to a vCJD epidemic of over 100 cases, politicians, lawyers, most health-care professionals and the public lose perspective. Currently, the risk of dying from vCJD is of the same order as the risk of dying from lightening strike. The risks of dying in a car accident or from smoking are orders of magnitude higher and yet there is no news value in this. The public does not hear a balanced debate on the risk of vCJD. We need one. HM

Alan Johnson
Consultant Ear, Nose and Throat Surgeon
Ear, Nose and Throat Department
Queen Elizabeth Hospital
Birmingham B15 2TH
Honorary Secretary, British Association of Otorhinolaryngologists, Head and Neck Surgeons

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KEY POINTS

- Variant Creutzfeldt–Jakob disease (vCJD) is, as yet, a rare disease.
- There has been massive exposure of the UK population to bovine spongiform encephalopathy (BSE) infected beef.
- The link between BSE and vCJD needs further definition.
- We have little measure of the likely size of the vCJD epidemic.
- The risk of spread of prion diseases on surgical instruments is a real one and the use of single-use instruments for surgery effectively removes this risk.
- Introduction of a change in surgical practice carries its own risks.
- A simple test to detect incubating cases of vCJD would be a huge breakthrough in the management of this disease.