AUTHOR'S REPLY: Dr Goodman's comment on 'dubious statistical reasoning' is unfortunately inaccurate. The probabilities in Table 2 of the paper are based on probability theory relating to conditional probability. The probability of an event A being true, given that event B is true, is equal to the probability of A and B being true divided by the probability of event B being true. Mathematically this is denoted:

$$P(A|B) = P(A \cap B)/P(B)$$

The argument was rehearsed in the paper for a sibship of three. We will demonstrate it here for a sibship of two. The possibilities for a sibship of two are: MM MF FM FF. Thus each of these has a probability of $\frac{1}{4}$. We need to calculate the probability of a sibship of two girls given that at least one sibling is a girl. We make no assumption about the position of the bulimic girl – a fact stated in the paper. Event A is 'all girl sibship' and event B is 'at least one sibling is a girl'. Applying the formula above we get:

$$P(\text{all girl sibship}|\text{at least one sibling is a girl}) = \tfrac{1}{4}/\tfrac{3}{4} \text{ i.e. } \tfrac{1}{3}.$$

The same argument applies for other sibship sizes. The probability of an all-girl sibship is only half if one assumes knowledge of the position of the sibling. Thus if you know the eldest sibling is a girl then the probability of an all-girl sibship is half.

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Transmissible dementias

SIR: The article by Harrison & Roberts (*Journal*, April 1991, **158**, 457–470) draws attention to the variability of pathological findings in the transmissible dementias.

Spongiform change has been held to be the main pathognomonic feature of several transmissible encephalopathies in both man and animals (Masters & Richardson, 1978). The transmissible encephalopathies are known to differ in the distribution and/or type of pathological lesion in the brain. For example, only 10% of Creutzfeldt-Jacob disease (CJD) brains show amyloid plaques, whereas they are seen in all cases of Gerstmann-Straussler syndrome. The clinical manifestations of these diseases are variable between and within disease. CJD may present either subacutely (rapid onset and progression, duration 6 months) or chronically (variable progression, duration 2–4 years), and it has been suggested that this variability could be accounted for, in some familial cases, by phenotypic expression of allelic variants of the prion protein gene (Hardy, 1989).

In the light of current interest in the transmissible dementias, and in establishing precise estimates of disease incidence, it is now important to note that diffuse Lewy body disease (DLB) may be both clinically and pathologically confused with CJD. DLB (also called Lewy body variant of Alzheimer's disease (AD) (Hansen *et al*, 1990), and senile dementia of the Lewy type (Perry *et al*, 1990)) has been clearly shown to exhibit spongiform change in pathological studies. The spongiform change in DLB has been clearly distinguished from the microvacuolation which occurs in the upper layers of cerebral cortex in AD and, in all cases so far reported, it is confined to mesial temporal lobe structures, throughout all cortical layers (Hansen *et al*, 1989; Byrne *et al*, 1989).

In a minority of cases (e.g. Byrne *et al*, 1989) these changes are associated with clinical features suggestive of CJD and in one of our cases (Byrne *et al*, 1989), these were associated with serial electroencephalographic changes consistent with the diagnosis of CJD. One attempt to transmit DLB with spongiform change failed (Hansen *et al*, 1989), and no estimation of prion protein has yet been performed.

Harrison & Roberts summarise the reports of spongiform change in AD, but those cases reported by Smith *et al* (1987) had symptoms suggestive of CJD, although myoclonus has been reported in AD.

In view of the fact that DLB is increasingly recognised as a common disease accounting for up to one-third of cases of dementia, with associated spongiform change in up to 6% of cases (Hansen *et al*, 1989; Byrne *et al*, 1989), the number of cases in which CJD is suspected, because of heightened clinical awareness, will be numerically very significant and is likely to be a confusing variable in studies of transmissible encephalopathies.

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https://doi.org/10.1192/S0007125000142217 Published online by Cambridge University Press