

spina bifida. In studies of births in Great Britain prior to 1979, the prevalence of spina bifida at birth was within the range of 1.5–4.1 per 1,000 births, and, in studies in the early 1980s, the prevalence was within the range of 0.7–1.9 per 1,000 births (Little and Elwood 1992). Some of these infants would have been stillborn. For example, in Northern Ireland during the period 1974–79, 15.5% of 569 cases of spina bifida or encephalocoele were stillborn (Little and Nevin 1989). In Glasgow and Liverpool during the period 1980–92, when fetuses from terminated pregnancies were excluded, 16% of 262 cases of spina bifida were recorded to have resulted in fetal deaths (EUROCAT Working Group 1995). Therefore, it appears that the proportion of cases of childhood cancer with neural tube defects is similar to what would be expected on the basis of data on the prevalence of these defects at birth, in Great Britain.

In the study by Narod et al. (1997), eight of the children with tumors of the brain or of the spinal cord were recorded as having spina bifida, compared with the 5.6 expected on the basis of the frequency of spina bifida among children with other types of cancer in Great Britain and with the 2.4 expected on the basis of the data for British Columbia. Again, the proportion of children with tumors of the brain or of the spinal cord who were recorded as having spina bifida (1.7 per 1,000 births) would appear to be within the range of prevalences at birth reported for Great Britain during the period in which the children included in the study by Narod et al. would have been born. Thus, the study by Narod et al. does not appear to support the hypothesis of a common maternal factor for brain tumors and spina bifida.

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Reply to Little

To the Editor:

My colleagues and I thank Dr. Little (1997 [in this issue]) for his important data. We saw an excess of neural tube defects in children with cancer, in the United Kingdom, compared with healthy controls from British Columbia. It is unclear to what extent the control group from British Columbia was comparable to the children from Britain, and our approach is inadequate when the baseline rates of disease differ for the two countries. Unlike the rates of other malformations, the rate of spina bifida was not significantly greater in children with solid tumors than in those with leukemia. We agree that our data do not allow us to conclude that there is an excess of cancer among children with neural tube defects.

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Lafora Progressive Myoclonus Epilepsy: Narrowing the Chromosome 6q24 Locus by Recombinations and Homozygosities

To the Editor:

Lafora disease (LD) is an autosomal recessive and rare but fatal epilepsy syndrome characterized by stimuli-sensitive myoclonus, absence and grand mal seizures, progressive intellectual and neurological deterioration, and periodic acid Schiff (PAS) stain-positive intracellular inclusion bodies. Eighty-four years after Gonzalo Lafora