Incidence of neural tube defects in liveborn and stillborn infants in British Columbia over a 10-year period

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Materials and methods

Since its inception in 1952 the BCHSR has kept records on virtually all liveborn infants with NTDs.⁷ Data on incidence are also available for stillbirths, although they are not as complete. The BCHSR uses over 80 sources of registration; those important for the identification of infants with NTDs include the physician's notice of birth, vital registration of death and diagnosis at the time of hospital discharge of children under 7 years of age.⁶

Between 1971 and 1980 inclusive the number of live births per year in British Columbia averaged 36 000. Data on the number of liveborn and stillborn infants with NTDs were obtained from the BCHSR's annual report⁸ and were analysed for total variation in incidence rate and for linear trends.⁹ Linear trends were examined for the years 1976 to 1980 inclusive, a period that corresponds with that for similar data from various regions in a recent study by Danks and Halliday.³ Differences were considered significant at the 5% level.

Results

Table I lists the numbers of liveborn and stillborn infants with anencephaly and spina bifida and the total number of infants born with NTDs in each year of the study period. The group of infants with anencephaly also includes a small number of infants with cranioschisis and iniencephaly. In addition to anencephaly these infants may also have had spina bifida or other congenital malformations or both. Infants classified as having spina bifida may also have had other congenital malformations but did not have anencephaly.

Between 1971 and 1980 there was no significant annual variation in the incidence rates for NTDs except for the rate of anencephaly in stillborn infants ($\chi^2 =$ 20.5; p < 0.025; degrees of freedom [df] = 9). Between 1976 and 1980 there was a significant trend towards a linear decline in this rate ($\chi_0^2 = 5.3$; p < 0.01; df = 1). During the latter period eight pregnancies were terminated because prenatal examination had identified anencephaly; however, when these data are included the trend persists and is still significant ($\chi_0^2 = 3.9$; p < 0.05; df = 1).

Discussion

Our results are similar to those from other areas in that the incidence rates appear to be declining more rapidly for an encephaly^{1,3,5} than for spina bifida and for NTDs in stillborn compared with liveborn infants.⁴

Reports of an apparent decline in the incidence of neural tube defects (NTDs) have come from various parts of the world. If these findings are consistent they would have an important impact on prenatal diagnosis and on screening programs. The incidence of NTDs over a 10-year period was examined in British Columbia, a province that has a population-based health surveillance registry through which there is virtually complete ascertainment of liveborn infants with NTDs. The results showed a significant decrease in incidence only for stillborn infants with anencephaly. The increased use of ultrasonography and the subsequent termination of pregnancies in which the fetus has been found to have anencephaly may explain this observation.

Des rapports signalant une baisse apparente de l'incidence des anomalies du tube neural (ATN) sont parvenus de différents points du globe. Si ces résultats devaient s'avérer, ils auraient un impact important sur le diagnostic prénatal et sur les programmes de dépistage. On a étudié l'incidence des ATN sur une période de 10 ans en Colombie-Britannique, une province qui possède un bureau de surveillance sanitaire grâce auquel on peut virtuellement s'assurer de toutes les naissances vivantes d'enfants porteurs d'une ATN. Les résultats ont montré une baisse significative de l'incidence seulement pour les mort-nés avec l'anencéphalie. L'utilisation accrue de l'échographie ultrasonique et les arrêts subséquents de la grossesse lorsque le foetus se révèle atteint d'anencéphalie peuvent expliquer cette observation.

Several centres in various parts of the world have recently reported a trend towards a decrease in the incidence of neural tube defects (NTDs).^{1.5} These observations, if universal, would have a significant impact on programs for both prenatal diagnosis and population screening. British Columbia, with a population of approximately 2.5 million, provides a good opportunity to study this trend because it has an excellent populationbased registry — the BC Health Surveillance Registry (BCHSR).⁶ We studied the incidence of neural tube defects in this province during the 10-year period 1971 through 1980.

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| Year | No. of neural tube defects (and rate per 1000 stillbirths, live births or total births) | | | | | | |
|------|---|-----------|----------|--------------|-----------------|-----------------|-----------------------------|
| | Anencephaly | | | Spina bifida | | | |
| | Stillborn | Liveborn | Total | Stillborn | Liveborn | Total | All neural tube defects* |
| 1971 | 13 (29) | 3 (0.09) | 16 (0.5) | 6 (14) | 22 (0.6) | 28 (0.8) | 44 (1.2) |
| 1972 | 9 (25) | 4 (0.12) | 13 (0.4) | 1 (3) | 25 (0.7) | 26 (0.7) | 39 (1.1) |
| 1973 | 18 (53) | 7 (0.20) | 25 (0.7) | 4 (18) | 32 (0.9) | 36 (1.0) | 61 (1.8) |
| 1974 | 24 (66) | 7 (0.20) | 31 (0.9) | 7 (19) | 29 (0.8) | 36 (1.0) | 67 [68] (1.9) |
| 1975 | 15 (36) | 10 (0.28) | 25 (0.7) | 11 (27) | 36 (1.0) | 47 (1.3) | 72 [75] (2.0) |
| 1976 | 15 (42) | 7 (0.20) | 22 (0.6) | 6 (17) | 25 (0.7) | 31 (0.9) | 53 (1.5) |
| 1977 | 16 (48) | 5 (0.14) | 21 (0.6) | 5 (15) | 21 (0.6) | 26 (0.7) | 47 [49] (1.3) |
| 1978 | 15 (45) | 12 (0.32) | 27 (0.7) | 6 (18) | 22 (0.6) | 28 (0.7) | 55 [58] (1.5) |
| 1979 | 10 (31) | 7 (0.18) | 17 (0.4) | 7 (22) | 27 (0.7) | 34 (0.9) | 51 [53] (1.3) |
| 1980 | 3 (10) | 10 (0.25) | 13 (0.3) | 1 (3) | 22 (0.6) | 23 (0.6) | 36 [37] (0.9) |

One factor that may cause a decrease in the incidence of NTDs among infants is the prenatal detection of affected fetuses and the subsequent termination of pregnancy. However, the overall results for British Columbia do not change when data from the University of British Columbia's prenatal diagnosis clinic are included. This clinic provides most of the prenatal diagnostic services for the province. When we include the fetuses with an NTD detected by prenatal diagnosis that were subsequently aborted, 1978 is the only year in which the total incidence rate for these defects changes, from 1.3 to 1.4/1000 total births, and this change is not significant.

The data suggest that the trend towards a linear decline in the incidence of anencephaly among stillborn infants may be less significant once the data from the prenatal diagnosis clinic are incorporated. However, these results must be interpreted with caution. Owing to the small numbers in our study, it was impossible to predict the annual number of pregnancies terminated because of anencephaly that would have resulted in a stillbirth. To allow data analysis, we therefore assumed that all eight of the fetuses aborted between 1976 and 1980 would have been stillborn. However, according to data for British Columbia⁷ one would expect six of these fetuses to have been stillborn and two to have been liveborn.

Another consideration is that in recent years in British Columbia approximately 50% to 60% of pregnancies have been monitored by ultrasonography for various reasons, such as to establish the gestational age of the fetus. This procedure would identify most fetuses with an encephaly but would be likely to miss those with spina bifida. When an affected fetus is identified by ultrasonography, the pregnancy is terminated under the direction of the physician who requested the ultrasonography and would therefore not necessarily be ascertained by the BCHSR, unlike the terminations associated with the prenatal diagnosis clinic. Province-wide statistics on such terminations are unavailable. The increased use of ultrasonography may therefore be partially responsible for the apparent decline in the incidence of an encephaly.

The incidence rate for NTDs is relatively low in British Columbia, at 1.55/1000 births.⁷ This may be

because the proportion of cases due to environmental factors, such as poor diet,¹⁰ is low. Therefore, if the decreasing incidence rates elsewhere are due to changes in environmental factors, we would not expect to see the same decrease in areas where the incidence is already low. This is borne out by the fact that the most dramatic decreases in the incidence of NTDs have been in areas where there was a fairly high incidence to start with. For example, between 1961 and 1979 in the Liverpool and Bootle areas in England the incidence of anencephaly and spina bifida per 1000 total births decreased from 3.67 to 0.38 and from 4.24 to 2.02 respectively,¹ a much more striking decline than that observed in regions where the incidence rates are considerably lower.²⁴

In summary, we found that the only category of NTDs for which there was a significant decrease in incidence in British Columbia was an encephaly among stillborn infants, the decrease occurring between 1976 and 1980 inclusive.

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