Mortality in Foetal Alcohol Syndrome

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Many papers about foetal alcohol syndrome (FAS) have followed the original publications, and much is now known about how maternal alcohol consumption during pregnancy may affect the foetus. These effects include a wide variety of malformations, intellectual problems including mental retardation and learning disabilities, and miscarriage and foetal death. Little has been written, however, about postnatal mortality. A literature review found only three papers in which mortality is mentioned. Spohr et al. followed 72 patients for 10 years and found that three patients (4%) had died. Two of 40 patients (5%) in a Scottish study had died, though all patients in this cohort were less than 12 years of age. The first 11 patients with FAS reported in the English language literature were reviewed 10 years later by Streissguth et al., the oldest survivor being 14.7 years. Two of the 11 (18%) were dead.

In a cohort of 207 patients whom we have studied, 12 have died. This paper reviews these cases.

METHODS

We carried out an epidemiological study of FAS in Saskatchewan, in which we attempted to identify all known cases of FAS in that province. We restricted our study to those with the full foetal alcohol syndrome. Details of the study have been published in this journal. Briefly, each patient was born in Saskatchewan before January 1, 1993, satisfied standard diagnostic criteria for the syndrome, and had a history of excessive maternal alcohol ingestion in pregnancy, either by self-report of the mother or by report of reliable observers in the mother’s community. Most of the patients were clients of the Alvin Buckwold Child Development Program in Saskatoon, the major referral centre for handicapped children in central and northern Saskatchewan, but cases were referred from other centres and from paediatricians, geneticists, and early childhood psychologists. Many were followed for a number of years and were seen on several occasions. We examined 192 personally at least once and reviewed the records of the other 15, each of whom had been diagnosed by paediatricians experienced in FAS. All except 13 had been born since 1972. We had direct access to records about the death of nine patients. In the others, information was given by correspondents. The cause of death as recorded on the death certificate for each case was confirmed by the Vital Statistics Division of Saskatchewan Health.

The expected number of deaths in the time period reviewed was calculated as follows. We noted the number of patient-years for each year from 1962 to 1992 for the age groups under 1 year, 1 to 14 years, and 15 to 30 years in our cohort. Since 86% of the cohort, and all of those who died, were North American Indians, we used the age-specific death rates for North American Indians to calculate the number of deaths that would be expected to occur in the patient-years in each of the three age-groups. These numbers were then summed to provide an overall expected number of deaths. In order to compare the observed deaths with the expected deaths, the observed deaths were assumed to have a Poisson distribution, the mean being equal to the expected value. A goodness-of-fit statistic based on the chi-square test was used for the comparison.

RESULTS

There were 12 deaths among the 207 identified cases.

Table I shows the patient-years and expected number of deaths in each of the three age groups. The overall expected number of deaths was 6.71 (p=0.04).

A summary of the cases and causes of death is given in Table II. The age at death ranged from 13 days to 29 years and 10 months. Seven were males. All patients were North American Indian. Congenital heart disease was the major factor in seven and a contributing factor in another. Two died from liver disease, two from systemic infection, and one from congenital abnormalities of the respiratory tract.

DISCUSSION

Our mortality rate of 5.8% is in keeping with the high rate seen in previously reported studies. The actual death rate in our patients is 3.5 times what would be expected in the general population. However, the majority of our patients were North American Indians, and mortality rates for that group have been consistently higher than for the rest of the population. Therefore, we calculated the expected

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This study was conducted with the aid of a research grant from the Health Services Utilisation and Research Commission of Saskatchewan.
The high mortality rate in our cohort could be an underestimate for two reasons. First, though we have been able to confirm the vital status of 185 patients in our cohort, we do not know the current status of 22. Some have left the province or been adopted, and their new addresses are not available to us. Second, some deceased FAS patients could have died before the diagnosis had been established or before referral to us or to one of our referral sources.

The deaths in this series were due primarily to the congenital anomalies caused by FAS. Eight of our patients in this series had a heart defect, in seven it was the direct cause of death, and in another it was a major contributory factor. Heart malformations were present in 34 (17%) of our total cohort of 207 patients, a figure very similar to the 18% of 550 case reports of FAS compiled by Abel.9 Congenital cardiac lesion in our cohort was an isolated ventricular septal defect, which occurred in 10 patients, while tetralogy of Fallot was seen in 7, including 5 reported here. This is a higher incidence of tetralogy of Fallot than the 0.9% reported in Abel’s series.8 Two patients died from liver disease, one secondary to extrahepatic biliary atresia and the other from the effects of what was diagnosed as “congenital cirrhosis”. This latter patient and two others, still alive, were previously described by us.19 One other patient died from the complications of congenital defects of the upper respiratory tract. Only one patient of the 12 did not have any congenital malformations (other than the facial abnormalities seen as part of the syndrome). Most of the patients described here had several surgical procedures, and a very complicated medical and social history. We could find no evidence that congenital malformations are commoner in American Indians than in other races,20-22 which suggests that the high proportion of patients with congenital malformations in our cohort is a consequence of FAS, and not of racial factors alone.

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Date of Death</th>
<th>Age</th>
<th>Underlying Cause of Death</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>5/77</td>
<td>5.5ms</td>
<td>Tetralogy of Fallot</td>
<td>Waterston procedure</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>2/80</td>
<td>4yr4ms</td>
<td>Tetralogy of Fallot</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>6/80</td>
<td>5yr3ms</td>
<td>Tetralogy of Fallot</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>9/80</td>
<td>1yr3ms</td>
<td>H. Influenzae meningitis</td>
<td>H. Influenzae meningitis</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>11/83</td>
<td>5yr9ms</td>
<td>Tetralogy of Fallot</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>3/84</td>
<td>1yr11ms</td>
<td>Bronchopneumonia, septicaemia</td>
<td>Bronchopneumonia, septicaemia</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>9/84</td>
<td>10yr7ms</td>
<td>Tetralogy of Fallot</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>8/86</td>
<td>5ms</td>
<td>Congenital extra-hepatic biliary atresia</td>
<td>Congenital extra-hepatic biliary atresia</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>2/88</td>
<td>7yr6ms</td>
<td>Ventricular and atrial septal defect</td>
<td>Ventricular and atrial septal defect</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>7/89</td>
<td>13days</td>
<td>Hypoplastic left heart syndrome</td>
<td>Hypoplastic left heart syndrome</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>9/92</td>
<td>29yr10ms</td>
<td>Congenital cirrhosis</td>
<td>Congenital cirrhosis</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>4/93</td>
<td>2yr2ms</td>
<td>Tracheoerythromalacia; laryngeal webbing</td>
<td>Tracheoerythromalacia; laryngeal webbing</td>
</tr>
</tbody>
</table>

### Table I
Calculation of Expected Number of Deaths in a Cohort of 207 Cases of FAS

<table>
<thead>
<tr>
<th>Period</th>
<th>Patient -Years</th>
<th>&lt; 1 Year M.R.*</th>
<th>Expected Deaths</th>
<th>Age Group 1 - 14 Years</th>
<th>Patient -Years</th>
<th>M.R.*</th>
<th>Expected Deaths</th>
<th>Age Group 15 - 30 Years</th>
<th>Patient -Years</th>
<th>M.R.*</th>
<th>Expected Deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>1962-71</td>
<td>10</td>
<td>48</td>
<td>0.48</td>
<td>44</td>
<td>1</td>
<td>0.044</td>
<td></td>
<td></td>
<td>18</td>
<td>4.5</td>
<td>0.08</td>
</tr>
<tr>
<td>1972-81</td>
<td>87</td>
<td>32</td>
<td>2.78</td>
<td>397</td>
<td>0.74</td>
<td>0.29</td>
<td></td>
<td></td>
<td>193</td>
<td>2.0</td>
<td>0.39</td>
</tr>
<tr>
<td>1981-92</td>
<td>110</td>
<td>15</td>
<td>1.65</td>
<td>1353</td>
<td>0.74</td>
<td>1.00</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0.47</td>
</tr>
<tr>
<td>TOTAL</td>
<td>4.91</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Expected Number of Deaths = 6.71 (p=0.04)

* Age-specific mortality rates in Saskatchewan Indians, per 1,000
The sequelaes of FAS are considerable for the patients, their families, and for society. To the long list of consequences of a mother drinking excessively in pregnancy must be added the possibility of the premature death of her child.

ACKNOWLEDGEMENTS

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REFERENCES


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