Neural tube defects 1974-94—down but not out

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Abstract

Aims—To describe accurately the total prevalence of neural tube defects (NTDs) in England and Wales over time, and to provide a benchmark up to 1994.

Methods—National data about NTDs reported as births or terminations are available from 1974-94, but reporting is incomplete. A local register of NTDs covering Oxfordshire/west Berkshire from 1965-94 was used to validate national data for the locality, using the method of capture and recapture, and hence to estimate incompleteness of reporting nationally.

Results—National underreporting is consistent at about two thirds of the true number of cases reaching at least the second trimester. The local register is much more complete, but time trends locally and nationally are similar. In England and Wales total prevalence declined from about 34 per 10000 live and stillbirths in 1974 to a plateau of just under 8 per 10000 in the 1990s.

Conclusions—The decline in NTD prevalence is real and seems to have stopped. How this relates to changes in diet or the practice of vitamin supplementation is unknown, and the implications of the plateau are uncertain. OPCS figures of 500 NTDs annually in England and Wales represent about two thirds of the true number of cases.

Keywords: neural tube defects, time trends, capture-recapture, folate supplementation.

Methods

The national data were obtained from the abortion notification system (statutory reporting) and the congenital malformation surveillance system (voluntary reporting) at the Office of Population Censuses and Surveys (OPCS). Cases were defined as those records coded as NTD according to the eighth and ninth revisions of the international classification of disease (ICD 8: 740, 741, 743.0, 756.1; ICD 9: 740, 741, 742.0, 756.1, V28.1). Data for Oxfordshire and west Berkshire were obtained from a register of NTDs retrospectively assembled from multiple sources over the past few years at the Oxford Record Linkage Study (ORLS). Population at risk data were all live and stillbirths registered in the corresponding years, available nationally from OPCS. Home and hospital births to Oxfordshire and west Berkshire residents in those two districts are held at the ORLS. As for 1974-94, we used the method of capture-recapture (assuming independence of reporting to the two data collection systems) to compare OPCS counts of NTD terminations/births notified from Oxfordshire/west Berkshire between 1991-4 with the cases known to the initial local register for that period, to assess the relative completeness of each source. We then merged the two to produce the enhanced local register of verified NTD cases.

Results

For the period 1974-90 we had already estimated the initial local register to be 96% complete, OPCS data to be 66% complete, and the final, enhanced local ascertainment (initial register plus OPCS) to be 99% of the true number of cases. For 1991-4, the figures are 77% for the initial register (predictably low because a major source of local information for 1974-90 was not available for 1991-4) and 60% for OPCS, with a local completeness of 91% after enhancement. We have no way of estimating completeness for the local register from 1965-73.

Table 1 shows total prevalence rates of NTDs in England and Wales from 1974-94 and among residents of Oxfordshire/west Berkshire on the enhanced local register who delivered anywhere in these two districts between 1965-94. Over the period 1975-90, national rates declined steeply and continuously from about 34 per 10000 live and stillbirths to reach a plateau prevalence in the 1990s of just under 8 per 10000. In Oxfordshire and west Berkshire, the local rates over the same period varied more widely, because they were based on a much smaller population.
and were almost always higher than the national rates, but they exhibited the same decline to a plateau.

**Discussion**

Although both the national and local datasets are incomplete, the evidence suggests that underascertainment has been consistent for both throughout the period (OPCS > 60%, ORLS > 90% complete), and the estimated trends are unbiased. In both there has been a decline in total prevalence of NTDs from 1974 to an apparent plateau in the 1990s. This picture contrasts starkly with that in South Australia, for instance, where no decline at all occurred between 1966 and 1991, the stable background rate of which we have now achieved. Genetic differences between women reproducing are an implausible explanation for our national trend, but might explain the international differences. Various enzyme defects are now hypothesised to represent the folate-dependent abnormality underlying NTD occurrence. Folate supplementation may universally reduce NTD occurrence, but there is little evidence that changes in diet or vitamin supplementation occurred in ways which can explain the different trends in risk in the two countries. Information about response to folate in populations with different historical patterns of risk will help us to judge whether folate supplementation alone will prevent the occurrence of most NTDs.

Our study provides accurate benchmark levels of NTD occurrence in England and Wales against which to assess the impact of the current folate supplementation campaign. Currently about 500 total cases a year are reported to OPCS, and this is likely to be about two thirds of the true number of cases in which the pregnancy reaches at least the second trimester.

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