STUDIES OF CEREBRAL PALSY IN THE CHILDHOOD POPULATION OF EDINBURGH.

by

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Part 4.

The prevalence of cerebral palsy and its social aspects in Edinburgh.
CHAPTER 1.

The prevalence of cerebral palsy amongst children in the City of Edinburgh and the problem of cerebral palsy in the community.

As a result of the survey it is possible to present some estimate of the prevalence of cerebral palsy amongst children in Edinburgh born between 1938 and 1952 inclusive, and indicate the extent to which patients were handicapped.

The prevalence of cerebral palsy by year of birth. The numbers of patients and the prevalence of cerebral palsy by year of birth are shown in Table 174. The prevalence of ascertained cerebral palsy amongst all children born between 1938 and 1952 was 1.991 cases per 1000. It will be observed, however, that there are considerable differences in the prevalence between the various three-year period. For example, whilst 2.546 cases per 1000 were found in the child population born between 1944 and 1946, only 1.132 cases per 1000 were ascertained amongst children born between 1950 and 1952.

The apparently lower prevalence of cerebral palsy amongst younger children is partly the result of their being a lower proportion of acquired cases than in older age groups. Though the majority of children whose cerebral palsy is of post natal origin acquire the condition within the first three years of life, it is to be expected that a few children born between 1950 and 1952 acquired cerebral palsy after the completion of the survey. (In fact 4 children are known to have acquired hemiplegia and one each ataxia and ataxic diplegia/
Table I74.

Prevalence of cerebral palsy by year of birth in Edinburgh.

<table>
<thead>
<tr>
<th>Year of Birth</th>
<th>Population</th>
<th>Number of patients</th>
<th>Prevalence per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Hemip.</td>
<td>Diplegia</td>
</tr>
<tr>
<td>1938</td>
<td>6749</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>1939</td>
<td>6574</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>1940</td>
<td>6209</td>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>1941</td>
<td>6099</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>1942</td>
<td>6388</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>1943</td>
<td>6538</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>1944</td>
<td>6533</td>
<td>7</td>
<td>11</td>
</tr>
<tr>
<td>1945</td>
<td>5993</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>1946</td>
<td>7902</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>1947</td>
<td>8900</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>1948</td>
<td>7700</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>1949</td>
<td>7500</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>1950</td>
<td>7200</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>1951</td>
<td>7100</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>1952</td>
<td>6900</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>1938-52, 144285</td>
<td>75</td>
<td>79</td>
<td>208</td>
</tr>
</tbody>
</table>

* Based on figures obtained from Edinburgh Education Authority and the Registrar General.
diplegia since this time). This lack of acquired cases is less important in reducing the apparent numbers of patients with cerebral palsy than is defective ascertainment. Ascertainment was relatively defective in this age group for a number of reasons. It was impossible to survey such large numbers of preschool children than those who were more readily accessible in schools. Ascertainment had to be on the basis of second hand reports much more frequently. Moreover since there is a considerable delay in the diagnosis of cerebral palsy and a further lapse of time before children are referred to treatment centres, it was inevitable that in many instances affected children did not become known to the survey. The delay in diagnosis and the institution of treatment was well illustrated by the histories of the 73 patients suffering from congenital diplegia. Sixty-four were noted to have symptoms by their parents before the age of 18 months, and in 58 they were taken to their doctors before this age. Yet the majority of cases had to wait for between 18 months and 2 years before the diagnosis was established. In 14 diplegia was recognised only after the age of 4 years.

The apparently lower prevalence of cerebral palsy amongst children born between 1933 and 1943 than between 1944 and 1949 appears to be due to a variety of factors. The same sources of information were available in both groups, and since the number of routine examinations was greater in the older school children than the younger, it might be expected that there /
there was a rather greater chance of cerebral palsy being diagnosed. It is difficult to attribute the smaller prevalence of cerebral palsy in older school children born between 1933 and 1943 to defective ascertainment. It seems probable that there is a real difference in prevalence amongst children born between 1933 and 1943 and those born between 1944 and 1949.

One possible reason for this difference is that patients suffering from cerebral palsy died before reaching the age of 10 years, and thus the numbers of patients above this age was smaller than they would have been had they survived. Unfortunately it is impossible to obtain any accurate information about the death rate amongst children suffering from cerebral palsy. It has been suggested that one in 7 patients perish in infancy, (Wyllie, 1951).

Five patients of the 203 included in this survey are known to have died in the 6 years since it was completed, all between the ages of 4 and 14 years. Pneumonia caused the deaths of three and status epilepticus one. In one the cause was unknown. The commonest causes of death appear to be complicating respiratory infections which occur most commonly in severely affected patients suffering from bilateral cerebral palsy, and epilepsy. It is interesting, therefore, to observe that there is a higher proportion of patients suffering from severe bilateral cerebral palsy amongst those born between 1944 and 1949 than there is between 1933/
1938 and 1943. It seems likely that improved treatment of complicating respiratory infections with antibiotics, especially penicillin, may have permitted the survival of severely affected patients who would have died before antibiotics were available.

Another cause of the apparent excess of patients born between the years 1944 and 1949 compared to 1938 and 1943 appears to be the improved chances of survival of children suffering from conditions known to cause, or be associated with cerebral palsy. Thus there is a higher proportion of patients suffering from acquired cerebral palsy amongst those born after 1943, (Table 175). Patients suffering from meningitis, septic thrombophlebitis, parainfectious encephalopathy and traumatic head injury would have died in the pre-antibiotic era now survive to show acquired hemiplegia, ataxic diplegia or ataxia.

Similarly, survival rates of premature infants have improved greatly in recent years. The infant mortality in Edinburgh halved between 1938 and 1952 and the mortality of premature infants weighing between 3 and 5½ pounds was reduced by 39% in one Edinburgh hospital. These changes are reflected in a higher proportion of prematurely born diplegic patients born between 1944 and 1949 compared to earlier years, Table 176. As increasing numbers of very small premature infants survive, the numbers of diplegic patients may also be expected to increase for the incidence of diplegia appears to increase.
Table 175.

Relative numbers of cases of acquired hemiplegia in 1935-1943 and 1944-1952 compared.

<table>
<thead>
<tr>
<th>Period</th>
<th>Number of cases of hemip.</th>
<th>Number of cases of acquired hemip.</th>
<th>Number of cases of acquired hemip. treated with penicillin.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1939-43</td>
<td>28</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>1944-52</td>
<td>47</td>
<td>25</td>
<td>17</td>
</tr>
<tr>
<td>1938-52</td>
<td>75</td>
<td>33</td>
<td>19</td>
</tr>
</tbody>
</table>

* Both developed infections which were treated with penicillin after 1943.

Table 176.

Numbers of diplegic patients and prematurely born diplegic patients ascertained in Edinburgh who were born in the years 1938-43 and 1944-49.

<table>
<thead>
<tr>
<th>Date of birth</th>
<th>Number of diplegic patients</th>
<th>Number of diplegic patients prematurely born</th>
</tr>
</thead>
<tbody>
<tr>
<td>1938-43</td>
<td>22</td>
<td>5</td>
</tr>
<tr>
<td>1944-49</td>
<td>45</td>
<td>19</td>
</tr>
</tbody>
</table>
increase inversely with birth weight amongst premature babies. (Ingram and Kerr, 1954). Two cases of hydrocephalus born within 1944 and 1949 would probably have died had recently developed neurosurgical techniques not been employed. One survived to show ataxia, the other ataxic diplegia.

Had there been a similar proportion of diplegic patients born prematurely, a similar proportion of children suffering from severe bilateral cerebral palsy and of cases of acquired hemiplegia and a similar proportion of patients suffering from the after effects of meningitis in the two six-year periods, the prevalence of cerebral palsy would have been very little lower amongst children born between 1933 and 1943 compared to 1944 and 1949.

It seems probable, therefore, that the observed difference in prevalence is a real one. It must be attributed to improved survival of severely affected patients suffering from cerebral palsy and the lower mortality in conditions which cause or are associated with cerebral palsy. The major part of these changes appear to be the result of the introduction of penicillin into clinical practice towards the end of the second world war which is the period, in fact, when the prevalence of cerebral palsy appears to increase.

The prevalence of cerebral palsy in different wards in the City of Edinburgh. The numbers of patients suffering from cerebral palsy in each of the 23 Wards in the City of Edinburgh were noted. Since approximate populations of children under the age of 15 could be obtained from the Census carried /
The prevalence of cerebral palsy in Edinburgh wards grouped according to the infant mortality, into good, intermediate and poor wards.


<table>
<thead>
<tr>
<th>Ward</th>
<th>Infant mortality</th>
<th>Population aged 0-15 yrs</th>
<th>Hemiplegia</th>
<th>Diplegia</th>
<th>Cerebral palsy</th>
<th>Incidence per 1000 under 16 yrs</th>
<th>All ages</th>
<th>Incidence of infectious disease for 1950, 51, 52 per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Poor wards</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>St. Giles</td>
<td>34</td>
<td>4,193</td>
<td>6</td>
<td>4</td>
<td>15</td>
<td>1,221</td>
<td>.814</td>
<td>3,053</td>
</tr>
<tr>
<td>Liberton</td>
<td>35</td>
<td>4,139</td>
<td>4</td>
<td>4</td>
<td>12</td>
<td>.652</td>
<td>.62</td>
<td>1,955</td>
</tr>
<tr>
<td>Collinon</td>
<td>31</td>
<td>4,188</td>
<td>3</td>
<td>5</td>
<td>14</td>
<td>.716</td>
<td>1,125</td>
<td>3,343</td>
</tr>
<tr>
<td>Pilten</td>
<td>33</td>
<td>3,772</td>
<td>10</td>
<td>3</td>
<td>17</td>
<td>1,057</td>
<td>.534</td>
<td>1,814</td>
</tr>
<tr>
<td>St. Bernards</td>
<td>38</td>
<td>3,391</td>
<td>4</td>
<td>3</td>
<td>9</td>
<td>1,215</td>
<td>.912</td>
<td>2,755</td>
</tr>
<tr>
<td>Portobello</td>
<td>31</td>
<td>5,630</td>
<td>5</td>
<td>1</td>
<td>8</td>
<td>.888</td>
<td>.179</td>
<td>1,421</td>
</tr>
<tr>
<td>Craigmillar</td>
<td>30</td>
<td>3,866</td>
<td>5</td>
<td>5</td>
<td>14</td>
<td>.898</td>
<td>.835</td>
<td>2,515</td>
</tr>
<tr>
<td>Colinton</td>
<td>32</td>
<td>3,730</td>
<td>1</td>
<td>3</td>
<td>6</td>
<td>.288</td>
<td>.805</td>
<td>1,609</td>
</tr>
<tr>
<td>Holyrood</td>
<td>32</td>
<td>4,100</td>
<td>3</td>
<td>5</td>
<td>12</td>
<td>.652</td>
<td>1,085</td>
<td>2,603</td>
</tr>
<tr>
<td>Totals</td>
<td>33.2</td>
<td>47,439</td>
<td>41</td>
<td>35</td>
<td>107</td>
<td>.850</td>
<td>.735</td>
<td>2,255</td>
</tr>
<tr>
<td>Intermediate wards</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Newington</td>
<td>29</td>
<td>3,763</td>
<td>6</td>
<td>3</td>
<td>11</td>
<td>1,593</td>
<td>.797</td>
<td>2,923</td>
</tr>
<tr>
<td>St. Andrews</td>
<td>29</td>
<td>3,303</td>
<td>1</td>
<td>2</td>
<td>6</td>
<td>.301</td>
<td>.605</td>
<td>1,817</td>
</tr>
<tr>
<td>Broughton</td>
<td>26</td>
<td>3,292</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>.301</td>
<td>.602</td>
<td>.902</td>
</tr>
<tr>
<td>West Leith</td>
<td>27</td>
<td>3,764</td>
<td>2</td>
<td>3</td>
<td>7</td>
<td>.331</td>
<td>.777</td>
<td>1,850</td>
</tr>
<tr>
<td>Grangemouth</td>
<td>27</td>
<td>3,860</td>
<td>4</td>
<td>2</td>
<td>12</td>
<td>.699</td>
<td>1,281</td>
<td>2,755</td>
</tr>
<tr>
<td>Merchiston</td>
<td>36</td>
<td>3,599</td>
<td>0</td>
<td>2</td>
<td>4</td>
<td>.625</td>
<td>.770</td>
<td>1,559</td>
</tr>
<tr>
<td>Totals</td>
<td>31.2</td>
<td>31,443</td>
<td>21</td>
<td>28</td>
<td>60</td>
<td>.672</td>
<td>.832</td>
<td>1,909</td>
</tr>
<tr>
<td>Good wards</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>George Square</td>
<td>21</td>
<td>2,629</td>
<td>5</td>
<td>2</td>
<td>7</td>
<td>1,767</td>
<td>.707</td>
<td>2,474</td>
</tr>
<tr>
<td>Morningside</td>
<td>14</td>
<td>2,590</td>
<td>0</td>
<td>3</td>
<td>5</td>
<td>1,158</td>
<td>.912</td>
<td>1,931</td>
</tr>
<tr>
<td>Sighthill</td>
<td>17</td>
<td>7,813</td>
<td>4</td>
<td>7</td>
<td>14</td>
<td>.494</td>
<td>.951</td>
<td>1,922</td>
</tr>
<tr>
<td>George/Balfron</td>
<td>17</td>
<td>3,629</td>
<td>2</td>
<td>2</td>
<td>7</td>
<td>.632</td>
<td>.452</td>
<td>1,583</td>
</tr>
<tr>
<td>Gorgie/Shankly</td>
<td>21</td>
<td>3,309</td>
<td>2</td>
<td>2</td>
<td>6</td>
<td>.604</td>
<td>.604</td>
<td>1,813</td>
</tr>
<tr>
<td>Linlith/Comely</td>
<td>23</td>
<td>3,304</td>
<td>0</td>
<td>2</td>
<td>2</td>
<td>.835</td>
<td>.835</td>
<td>2,846</td>
</tr>
<tr>
<td>Totals</td>
<td>19.9</td>
<td>22,738</td>
<td>13</td>
<td>18</td>
<td>41</td>
<td>.52</td>
<td>.72</td>
<td>1,803</td>
</tr>
<tr>
<td>City of Edinburgh</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1951 census returns</td>
<td>28.3</td>
<td>101,625</td>
<td>75</td>
<td>79</td>
<td>208</td>
<td>0.735</td>
<td>0.704</td>
<td>2.046</td>
</tr>
</tbody>
</table>
carried out by the Registrar General in 1951, it was possible to calculate the prevalence for each Ward.

It will be seen that the prevalence varies from .368 cases per 1000 of the child population in Murrayfield/Gramond to 3.343 per 1000 in St. Giles. (Table 177.) Because of the small number of patients in individual wards considerable differences in prevalence were to be expected. Nevertheless intensive efforts were made to find reasons which might have made ascertainment less adequate in districts showing relatively few cases. The possibility that cases were being missed in outlying wards, or in districts in which a high proportion of the children went to private schools were considered. It was not possible to find any consistent trend in the prevalence figures by Wards to support the hypothesis that ascertainment had been uneven. It seemed likely that there were quite large actual differences in the prevalence in different Wards.

Reasons for these differences will be discussed in more detail when the social etiology of cerebral palsy is considered.

Comparison of the prevalence of cerebral palsy found in the present survey with those found in other surveys. An attempt has been made to compare the prevalence of cerebral palsy obtained as a result of this survey with those found in other surveys of urban areas in England, and with figures obtained from the British Council for the Welfare of Spastics for Wales and England. It will be seen that these vary /
<table>
<thead>
<tr>
<th>Authority</th>
<th>Town</th>
<th>Prevalence of Cerebral Palsy per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>British Council for the Welfare of Spastics, (1948) Selected urban areas</td>
<td>Wallasey</td>
<td>2.4</td>
</tr>
<tr>
<td></td>
<td>Norwich</td>
<td>2.0</td>
</tr>
<tr>
<td></td>
<td>Salford</td>
<td>2.0</td>
</tr>
<tr>
<td></td>
<td>Oldham</td>
<td>1.9</td>
</tr>
<tr>
<td>Cohen, (1954)</td>
<td>Sheffield</td>
<td>1.9</td>
</tr>
<tr>
<td>Smallwood, (1953)</td>
<td>Bristol</td>
<td>2.0</td>
</tr>
<tr>
<td>This survey (1952-53)</td>
<td>Edinburgh</td>
<td>2.3</td>
</tr>
</tbody>
</table>
vary from 1.0 case of cerebral palsy per 1,000 children of school age to 2.4 cases per 1,000.

The reasons for the variations in the figures for prevalence obtained are probably related more closely to the efficiency of ascertainment in the surveys than to differences in the actual prevalence of cerebral palsy. But this is by no means as certain as has been suggested by some recent authors. For example, the relationship of the prevalence of cerebral palsy of congenital origin to variations in the infant mortality rate is by no means clear. It is likely that places with a relatively high infant mortality rate, and presumably, therefore, a lower standard of maternal and infant care may show differences in the incidence of cerebral palsy from those where the standard of care is higher.

Nevertheless it is probable that the figures in Table 173, for Wallasey, Norwich, Oldham, Salford, Sheffield and Bristol, are roughly comparable to those of the present survey. They were all obtained from urban areas and were the result of special efforts to ascertain the prevalence of cerebral palsy in the populations of school age. It will be seen that they all give prevalence of between 1.9 and 2.4 cases of cerebral palsy per 1,000 children of school age.

Comparison of the prevalence obtained in this survey with that obtained in other studies in which fewer sources were contacted. An attempt was made to make the survey comparable /
Table 179.
Sources of ascertainment in 208 patients with cerebral palsy in Edinburgh.

<table>
<thead>
<tr>
<th>Source</th>
<th>Number of patients</th>
<th>Cumulative totals</th>
<th>Prevalence per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>School Medical Service and School for spastics</td>
<td>97</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Public Health Department</td>
<td>16</td>
<td></td>
<td></td>
</tr>
<tr>
<td>General Practitioners</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total known to Authorities before present survey</td>
<td>114</td>
<td>II4</td>
<td>1.094</td>
</tr>
<tr>
<td>Hospitals</td>
<td>52</td>
<td>I56</td>
<td>1.59</td>
</tr>
<tr>
<td>Institutions for the mentally defective</td>
<td>8</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Personal cases</td>
<td>34</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Grand total</td>
<td>208</td>
<td></td>
<td>1.991</td>
</tr>
</tbody>
</table>

Table 180.
Prevalence of cerebral palsy in school children ascertained by Local Authorities and by recent surveys.

<table>
<thead>
<tr>
<th>Author</th>
<th>Source of data.</th>
<th>Place.</th>
<th>Prevalence per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asher and Schonell(1950)</td>
<td>School M.Os, Public Health Authority and General practitioners</td>
<td>Birmingham.</td>
<td>1.0</td>
</tr>
<tr>
<td>Dunsdon, (1952)</td>
<td>Local Authorities</td>
<td>Various.</td>
<td>1.3</td>
</tr>
<tr>
<td>Holoran, (1952)</td>
<td>School M.Os, Public Health Authorities and Leeds.</td>
<td>Leeds.</td>
<td>1.6</td>
</tr>
</tbody>
</table>
comparable to studies in which only the cases known to public health and school medical authorities and to general practitioners were included. In every case the way in which the patient came to the notice of the survey for the first time was recorded. This is shown in Table 179. It is thus possible to compare, in a rough way, the incidence obtained in different places where only a proportion of the possible sources of cases of cerebral palsy were contacted.

If one had relied on questionnaires distributed to Edinburgh public health and school medical authorities and to general practitioners, 114 patients with cerebral palsy would have become known. This is equivalent to a prevalence of 1.094 cases per 1,000 children born between 1933 and 1952, or approximately half the figure for prevalence found during the more complete survey. The figure of 1.094 patients per 1,000 children in the population agrees very closely however with figures obtained as a result of questionnaires to various unselected urban authorities in England, by the British Council for the Welfare of Spastics. It is similar also to figures obtained as a result of a number of other investigations. Table (Asher and Schonell, 1950; Dunsdon, 1952).

Similarly, if cases obtained as a result of contacting hospitals are added to those obtained from general practitioners and public health and school medical authorities, a prevalence for cerebral palsy amongst Edinburgh children of 1.59 cases per 1,000 of the population is obtained. This estimate /
Table I8I.

PROPORTIONAL DISTRIBUTION OF VARIOUS TYPES OF CEREBRAL PALSY IN RECENT SURVEYS

<table>
<thead>
<tr>
<th>Year</th>
<th>No. of patients</th>
<th>This Survey</th>
<th>Asher and Schonell</th>
<th>Evans</th>
<th>Hellebrandt</th>
<th>Dunsdon</th>
<th>Special Schools</th>
<th>Selected Areas</th>
</tr>
</thead>
<tbody>
<tr>
<td>1953</td>
<td>208</td>
<td>36.65%</td>
<td>29%</td>
<td>9%</td>
<td>24%</td>
<td>13%</td>
<td>35.5%</td>
<td></td>
</tr>
<tr>
<td>1950</td>
<td>349</td>
<td>37.95%</td>
<td>29%</td>
<td>9%</td>
<td>24%</td>
<td>13%</td>
<td>35.5%</td>
<td></td>
</tr>
<tr>
<td>1948</td>
<td>112</td>
<td>36.7%</td>
<td>29%</td>
<td>9%</td>
<td>24%</td>
<td>13%</td>
<td>35.5%</td>
<td></td>
</tr>
<tr>
<td>1950</td>
<td>531</td>
<td>13%</td>
<td>29%</td>
<td>9%</td>
<td>24%</td>
<td>13%</td>
<td>35.5%</td>
<td></td>
</tr>
<tr>
<td>1952</td>
<td>780</td>
<td>6%</td>
<td>29%</td>
<td>9%</td>
<td>24%</td>
<td>13%</td>
<td>35.5%</td>
<td></td>
</tr>
</tbody>
</table>

Hemiplegia
Diplegia
Type not stated
Paraplegia
Tetraplegia
Trilateral hemiplegia
Diplegia and ataxia
Ataxia
Dyskinesia
Other
Mixed forms

100% 99.9% 100% 100% 100% 100%
estimate agrees with that of certain other investigations utilising these sources. (Helenan, 1952).

On the basis of these comparisons, and more significantly on the comparisons with the more extensive surveys of cerebral palsy in childhood, it seems justifiable to state that the overall prevalence of the condition appears not to vary very greatly from urban area to urban area in Great Britain.

Comparison of the proportional distribution of cases suffering from various types of cerebral palsy in different surveys. It is difficult to compare the relative frequency of the different forms of cerebral palsy in different surveys because of the great variations in terminology found in them. In Table 131 an attempt has been made to compare the figures obtained in different investigations. It cannot be regarded as strictly accurate because it has been necessary to depart from the classifications adopted by the investigators themselves and only in some were sufficient details of cases given to make possible reclassification with any confidence. Nevertheless, it is probable that the cases in the table give some idea of the frequency of the various types of cerebral palsy in the different surveys.

It will be observed that there are considerable variations in the proportion of cases of each type of cerebral palsy encountered. Thus in the survey of Evans, the proportion of cases of hemiplegia is low. This is probably because his survey was based on the results of the examination of children who were potential admissions to a special school for /
for cerebral palsy. Since a high proportion of hemiplegic patients can manage at normal schools few might be expected to be seen in this survey. On the other hand the number of patients with dyskinesia who can cope with normal schooling is smaller, and these children, because they tend to suffer less intellectual impairment than those with other forms of cerebral palsy would be encountered. The proportion of cases of dyskinesia was higher and of hemiplegia lower in Dunsdon's survey of children who were candidates for special schools and contrasts with her figures for selected areas, in which cases were unselected.

It is of interest that the figures obtained in this survey are approximately similar to those found in the regional surveys of Asher and Schonell and Dunsdon, in which cases were unselected.

It seems probable that a proportion of cases suffering from the dystonic stage of diplegia were classed as diplegia in the present survey and as dyskinesia in the others. This would clearly tend to result in a higher incidence of diplegia and a lower incidence of dyskinesia in the present survey.

It is interesting that the only survey reporting a comparable proportion of ataxic patients was that of Hellebrandt, which was based on the examination of 531 cases of ataxia in Phelp's clinic. It is possible that ataxia, being the most difficult form of cerebral palsy to diagnose unless the condition is gross, has been missed in a number of patients in the/
### Table I82a.

#### SEVERITY OF CEREBRAL PALSY IN 208 PATIENTS

<table>
<thead>
<tr>
<th>Type of Palsy</th>
<th>No. of Mild Cases</th>
<th>Moderately Severe</th>
<th>Severe</th>
<th>Totals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia   ...</td>
<td>27</td>
<td>26</td>
<td>22</td>
<td>75</td>
</tr>
<tr>
<td>Diplegia     ...</td>
<td>13</td>
<td>35</td>
<td>31</td>
<td>79</td>
</tr>
<tr>
<td>Diplegia and ataxia</td>
<td>4</td>
<td>8</td>
<td>3</td>
<td>15</td>
</tr>
<tr>
<td>Ataxia       ...</td>
<td>4</td>
<td>8</td>
<td>3</td>
<td>15</td>
</tr>
<tr>
<td>Dyskinesia   ...</td>
<td>4</td>
<td>6</td>
<td>7</td>
<td>17</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>Other         ...</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>52</strong></td>
<td><strong>80</strong></td>
<td><strong>76</strong></td>
<td><strong>208</strong></td>
</tr>
</tbody>
</table>

(25%) (38%) (37%) (100%)

### Table I82b.

#### EXTENT OF CEREBRAL PALSY IN 208 PATIENTS

<table>
<thead>
<tr>
<th>Type of Palsy</th>
<th>Two Useful Upper Limbs</th>
<th>One Useful Upper Limb</th>
<th>No Useful Upper Limb</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia   ...</td>
<td>27</td>
<td>48</td>
<td>0</td>
<td>75</td>
</tr>
<tr>
<td>Diplegia     ...</td>
<td>33</td>
<td>19</td>
<td>27</td>
<td>79</td>
</tr>
<tr>
<td>Diplegia with ataxia</td>
<td>4</td>
<td>6</td>
<td>2</td>
<td>12</td>
</tr>
<tr>
<td>Ataxia       ...</td>
<td>15</td>
<td>0</td>
<td>0</td>
<td>15</td>
</tr>
<tr>
<td>Dyskinesia   ...</td>
<td>3</td>
<td>3</td>
<td>11</td>
<td>17</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>Other         ...</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>83</strong></td>
<td><strong>76</strong></td>
<td><strong>49</strong></td>
<td><strong>208</strong></td>
</tr>
</tbody>
</table>

(40%) (37%) (23%) (100%)
the other surveys. Unless ataxia is tested for extremely carefully, it is very easy to miss its presence and, as has been noted, a number of ataxic patients show negative Romberg tests though they may be quite severely affected. Yet the Romberg test has been regarded as being the soundest and most convenient means of testing for ataxia by many text books on clinical neurology. (Monrad-Krohn, 1943).

The sex distribution of children suffering from cerebral palsy. In the series of 203 children suffering from cerebral palsy, 126 or 62.5% were male, and 52 or 29.5% female. This distribution is similar to that reported by a number of recent surveys, (Asher and Schonell, 1950; British Council for the Welfare of Spastics, 1943). In other surveys a more equal sex distribution has been reported, (Cohen, 1954; Floyer, 1954).

Females outnumbered males in only two forms of cerebral palsy, bilateral hemiplegia and dyskinesia. The excess in dyskinesia appeared to be due to there being more girls than boys suffering from the after effects of kernicterus.

The severity of the cerebral palsy. Different criteria were used to assess severity in each type of cerebral palsy. This means that a child considered to suffer from a severe form of one type of cerebral palsy may be able to lead a much more normal life than a child who suffers from a different type.
type of cerebral palsy, which may be considered to be only
moderately severe. For example, children with severe hemi-
plegia or ataxia are likely to be considerably more independent
than a patient with moderately severe diplegia or bilateral
hemiplegia.

By taking account of the extent of the disability as well
as its severity, however, it is possible to obtain some idea
of the total physical handicap resulting from impairment of
the use of the limbs.

The severity and extent of the cerebral palsy in the
different categories is shown in Tables 182a, 182b. The
majority of the 52 children considered to be mildly affected
were able to take part in most normal activities provided
associated disabilities such as mental defect, blindness, or
hearing impairment were not present. Patients considered to
be moderately severely affected were unable to lead anything
approaching normal lives and those with severe cerebral palsy
were very dependent and handicapped to the extent that even
self care was impossible. It will be seen from the table
that 25% of patients were considered to be mildly physically
handicapped and that over half of them suffered from hemiplegia.
38% were classified as being moderately severely handicapped.
Walking was possible or was expected to be achieved eventually
in all of the 45 patients in this category. All the patients
had at least one functionally useful upper limb, and in 26 the
upper limbs were either functionally normal or only slightly
affected. Forty-five patients showed unilateral upper limb
involvement /
involvement of such severity that normal activities were impossible with the affected hand. In 9 patients there was considerable impairment of manipulation bilaterally. Though physically capable of self care eventually, the majority of the moderately severely affected patients were very dependent upon their parents in early childhood, and required special education provision. It was considered that very few would later be capable of finding employment in a competitive labour market.

Severely affected patients numbered 76 or 37% of the series. In approximately one-third, not even supported walking was possible or likely to be achieved, and even hemiplegic children walked very late and were often unsteady for many years. Five of the 76 patients considered to be severely affected had two useful hands, though there was some neurological involvement in the upper limbs in them all. In 31 there was one functionally useful upper limb and in 40 normal manipulation was impossible with either hand. Only the hemiplegic patients were likely to achieve independence in self care. The majority of children in this category required some help in washing, dressing, toileting or feeding and were likely to need it for the rest of their lives. Only the hemiplegic patients were physically capable of even sheltered employment.

Associated disabilities in children suffering from cerebral palsy. If paresis of the limbs was the only manifestation of cerebral palsy the vast majority of patients would /
A comparison of the percentage distribution of intelligence quotients in this survey with that found in other recent investigations, and in a group of unaffected Scottish children.

### A. Intelligence quotients

<table>
<thead>
<tr>
<th>130 or more</th>
<th>115 to 129</th>
<th>100 to 114</th>
<th>85 to 99</th>
<th>70 to 84</th>
<th>55 to 69</th>
<th>Under 55</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dunsmon 1952</td>
<td>0.5</td>
<td>2</td>
<td>6.2</td>
<td>15.5</td>
<td>17.2</td>
<td>23.6</td>
</tr>
<tr>
<td>This survey</td>
<td>4.9</td>
<td>12.8</td>
<td>14.3</td>
<td>24.2</td>
<td>21.9</td>
<td>21.9</td>
</tr>
</tbody>
</table>

### B. Intelligence quotients

<table>
<thead>
<tr>
<th>130 or more</th>
<th>110 to 129</th>
<th>90 to 109</th>
<th>70 to 89</th>
<th>50 to 69</th>
<th>25 to 49</th>
<th>Under 25</th>
<th>Untestable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asher and Schonell 1950</td>
<td>0.6</td>
<td>3.4</td>
<td>20.1</td>
<td>26.8</td>
<td>22.9</td>
<td>10.7</td>
<td>11.6</td>
</tr>
<tr>
<td>Hologen 1952</td>
<td>-</td>
<td>2.8</td>
<td>22.0</td>
<td>36.2</td>
<td>21.3</td>
<td>8.5</td>
<td>3.5</td>
</tr>
<tr>
<td>Unaffected children</td>
<td>4.7</td>
<td>21.2</td>
<td>43.6</td>
<td>24.3</td>
<td></td>
<td>1.3</td>
<td></td>
</tr>
</tbody>
</table>
would be educable. It is because of the associated disabili-
ties, in particular intellectual impairment, epilepsy, speech
defects, impaired vision and hearing, and specific educational
difficulties that the majority required special educational
care. In Table I85 are shown some of the more important disa-
bilities encountered.

Intellectual impairment. The majority of patients
suffering from cerebral palsy are of subnormal intelligence.
The results of a variety of different tests employed by a
number of different psychologists who examined 171 of the
children in the present series are shown in Table 183. Since
it was predominantly the younger patients who were untested,
those who were examined are probably representative of the
series. It would be wrong to place too much reliance upon
the results though their similarity to those of other recent
surveys suggests that they are reasonably accurate. (Dunadon,
1952; Floyer, 1954; Illingworth, 1953) Table 184.

It will be observed that 43.3% of the tested patients had
intelligence quotients of 69 or less and were, therefore, so
mentally handicapped that they required special educational
provision if educable, irrespective of their physical disabil-
ities. 21.9% of those tested had quotients of less than 55
and were quite ineducable. 43 or 24.2% of the tested patients
had intelligence quotients between 70 and 84. A few of them
whose cerebral palsy was mild were attending normal schools,
but the majority were attending schools for the mentally or
physically /
<table>
<thead>
<tr>
<th>Intelligence Quotient</th>
<th>Over 115</th>
<th>100-114</th>
<th>85-99</th>
<th>70-84</th>
<th>55-69</th>
<th>Under 55</th>
<th>Untested</th>
<th>Totals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>1</td>
<td>2</td>
<td>4</td>
<td>4</td>
<td>8</td>
<td>5</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Severe</td>
<td>0</td>
<td>4</td>
<td>4</td>
<td>9</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>Total hemiplegia</td>
<td>3</td>
<td>10</td>
<td>10</td>
<td>17</td>
<td>17</td>
<td>11</td>
<td>7</td>
<td>75</td>
</tr>
<tr>
<td>Diplegia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paraplegic</td>
<td>1</td>
<td>2</td>
<td>5</td>
<td>6</td>
<td>7</td>
<td>1</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>Tetraplegic</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>4</td>
<td>3</td>
<td>11</td>
<td>7</td>
<td>27</td>
</tr>
<tr>
<td>Total diplegia</td>
<td>3</td>
<td>7</td>
<td>6</td>
<td>15</td>
<td>13</td>
<td>15</td>
<td>20</td>
<td>79</td>
</tr>
<tr>
<td>Diplegia with ataxia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ataxia</td>
<td>0</td>
<td>2</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>12</td>
</tr>
<tr>
<td>Dipskinesia</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>6</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>15</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>Other</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Totals</td>
<td>8</td>
<td>22</td>
<td>24</td>
<td>43</td>
<td>37</td>
<td>37</td>
<td>37</td>
<td>208</td>
</tr>
<tr>
<td>Percentage distribution</td>
<td>3-8</td>
<td>10-6</td>
<td>11-5</td>
<td>20-7</td>
<td>17-8</td>
<td>17-8</td>
<td>17-8</td>
<td>100</td>
</tr>
<tr>
<td>Percentage distribution</td>
<td>4-9</td>
<td>12-8</td>
<td>14-3</td>
<td>24-2</td>
<td>21-9</td>
<td>21-9</td>
<td>—</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 183.

INTELLIGENCE QUOTIENTS OF 208 PATIENTS WITH CEREBRAL PALSY

Table 184.

PERCENTAGE DISTRIBUTION OF INTELLIGENCE QUOTIENTS IN THIS SURVEY COMPARED WITH OTHER RECENT INVESTIGATIONS AND A GROUP OF UNAFFECTED SCOTTISH CHILDREN

<table>
<thead>
<tr>
<th>Intelligence Quotients</th>
<th>130 or More</th>
<th>115 to 129</th>
<th>100 to 114</th>
<th>85 to 99</th>
<th>70 to 84</th>
<th>55 to 69</th>
<th>Under 55</th>
<th>Under 25</th>
<th>Untestable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dunsdon (1952)</td>
<td>0-5</td>
<td>2</td>
<td>6-2</td>
<td>15-5</td>
<td>17-2</td>
<td>23-6</td>
<td>35</td>
<td>10-7</td>
<td>11-6</td>
</tr>
<tr>
<td>This survey</td>
<td>4-9</td>
<td>12-8</td>
<td>14-3</td>
<td>24-2</td>
<td>21-9</td>
<td>21-9</td>
<td>21-9</td>
<td>10-7</td>
<td>11-6</td>
</tr>
<tr>
<td>Asher and Schonell (1950)</td>
<td>0-6</td>
<td>3-4</td>
<td>20-1</td>
<td>26-8</td>
<td>22-9</td>
<td>10-7</td>
<td>11-6</td>
<td>1-3</td>
<td>3-9</td>
</tr>
<tr>
<td>Holoran (1952)</td>
<td>0-6</td>
<td>3-4</td>
<td>20-1</td>
<td>26-8</td>
<td>22-9</td>
<td>10-7</td>
<td>11-6</td>
<td>1-3</td>
<td>3-9</td>
</tr>
<tr>
<td>Unaffected children (Macneeken, 1939)</td>
<td>0-6</td>
<td>3-4</td>
<td>20-1</td>
<td>26-8</td>
<td>22-9</td>
<td>10-7</td>
<td>11-6</td>
<td>1-3</td>
<td>3-9</td>
</tr>
</tbody>
</table>
physically handicapped. 54 children, 32% of those tested had intelligence quotients of 35 or more and may charitably in some cases be regarded as being of average intelligence. Approximately two-thirds of them attended normal schools, but the remainder were in schools for the physically handicapped.

The distribution of patients by type of cerebral palsy and intelligence quotients is similar to that observed in most recent surveys, (Dunsdon, 1952; Floyer, 1954). Children suffering from bilateral hemiplegia or diplegia with tetraplegic involvement tend to be less intelligent than those with hemiplegia, paraplegia or dyskinesia. Within the individual categories of cerebral palsy children with extensive or severe physical defects tended to be less intelligent than those whose limb involvement was less marked. This is well shown in diplegic patients. Whereas 10 of the 29 paraplegic cases were of average intelligence, only 2 of the 27 tetraplegic patients were.

**Aphasia.** Aphasia is difficult to diagnose with certainty in cerebral palsied children. In the early years its predominant manifestation is retardation of speech development. It is often associated with mental defect and in a proportion of cases there are quite gross articulatory defects in association.

Aphasia was evident in 16 of the patients, or in 7.7% of the series. Twelve were hemiplegic and of these in only one was the paresis left-sided. The remaining patients were diplegic /
diplegic and it is of interest that three of the four showed tripligic involvement, the right upper limb being affected in all. Some degree of mental impairment was present in 9 of the patients. In all, however, the degree of language difficulty was out of all proportion to the severity of the mental retardation or of the retardation of speech development. Difficulties in word finding, the replacement of a wanted word by an irrelevant one, inversion of word order were the most marked features. In 3 there were additional difficulties in comprehension.

It is likely that more detailed psychological examination would have revealed aphasia in a higher proportion of patients in the series.

Speech defects in children suffering from cerebral palsy. Speech was considered to be defective only when it was unintelligible in whole or part. Minor abnormalities of articulation which did not impair comprehension of what he said were not considered to be significant. Generous allowance was made for regarded speech development for many of the patients were mentally retarded, and slow speech development was commoner than normal speech development.

102 patients or 48.3% of the present series were considered to have defective articulation. This is probably comparable with the figure of 79% (which included minor defects) given by Dunsdon, (1952). All the patients with bilateral hemiplegia showed speech defects, but since most of them were grossly /
### Table 185.

**ASSOCIATED DISABILITIES AND SCHOOLING IN 208 PATIENTS SUFFERING FROM CEREBRAL PALSY**

<table>
<thead>
<tr>
<th>Classification of Palsy</th>
<th>No. of Cases</th>
<th>Epilepsy</th>
<th>Aphasia</th>
<th>Visual Defect</th>
<th>Speech Defect</th>
<th>Overactivity</th>
<th>Schooling</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Grand Mal Jacksonian</td>
<td>Petit Maland Other</td>
<td></td>
<td></td>
<td></td>
<td>Normal School</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>27</td>
<td>6</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>11</td>
<td>9</td>
</tr>
<tr>
<td>Moderately severe</td>
<td>26</td>
<td>10</td>
<td>4</td>
<td>6</td>
<td>4</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Severe</td>
<td>22</td>
<td>8</td>
<td>0</td>
<td>4</td>
<td>6</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>75</td>
<td>22</td>
<td>4</td>
<td>12</td>
<td>11</td>
<td>21</td>
<td>9</td>
</tr>
<tr>
<td>Diplegia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paraplegia</td>
<td>29</td>
<td>6</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Triplegia</td>
<td>22</td>
<td>9</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>14</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>79</td>
<td>22</td>
<td>9</td>
<td>4</td>
<td>13</td>
<td>35</td>
<td>0</td>
</tr>
<tr>
<td>Diplegia with ataxia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Triplegia</td>
<td>9</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>Tetraplegia</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
<td>1</td>
<td>4</td>
<td>0</td>
<td>1</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>Ataxia</td>
<td>15</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>11</td>
<td>4</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>17</td>
<td>0</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>15</td>
<td>1</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>8</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>8</td>
<td>8</td>
<td>1</td>
</tr>
<tr>
<td>Other forms of cerebral palsy</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Grand total</td>
<td>208</td>
<td>53</td>
<td>22</td>
<td>16</td>
<td>35</td>
<td>102</td>
<td>16</td>
</tr>
<tr>
<td>Percentage</td>
<td></td>
<td>28.4</td>
<td>10.5</td>
<td>7.7</td>
<td>16.8</td>
<td>48.8</td>
<td>7.7</td>
</tr>
<tr>
<td>Percentage of school age</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Table 186.

**NO. OF PATIENTS WITH CEREBRAL PALSY BETWEEN AGES OF 6 AND 11 FOUND REVERSING LETTERS WHEN READING OR WRITING IN A SMALL EDINBURGH SURVEY**

<table>
<thead>
<tr>
<th>Type of Palsy</th>
<th>No. of Patients Questioned</th>
<th>Reading Only</th>
<th>Writing Only</th>
<th>Both Reading and Writing</th>
<th>Totals</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right hemiplegia</td>
<td>12</td>
<td>1</td>
<td>4</td>
<td>3</td>
<td>8</td>
</tr>
<tr>
<td>Left hemiplegia</td>
<td>13</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Diplegia</td>
<td>24</td>
<td>0</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Ataxia and diplegia</td>
<td>5</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Totals</td>
<td>57</td>
<td>1</td>
<td>10</td>
<td>6</td>
<td>17</td>
</tr>
</tbody>
</table>
grossly mentally defective and had at best only single words, these were probably not a major handicap. Speech was much less intelligible in only two of the 17 dyskinetic patients, 2 of the 12 suffering from ataxic diplegia and 4 of those with ataxia. Patients in these categories and those suffering from diplegia showed paresis and incoordination of the articulatory organs as their major disability. Their speech defects were predominantly dysarthric in type, though hearing impairment and involvement of respiratory muscles were important in dyskinesia. The speech defects in hemiplegic patients tended to be developmental in type, articulation being immature in pattern rather than abnormal because of local abnormalities. True dysarthria was recognised in only 4 of the 21 hemiplegic patients with speech defects.

The importance of speech defects in children suffering from cerebral palsy is increased because of their restricted opportunities for self-expression by other means. This is especially true of severely affected dyskinetic patients, a considerable proportion of whom are of average intelligence. The presence of speech abnormalities in addition to severe physical handicaps still further limits the scope of education and the possibilities of finding employment in a significant number of patients suffering from cerebral palsy.

Specific learning difficulties in children suffering from cerebral palsy. Difficulties with the written word occur more frequently in patients suffering from cerebral palsy than
does aphasia. The difficulties were not fully investigated in patients in the present series though 57 children between the ages of 6 and 11 who were considered to be of average intelligence were examined from this point of view. (Table 185).

It will be observed that significant difficulties in learning to read or write were present in 17 at the time they were seen. The difficulties tended to be rather characteristic. The children found specific difficulty in recognising the shape, orientation and relationship of letters and small words. "Mirror image letters" such as b and d, or p and q were frequently confused as were "Reversible words" like 'god' and 'dog', 'saw' and 'was'. There is a tendency to reverse the order of words in a sentence when reading and to omit small words. Large words with definite shape were often read more easily than small words. Even when they recognised the written symbols patients often had considerable difficulty in finding the phonic equivalents and in forming words from them. In writing there was the same tendency to letter and word reversal. Conjunctions, prepositions, auxiliary verbs and pronouns were often omitted, and the order of syllables in large words confused. In children with severe dysgraphia spontaneous composition was often quite unintelligible. The abnormalities in both reading and writing are very similar to those encountered in specific dyslexia and dysgraphia found in physically normal children.
Of the relatively small number of patients examined who showed dyslexia or dysgraphia, 10 were hemiplegic, and in 3 of these the hemiplegia was on the right. Two of the diplegic patients showed triplegic involvement, the right arm being affected in both cases. The other 3 were tetraplegic. Speech development had been markedly retarded in the majority of those who showed specific dyslexia or dysgraphia and 4 showed definite aphasia at the time of examination. Three of these were suffering from right hemiplegia and one diplegia.

Clearly educational disabilities of this type are a major handicap in patients who show so many other associated abnormalities in their physical and mental activities. (Table 186).

Overactive Behaviour. Sixteen patients showed overactivity which was characterised by an intense preoccupation with the immediate surroundings, marked restlessness, an almost compulsive desire to explore everything in the vicinity by touching it and often putting it to the mouth, and very limited attention span. Often there was an almost complete lack of insight or consideration for other people, and complete inability to accept the least frustration, so that any checking of his activities would often result in the child attacking whoever tried to stop him gratifying his momentary whim. The disordered behaviour makes it almost impossible to assess intelligence with any accuracy, but in many cases the overactivity diminishes as the child matures (commonly at the age of /
of between 7 and 10 years) and he may then be found to be not grossly defective. Formal schooling is impossible while the child shows marked overactivity of this type and even training and therapy are very limited in scope.

Nine of the 16 patients were hemiplegic and 4 ataxic. Seven of the 9 hemiplegic patients were cases of acquired hemiplegia and 6 were epileptic. In all cases the overactivity, and not the physical defect, was the clinical manifestation which most worried the parents.

Hearing impairment. Impairment of hearing sufficient to necessitate the patient being taught as a deaf child was present in only one case. Seven dyskinetic patients were shown to suffer from high frequency deafness as a result of audiometry and another 2 were suspected of having some hearing impairment.

Four patients were hard of hearing as a result of meningitis which had also caused their cerebral palsy, and 3 as a result of chronic bilateral otitis media.

Loss of vision. Some loss of vision, not due to refractive errors was found in 35 patients in the series. In 7 patients considered to be educable the impairment of vision was sufficiently severe to necessitate education by non-visual means. Four of these were diplegic patients who suffered from retrolental fibroplasia.

Visual field defects were practically confined to hemiplegic patients and were observed in 9 cases. The very good compensation /
compensation achieved even when hemianopia was complete was noteworthy. Unilateral optic atrophy was noted in only two hemiplegic patients.

The majority of the patients with serious bilateral loss of vision were found to suffer from severe bilateral cerebral palsy associated with mental defect. The blindness was almost certainly cortical in origin in the majority. Thus all 8 patients suffering from bilateral hemiplegia appeared to have very impaired visual acuity, but in none of them could defective vision be held responsible for the child being ineducable or unemployable.

**Epilepsy.** The numbers of patients in each category who were epileptic are shown in Table 135. Sixteen patients suffered from petit mal or myoclonic jerks, involving only relatively brief impairment of consciousness. Some of these also had grand mal seizures. Myoclonic jerks and petit mal tended to occur with greatest frequency in patients with severe bilateral cerebral palsy associated with mental defect and they were, therefore less important in restricting activities than would otherwise have been the case.

Grand mal and attacks of Jacksonian type with more prolonged periods of loss or impairment of consciousness occurred in 53 patients, or approximately 25%. They were most frequent in hemiplegia (29%), diplegia (27.5%), bilateral hemiplegia (62.5%), and occurred in none of the patients suffering from dyskinesia. This is in agreement with most recent surveys, (Skatvedt, 1958). In the majority of patients, grand mal or Jacksonian /
Jacksonian attacks occurred between once a month and once a year in spite of anti-epileptic drugs being given. Attacks were rarely controlled completely by anti-epileptic medication. Within the respective categories, grand mal and Jacksonian attacks occurred almost as frequently when the cerebral palsy was mild as when it was severe. Thus in many patients, especially those suffering from hemiplegia, epilepsy was a major disability further limiting the possibilities of education and employment. Epilepsy had necessitated the transfer of a number of hemiplegic children from normal schools to those for the physically handicapped.

The education of children with cerebral palsy. In Table 185 are shown the numbers of children of school age in the series who attended various types of school or were classified as ineducable. The figures obtained during this survey were compared with those of other similar surveys. The proportions of children in different schools vary in different surveys. This is probably a reflection on the selective nature of some series and of the differences in the types of schools available in different districts rather than a measure of differences in the types of handicap suffered by children in different areas. Nevertheless, the surveys show that not more than one quarter of the children who suffer from cerebral palsy are capable of benefiting from normal educational methods. Only three patients in the present series were thought to be capable of taking their higher leaving certificates. Over half the patients in normal schools suffered from hemiplegia and, therefore,

therefore, had at least one functionally normal upper limb. In contrast, only 11 patients with diplegia were in normal schools and there was tetraplegic paresis on only one of these. In general, types of cerebral palsy which tended to involve all limbs resulted in a smaller proportion of children in normal schools. Thus, in spite of the relatively less severe mental impairment than in other forms of cerebral palsy, only two patients with dyskinesia were in normal schools. Of the 12 patients of school age who showed ataxia, there were only 2 in normal schools.

Thirty-three of the patients of school age (approximately 21.5%) were classified as ineducable. In practice this category was found to consist almost entirely of severely mentally defective children. Thirteen of the patients classified as ineducable were in institutions for the mentally defective. The remainder were in occupation centres or at home. Twenty-one were bedridden and in need of constant care and attention. Eighty-one of the patients of school age (52.6%) attended special schools for the mentally or physically handicapped. The decision as to which type of school was best for the child was decided on the basis of the relative severity of the palsy and of the intellectual impairment. Though there was some overlapping of the types of case found in these schools; in general, those with intelligence quotients of under 70 were found in schools for the mentally handicapped and those with intelligence quotients of over 70 were in schools for the physically handicapped. Forty-three children, approximately
### Table 187

**Assessment of Prospects of Employment in Present Circumstances of 154 Children of School Age with Cerebral Palsy**

<table>
<thead>
<tr>
<th>Unemployable Criteria</th>
<th>Nos. of Patients</th>
<th>Nos. of Patients</th>
<th>Probably Employable Criteria</th>
<th>Nos. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intelligence quotient below 70</td>
<td>Hemiplegia with epilepsy</td>
<td>5</td>
<td>Hemiplegia Normal schools.</td>
<td>23</td>
</tr>
<tr>
<td></td>
<td>Hemiplegia</td>
<td>22</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Diplegia</td>
<td>25</td>
<td>Mild hemiplegia M.H. schools.</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Other</td>
<td>15</td>
<td>Paraplegia P.H. schools.</td>
<td>7</td>
</tr>
<tr>
<td>Total I.Q. under 70</td>
<td>62</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intelligence quotient between 70 and 85, but severely affected</td>
<td>Triplegia Normal schools</td>
<td>1</td>
<td>Triplegia Normal schools</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Hemiplegia P.H. &quot;</td>
<td>2</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>M.H. &quot;</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tetraplegia Normal schools</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Others</td>
<td>12</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total I.Q. between 70 and 85</td>
<td>24</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total unemployable</td>
<td>86</td>
<td>25</td>
<td>43</td>
<td></td>
</tr>
<tr>
<td>Percentage</td>
<td>56</td>
<td>Percentage possibly employable</td>
<td>16</td>
<td>Percentage probably employable</td>
</tr>
</tbody>
</table>
of those of school age, were in schools for the physically handicapped, of whom six were in residential schools.

Prospects of regular employment for children with cerebral palsy. During the survey some experience was gained of the employment difficulties encountered by a number of patients between the ages of 16 and 18 who suffered from cerebral palsy. On the basis of this experience an attempt was made to assess the prospects of regular employment and the need for the care of the cases of school age ascertained during the survey. Patients were placed in three categories, probably employable, possibly employable, or unemployable (Table 187).

The majority of patients who attended normal schools were considered to be probably employable with the exceptions of two relatively severely affected patients with diplegia, who were placed in the 'possibly employable' group. Three patients with hemiplegia and two with diplegia in schools for the physically handicapped were felt to be probably employable. In all 43 patients, or 28% of those of school age, were considered to be probably employable.

Patients with intelligence quotients of less than 70 were automatically classified as unemployable. Because of intellectual impairment they were only fitted for unskilled manual work, which they were unable to do on account of their physical handicaps. Patients with intelligence quotients of between 70 and 85 might be expected to be suitable for unskilled work, but the majority would only be capable of this if the
palsy was of a relatively slight degree. Therefore, those who were moderately severely or severely affected were classified as unemployable, and patients with mild cerebral palsy were classified, somewhat optimistically, as being possibly employable. In all 86 patients, approximately 56% of those of school age, were classified as unemployable.

Twenty-five patients were classified as possibly employable, 16% of those of school age. Two cases in this category came from normal schools, 20 from schools for the physically handicapped, including five patients with ataxia and six with dyskinesia. Three patients came from schools for the mentally handicapped. They had intelligence quotients of between 70 and 85 and relatively mild cerebral palsy.

It seems clear from this somewhat speculative assessment of the chances of regular employment for children with cerebral palsy that not more than one-third are likely to be self-supporting in present conditions. A number of the patients who do obtain regular work will obtain only jobs in which prospects are poor. In present labour conditions relatively few of the 16% of children in the 'possibly employable' group are likely to obtain regular employment, though in favourable conditions, and under some form of supervision many might make a contribution to their own maintenance.

The 56% of patients in the unemployable group are unlikely to be able to make any contribution to their own maintenance. The majority will be in need of constant care for many years.

Twenty-one of the patients in this group are bedridden and probably will all eventually be in need of constant nursing attention in institutions.
CHAPTER 2

Social conditions of patients suffering from cerebral palsy.

Unfortunately it was not possible in the conditions of the survey to obtain all the information about social conditions one would have wished to have. Thus, no full accounts can be presented of the degree of overcrowding, the nutrition of mothers, diet of their children, or the prevalence of infections in the families studied.

Such information about the social backgrounds of patients as was obtained will be presented.

Distribution of patients by the social class of father.

The distribution of 204 of the 205 patients by the social class of their fathers, using the classification of the Registrar General, is shown in Table 188. It will be noted that a comparatively high proportion are in social classes I and II and IV and V.

In Table 189 the percentage distribution of patients in the different categories of cerebral palsy is compared to that given by the Registrar General for all live births in Counties of Cities, Scotland, 1951, since figures for the City of Edinburgh are not available.

It will be observed that there is an excess of patients in social classes I and II and in IV and V, at the expense of social class III compared to the figures for all live births.
### TABLE 183

Distribution of patients with cerebral palsy by social class of Father.

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Unknown</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of C.P.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hemiplegia.</td>
<td>3</td>
<td>7</td>
<td>29</td>
<td>19</td>
<td>15</td>
<td>2</td>
<td>75</td>
</tr>
<tr>
<td>Bilat. Hemiplegia</td>
<td>1</td>
<td>0</td>
<td>6</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>Diplegia.</td>
<td>5</td>
<td>13</td>
<td>28</td>
<td>15</td>
<td>17</td>
<td>1</td>
<td>79</td>
</tr>
<tr>
<td>Ataxic Diplegia</td>
<td>0</td>
<td>1</td>
<td>6</td>
<td>2</td>
<td>3</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>Ataxia.</td>
<td>0</td>
<td>1</td>
<td>8</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>15</td>
</tr>
<tr>
<td>Dyskinesia.</td>
<td>3</td>
<td>4</td>
<td>6</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>17</td>
</tr>
<tr>
<td>Other.</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td><strong>Total.</strong></td>
<td>12</td>
<td>26</td>
<td>85</td>
<td>43</td>
<td>38</td>
<td>4</td>
<td>208</td>
</tr>
</tbody>
</table>
Comparison of distribution by social class of father of patients suffering from cerebral palsy and all live births in Counties of Cities. (Registrar General for Scotland 1951).

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Percentage by Social Class</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>I</td>
</tr>
<tr>
<td>% of Live Births in Scotland</td>
<td>7.7</td>
</tr>
<tr>
<td>% of cerebral palsy</td>
<td>of cases</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>73</td>
</tr>
<tr>
<td>Bilat. Hemiplegia</td>
<td>8</td>
</tr>
<tr>
<td>Diplegia</td>
<td>78</td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>12</td>
</tr>
<tr>
<td>Ataxia</td>
<td>14</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>17</td>
</tr>
<tr>
<td>Other</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>204</td>
</tr>
</tbody>
</table>
The proportional excess of patients in social classes IV and V is greatest in hemiplegia of all the categories. The proportional excess in Social Classes I and II is greatest in dyskinesia and diplegia. These differences from the distribution of live births are sufficiently great to be of statistical significance in hemiplegia in spite of the small number of cases.

The prevalence of cerebral palsy by social class. In Table 90 the prevalence of cerebral palsy has been calculated using an estimated population by social class. To obtain this estimate the population under the age of 16 was divided according to the social class distribution of births in Counties of Cities. This necessarily involves the possibility of quite a large error, but is probably the most accurate estimate available in the absence of figures for the actual population of Edinburgh by age and social class, which the Registrar General was unable to provide.

It will be observed that the figures obtained support the conclusion already offered, that cerebral palsy has a greater prevalence in the upper and lower social classes than in social class III. All types of cerebral palsy do not behave in the same way, however. Dyskinesia and diplegia is markedly more prevalent in social class I and II than in the others. Hemiplegia is more prevalent in social classes IV and V.

It seems likely that the remarkably high prevalence of diplegia in social classes I and II may be related to the high /
The prevalence of cerebral palsy by social class in an estimated population under the age of fifteen years in Edinburgh in 1951. (Cases per 1000)

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Estimated Population</th>
<th>Hemiplegia</th>
<th>Diplegia</th>
<th>Dyskinesia</th>
<th>Other</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>3760</td>
<td>.8</td>
<td>1.4</td>
<td>.8</td>
<td>.3</td>
<td>3.3</td>
</tr>
<tr>
<td>II</td>
<td>8943</td>
<td>.8</td>
<td>1.5</td>
<td>.5</td>
<td>.2</td>
<td>3.0</td>
</tr>
<tr>
<td>III</td>
<td>59248</td>
<td>.5</td>
<td>.5</td>
<td>.1</td>
<td>.3</td>
<td>1.7</td>
</tr>
<tr>
<td>IV</td>
<td>14736</td>
<td>1.3</td>
<td>1.0</td>
<td>.1</td>
<td>.4</td>
<td>2.8</td>
</tr>
<tr>
<td>V</td>
<td>14938</td>
<td>1.0</td>
<td>1.1</td>
<td>.1</td>
<td>.3</td>
<td>2.5</td>
</tr>
<tr>
<td>All</td>
<td>101625</td>
<td>.74</td>
<td>.70</td>
<td>.17</td>
<td>.36</td>
<td>2.0</td>
</tr>
</tbody>
</table>
Table 191

Distribution of hemiplegic patients by social class of father compared to live births in Counties of Cities, Scotland, 1951.

<table>
<thead>
<tr>
<th>Class</th>
<th>Congenital No.</th>
<th>Unknown No.</th>
<th>Acquired No.</th>
<th>Total No.</th>
<th>% Live Births</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>3</td>
<td>3.7</td>
</tr>
<tr>
<td>II</td>
<td>3</td>
<td>2</td>
<td>6</td>
<td>7</td>
<td>8.8</td>
</tr>
<tr>
<td>III</td>
<td>12</td>
<td>3</td>
<td>43</td>
<td>29</td>
<td>58.3</td>
</tr>
<tr>
<td>IV</td>
<td>6</td>
<td>2</td>
<td>13</td>
<td>19</td>
<td>14.5</td>
</tr>
<tr>
<td>V</td>
<td>7</td>
<td>3</td>
<td>15</td>
<td>15</td>
<td>14.7</td>
</tr>
<tr>
<td>Unknown</td>
<td>1</td>
<td>0</td>
<td></td>
<td>2</td>
<td>0</td>
</tr>
</tbody>
</table>

*Table showing distribution of hemiplegic patients by social class of father compared to live births in Counties of Cities, Scotland, 1951.*
high maternal age of mothers in these classes for the maternal age tends to be high in diplegia. The excess in social classes IV. and V. may be associated with the relatively high incidence of premature birth which prevails.

The high prevalence of dyskinesia is more difficult to account for. It seems possible that it may be related to the rather high proportion of first born children to elderly mothers who suffered from birth injury in this group. An additional factor may be that the survival rate of kernicteric babies was higher, due to better infant care in social classes I. and II. than in the lower social classes.

It is interesting to note that the prevalence of both congenital and acquired hemiplegia is greater in social classes IV. and V. than in social classes I. II. and III. though the aetiological factors involved are probably different. (Table 191). In acquired hemiplegia the excess of cases in social classes IV. and V. is probably the result of the greater exposure to infections of children during their early years, due to overcrowding and the prevalence of communicable infections in the often numerous older siblings. In congenital hemiplegia the greater prevalence may be the result of a higher frequency of abnormal pregnancies, labours and deliveries in social classes IV and V. than in I. II. or III.

The prevalence of cerebral palsy in Wards of the City of Edinburgh. In order to investigate the effects of living conditions /
conditions on the prevalence of cerebral palsy, Wards were rather arbitrarily classified as "Good", "Intermediate", or "Poor", using figures for their respective Infant Mortalities as criteria for classification. Wards with an infant mortality of under 25 per 1000 were classified as "good"; wards with an infant mortality of 31 or more as "poor". Intermediate wards were those with mortalities of between 26 and 30. It was realised that the infant mortality was only a rough indication of prevailing social conditions, but it appeared to be the best simple available measure as suggested by Crew, (1943).

In general the grouping by Infant Mortality does appear to result in Wards with similar social characteristics being placed in the same category. The total populations of all ages were larger in poor Wards than in intermediate or good Wards. Largely owing to the relatively higher birth rates in "poor" Wards than in "intermediate", or "good" Wards, the size of the child population was proportionately higher still. Thus, the average child population in "poor" Wards was over 5,000, and in "intermediate" or "good" Wards between 3,500 and 4,000. Overcrowding was more marked in "poor" wards than in those in the other groups. Shared lavatories were found more commonly, notifiable infectious diseases occurred more frequently, and probably in younger children. At the same time it must be recognised that social conditions within the Wards were far from uniform. Some "intermediate" Wards in /
in particular have a very mixed population, with a relative excess of fathers in social classes I. and II. and IV. and V. at the expense of class III. St. Andrews, with its mixture of Georgian squares and slum property is a good example of this lack of uniformity of social conditions in the population of individual Wards.

As will be seen from Table 17\textsuperscript{7}, the prevalence of cerebral palsy was found to be higher in "poor" wards than in "intermediate" wards and higher in "intermediate" than in "poor" wards. Within each group of Wards the prevalence varied greatly, but within the "poor" wards there were none with less than 1.4 cases per 1000 of the child population, and 5 of the 9 wards had more than 2 cases per 1000. Three of the 8 "intermediate" wards and only one of the 6 "good" wards showed a prevalence of more than 2 cases per 1000.

The prevalence of hemiplegia in each group of Wards was almost exactly proportional to the average infant mortality. The prevalence of diplegia did not follow this pattern. Diplegia occurred more frequently in "intermediate" Wards than in "poor" or "good" Wards. The numbers of cases are too small for reliable conclusions to be drawn about the causes of these differences in prevalence. It seems likely, however, that "intermediate" Wards may contain an excess of families in social classes I. and II. in which maternal age is relatively high, and an excess of families in social classes IV. and V. in which premature birth is frequent. In "good" Wards relatively more older mothers in social classes I. and II. might /
### Table 192

Employment of parents suffering from cerebral palsied offspring.

#### a. Father or adoptive father.

<table>
<thead>
<tr>
<th>Category</th>
<th>Unemployed</th>
<th>Occasional employment</th>
<th>Regular, Father absent</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia</td>
<td>5</td>
<td>12</td>
<td>53</td>
<td>5</td>
</tr>
<tr>
<td>Bilat. Hemiplegia</td>
<td>0</td>
<td>0</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Diplegia</td>
<td>5</td>
<td>8</td>
<td>49</td>
<td>15</td>
</tr>
<tr>
<td>Ataxic Diplegia. I</td>
<td>1</td>
<td>1</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>Ataxia</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>0</td>
<td>0</td>
<td>17</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
<td>22</td>
<td>144</td>
<td>28</td>
</tr>
<tr>
<td>Approx. %</td>
<td>6</td>
<td>11</td>
<td>69</td>
<td>14</td>
</tr>
</tbody>
</table>

#### b. Mothers or adoptive mothers.

<table>
<thead>
<tr>
<th></th>
<th>At home only</th>
<th>Part time employ.</th>
<th>Full-time employ.</th>
<th>Not at home. (Dead, hospitalised etc)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia</td>
<td>51</td>
<td>1</td>
<td>3</td>
<td>5</td>
</tr>
<tr>
<td>Bilat. Hemiplegia</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Diplegia</td>
<td>55</td>
<td>4</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>Ataxic Diplegia. I</td>
<td>0</td>
<td>4</td>
<td>0</td>
<td>8</td>
</tr>
<tr>
<td>Ataxia</td>
<td>13</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>15</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>140</td>
<td>35</td>
<td>13</td>
<td>17</td>
</tr>
<tr>
<td>Approx. %</td>
<td>65</td>
<td>17</td>
<td>6.5</td>
<td>8.5</td>
</tr>
</tbody>
</table>
Table 193.

Employment of mothers and unemployment of fathers of 76 diplegic patients by Social Class.

<table>
<thead>
<tr>
<th>Mother</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Full-time employment</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>4</td>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>Part time employment</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>6</td>
<td>2</td>
<td>II</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Father</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Permanent employment</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Frequent unemployment</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>5</td>
</tr>
</tbody>
</table>
might be expected, but the incidence of premature birth will be less as there will be fewer families in social classes IV. and V. In "poor" wards the effect of the excess of mothers in social classes IV. and V. who have premature babies will probably be offset by the lower proportion of older mothers in social classes I. and II.

**Employment of parents and poverty in the families of patients suffering from cerebral palsy.** The care received by patients in intact families before and after birth depended on a very large number of different factors. In conditions of severe poverty it is not to be expected that even the best mother could adequately care for a handicapped child. Whereas severe poverty was rare, relative poverty was found not uncommonly. It was usually due to unemployment of the father due to physical illness, poor intelligence or alcoholism.

The employment of fathers is indicated in Table 192. It will be observed that only in 69% of cases was the father at home and in regular employment. In 14% of cases the father of the child was dead or not at home. In 17% he was unemployed, either permanently or for average periods of over 4 months in the year. The majority of the fathers not in regular employment were in social classes IV. and V. and in many of these cases relative poverty was present. As might be expected, the category of cerebral palsy with the highest proportion of fathers in social classes IV. and V. also showed the highest proportion not in regular employment.

No /
No fathers were unemployed in the category of dyskinesia, in which there was a high proportion in social classes I. and II.

In many cases the effects of persistent or intermittent unemployment were increased in the home by the fact that the parents were "poor managers". Thus, in 8 cases of hemiplegia in social classes IV and V, the parents were both considered to be of subnormal intelligence, and in another 7, one or both were chronic alcoholics. "Poor managing" by the parents also resulted in lack of care for the child with cerebral palsy even when the father was in regular employment.

A high proportion of the mothers of illegitimate patients and those whose husbands were not fully employed themselves, took part-time or full-time employment. Whilst this often greatly relieved the financial stress of families it was liable to have adverse effects on the care the patients received. (Table 193). It will be observed that a total of 32% were deprived of the full-time maternal attention by death, illness, disappearance, or employment of their mothers. Again the highest proportion is hemiplegia, though that in diplegia is almost as high. Of the 9 fully employed mothers in the category of diplegia, 6 had worked until the seventh month of gestation whilst carrying the patients, as had 8 of the 11 in part-time employment. The distribution of working mothers in this category by social class of husband is shown in Table 193. As in all the categories, a higher proportion of mothers in social class IV, than in social class V, obtained employment — possibly /
possibly a reflection of their greater ability and initiative. The only working mother in social class I. was a medical practitioner.

Marital status of parents and legitimacy of patients.
Data on the legitimacy of patients and the marital state of parents was requested but not sought for with the energy employed in some of the other enquiries of the survey. Parents frequently felt that legitimacy of the child and their marital relationships were of moral rather than medical importance and this view had to be respected.

A total of 15 of the 205 patients about whose social background some information is available were known to be illegitimate. A further 6 were conceived before marriage and subsequently legitimised. It was of interest that all but one of the latter had fathers in social class III, whilst all but 2 of the illegitimate children were placed in social classes IV or V. It seemed that more importance was attached by parents in social class III than by those in social classes IV and V. to their children being born in wedlock. Thus, in Case 60, the mother of a diplegic child never saw her husband again after returning from maternity hospital, but felt she "had done the right thing by her". In another case of diplegia (Case 99), the grandparents insisted on the mother marrying, but she refused further consummation after the birth of the affected child.
Only 2 of the illegitimate children were adopted, in one case by the mother's sister and her husband. The remaining 13 patients were living with one parent or in institutions. Obviously the illegitimate child with significant handicaps, the result of cerebral palsy, is less likely to find himself in a favourable social setting than a normal illegitimate child.

It will be seen from Table 196, that a total of 39 patients in the series (approximately 19%) were living in broken homes at the time of examination. The reasons for the broken homes are indicated. It was interesting to observe the rather high mortality amongst parents of children with cerebral palsy, predominantly in social classes IV and V. This raised the question of the extent to which illness had contributed to the father being unable to take regular employment, and therefore, in his social status being lower than would otherwise have been the case.

The children coming from broken homes were not necessarily less well cared for materially than those with intact families. But in the majority care was less adequate. When the child was living with the mother only it was usually found either she was not working and there was real poverty, or that she was working whole or part-time. When mothers were working it was found that patients without useful grandmothers were liable to considerable neglect.

Emotional deprivation of children in broken homes was common /
Table 194.
Legitimacy of patients with cerebral palsy and their social situation.

<table>
<thead>
<tr>
<th>Category of Cerebral Palsy</th>
<th>Illegit.</th>
<th>Known Ante-marital concept</th>
<th>With parents</th>
<th>With one parent</th>
<th>With relatives or in institutions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia</td>
<td>4</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Bilat. Hemipl.</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Diplegia</td>
<td>5</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Ataxic Diplegia</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ataxia</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>6</td>
<td>6</td>
<td>12</td>
<td>4</td>
</tr>
</tbody>
</table>

(A number of institutionalised children are included in this table according to their social situation before leaving home.)
TABLE 195

Legitimacy of patients with cerebral palsy and their social situation

A number of institutionalised children are included and one classified according to social situation before institutionalisation.

<table>
<thead>
<tr>
<th>Category of cerebral palsy</th>
<th>Illegitimate</th>
<th>Known antemarital conception</th>
<th>With two &quot;parents&quot;</th>
<th>With one &quot;parent&quot;</th>
<th>Relatives, adopted or institutions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegia</td>
<td>4</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Diplegia</td>
<td>5</td>
<td>3</td>
<td>3</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ataxia</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>6</td>
<td>6</td>
<td>11</td>
<td>4</td>
</tr>
<tr>
<td>Category of cerebral palsy</td>
<td>Total cases known</td>
<td>Single mother</td>
<td>Separated or divorced</td>
<td>Mother dead</td>
<td>Father dead</td>
</tr>
<tr>
<td>---------------------------</td>
<td>------------------</td>
<td>--------------</td>
<td>----------------------</td>
<td>------------</td>
<td>------------</td>
</tr>
<tr>
<td>Hemiplegia.</td>
<td>75</td>
<td>4</td>
<td>5</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Bilateral hemiplegia.</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Diplegia.</td>
<td>76</td>
<td>5</td>
<td>6</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Ataxic diplegia.</td>
<td>12</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Ataxia.</td>
<td>15</td>
<td>4</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Dyskinesia.</td>
<td>17</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Other.</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>205</strong></td>
<td><strong>15</strong></td>
<td><strong>12</strong></td>
<td><strong>4</strong></td>
<td><strong>6</strong></td>
</tr>
</tbody>
</table>

A total of 39 children, approximately 19% were deprived of one or both parents.
common neither because the child was illegitimate, or because his mother resented the burden that having an abnormal child who nobody wanted to adopt, placed on her. Certainly the chances of subsequent marriage or remarriage of mothers with handicapped children do appear to be lessened.

In some cases the birth of the handicapped child, with the consequent physical and emotional stresses that resulted in the family, appeared to be the precipitating factor leading to separation or divorce of the parents. Important factors in this were maternal exhaustion and consequent inability to perform her wifely duties to the husband's satisfaction, the parents' feelings of guilt about the child's handicap and sometimes their tendency to blame each other for it. Parental illness, mental instability, alcoholism, unemployment and poor social conditions tended to make the burden on the parents of the cerebral palsied child greater in social classes IV. and V. in which parental death, separation and divorce were commoner.

The only categories of cerebral palsied which contained sufficient patients to merit further statistical analysis by social class were hemiplegia and diplegia.

Health of siblings by social class. The distribution of hemiplegic and diplegic patients, of siblings suffering from physical and mental disorders by social class are shown in Table 197. It will be observed that there is a higher proportion of abnormal siblings in social classes III than in I and II. and in IV. and V. than in /
in III. The higher proportion is almost entirely due to the excess of mentally handicapped patients in the lower social classes. This is illustrated for diplegia in Table 198. The numbers of siblings are too small for the differences between the categories of congenital hemiplegia and diplegia on the one hand, and acquired hemiplegia on the other, to be statistically significant.

The etiological significance of the higher proportion of abnormal siblings in social classes IV. and V. is difficult to assess. A high proportion of parents in these classes were mentally retarded and might be expected to produce a relatively high number of mentally retarded children. On the other hand it is possible that there are common etiological factors in these social classes which predispose to the production of cerebral palsied or mentally retarded offspring.

Health of parents by social class. The numbers and proportions of hemiplegic and diplegic patients (the only groups large enough for statistical analysis) suffering from physical or mental disease or severe psychiatric disorder are shown in Tables 199a + b. Since there are no significant differences between the proportions of abnormal parents in acquired or congenital hemiplegia, or in hemiplegia of unknown origin, all cases of hemiplegia will be considered together. The proportion of abnormal parents increases from social classes I. to social class V. in both hemiplegia and diplegia. The proportion of abnormal parents is higher in diplegia than in/
TABLE 197

Number and percentages of abnormal siblings of hemiplegic patients by social class

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Congenital hemiplegia</th>
<th>Acquired hemiplegia</th>
<th>Diplegia</th>
<th>Congenital hemiplegia</th>
<th>Acquired hemiplegia</th>
<th>Diplegia</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>II.</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>III.</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>7</td>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>IV.</td>
<td>4</td>
<td>5</td>
<td>2</td>
<td>31</td>
<td>15</td>
<td>7</td>
</tr>
<tr>
<td>V.</td>
<td>3</td>
<td>3</td>
<td>13*</td>
<td>30</td>
<td>17</td>
<td>39</td>
</tr>
</tbody>
</table>

* Excluding cases 153 and 159 who are diplegic twins.
### Table (a)

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients in whom histories known.</td>
<td>4</td>
<td>12</td>
<td>27</td>
<td>14</td>
<td>16*</td>
<td>73</td>
</tr>
<tr>
<td>Number with only physically abnormal siblings.</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0*</td>
<td>2*</td>
</tr>
<tr>
<td>Number of patients with only mentally abnormal siblings.</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Number with mentally and physically abnormal siblings.</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Total number with abnormal siblings.</td>
<td>1</td>
<td>0</td>
<td>3</td>
<td>2</td>
<td>6*</td>
<td>12*</td>
</tr>
</tbody>
</table>

### Table (b)

| Total number of siblings. | 2 | 12 | 34 | 29 | 23 | 110 |
| Number of physically abnormal siblings. | 0 | 0 | 1 | 1 | 2* | 4* |
| Number of mentally abnormal siblings. | 1 | 0 | 2 | 1 | 9 | 12 |
| Number of physically and mentally abnormal siblings. | 0 | 0 | 0 | 0 | 2 | 2 |
| Total number of abnormal siblings. | 1 | 0 | 3 | 2 | 13* | 18* |

* Excluding cases 158 and 159 - diplegic twins.
<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of cases</td>
<td>5</td>
<td>13</td>
<td>28</td>
<td>15</td>
<td>15</td>
<td>76</td>
</tr>
<tr>
<td>Parents healthy</td>
<td>5</td>
<td>9</td>
<td>11</td>
<td>2</td>
<td>1</td>
<td>23</td>
</tr>
<tr>
<td>One or both parents mentally subnormal</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>6</td>
<td>10</td>
<td>17</td>
</tr>
<tr>
<td>One or both parents with psychiatric disorders</td>
<td>0</td>
<td>1</td>
<td>5</td>
<td>5</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td>One or both parents suffering from physical disorders</td>
<td>0</td>
<td>3</td>
<td>11</td>
<td>7</td>
<td>7</td>
<td>28</td>
</tr>
<tr>
<td>Total number of cases with unhealthy parents</td>
<td>0</td>
<td>4</td>
<td>17</td>
<td>13</td>
<td>14</td>
<td>48</td>
</tr>
<tr>
<td>Approx. % of patients with abnormal parents</td>
<td>24</td>
<td>61</td>
<td>90</td>
<td>63</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number of cases</td>
<td>3</td>
<td>7</td>
<td>29</td>
<td>19</td>
<td>15</td>
<td>73</td>
</tr>
<tr>
<td>One or both parents mentally subnormal</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>5</td>
<td>7</td>
<td>12</td>
</tr>
<tr>
<td>One or both parents with psychiatric disorders</td>
<td>0</td>
<td>1</td>
<td>4</td>
<td>3</td>
<td>4</td>
<td>12</td>
</tr>
<tr>
<td>Physical disorders in one or two parents</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Total abnormal</td>
<td>1</td>
<td>1</td>
<td>6</td>
<td>9</td>
<td>15</td>
<td>32</td>
</tr>
<tr>
<td>Approx. % with abnormal parents</td>
<td>20%</td>
<td>20%</td>
<td>48%</td>
<td>43%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
In hemiplegia though there are relatively fewer fathers in social classes IV. and V. than in hemiplegia.

In both groups the greatest increase in the proportions of abnormal parents in the lower social classes is due to the higher number of mentally abnormal and psychiatrically disturbed parents. Physical disease is also commoner in the lower social classes, however. It is possible to interpret these figures in different ways. It may be that the abnormal parents are in social classes IV. and V. because of their inability to compete in the employment market, and/or they may be ill directly or indirectly because of poor social conditions. The figures give no indication as to whether the fact that the parents were abnormal, or the fact that they lived in poor social conditions were of greater aetiological importance in their children's cerebral palsy. The health of parents will be discussed in more detail when family history is discussed.

The size of families of parents with children suffering from hemiplegia and diplegia. The only categories of cerebral palsy which contained sufficient patients to merit further statistical analysis of the effects of social factors were hemiplegia and diplegia.

The size of families by social class. In Table 200 is shown the number of surviving children born to the parents of patients suffering from congenital and acquired hemiplegia and diplegia. It will be observed that for all three conditions the average number of children is greater in the lower social classes /
TABLE 200

The number of children born to the parents of patients suffering from hemiplegia (congenital and acquired), and diplegia.

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Number of cases</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>1</td>
<td>2</td>
<td>14</td>
<td>11</td>
<td>5</td>
<td>33</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>1</td>
<td>3</td>
<td>12</td>
<td>6</td>
<td>7</td>
<td>29</td>
</tr>
<tr>
<td>Diplegia</td>
<td>4</td>
<td>12</td>
<td>27</td>
<td>15</td>
<td>17</td>
<td>75</td>
</tr>
<tr>
<td><strong>Number of children</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>2</td>
<td>3</td>
<td>44</td>
<td>42</td>
<td>22</td>
<td>113</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>3</td>
<td>5</td>
<td>26</td>
<td>19</td>
<td>17</td>
<td>70</td>
</tr>
<tr>
<td>Diplegia</td>
<td>6</td>
<td>25</td>
<td>62</td>
<td>47</td>
<td>46</td>
<td>186</td>
</tr>
<tr>
<td><strong>Average number of children per family</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>1.7</td>
<td>1.7</td>
<td>3.1</td>
<td>3.1</td>
<td>4</td>
<td>3.4</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>2.0</td>
<td>2.2</td>
<td>2.7</td>
<td>2.7</td>
<td>2.4</td>
<td>2.4</td>
</tr>
<tr>
<td>Diplegia</td>
<td>2.1</td>
<td>2.3</td>
<td>2.8</td>
<td>2.8</td>
<td>2.5</td>
<td>2.5</td>
</tr>
</tbody>
</table>
TABLE 201
Foetal and infant loss to mothers of patients with hemiplegia and diplegia

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I.</th>
<th>II.</th>
<th>III.</th>
<th>IV.</th>
<th>V.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. of Preg.</td>
<td>Loss</td>
<td>No. of Preg.</td>
<td>Loss</td>
<td>No. of Preg.</td>
</tr>
<tr>
<td>Acquired hemiplegia.</td>
<td>2</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>51</td>
</tr>
<tr>
<td>Congenital hemiplegia.</td>
<td>3</td>
<td>0</td>
<td>5</td>
<td>0</td>
<td>32</td>
</tr>
<tr>
<td>Diplegia.</td>
<td>7</td>
<td>1</td>
<td>27</td>
<td>2</td>
<td>65</td>
</tr>
</tbody>
</table>

% Loss

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>6</td>
<td>0</td>
<td>6</td>
</tr>
</tbody>
</table>
### TABLE 202

Abortions, stillbirth, neonatal deaths and infant deaths amongst siblings of diplegic patients by social class of father

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I.</th>
<th>II.</th>
<th>III.</th>
<th>IV.</th>
<th>V.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abortion</td>
<td>0</td>
<td>1</td>
<td>6</td>
<td>3</td>
<td>3</td>
<td>13</td>
</tr>
<tr>
<td>Stillbirths</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Neonatal deaths</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Infant deaths</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Total number with full histories</td>
<td>4</td>
<td>12</td>
<td>27</td>
<td>14</td>
<td>16</td>
<td>73</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Number of:</th>
<th>I.</th>
<th>II.</th>
<th>III.</th>
<th>IV.</th>
<th>V.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abortions</td>
<td>0</td>
<td>2</td>
<td>7</td>
<td>3</td>
<td>8</td>
<td>20</td>
</tr>
<tr>
<td>Stillbirths</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Neonatal deaths</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Infant deaths</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Total foetal and infant wastage</td>
<td>1</td>
<td>2</td>
<td>8</td>
<td>5</td>
<td>16</td>
<td>32</td>
</tr>
<tr>
<td>Total number of pregnancies other than those of patients</td>
<td>3</td>
<td>15</td>
<td>33</td>
<td>33</td>
<td>50</td>
<td>139</td>
</tr>
<tr>
<td>Percentage foetal and infant wastage in other pregnancies</td>
<td>33</td>
<td>13</td>
<td>21</td>
<td>15</td>
<td>32</td>
<td>23</td>
</tr>
</tbody>
</table>

Difference in the percentage mortality by social class is not significantly different statistically.
### History of the pregnancies and deliveries resulting in the birth of 75 diplegic patients by social class of father

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Pregnancy and delivery normal</th>
<th>Pregnancy abnormal</th>
<th>Both abnormal</th>
<th>General Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td>Delivery normal</td>
<td>Total</td>
<td></td>
</tr>
<tr>
<td>I.</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>II.</td>
<td>0</td>
<td>2</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>III.</td>
<td>2</td>
<td>6</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>IV.</td>
<td>2</td>
<td>3</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>V.</td>
<td>1</td>
<td>3</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>5</td>
<td>15</td>
<td>20</td>
<td>6</td>
</tr>
</tbody>
</table>
Tables of abnormalities of pregnancy and delivery by social class of father in 75 diplegic patients.

**TABLE 204**

Abnormality of pregnancy by social class

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Normal pregnancy</th>
<th>Abnormal pregnancy</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.</td>
<td>7</td>
<td>10</td>
<td>17</td>
</tr>
<tr>
<td>II.</td>
<td>12</td>
<td>15</td>
<td>27</td>
</tr>
<tr>
<td>III.</td>
<td>16</td>
<td>15</td>
<td>31</td>
</tr>
<tr>
<td>IV.</td>
<td>17</td>
<td>11</td>
<td>27</td>
</tr>
<tr>
<td>V.</td>
<td>13</td>
<td>13</td>
<td>31</td>
</tr>
</tbody>
</table>

**TABLE 205**

Abnormality of delivery by social class

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Normal pregnancy</th>
<th>Abnormal pregnancy</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.</td>
<td>6</td>
<td>11</td>
<td>17</td>
</tr>
<tr>
<td>II.</td>
<td>16</td>
<td>11</td>
<td>27</td>
</tr>
<tr>
<td>III.</td>
<td>13</td>
<td>13</td>
<td>31</td>
</tr>
</tbody>
</table>
Tables of abnormalities of pregnancy and delivery by social class of father in 75 diplegic patients.

**TABLE 206**

Abnormality of pregnancy and labour or delivery by social class

<table>
<thead>
<tr>
<th>Social class</th>
<th>Pregnancy and labour and delivery normal</th>
<th>Pregnancy and labour and/or delivery abnormal</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.</td>
<td>3</td>
<td>14</td>
<td>17</td>
</tr>
<tr>
<td>II.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>III.</td>
<td>8</td>
<td>19</td>
<td>27</td>
</tr>
<tr>
<td>IV.</td>
<td>9</td>
<td>22</td>
<td>31</td>
</tr>
<tr>
<td>V.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**TABLE 207**

Prematurity by Social Class

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Pregnancy and delivery normal</th>
<th>Pregnancy and labour or delivery abnormal</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.</td>
<td>3</td>
<td>14</td>
<td>17</td>
</tr>
<tr>
<td>II.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>III.</td>
<td>8</td>
<td>19</td>
<td>27</td>
</tr>
<tr>
<td>IV.</td>
<td>9</td>
<td>22</td>
<td>31</td>
</tr>
<tr>
<td>V.</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
classes than in the higher. The Table gives only a partial demonstration of the difference in the size of families. A high proportion of mothers in social classes I. and II. were over the age of thirty compared to the proportion in social classes IV. and V. Whilst the mothers in the upper social classes might be expected to have few further children, those in the lower social classes might be expected to have many. Another factor of smaller importance is that the foetal and infant loss is rather higher to mothers in social classes IV. and V. than in I. II. or III. The difference in the fertility of mothers by social class is rather greater than the differences indicated when the numbers of surviving children are considered.

The foetal and infant loss by social class. The foetal and infant loss by social class to mothers with diplegic and hemiplegic patients is shown in Table 201. It will be observed that the loss is greater for all categories in social classes IV. and V. than in III. and greater in III. than for I. and II. combined. Unfortunately the numbers of patients are too small for these differences to be of statistical significance, but the trend is in accordance with the other evidence presented. A more detailed account of the causes of the foetal and infant loss in the other pregnancies of the mothers of diplegic patients is shown in Table 202.

The tendency for mothers of hemiplegic and diplegic patients to have abnormal pregnancies, labours and deliveries by social class. Since all but one of the pregnancies, labours /
or deliveries resulting in the birth of patients suffering from congenital hemiplegia were abnormal, no study of these by social class was made. The pregnancies, other than those resulting in the birth of patients were studied. It was found that mothers of children with acquired hemiplegia appeared to be more liable to have abnormalities of parturition if they were in social classes IV. or V. than in I. II. or III. The figures were too small for the differences to be of statistical significance but the pattern is similar to that observed in the general population by Baird, (1945).

Mothers with children suffering from congenital hemiplegia had a similar proportion of abnormal other pregnancies, labours, or deliveries whatever their social class. This proportion was higher than for mothers with patients suffering from congenital hemiplegia, but there was no greater prevalence of abnormal offspring. (Tables 204-206)

Since abnormal pregnancy, labour and delivery was less constant in the conceptions which resulted in the births of diplegic patients, these could be analysed for study by social class. It was found that the slightly greater proportion of abnormal parturitions in social classes IV. and V. than in I. II. and III. was not statistically significant. On the other hand, the higher proportion of prematurely born patients in social classes III. IV. and V. compared to I. and II. was possibly significant (being approximately .05) (Table 207.

This suggests that the abnormalities of parturition encountered in social classes I. and II. though no less frequent than those in /
### TABLE 208

Distribution of 79 diplegic patients by age of mother at time of delivery and by social class of father

<table>
<thead>
<tr>
<th>Maternal age</th>
<th>I.</th>
<th>II.</th>
<th>III.</th>
<th>IV.</th>
<th>V.</th>
<th>Unknown</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 20</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>20 - 24</td>
<td>0</td>
<td>2</td>
<td>8</td>
<td>4</td>
<td>6</td>
<td>0</td>
<td>20</td>
</tr>
<tr>
<td>25 - 29</td>
<td>1</td>
<td>2</td>
<td>2</td>
<td>4</td>
<td>3</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>30 - 34</td>
<td>2</td>
<td>7</td>
<td>6</td>
<td>5</td>
<td>4</td>
<td>0</td>
<td>24</td>
</tr>
<tr>
<td>35 - 39</td>
<td>1 (75%)</td>
<td>6 (66%)</td>
<td>6 (52%)</td>
<td>2 (47%)</td>
<td>0 (29%)</td>
<td>0</td>
<td>10</td>
</tr>
<tr>
<td>40+</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Unknown</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>5</td>
<td>13</td>
<td>28</td>
<td>15</td>
<td>17</td>
<td>1</td>
<td>79</td>
</tr>
</tbody>
</table>

Average age of mother: 32 30 29 29 26 - 29

### TABLE 209

Distribution of 35 first born diplegic patients by age of mother at time of delivery and by social class of father

<table>
<thead>
<tr>
<th>Maternal age</th>
<th>(I. II)</th>
<th>III.</th>
<th>(IV. V.)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>15 - 19</td>
<td>0</td>
<td>3</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>20 - 24</td>
<td>2</td>
<td>6</td>
<td>6</td>
<td>14</td>
</tr>
<tr>
<td>25 - 29</td>
<td>3</td>
<td>0</td>
<td>3</td>
<td>6</td>
</tr>
<tr>
<td>30 - 34</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>35+</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>10</td>
<td>10</td>
<td>15</td>
<td>35</td>
</tr>
</tbody>
</table>

Average age: 30 22 25 26
in social classes III, IV, and V. result in premature delivery less commonly, and may be of different aetiological significance. The mothers who gave birth to premature diplegic patients (predominantly in classes III, IV, and V.) were also found to have a higher proportion of abnormal other parturi¬tions than those giving birth to mature patients. Unfortunately there were too few other pregnancies in social classes I. and II. for these to be analysed statistically for difference by social class.

Maternal age and social class. Only in the category of diplegia were there sufficient cases for an analysis of maternal age by social class to be rewarding. (Table 208)

There was a higher proportion of mothers over the age of thirty in social classes I. and II. than in IV. or V. This difference in age was statistically significant using the X2 test. The differences in maternal age between social class III. and IV. and V. and social classes I. and II. and III. respectively were not significant.

Since family size tended to be smaller in social classes I. and II. than in III, IV, or V. there is a relative excess of first and second born children in social classes I. and II. and a relative excess of later born children in the other social classes. This tends to minimise the actual differences in maternal age. In order to obtain a more valid comparison, the ages of the mothers at the time of the births of first born diplegic patients was studied, Table 209. It will be noted that the differences in age between the classes are /
TABLE 210

Abnormalities of parturition in conceptions other than those resulting in the birth of patients with congenital and acquired hemiplegia.

Percentage of abnormal pregnancies.

<table>
<thead>
<tr>
<th>Social Class</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
<th>V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia</td>
<td>0</td>
<td>100</td>
<td>40</td>
<td>32</td>
<td>42</td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>0</td>
<td>0</td>
<td>5</td>
<td>8</td>
<td>23</td>
</tr>
</tbody>
</table>
are much greater and are of statistical significance in spite of the small number of patients. The low maternal age in Social Class III. is noteworthy. Mothers in this class tend to have more children than those in Social Classes I. & II. and fewer spaced at longer intervals than those in IV. and V.

It is tempting to speculate that the risk of having a diplegic child may be increased when the mother is old and relatively infertile, as in Social Classes I. and II. or when she has children rapidly one after the other as in Social Classes IV. and V. The relatively low prevalence of diplegia in Social Class III. might be due, in part, to neither of these features being so marked.

The antenatal care of mothers and place of birth of patients. The possibility that the excess of patients suffering from cerebral palsy whose fathers were in Social Classes IV. or V. might partly be attributable to inadequate antenatal and obstetric care required investigation.

The place of birth of the patients and whether they were booked in hospital or not is shown in Table 211. It will be observed that approximately half the mothers were booked and delivered in hospital and that a further 19 were delivered in nursing homes. All these appeared to have had regular adequate antenatal attention. It was much more difficult to judge the standard of the antenatal care and obstetrics employed by doctors looking after the 63 mothers who were delivered at home. Only 14 of the patients had to be admitted to hospital as emergencies - a low figure judged in the light /
### TABLE 213
The place of birth of hemiplegic patients

#### A.

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>Congenital First Subsequent</th>
<th>Acquired First Subsequent</th>
<th>Unknown First Subsequent</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Booked in hospital.</td>
<td>7</td>
<td>7</td>
<td>6</td>
<td>11</td>
</tr>
<tr>
<td>Not booked but born in hospital.</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Nursing Home.</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Born at home.</td>
<td>3</td>
<td>7</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Unknown.</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>15</td>
<td>13</td>
<td>20</td>
</tr>
</tbody>
</table>

#### B.

By Social Class

<table>
<thead>
<tr>
<th>Aetiology</th>
<th>I. First Sub.</th>
<th>II. First Sub.</th>
<th>III. First Sub.</th>
<th>IV. First Sub.</th>
<th>V. First Sub.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Booked in hospital.</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Not booked but born in hospital.</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Nursing Home.</td>
<td>2</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Born at home.</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Unknown.</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Total</td>
<td>4</td>
<td>0</td>
<td>5</td>
<td>2</td>
<td>12</td>
<td>15</td>
</tr>
</tbody>
</table>
TABLE 214

Antenatal and obstetric care of the mothers of the 74 patients with diplegia

<table>
<thead>
<tr>
<th>Antenatal care</th>
<th>I.</th>
<th>II.</th>
<th>III.</th>
<th>IV.</th>
<th>IV.</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Attended hospital or</td>
<td>4</td>
<td>13</td>
<td>26</td>
<td>15</td>
<td>16</td>
<td>74</td>
</tr>
<tr>
<td>local clinics.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Attended own doctor.</td>
<td>3</td>
<td>3</td>
<td>8</td>
<td>4</td>
<td>1</td>
<td>19</td>
</tr>
<tr>
<td>Inadequate antenatal</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>5</td>
<td>8</td>
</tr>
<tr>
<td>care.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>4</td>
<td>13</td>
<td>26</td>
<td>15</td>
<td>16</td>
<td>74</td>
</tr>
</tbody>
</table>
light of the prevailing opinion that the majority of cases of cerebral palsy must be ascribed to birth injury.

The standard of antenatal care appeared to be quite adequate in the majority of patients, Table 212. Only in 14% did mothers fail to arrange for regular supervision during their pregnancies. The high proportion of these in the category of dyskinesia is slightly surprising in view of the relatively high proportion of fathers in Social Classes I and II. Examination of the finding showed that all five of the mothers had had previous children; five to two mothers, and four in a third. One of the cases was in Social Class II, three in III, and one in IV. All but one of them had their babies at home. The impression gained was that these women were somewhat over-confident about their reproductive powers, and blase about the business of childbirth, rather than ignorant or lazy.

In view of the probability that the degree to which birth injury is responsible for the conditions is different in different types of cerebral palsy, the two larger groups, hemiplegia and diplegia were studied in more detail individually.

In Table 213, are compared the proportions of mothers with patients showing congenital and acquired hemiplegia who had their babies in hospital, at home, and in nursing homes. No statistically significant difference in the figures was found. Nor could any differences be found between the standard of antenatal and obstetric care per se in these groups.
groups. On the other hand, all but one of the 8 cases in
which the mothers of hemiplegic patients appeared to have
received inadequate antenatal care were in Social Classes IV.
and V.

The rather similar findings were noted in the mothers
of patients suffering from diplegia, Table 214. It will be
observed that all the 3 patients in whom antenatal care was
thought to have been inadequate were in Social Classes IV.
and V. Six of the eight mothers requiring emergency admis¬
sion to hospital for delivery were in the same classes. The
other two were in Social Class III.

Taking all the figures together it does not seem possible
to make out a case for the majority of cases of cerebral palsy
being due directly or indirectly to inadequate antenatal or
obstetric care. As might be expected, however, there appears
to be a greater deficiency of antenatal care in Social Classes
IV. and V. than in I, II. or III.
SECTION 5

THE ASSOCIATION OF BIRTH INJURY WITH CEREBRAL PALSY.
The association of birth injury with cerebral palsy.

Historical. It is almost required by tradition that any discussion of birth injury and cerebral palsy should begin by paying respects to Shakespeare, if not to Richard the Third himself, (Little, 1893; Collier, 1899, 1924; Sachs and Haumann, 1926). A more graphic and typically English description of the dangers of parturition to the child is to be found in Tristram Shandy, and perhaps Sir Walter Scott may be criticised for his failure to present, with his customary detail, the birth history of Nick Strumpf.

In spite of these works it is difficult to assess the extent to which it was generally realised before the medical investigations of the past 130 years, that birth injury could result in permanent damage to the nervous system.

In this country, credit for having realised the aetiological importance of birth injury in cerebral palsy is usually given to Little (1953, 1962). But his studies had been preceded by those of a number of authors on the continent, most of whom were particularly interested in the pathological aspects of cerebral palsy and birth injury.

In this chapter the literature on birth injury as a cause of cerebral palsy will be reviewed and in later chapters the evidence obtained from other studies of birth injury about the effects of abnormal parturition on the child will be considered.

Various phases of interest in cerebral palsy are reflected in the ways in which the relationship to birth injury has been studied.
studied. French and German pathologists were responsible for much of the early work on etiology between 1927 and the 1860s. The phase of pathological research was succeeded by one in which clinical interest was predominant in America, Great Britain and on the continent. In the past thirty years increasing attention has been paid to the problems of treatment and rehabilitation of the child with cerebral palsy, and speculation about possible prevention of the condition has intensified interest in the problem of its etiology.

The Pathologists. That a proportion of the cases of cerebral palsy (especially hemiplegia) which he studied from the pathological point of view were suffering from the effects of birth injury was suspected by Casauvielh, (1927). He attempted to classify the atrophic changes he observed in the brains of six cases diagnosed during life as suffering from cerebral atrophy, as either "L'agénésie cérébrale" or "l'atrophie consecutif" depending on whether there had been defects in development due to constitutional factors or damage to the growing brain as a result of trauma or disease. He complained that the current knowledge of disease in the first few months of life and prenatal life was scanty and that this made assessment of the timing and nature of the causal factors of cerebral damage at this age impossible to determine. Nevertheless, by his classification he established the lines which controversy

about the nature of the atrophic changes found in the brain in children suffering from /
from cerebral palsy would follow.

During the next twenty years series of cases were published in which atrophic changes were attributed to inflammatory disease in foetal life, "traumatic encephalitis", or primary failures of development unrelated to acquired disease, (Dugas, 1826; Breshet, 1824). Very detailed study of the various forms of atrophy was made. By classifying them on the basis of the distribution and extent of the atrophy the associated findings such as fibrous plaques, cystic cavities and yellow staining attempts were made to determine the nature of the initial disease process, (von Lallemand, 1834; Henoch, 1942).

Cruvehlier recognised seven pathological categories of cerebral atrophy. He thought that it was impossible to distinguish between the changes which were due to arrests or abnormalities of development due to constitutional factors and those due to disease in prenatal life. On the other hand, it was quite often possible to suggest on the basis of the late pathological changes the nature of the original disease process, haemorrhage, "red softening" (on the basis of venous obstruction), encephalitis, meningitis, meningoencephalitis (chronic pachymeningitis) could be postulated as having occurred on the basis of the yellow staining of tissue, loss of cerebral substance or diffuse alterations of cellular content widespread amongst many convolutions. By implication rather than /
than direct statement, Cruvehlier gives the impression of regarding haemorrhage as being due to birth trauma in the majority of congenital cases, (Cruvehlier, 1862).

Cotard was more doubtful than Cruvehlier about the extent to which late pathological syndromes could be related to initial disease processes and classified cerebral atrophy into three main categories. The first was chronic meningoencephalitis with shrinkage of the underlying convolutions. This might be the result of infection in the meningitis, or due to the organisation of blood clot around the base of the brain which had been extravasated as a result of injury, often at the time of birth. The second type of atrophy was lobar sclerosis in which there was shrinkage of individual convolutions. The third was atrophy accompanied by loss of cerebral substance (later called porencephaly). He thought that the common causes of these disorders were traumatic encephalitis, especially after instrumental delivery, brain softening, especially after venous thrombosis which he stated was common in the early months of life, cerebral haemorrhage and meningeal haemorrhage. He made no attempt to correlate the various pathological findings to specific abnormalities of parturition or the neonatal period.

Further study of Cotard's second group of atrophies was made by Kundrat, (1882) following the work of Heschl, (1854). He demonstrated that atrophy with loss of cerebral substance could occur before labour had commenced for it could be found in /
in stillborn babies. Histological studies showed that it never seemed to occur before the fifth month of gestation. The loss of substance occurred most commonly in the region of the middle cerebral artery. On this basis he suggested that as well as occasional cases of embolism or obstruction of the artery in the course of labour, interference with placental blood supply during late pregnancy or prolonged labour might also be a cause of porencephaly. The distribution of the porencephaly was on the basis of comparative inadequacy of the circulation in the distribution of the middle cerebral artery. A form of anaemic necrosis resulted.

At this time considerable controversy occurred as to the extent to which developmental abnormalities of the arteries or acquired disease in them might be determining factors in porencephaly and other forms of localised cerebral atrophy. Cases were presented in which hemiatrophy of the brain was attributed to congenital abnormalities of the arteries by Mühr, (1876), and Klebs, (1876).

Abercrombie thought that the incidence of foetal endocarditis had been underestimated and that embolism of cerebral arteries, most commonly the middle cerebral was commoner than had been thought, (Abercrombie, 1897). The predisposing effect of syphilitic endarteritis to thrombosis in the intracranial arteries was stressed by Osler (1899) and Freud, (1897). It is true to say, however, that the contentions of Kundrat remained unshaken. So far as porencephaly was concerned the frequent /
frequent origin of the condition in a disturbance of foetal circulation at some time in late pregnancy or during labour was established.

Unfortunately, however, the attention devoted to Kundrat's labours was less than that devoted to less soundly based contemporary work. McNutt emphasised the frequency with which subdural haemorrhage occurred following birth trauma and regarded this as the major cause of cerebral palsy, (McNutt, 1835). At the same time, Strumpell's suggestions that all forms of cerebral atrophy could be explained on the basis of poliomecephalitis analogous to poliomyelitis and probably due to the same aetiological factors made a deep impression, (Strumpell, 1839).

He suggested that differences in the extent of cerebral atrophy and in the severity of the related loss of brain tissue might be attributed to different intensities of the original prenatal poliomecephalitis. The late pathological changes could vary from small areas of cerebral atrophy to large porencephalic defects. This view met with a great deal of uncritical acceptance though the pathological evidence on which it was based was shown to be quite inadequate by Freud, (1897).

It was largely as a result of Freud's work that the limitations of the amount of information to be obtained from a study of late pathological pictures in cerebral palsy were finally realised. Freud pointed out that firm inferences about /
about aetiology could not be made as a result of studying late pathological changes. In the first place not enough was known about the early stages of the processes which eventually resulted in the final pathological pictures. In the second, not enough was known about the nature or mode of action of the aetiological factors which caused brain damage.

To a limited extent the gaps in knowledge of the development of the final pathological pictures found in cerebral palsy have been filled since the time of Freud. Experimental hypoxia in newborn animals has produced pathological pictures comparable to those found in cerebral palsy, (Windle and Becker, 1942, 1943; Windle, Becker and Weil, 1944). The correlation of numerous neuropathological observations made on patients who have survived severe hypoxia for varying periods has led to increased understanding of the sequence of events in the human brain. Thus, the ways in which cerebral hemiatrophy, diffuse cortical atrophy, partial convolutional atrophy, atrophic cortical sclerosis (mantle sclerosis) atrophic lobar sclerosis, hydrancephaly and porencephaly may result from hypoxic injury have been described in detail by Courville and Marsh, (1944) and Courville, (1947, 1950, 1952, 1953). Courville points out that the following effects of hypoxia may be recognised. "(1) Diffuse loss of nerve cells either cortical or striatal, (2) The gross sclerosing processes which deform the cortex generally or locally, (3) The disorders of myelination, either in the form of demyelination or hyperplasia, and (4) the formation of cysts in the cerebral centrum or/
or cortex and centrum", (Courville, 1952). Progressive cerebral degeneration may occur after asphyxia and Courville holds that a proportion of the cases of cerebral atrophy in early life are the result of asphyxia.

More stress has been put on the effects on the brain of vascular abnormalities than on anoxia by other authors, including Harburg and Casamajor, (1944). They point out the regions of the brain affected in stillbirths apparently resulting from birth injuries and the regions showing residual lesions in patients with cerebral palsy are often similar. In particular, (1) The area of the great central vein, including the whole centrum ovale. (2) The area of the superior longitudinal sinus. (3) Less significant, the area of the sinus cavernosus and some ventral branches of the surface veins, i.e. the area ventral to the Sylvian fissure. "Phlebothrombosis and phlebostasis are frequently the cause of severe disintegration of the brain in early childhood. Lesions of the veins and venous sinuses are predominantly caused by trauma before, during or after birth, but other causes such as infection (?) cannot be excluded". A case of postnatal trauma followed by progressive cerebral degeneration and a late appearance of cystic degeneration on pathological examination has been described by Benda, (1945). This author considers that it is impossible to distinguish between lesions due to circulatory disturbances which may be the result of trauma, and changes due to true hypoxia.

Further /
Further advances have also been made in the diagnosis of congenital malformations of the brain. The work of Gonel on the normal development of the foetal nervous system has enabled more accurate dating of the time at which deviations from the normal growth patterns occurred, (Gonel, 1927-1959). A number of pathological changes previously thought to be the result of birth injury have been shown to be attributable to cerebral malformations, (Minkowski, 1952). The differentiation of porencephaly (due to birth injury), from schizencephaly (due to congenital malformation) is an example of the more accurate aetiological diagnosis which is now possible on the basis of pathological findings, (Yakovlev and Wadsworth, 1946). Another example is the finding of Status Marmoratus which is now generally regarded as being due to kernicterus and/or hypoxic brain injury in the vast majority of cases, though until recently this conclusion was hotly disputed, (Norman, 1944, 1947; Malamud, 1950; Benda, 1952).

The Clinicians. The first writer to have realised the importance of "birth injury" as a cause of cerebral palsy from the clinical, rather than the more academic or pathological point of view was Little, (1943, 1962). It is the latter paper which has come to be regarded as the classical account of his findings: "On the influence of abnormal parturition, difficult labours, premature birth and asphyxia neonatorum on the mental and physical condition of the child, especially in relation to deformities".

In /
In it he suggests that the prevailing assumption that all children who appeared to suffer birth injury either died or, more commonly, recovered completely was erroneous. A relatively small proportion of children who were damaged survived in a permanently damaged condition. "Spastic rigidity" was especially liable to occur in these survivors. Not all the "deformities" liable to occur are clearly specified, but amongst the sixty-three cases whose summaries he presented may be recognised congenital hemiplegia and diplegia, dyskinesia and congenital torticollis.

The frequency with which these disorders followed abnormal parturition was striking. "The forms of abnormal parturition which I have observed to precede certain mental and physical derangements of the infant consist of difficult labours, i.e. unnatural presentations, tedious labours from rigidity of the maternal passages or aperture, instrumental labours in which turning has had recourse to, breech presentations, premature labours and cases in which the umbilical cord had been entangled around the neck or had fallen down before the head".

The mechanism by which these forms of abnormal parturition caused damage was believed to be similar to that present in stillborn infants, or in those dying in the neonatal period after asphyxia neonatorum. The pathological findings in such cases had been described in detail by a number of authors including Weber, (1951) and Hecker, (1953). These studies had /
had shown that in the majority of cases generalised congestion especially of brain, lungs and other viscera, petechial haemorrhages, sometimes ecchymoses and effusion of blood into the meninges were found. The changes had been interpreted as being due to asphyxia, rather than to trauma. Hence, "- the more the facts I adduce are studied, the more apparent, in my opinion, will it be that a large proportion of infants either dead, stillborn, apoplectic or asphyxiated at birth have been rendered so by interruption of the proper placental relation of the foetus to the mother and non-substitution of pulmonary respiration then from direct mechanical injury to the brain and spinal cord". Little does not deny the possibility of traumatic damage, including "rupture of the longitudinal and transverse sinuses of the brain" occurring, but stresses that blood in the linings of the brain is more often the result of asphyxia. When the blood was present at the base of the brain the medulla oblongata might be affected permanently and difficulty in swallowing, speaking and other "functions of organic life" could result. He appears to have regarded damage to the spinal cord and medulla as being of at least equal importance to the effects on the brain itself. Failure to breathe after birth might be due to brain damage, but persistent failure to establish respiration was an additional damaging agent.

The importance of Little's paper was not fully realised at the time. In fact, however, he had not only noted the association /
association of abnormal parturition with cerebral palsy, but had defined the disorders of the birth process most likely to cause damage to the infant..... the factors of Little, Freud, (1891). Subsequent workers tended to emphasise first one of these factors, then another, but no such comprehensive account of the relationship between the different abnormalities of parturition and cerebral palsy was written for many years.

Ten cases of cerebral palsy were presented by Ross (1832). He distinguished between acquired hemiplegia and congenital (birth) palsies. Amongst the latter, hemiplegia, tetraplegia, paraplegia and cases of "bilateral athetosis" had to be distinguished. Ross emphasised the importance of "asphyxial labour" in causing damage to the brain, the developing pyramidal tracts, and in some cases the spinal cord. He thought that even in children showing spastic paraplegia the cerebrum was usually the primary site of damage. In some cases of bilateral spastic paralysis, developmental defects and not birth injuries were present. Neither Ross nor Hadden (1834) specified the precise abnormalities of parturition which might be expected to result in injury to the nervous system.

In 1835 Sarah McNutt described a case in which, after an assisted breech delivery with forceps to the aftercoming head, a girl had recurrent convulsions. The convulsions lasted from nine days, and after them there was "complete paralysis of both sides of the body, not including the face". She /
She died of pneumonia and at autopsy, atrophic areas were present in both cerebral hemispheres and the meninges were thickened. She also described a series of cases of stillbirth and neonatal death in which subdural haemorrhages were found following breech and forceps delivery, (McNutt, 1885a, 1885b). In spite of the fact that Welch, the pathologist who examined her case of “complete paralysis of both sides of the body” thought the atrophic areas were certainly of prenatal origin, McNutt concluded that subdural haemorrhages were the commonest causes of congenital cerebral palsy. They were most often the result of breech or traumatic forceps deliveries and by pressing on the surface of the brain they caused cerebral atrophy.

McNutt’s work was widely accepted and the emphasis in contemporary writings shifted from the ill-effects of asphyxia on the brain to the dangers of trauma and especially breech and instrumental deliveries. For example, Gowers noted that in cases of palsy which depended on an injury to the brain “you find a strong presumption that an injury during birth has been their cause .... In most cases there is a history of difficult parturition, sometimes of a labour that was only ended by forceps; in many cases the head of the child presented unmistakable external signs of the compression it had endured”. Sixteen of twenty-six of his patients with cerebral palsy were first born, and of the other ten, six were delivered by the breech. He accepted McNutt’s explanation of the disorder ...” — whose investigations constitute,
I think, by far the most valuable contribution to medical science that the profession has yet received from a member of her sex. The explanation appears to have been accepted equally for all forms of cerebral palsy, (Gower, 1883). With improvements in the classification of the various forms of cerebral palsy it became possible to compare and contrast the birth histories in the different categories of disease. Unfortunately, since the classifications and thoroughness of history-taking varied there was considerable disagreement about the significance of birth injury in different series. For example, birth injury as a cause of either congenital hemiplegia or "bilateral hemiplegia" (diplegia) was thought to have been over-emphasised by Lovett, (1883). Osler had 15 cases of congenital hemiplegia of whom 9 were delivered by forceps and 2 were premature, (Osler 1889). He considered birth injury, and more particularly trauma, to be important in congenital hemiplegia and in "bilateral hemiplegia". Of his 20 patients in the latter category, five showed some involuntary movements and would probably be classified as suffering from dyskinesia now. Of the 5, 3 had a history of abnormal labour and one of neonatal jaundice. Of the 15 who would now be classified as suffering from tri- or tetraplegic diplegia, 3 were born prematurely, 2 were delivered by forceps, 2 after abnormal labour and one after the mother had had a heavy fall during pregnancy. In 6 cases the birth had been normal, and in one case there was no birth history. Osler thought that birth injury was less important in spastic paraplegia.
paraplegia. Six of his 11 cases in this category had no history of abnormal labour or delivery.

Rather similar figures to those of Osler were presented in an analysis of 49 cases of congenital cerebral palsy by Sachs and Peterson, (1890). They found there had been "some difficulty in labour, simple delay, or instrumental delivery" in approximately one-third of their cases. They were probably the first authors to draw attention to the fact that prolonged labour was probably a rather more important cause of trauma than was instrumental delivery.

The possibility of hereditary factors being important in predisposing children to cerebral lesions of congenital origin was discussed by Wallenberg, (1896). He found that only 6 of 19 patients with congenital hemiplegia had a history of forceps delivery or difficult labour, and regarded birth injury as being merely a precipitating factor producing the clinical picture in a predisposed individual.

Prematurity, another of the factors of Little was thought to be the important aetiological agent in diplegia by Brissaud, (1894). He postulated that the greater severity of the involvement in the lower limbs than the upper could be explained on the basis of agenesis of the pyramidal tracts resulting from the changed environment to which they were prematurely subjected. If the child were very premature then both upper and lower limbs would be affected, but if less premature than the further developed pyramidal fibres destined to supply the upper
upper limbs would be largely spared. He attempted to restrict the term "Little's disease" to prematurely born children with abnormalities in the pyramidal tracts. For this he was later taken severely to task by Freud, (1897).

The work of Freud summarised the previous opinions about the importance of birth trauma and criticised them with a relentlessness and clarity which still compels admiration, (Freud, 1893, 1897). Freud has rather little to say about the clinical study of the aetiology of congenital hemiplegia in childhood, but thought birth injury more important than in diplegia.

Freud is often quoted as having stated that birth injury is unimportant as a cause of diplegia, (Collier, 1999, 1924; Ford, 1926). This appears to be the result of misinterpreting his somewhat involved and complex argument. Freud described two series of cases of diplegia, (in which category are included "paraplegic rigidity", "generalised rigidity", "bilateral hemiplegia", and "choreic forms"). In 34% of 270 cases taken from the literature there seemed to be no obvious cause of the condition. In 7%, prenatal factors appeared to be important, and extrauterine factors in 7%. Little's factors were present in up to 62%. In Ganghofner's less selected series, 42.7% were without apparent aetiological factors, maternal factors were present in 31.4%, Little's factors in 9.2% and extrauterine causes in 6.2%. More detailed analysis showed that in cases of generalised rigidity the cause was unknown in about one-third /
one-third of cases, difficult birth was present in another third, and premature birth in about one-sixth. In paraplegic rigidity premature birth was present in about half of the cases and a quarter were of unknown aetiology. In choreic forms the causal factors were unknown in about 50 per cent. of cases. Asphyxia was significant in about 25 per cent. and maternal factors (disease of the mother) were also important.

The associations between these single aetiological factors and the individual conditions were not exclusive. For example, a proportion of premature children showed generalised rigidity and a proportion of cases of paraplegic rigidity had shown asphyxia at the time of birth. Moreover it was important to distinguish between associations and cause and effect. For example, a proportion of patients suffering from cerebral palsy who had been born after abnormal labour or delivery could be shown to be suffering from prenatal disorders. "Difficult birth and premature birth are not always accidental happenings, but may often be the consequence of an underlying cause, or even by the expression of this without having any effect (on the child) per se. It is easy to conceive of those pathological factors which have made the intrauterine development abnormal extending their influence to birth process, so that abnormal birth is the crowning termination of an abnormal pregnancy". Freud goes on to lay stress on Little's observation that the majority of children born after abnormal labour recover apparently unharmed, and suggests that there must be some reason why a minority of children are affected whilst the majority /
majority are spared. Clinical observation lends little support to the assumption that the noxious factors present at the time of birth are more severe in the cases which are affected. He suggests that there is another explanation, "The supposition that other contributory factors come into play which render the abnormal birth pathogenic. What these factors are, whether in the development of the brain, or the resistance of the vascular system, or of the nervous tissue is not known. ... Falling ill with Little's syndrome amounts to a proof of congenital conditioning." He points out that when this conditioning is marked even the stress of normal birth may be enough to produce diplegia.

Eight years after Freud's paper, the category of congenital ataxia was added to the group of cerebral palsies by Batten, (1905). He described 3 cases in 3 of which, at least, there was a history of abnormal labour and delivery. Further cases were added by him in 1905 and Hunt described 4 more with a history of "birth injury" in 1913. Following Hunt's paper there was considerable controversy as to how many cases suffering from ataxia or ataxic diplegia in childhood were the result of developmental abnormalities, and how many to actual injury during labour and delivery, (Ford, 1952).

In 1899 Collier presented 7 patients suffering from cerebral diplegia. In some of these the disorder appears to have been progressive and rather atypical in its course. He noted the familial and hereditary occurrence of diplegia and /
and discussed its aetiology. "The attributed aetiological factors are here summarised:

1. Maternal pathological states during pregnancy; general ill-health, syphilis, specific fevers, eclampsia, pathological psychic states and injury to the abdomen.


3. Heredity and consanguinity of parents.

4. Postnatal pathological conditions, acute infectious diseases and acute fevers, vascular lesions, epilepsy, acute encephalitis."

He stated that more than one of the attributed causes might be present in a given case and that "it is probable that many of the abnormalities of birth stand not in causal relationship to the disease, but as concomitant effects of a morbid maternal state. It is certain also that some of the postnatal factors are immediate precipitating factors, bringing into evidence a congenitally installed disease."

In 1924 he re-stated his thesis with some modifications. He then believed that diplegia was the result of degenerative disease of the cortical neurones which was often familial and similar in type to amaurotic familial idiocy (which condition he felt should be included in the category of diplegia). He stated - "I would remove maternal ill-health from the causal factors, placing them as common, but by no means constant clinical associates of diplegia, indicative of something wrong with the foetus, or of something wrong with those mysterious /
mysterious relations between mother and offspring which determine a speedy delivery at full-time".

In an analysis of 200 cases of bilateral spastic paralysis, Ford found a history of abnormal labour in only 15%. 33% were prematurely born. He commented that the symmetry of the paresis, the small size of the head and the absence of pathological evidence of birth injury in the few available autopsies, were much in favour of birth injury being relatively unimportant in the aetiology of the condition. In contrast to these cases there was a history of abnormal labour in about 70% of patients suffering from asymmetrical cerebral palsy. Most of the heads were large rather than small, and there was evidence of focal brain damage on neurological examination. "From the material available the conclusion seems indicated that the congenital hemiplegias, monoplegias and double hemiplegias with large, normal or asymmetrical heads are in most cases to be considered true birth palsy. Intrauterine arrests of development and vascular occlusions undoubtedly constitute a part of this group, but probably a smaller part.''

Patten was also impressed with the inconstancy of the association between cerebral diplegia and birth injury. He could find few differences in the clinical courses, or in the pathological findings (of which he had few cases) of patients suffering from diplegia who had, or had not, a history of birth /
birth injury. He thought his findings "indicated more than
the effects of trauma or vascular accidents in the neurologic
conditions of the newborn infant". Intrinsic factors affecting
"the integrity of cortical cells or the proper myelination
of the corticospinal tracts and associated fibers" might
be postulated, (Patten, 1931).

In the same year Crothers described 3 patients suffering
from involuntary movements who had been abnormal from birth.
He attributed their disorder to disease of the basal ganglia
acquired as a result of birth injury, (Crothers, 1931).
Further cases of "congenital choreothetosis" attributed to
birth injury and especially to postnatal asphyxia were present¬
ed by Byers, (1942).

One hundred and eighty five cases of "cerebral spastic
paralysis" whose clinical condition was not further described
were reviewed by Gustafson and Garceau, (1941). The greatest
number of cases appeared to follow spontaneous vertex delivery.
Thirty per cent. were prematurely born. Posterior pituitary
extracts had been used in 13% of the series. "Cerebral spastic
paralysis was present in many infants showing no clinical
evidence of apnoea or asphyxia". The value of studies of this
kind is extremely limited by the failure of the authors to
attempt some correlation of the type of palsy with the birth
histories.

More recent surveys have tended to give similar findings
to those of Ford. To some extent the conclusions that abnor-
mal birth was not such an important cause of diplegia as had
been /
thought were strengthened by Penrose's finding that a proportion of cases appeared to be due to a Mendelian recessive genetic trait, (Penrose, 1933). Families of diplegic patients were found to have at least one other affected diplegic child in 6.5% of cases by Yannet, (1944). 85% of pregnancies were uneventful, and labour, delivery and the neonatal state were found to be normal in 75% of cases. In contrast, 48% of labours were abnormal in children suffering from congenital hemiplegia. Yannet considered that this evidence suggested birth injury to be more important in the aetiology of hemiplegia than in that of diplegia. Diplegia was often due to congenital malformations of genetic origin. The fact that a higher proportion of hemiplegic than of diplegic patients were first born supported this contention, (McGovern and Yannet, 1947).

A detailed clinical and pathological study was made of 50 cases suffering from congenital hemiplegia by Stewart, 1943). He tabulated his findings as follows.

**Etiological factors in Congenital Hemiplegia**

**Stewart, 1943. Fifty Cases**

<table>
<thead>
<tr>
<th>Factor</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ill health of mother during pregnancy</td>
<td>10</td>
</tr>
<tr>
<td>Injury to mother by fall during pregnancy</td>
<td>2</td>
</tr>
<tr>
<td>Difficult or abnormal labour</td>
<td>26</td>
</tr>
<tr>
<td>Cerebral angioma with facial naevus</td>
<td>2</td>
</tr>
<tr>
<td>Unascertained.</td>
<td>10</td>
</tr>
</tbody>
</table>

Unfortunately, his rather scanty description of the forms
of abnormal labour and the neonatal state of the babies makes further analysis of the types of birth injury difficult.

The early histories of 114 patients suffering from cerebral palsy were studied by Evans, (1943). Only his "spastic" and "athetoid" groups were large enough for detailed analysis of the aetiological importance of single factors in the birth histories. By a comparison of the histories of 38 "spastic" (diplegia) cases with those of 38 "athetoids" (dyskinesias) and a control group, he reached the following conclusions. There appeared to be no significant increase in the proportion of abnormal pregnancies, labours and deliveries in the spastic group as compared to the controls. There was a significant increase in the proportion of prematurely born patients. The "athetoid group" could be divided into those with jaundice (due to rhesus incompatibility) in the neonatal period, and those with a history of asphyxia, often associated with primogeniture, the use of anaesthesia and forceps. "Taken together these features suggest an association between difficult labour and the development of double athetosis". (See Table 216.)

Very similar conclusions were reached by Asher and Schonell, (1950) who studied 400 cases of cerebral palsy obtained during a regional survey of children suffering from the condition in the Midlands of England. Some of the differences in the findings of these authors and those of Evans are probably explicable on the different selection of cases, for many of those described by the latter were candidates for a /
a school for spastic children. Their findings are summarised in Table 215.

The numbers of cases with a history of birth injury or with a neonatal state suggestive of birth injury have been combined and are shown in Table 215. It will be observed that all the studies show a higher proportion of abnormal labours and deliveries amongst children with dyskinesia than amongst those with diplegia. All these authors agree that birth injury appears to be responsible for more cases of "athetosis" than of "spastic" paralysis. A higher proportion of diplegic than hemiplegic patients appear to be suffering from the effects of developmental anomalies of the brain.

No large series of patients of ataxic patients appears to have been published which studies aetiological factors.

Unfortunately many other recent studies involving much labour give information of limited value as the clinical conditions of the patients are inadequately described or classified, and in some of them patients suffering from cerebral palsy are grouped together with those who are mentally retarded, or epileptic, (Link, 1950; McKhann, Belnap and Beck, 1951; Denhoff and Holden, 1951; Latham, Anderson and Eastman, 1954).
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No. of cases</th>
<th>Abnormal Labour</th>
<th>Normal Labour **</th>
<th>Obstetric history unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Abnormal Labour</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>With Birth Injury</td>
<td>Without Birth Injury</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Total</td>
<td>17</td>
<td>12</td>
</tr>
<tr>
<td>Athetosis</td>
<td>26</td>
<td>12</td>
<td>60</td>
<td>5</td>
</tr>
<tr>
<td>Symmetrical quadriplegia</td>
<td>43</td>
<td>12</td>
<td>58</td>
<td>7</td>
</tr>
<tr>
<td>Asymmetrical quadriplegia</td>
<td>11</td>
<td>3</td>
<td>100</td>
<td>0</td>
</tr>
<tr>
<td>Paraplegia</td>
<td>50</td>
<td>16</td>
<td>63</td>
<td>6</td>
</tr>
<tr>
<td>Right hemiplegia</td>
<td>43</td>
<td>16</td>
<td>63</td>
<td>6</td>
</tr>
<tr>
<td>Left hemiplegia</td>
<td>28</td>
<td>11</td>
<td>82</td>
<td>2</td>
</tr>
<tr>
<td>Other spastics</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Others</td>
<td>17</td>
<td>12</td>
<td>66</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>221</td>
<td>87</td>
<td>65.5</td>
<td>30</td>
</tr>
</tbody>
</table>

** Including those where an anaesthetic was administered but labour was otherwise normal.
The percentage of patients in a recent series suffering from various forms of cerebral palsy who had a history of abnormal pregnancy, abnormal labour or delivery, or were premature.

<table>
<thead>
<tr>
<th>Type of Palsy</th>
<th>Hemiplegia</th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Author</td>
<td>Stewart 1943</td>
<td>Asher &amp; Schonell 1950</td>
<td>Yannet</td>
<td>Asher &amp; Schonell 1950</td>
<td>Evans 1943</td>
<td>Yannet</td>
<td>Asher &amp; Schonell 1950</td>
</tr>
<tr>
<td>Abnormal pregnancy</td>
<td></td>
<td>24</td>
<td>-</td>
<td>5</td>
<td>-</td>
<td>21</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td>Abnormal labour or delivery</td>
<td></td>
<td>52</td>
<td>55</td>
<td>48</td>
<td>43</td>
<td>36</td>
<td>17</td>
<td>73</td>
</tr>
<tr>
<td>Normal pregnancy and normal delivery</td>
<td></td>
<td>24</td>
<td>45</td>
<td>7</td>
<td>52</td>
<td>-</td>
<td>2</td>
<td>27</td>
</tr>
<tr>
<td>Prematurity</td>
<td></td>
<td>8</td>
<td>30</td>
<td>16</td>
<td>44</td>
<td>24</td>
<td>13</td>
<td>28</td>
</tr>
</tbody>
</table>
Birth Injury as a cause of cerebral palsy.

Introduction. Today birth injury has come to mean damage to the foetus sustained directly or indirectly as a result of the birth process. Birth injury must be distinguished from damage to the infant sustained after birth as in acquired (postnatal cerebral palsy). The distinction is not always easily made for the clinical effects of brain damage caused during pregnancy, or labour may be delayed for some hours or days. Birth injury must also be distinguished from abnormalities of the brain which are the result of developmental malformations dating from early pregnancy and which may be genetically determined, or due to a variety of teratogenic insults.

In practice it is convenient to consider the term "birth injury" to comprise damage sustained by the foetus in the last trimester of pregnancy, or during labour and the process of delivery. To determine whether cerebral abnormalities are due to birth injury or not in the individual patient suffering from cerebral palsy may be very difficult. Aetiological diagnosis in cerebral palsy is circumstantial in the majority of cases. In general, the clinical manifestations in cerebral palsy are those of immature patterns of motor behaviour, whether the underlying abnormality of the brain is due to birth injury or to developmental malformations. Pathological findings are available in only a minority of patients,
and even then may not distinguish conclusively between malformation, birth injury or damage acquired postnatally. (Stewart, 1942, 1948; Normen, 1944). The presence of abnormalities of pregnancy, labour or delivery does not necessarily mean that these have damaged the child. Children with developmental malformations of many types are more frequently born after abnormal pregnancy, labour and delivery than are normal children. (Landtman, 1948). Yet, whereas cleft palate is not ascribed to birth injury, a causal relationship between abnormalities of parturition and cerebral palsy is often assumed on scanty evidence.

Assessments of the relative danger to the child resulting from particular abnormalities of parturition are largely dependent upon studies of series of stillbirths and neonatal deaths. Yet if these assessments are applied to patients suffering from cerebral palsy it is immediately implied that the harmful factors which cause foetal or neonatal death are the same as those which cause brain damage compatible with survival in cerebral palsied patients. There is much evidence to suggest that this is a justified assumption, but it is important to remember that an assumption has been made, (Lilienfeld and Parkhurst, 1951). In a given Western community with a relatively high standard of living there are likely to be about ten stillbirths and between 3 and 10 neonatal deaths for every case of congenital cerebral palsy. Clearly there may be factors other than the severity of the damage /
damage caused by abnormalities of parturition to account for the fact that the majority of children survive apparent birth injury and seem healthy, a minority perish, and a still smaller minority survive to show cerebral palsy. Little or nothing is known of these possible factors determining whether a child survives unharmed, dies, or survives in a damaged state.

It is still true, however, that the largest body of evidence about the effects on the child of the various abnormalities of parturition is that derived from studies of series of children who have been stillborn or died shortly after birth. This evidence may be supplemented by clinical observations, e.g. the presence or absence of foetal distress or neonatal apnoea when complications of pregnancy, labour or delivery have occurred. More recently oxygen saturation in the cord blood has been directly estimated in large series of cases with and without disorders of parturition. The results of these studies provide valuable but limited confirmatory evidence about the effects of the various disorders of pregnancy, labour and delivery on the offspring. Further evidence is provided by animal experiments in which the effects of artificially simulated disorders of parturition on the offspring have been observed. In the following pages an attempt is made to assess the dangers to the child of the commoner disorders of parturition. For convenience these are considered in terms of their likely effect on the child. They /
They are classified as "Hypoxic", "Traumatic" and "Toxic". This classification is somewhat arbitrary and artificial. For example, in the last resort the majority of toxins act by interfering with the nutrition or oxygen exchange of nerve cells, and there are many abnormalities of parturition such as very prolonged labour which may be demonstrated to cause both hypoxia and trauma.

Nevertheless there are pathological findings which are accepted by pathologists as indicating "hypoxic" or "traumatic" death in children dying before or shortly after birth. Specific toxic factors or infections may also produce characteristic pathological appearances. By correlating the birth histories with the pathological findings in large series of cases it is possible to study the effects of the various disorders of parturition on the child. These disorders are then classified, rather crudely according as to whether they are "toxic", "hypoxic" or "traumatic", from the child's point of view.

Finally, using the information obtained from published studies, an attempt is made to interpret the birth histories of the patients suffering from cerebral palsy, studied during the survey, in terms of their likely hypoxic, traumatic and toxic effects.

The criteria and causes of hypoxic birth injury.

The criteria of hypoxia. — Though sudden severe hypoxia
CHAPTER 2b.
The criteria and causes of hypoxic birth injury.

The criteria of hypoxia. Though sudden severe hypoxia may result in foetal or neonatal death without there being recognisable pathological changes in the nervous system or elsewhere, hypoxic death usually results in pathological appearances which are diagnostic, (Parrot, 1873; Hutinel, 1877; Browne, 1921; Potter, 1951; Courville, 1954; Morison, 1946). They have been well summarised by Macgregor, (1943).

"At autopsy, evidence of foetal asphyxia is characteristic and easily recognised: profound lividity of the skin; a dusky cyanotic hue of all the tissues and organs; extreme engorgement of veins everywhere; dark fluid blood; and petechial haemorrhages under the serous membranes, especially in the thorax and sometimes elsewhere, notably in the thymus gland. This may be all that is to be found, but other results are not uncommon. Among these the most important are severe and possibly fatal haemorrhage in various situations, and certain effects on the lungs and respiratory function.

Reference has already been made to asphyxia as probably the usual cause of intraventricular and subarachnoid forms of intracranial haemorrhage. Other situations in which it may be concerned in producing disastrous haemorrhages are the suprarenal glands and the surface of the liver. In the former situation it may destroy one or both suprarensals, with immediately fatal result".

Though not constant, these changes appear to constitute the best criteria of hypoxia available. Certainly clinical criteria, such as slowing or irregularity of the foetal heart and /
and the passage of meconium are less reliable, (Soidenhoff and Brill, 1954). Studies of the oxygen saturation in the umbilical vein are not yet numerous enough, or well enough controlled to be considered as sure indications of the degree of hypoxia to be expected in various disorders of pregnancy and labour, (Walker, 1953; Tizard, 1956).

Studies of hypoxic stillbirth and neonatal death. In Tables 217, to 219, the numbers of hypoxic stillbirths and neonatal deaths are shown by the complications of parturition which occurred in two large American series, and a smaller, more closely studied Scottish one, (Nesbitt and Anderson, 1953; Potter and Adair, 1939; Drillien, 1947).

The most detailed analysis of the complications of parturition was that of Potter and Adair, (1939). They studied both the apparent cause of the hypoxia and the mode of delivery, recognising that in a child that was already hypoxic, stressful forms of delivery were much more dangerous than when the foetal oxygenation was adequate.

From their tabulated findings it will be observed that 109 of the 136 cases suffered from various complications of pregnancy, and in only 21 were born by natural cephalic delivery. In the remaining 115, labour or delivery as well as pregnancy, were abnormal. This illustrates the difficulty in isolating single disorders of parturition for study.

In all the series, however, the major apparent causes of hypoxia were similar. There was placental separation complicating/
### Chicago Lying-in Hospital - 221 stillbirths in 17500 births

<table>
<thead>
<tr>
<th>Anoxia</th>
<th>Before labour</th>
<th>During labour</th>
<th>Total</th>
<th>Approx.</th>
<th>Before labour</th>
<th>During labour</th>
<th>Total</th>
<th>Approx.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>prem. term</td>
<td>prem. term</td>
<td></td>
<td></td>
<td>prem. term</td>
<td>prem. term</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Abruptio placenta</td>
<td>13 1</td>
<td>10 7</td>
<td>31</td>
<td>37</td>
<td>6 11</td>
<td>4 10</td>
<td>31</td>
<td>29</td>
</tr>
<tr>
<td>Placenta previa</td>
<td>1 1</td>
<td>2 0</td>
<td>4</td>
<td>5</td>
<td>1 2</td>
<td>2 2</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Cord obstruction</td>
<td>5 8</td>
<td>4 10</td>
<td>27</td>
<td>32</td>
<td>1 1</td>
<td>7 1</td>
<td>10</td>
<td>9.5</td>
</tr>
<tr>
<td>Complications of preg. labour and delivery, including anoxic deaths from toxemia.</td>
<td>0 0</td>
<td>1 7</td>
<td>3 10</td>
<td>1 10</td>
<td>5 27</td>
<td>43</td>
<td>41</td>
<td></td>
</tr>
<tr>
<td>Abdominal preg.</td>
<td>2 0</td>
<td>0 0</td>
<td>2</td>
<td>2</td>
<td>0 1</td>
<td>0 0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Others</td>
<td>3 3</td>
<td>3 3</td>
<td>12</td>
<td>14</td>
<td>2 1</td>
<td>1 9</td>
<td>10</td>
<td>12.5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>24 13</strong></td>
<td><strong>20 27</strong></td>
<td><strong>84</strong></td>
<td><strong>100</strong></td>
<td><strong>11 26</strong></td>
<td><strong>19 49</strong></td>
<td><strong>105</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>

### Jules Holmes - 224 cases in 11248 births

<table>
<thead>
<tr>
<th>Anoxia</th>
<th>Before labour</th>
<th>During labour</th>
<th>Total</th>
<th>Approx.</th>
<th>Before labour</th>
<th>During labour</th>
<th>Total</th>
<th>Approx.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>prem. term</td>
<td>prem. term</td>
<td></td>
<td></td>
<td>prem. term</td>
<td>prem. term</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Abruptio placenta</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>6 11</td>
<td>4 10</td>
<td>31</td>
<td>29</td>
</tr>
<tr>
<td>Placenta previa</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 2</td>
<td>2 2</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>Cord obstruction</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 1</td>
<td>7 1</td>
<td>10</td>
<td>9.5</td>
</tr>
<tr>
<td>Complications of preg. labour and delivery, including anoxic deaths from toxemia.</td>
<td>0 0</td>
<td>1 7</td>
<td>3 10</td>
<td>1 10</td>
<td>5 27</td>
<td>43</td>
<td>41</td>
<td></td>
</tr>
<tr>
<td>Abdominal preg.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0 1</td>
<td>0 0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Others</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2 1</td>
<td>1 9</td>
<td>10</td>
<td>12.5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>11 26</strong></td>
<td><strong>19 49</strong></td>
<td><strong>105</strong></td>
<td><strong>100</strong></td>
<td><strong>11 26</strong></td>
<td><strong>19 49</strong></td>
<td><strong>105</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>
### TABLE 218

**Causes of asphyxia in 63 cases of 372 cases of stillbirths. (Drillien, 1947)**

<table>
<thead>
<tr>
<th>Asphyxia</th>
<th>Total</th>
<th>Approx. %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Due to toxaemia</td>
<td>11</td>
<td>17</td>
</tr>
<tr>
<td>&quot;&quot; ante partum haemorrhage</td>
<td>27</td>
<td>43</td>
</tr>
<tr>
<td>Associated with breech delivery</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>&quot;&quot; version extraction</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>&quot;&quot; forceps delivery</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>&quot;&quot; Caesarean section</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>&quot;&quot; prolonged labour</td>
<td>10</td>
<td>15</td>
</tr>
<tr>
<td>Others and cause unknown</td>
<td>7</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>63</td>
<td>100</td>
</tr>
</tbody>
</table>
### TABLE 219

<table>
<thead>
<tr>
<th>Complications of pregnancy and mode of delivery in 136 cases of stillbirths and neonatal deaths from anoxia in a series of 526 cases. Potter and Adair, 1939</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Anoxemia</td>
</tr>
<tr>
<td>Mat. compl.</td>
</tr>
<tr>
<td>Abruptio placentae</td>
</tr>
<tr>
<td>Abruptio &amp; toxemia</td>
</tr>
<tr>
<td>Toxaemia</td>
</tr>
<tr>
<td>Cord compl.</td>
</tr>
<tr>
<td>Medical compl.</td>
</tr>
<tr>
<td>No mat. compl.</td>
</tr>
<tr>
<td>All cases from all causes</td>
</tr>
</tbody>
</table>
complicating preeclampsia or placenta previa, preeclampsia or cord complications. These disorders of parturition were present in 73% of the stillbirths and 49% of the neonatal deaths in the series of Potter and Adair, (1939). They will now be considered in more detail.

**Hypoxia during pregnancy.**

The effects of preeclamptic toxaemia on the foetus. The evidence that preeclampsia is frequently associated with the occurrence of foetal injury or death is derived from a large number of different clinical and pathological studies. But as in the present series there is difficulty of finding a large number of cases in which preeclampsia appeared to be the only disorder of parturition.

In a study of 773 stillbirths and neonatal deaths it was found that fifty-nine per cent. were premature by weight, though only 6.3% of the total births from which the series was taken were premature, (Potter and Adair, 1939). These authors found maternal complications in 44.6% of their 773 cases. Toxaemia was second in frequency only to placental separation as a maternal complication. Only eighteen per cent. of the infants born to toxaemic mothers showed definite pathological evidence of hypoxia, but others had lesions, such as intracranial haemorrhage which are likely to have been hypoxic in origin. Maternal complications were noted in 9 per cent. of the infants born at term and later died and in 28 per cent. of those stillborn at term.

Rather /
Rather similar statistics were presented in pathological surveys of neonatal mortality and morbidity by Macgregor, (1946, 1948). She stressed the importance of maternal disorders of pregnancy as a cause of death from asphyxia before, during, or immediately after labour, and also noted the prominent part played by asphyxia in the aetiology of subarachnoid and intraventricular haemorrhage, conditions classified separately from asphyxia by the American author. Of 162 stillbirths attributed to asphyxia in Macgregor's series, ninety-four were full-time and sixty-eight premature.

In a study of 567 stillbirths in Johns Hopkins Hospital Obstetric Service it was found that 180 were associated with maternal toxaemia, of whom approximately 60% were premature, (Nesbitt and Anderson, 1953). These authors found that 9.5% of the stillborn infants of toxaemic mothers showed pathological signs of hypoxia only, and 36% showed no abnormalities on pathological examination. The remaining cases showed other lesions such as intracranial haemorrhage, pulmonary atelectasis and various congenital abnormalities. The authors commented on the difficulty of relating their pathological findings directly to the presence of preeclampsia because of the frequency with which abruptio placentae and heavy sedation were complications of the clinical picture. They commented "In fact in 42.7% of infants who were stillborn, or died in the neonatal period it was thought the deaths were unrelated directly to the toxaemia, but were the result /
result of allied conditions related to the treatment of the
disease or to obstetric intervention".

The fact that patients with preeclamptic toxaemia show
more extensive and frequent infarction of the placenta than
do those without toxaemia is an old observation but only
relatively recently has statistical study been made of the
chances of severe infarction, placental separation, ante-
partum haemorrhage and anoxia of the foetus been made. It
has been shown infarction in placentae of toxaemic mothers
tends to be more extensive than in mothers without toxaemia
and that the more severe the toxaemia the greater are the
chances of placental separation. It has been said that the
patient without toxaemia has only one chance in a hundred
and twenty of placental separation, but that if she has hyper-
tension, weight gain and albuminuria, her chances are one in
eighteen. In toxaemia of severe degree, the chances of
partial placental separation are probably in the region of
one in seven or eight cases, and in eclampsia, one in two or
three, (Hertig, 1953). Partial separation of the placenta
is not necessarily accompanied by antepartum haemorrhage,
and it is with sorrow that one must record that the details
noted of placentae in the present series are scanty.
Fortunately, however, the toxaemic cases were more frequently
delivered in hospital than were mothers with other disorders
of pregnancy. Hertig has shown convincingly that the risk
to the foetus in preeclampsia is increased when placental
separation, which is not always clinically recognisable, has
occurred, (Table 220).
### TABLE 220

<table>
<thead>
<tr>
<th></th>
<th>Mild</th>
<th>Severe</th>
<th>Eclampsia</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Per cent. foetal loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>without separation</td>
<td>7.0</td>
<td>27.0</td>
<td>56.0</td>
<td>11.0</td>
</tr>
<tr>
<td>Per cent. foetal loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>with separation</td>
<td>43.0</td>
<td>65.0</td>
<td>70.0</td>
<td>55.0</td>
</tr>
</tbody>
</table>

It has to be remembered, however, that the majority of the children of toxaemic mothers who succumb and are still-born, or die in the neonatal period, are premature.

In a carefully controlled survey of the effects of maternal ill-health, complications of pregnancy and delivery, and other factors on the incidence of prematurity, stillbirth and neonatal death in a hospital series, Drillien was unable to demonstrate any significant difference between the survival rates of children of similar weights born to toxaemic or non-toxaemic mothers, (Drillien, 1947). She concluded that the greater overall perinatal mortality amongst children born to mothers with preeclampsia was due to the fact that they gave birth to a much higher proportion of premature infants, 14.9% in contrast to 4.5% in the control group. In 31 cases of stillbirth attributed to asphyxia, fifty-nine weighed more than five and a half pounds at birth, and 24.4% of the mothers had shown evidence of toxaemia. (Table 221)

There is a growing amount of clinical evidence that preeclampsia places the foetus in danger of becoming hypoxic. The slow decline in oxygen concentration in the umbilical vein /
vein from the thirtieth to the thirtyninth week of gestation which is observed in normal pregnancies is greatly exaggerated in pregnancy complicated by preeclampsia, (Walker and Turnbull, 1953; Walker, 1954). This author found that in moderately severe preeclampsia the oxygen content in the vein may be low some weeks before term and the oxygen carrying capacity high. Even in mild preeclampsia, however, the saturation at birth may fall well below normal level. This danger is accentuated when the infant is at all postmature, a point also emphasised by Gruenwald, (1953).

On the basis of these findings it was felt justifiable to classify cases in which the mothers were known to have preeclampsia and being possibly hypoxic during pregnancy when the toxaemia was moderately severe and probably hypoxic when it was severe.

The aetiological importance of antepartum haemorrhage. The danger to the foetus resulting from antepartum haemorrhage with its attendant disturbance of placental nutrition is well recognised. Antepartum haemorrhage was found to be the commonest cause of perinatal death attributable to maternal disease in a survey of 773 stillbirths and neonatal deaths by Potter and Adair, (1939). Of the 117 cases in which foetal or neonatal death followed antepartum haemorrhage, approximately half showing no pathological lesions, one-third showed evidence of asphyxia and in the remainder "a variety of causes of death were found". In a similar survey of the causes of stillbirth and neonatal death, Macgregor (1947) noted /
### TABLE 221a

**Survey of Infants in Placenta Previa, 1931-45, Chicago Iving-In Hospital**  
*(Davis, Potter and Bruce, 1953)*

<table>
<thead>
<tr>
<th>Grams</th>
<th>Number</th>
<th>Stillbirths</th>
<th>Live births</th>
<th>Neonatal deaths</th>
<th>Total %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Over 2,500</td>
<td>181</td>
<td>9</td>
<td>172</td>
<td>10</td>
<td>89.5</td>
</tr>
<tr>
<td>1000 - 2500</td>
<td>119</td>
<td>24</td>
<td>95</td>
<td>33</td>
<td>52.1</td>
</tr>
<tr>
<td>Under 1000</td>
<td>29</td>
<td>19</td>
<td>10</td>
<td>9</td>
<td>3.4</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>329</td>
<td>52</td>
<td>277</td>
<td>52</td>
<td>68.4</td>
</tr>
<tr>
<td>%</td>
<td>100</td>
<td>15.8%</td>
<td>84.2%</td>
<td>15.8%</td>
<td></td>
</tr>
</tbody>
</table>

### TABLE 221b

**Treatment of Placenta Previa**  
*(Macafee 1945)*

<table>
<thead>
<tr>
<th>Cases</th>
<th>Stillbirths and neonatal deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>1932-36</td>
<td>39</td>
</tr>
<tr>
<td>1937-44</td>
<td>41</td>
</tr>
</tbody>
</table>

23.6%
noted that, "Taking foetal and neonatal deaths together, asphyxia ranked as the most frequent of the common causes of death ....." Amongst both stillborn and live born groups antepartum haemorrhage takes precedence as a cause of foetal asphyxia". In Scotland in 1939, antepartum haemorrhage was the attributed cause of stillbirth in approximately thirteen per cent. of 3,832 cases, though there is some doubt as to reliability of the returns on which these figures are based, (Baird, 1945).

In her studies of prematurity, stillbirth and neonatal death which have been referred to, Drillien found that the primary cause of stillbirth in sixteen per cent. of 373 cases was antepartum haemorrhage. She emphasised that antepartum haemorrhage caused a statistically significant increase in the incidence of stillbirth even when allowance was made for the higher incidence of premature birth following antepartum haemorrhage compared to a control group, (Drillien, 1947). In a review of the causes of stillbirth in 281 cases occurring in the Johns Hopkins Hospital between 1937-1949 it was found that 43 or 15.3% were attributable to antepartum haemorrhage, with or without the presence of preeclamptic toxaemia. "It was the greatest single cause of foetal death in utero of known causes", (Nesbitt and Anderson, 1953).

The hypoxia caused by antepartum haemorrhage is frequently rapidly lethal and premature onset of labour following it is common. It is not surprising, therefore, that in as many as 40% /
40% or more of the autopsies on stillborn foetuses following antepartum haemorrhage the classical pathological evidence of anoxia is lacking. In the remaining cases, however, hypoxic lesions are almost universally present, (Macgregor, 1943; Potter, 1953).

In a study of the survival rate after haemorrhage from placenta previa between 1931-1954 in Chicago Lying-in-Hospital it was found that while 89.5% of mature infants survived, only 52.1% of viable premature infants survived, Table 111A (Davis, Potter and Bruce, 1943). Even with modern conservative treatment of haemorrhage from placenta previa the combined foetal and infant mortality is very high, Table 111B (Macafee, 1945).

Thus, there is evidence that antepartum haemorrhage, whether due to placenta previa or to other causes, is an extremely lethal condition, resulting in high stillbirth and neonatal mortality rates. The majority of deaths resulting are attributable to hypoxia, occurring very frequently in the premature infant. For the purposes of this investigation, therefore, antepartum haemorrhage in the last trimester of pregnancy is regarded as a probable cause of foetal anoxia.

The effects on the foetus of threatened abortions in early pregnancy are largely unknown. A higher incidence of foetal abnormality in cases of threatened abortion successfully treated has been reported, but it seems more probable that the foetal abnormality is primary, rather than secondary to /
to any effect of the abortion. Since the hypoxia caused by threatened abortion is not strictly paranatal it will not be considered as a cause of hypoxia for present purposes.

Post-maturity. Post-maturity is not specified as an abnormality of pregnancy in the majority of studies of stillbirths and neonatal deaths. Yet there is evidence that post-maturity does predispose to foetal hypoxia.

Experimental work on rabbits and ewes has shown that whereas placental efficiency increases in the first two-thirds of pregnancy, it declines in the last third. The decline is most marked shortly before term and is accentuated if pregnancy is abnormally long, (Barcroft, Kennedy and Mason, 1939; Barcroft, 1946; Barcroft and Young, 1945; Snyder, 1949). Recent work has demonstrated that there is a similar sequence of events in the human. Blood taken from the umbilical vein in the course of elective Caesarian section at various stages of pregnancy has shown that placental function is optimum at about thirty-three weeks, and then shows a slow decline until thirty-nine weeks gestation. Thereafter, the decline is much more rapid and if gestation is prolonged to forty-three weeks foetal oxygenation is adequate only for current needs. There is no reserve of placental function should labour be stressful, (Walker and Turnbull, 1953; Walker, 1953; Gruenwald, 1953). These findings may be correlated with the appearance and progression of degenerative changes in the placentas, (Brews,
Recent clinical studies have confirmed the earlier observations that there was an increase in the incidence of intrauterine death and of complications of labour when pregnancy had been unduly prolonged, (Ballantyne and Browne, 1922). A rise of foetal mortality from 4.7% for those delivered between 294 and 300 days gestation to 10.9% for those delivered after a gestation of 308 days or more was reported by Clayton, (1941). An almost two-fold increase of the incidence of foetal distress during labour in post-maturity as compared to mature foetuses was reported by McKiddie, (1949).

For present purposes gestation of more than 41 weeks is considered to be a possible cause of hypoxia.

Maternal causes of foetal hypoxia. It has been demonstrated experimentally that although severe maternal anaemia has no significant effect on foetal oxygenation, decreased oxygen saturation or slowed circulation of the maternal blood is reflected in the foetal blood oxygen levels, (Barcroft, 1949; Snyder, 1942). Chronic maternal anoxaemia is most likely to occur during pregnancy as a result of cardiac or respiratory disease. For example, pneumonia accompanied by cyanosis was found to result in premature delivery of 69% of women who suffered from the condition after the sixth month of gestation, (Finland and Dublin, 1939).

Unfortunately the authors do not state how many of the
high proportion of stillbirths and neonatal deaths could be attributed to hypoxia. In this work respiratory diseases accompanied by maternal cyanosis are classified as being possible causes of foetal hypoxia.

In congenitive cardiac failure, foetal oxygenation is likely to be impaired by slow circulation of the blood as well as by diminished oxygenation of the maternal haemoglobin. In most large hospital series, however, the stillbirth and neonatal mortality rates of the offspring of mothers with heart disease are not much higher than those found amongst children of normal mothers, (MacRae, 1953, 1954). This is largely due to the fact that those with severe cardiac decompensation frequently have their pregnancies terminated during the first trimester. The dangers of severe maternal and foetal anoxaemia occurring during prolonged or difficult labour are stressed by a number of authors, (Hamilton, SE. and Thompson, 1941; Bramwell, 1953; MacRae, 1953). In this investigation decompensated heart disease is considered to be a possible cause of foetal hypoxia.

Maternal syncope during pregnancy has not been much studied by recent authors though the early writers on cerebral palsy noted its occurrence in a proportion of their cases. Prolonged loss of consciousness associated with reduction of blood pressure and slowed circulation as in vasovagal attacks seems likely to be a cause of foetal hypoxia. In severe epileptic attacks with loss of consciousness and respiratory embarrassment, maternal and foetal anoxaemia seems to be a possible /
possible complication. Thus for the present work, recurrent or prolonged syncope is considered to be a cause of possible hypoxia.
Hypoxia during labour and delivery.

Numerous studies show that there is an increase in stillbirth and neonatal death rates when labour is complicated or delivery is other than spontaneous and by the vertex. For example, in Table 222 are shown the percentage mortalities by mode of delivery in two recent hospital studies.

It is more difficult to elucidate what proportion of the deaths associated with abnormalities of labour or delivery are hypoxic. This is because abnormalities of labour and delivery are often multiple and may have been preceded by disorders of pregnancy which may also have damaged the child. For example, in the series of Potter and Adair, two-thirds of breech deliveries had been preceded by complications of pregnancy. In addition, some types of abnormal labour and delivery may have a number of different effects. For example, traumatic brain damage is relatively frequent in breech delivery, but breech delivery may also be attended by cord prolapse, or inhalation of liquor causing hypoxia. Moreover, a high proportion of breech deliveries are of premature infants, a relatively high proportion of whom may suffer from congenital malformations.

Similar difficulties are encountered when the cause of the excess of stillbirths and neonatal deaths following prolonged labour are studied. As pointed out by Baird, clinical findings would suggest that prolonged labour would result in trauma, but on pathological examination indications of hypoxic death /
Foetal mortality by type of delivery.

**TABLE 222**

<table>
<thead>
<tr>
<th>Mode of delivery</th>
<th>Drillien, 1947 % of deaths</th>
<th>Potter and Adair, 1939 % of deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Premature</td>
<td>Mature</td>
</tr>
<tr>
<td>Spontaneous vertex</td>
<td>33.8</td>
<td>1.6</td>
</tr>
<tr>
<td>Low forceps</td>
<td>(</td>
<td></td>
</tr>
<tr>
<td>Mid cavity forceps</td>
<td>(</td>
<td></td>
</tr>
<tr>
<td>High forceps</td>
<td>(</td>
<td></td>
</tr>
<tr>
<td>Version and extraction</td>
<td>25.0</td>
<td>4.4</td>
</tr>
<tr>
<td>Breech</td>
<td>52.8</td>
<td>15.1</td>
</tr>
<tr>
<td>Caesarean section</td>
<td>36.7</td>
<td>6.0</td>
</tr>
</tbody>
</table>

not including 20 dead born which were macerated

**TABLE 223**

Table showing the relationships between length of labour, length of gestation and foetal mortality.
(Baird, 1955)

<table>
<thead>
<tr>
<th>Length of Labour</th>
<th>38 wks.</th>
<th>38-39 wks.</th>
<th>40 and 41 wks.</th>
<th>42 wks. and over</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 24 hrs.</td>
<td>53</td>
<td>24.5</td>
<td>47</td>
<td>17.0</td>
</tr>
<tr>
<td>24 - 48 hrs.</td>
<td>20</td>
<td>25.0</td>
<td>47</td>
<td>10.6</td>
</tr>
<tr>
<td>48 hrs. +</td>
<td>8</td>
<td>12.5</td>
<td>20</td>
<td>10.0</td>
</tr>
</tbody>
</table>

|               | 23.4    | 13.1       | 7.09           | 13.7             |
Similar difficulties are encountered when the cause of the excess of stillbirths and neonatal deaths following prolonged labour are studied. As pointed out by Baird, clinical findings would suggest that prolonged labour would result in trauma, but on pathological examination indications of hypoxic death are found. Whether trauma has actually occurred and been complicated by hypoxia, or whether the child was already hypoxic before labour commenced is often unknown. Baird places stillbirths and neonatal deaths following prolonged labour without other obvious clinical abnormalities of parturition likely to damage the child in a category of "stress asphyxia". He groups them together with deaths due to trauma, however, for they appear to be associated with abnormalities of labour, and there seems to be considerable evidence that trauma is of major contributory importance, (Baird, 1955) Table 223.

Some evidence that hypoxia may precede rather than be initiated during prolonged labour is forthcoming from recent studies of cord blood oxygen saturations, (McKiddie, 1949; Walker, 1953). Baird's mortality figures by duration of labour and length of gestation also lend some support for this view. In this work, therefore, prolonged labour will be classified rather arbitrarily, as by Baird, as a possible cause of traumatic damage rather than a cause of hypoxia. (Table 223).

Similarly, though it may be demonstrated that there is an /
an excess of stillbirths and neonatal deaths apparently due to hypoxia after instrumental or breech deliveries compared to spontaneous vertex deliveries, abnormal forms of delivery more often result in traumatic death than hypoxia, (Drillien, 1947). They will, therefore, be classified as possible and probable causes of trauma in this work, rather than as causes of hypoxia. Other possible and probable causes of hypoxia during labour and delivery will now be considered.

Prolapse and Torsion of the Cord. 7.2 stillbirths per 1000 live, and stillbirths were attributed to torsion or prolapse of the cord by the Registrar General for Scotland (1950). Approximately 10 per cent. of all stillbirths appeared to be due to cord complications. In the two hospital series previously quoted, Table 223, cord complications were the cause of death in 32% and 9.5% stillbirths respectively. In a study of the causes of stillbirth and neonatal death in the Chicago Lying-in-Hospital from 1946-49, cord complications were found to have been present in 30%, (Davis, Potter and Bruce, 1953). These authors stress the difficulty of attempting to prove the high mortality from cord complications and comment on the increased incidence of cord prolapse in mal-presentations, premature delivery and foetal malformations. Though any obstruction to the circulation in the cord is liable to result in severe and sudden foetal hypoxia, the effect of the cord around the neck, will obviously be to increase the degree of cerebral venous engorgement to a greater degree than if hypoxia alone had been present.
To some extent, therefore, birth with the cord around the neck might be regarded as being traumatic as well as anoxic in its effects upon the foetus. From the point of view of the survey, cord prolapse and/or torsion are regarded as probable causes of foetal hypoxia.

Maternal anaesthesia and analgesia as causes of foetal hypoxia. The significance of analgesia and anaesthesia given during labour as a cause of foetal hypoxia and mortality is very controversial. Its assessment is rendered extremely difficult by a number of considerations. The action of anaesthetic and analgesic agents in causing foetal hypoxia is indirect. Foetal hypoxia may be caused by maternal anoxaemia if the anaesthetic is given by an unskilled person, or in excessive quantity so that respiratory depression occurs. Secondly, the foetal respiratory centre may be poisoned directly by the anaesthetic agent especially if it is given for a prolonged period or in excessive amount. These effects may occur independently or in association. The toxic effect may predominate and cause apnoea with good oxygenation of the foetus or the anoxaemic effect may predominate in which case low levels of anaesthetic are likely to be found in the foetal blood, (Smith, 1943).

The fact that the conditions for which strong analgesics and general anaesthesia are required are those which themselves are liable to result in foetal damage and anoxia makes assessment still more difficult. It is easy to show that neonatal apnoea /
apnoea occurs more frequently following the use of anaesthetics during labour or delivery than when they have not been employed. But there are few investigations in which matched series of similar cases are compared. However, there is a certain amount of information derived from clinical observation and animal experiments which allows tentative conclusions to be drawn.

**General anaesthetics.** There have been a number of surveys into the foetal and neonatal mortality and incidence of apnoea in cases in which local, regional, caudal and spinal anaesthetics have been given in conditions which would normally be treated under general anaesthesia, (Torrie, 1945; Tucker and Benaron, 1934; Hingson et al, 1948; Greenhill, 1953; Dieckmann, 1945). The majority of these reports indicate a slightly lower stillbirth and neonatal mortality than comparable series with general anaesthesia and a considerably smaller incidence of neonatal apnoea. (Table 224). (Snyder, 1949; Donnelly, 1944; Lull and Hingson, 1944). "In general, they diminish the direct anaesthesia hazard to the foetus, but at least in their use to date, seem to transfer the risk to the mother". (Smith, 1948).
Thus it seems to be established that general anaesthetics do contribute to foetal and neonatal mortality, and more especially to the incidence of hypoxic death. This is in accordance with the findings of a very large series of patients investigated by Cole, Kinball and Daniels, (1939).

In rabbits it is possible to expose the uterus and observe the foetuses directly without interrupting the umbilical circulation, (Rosenfeld and Snyder, 1939). These authors laid stress on the suppression of the normal intrauterine respiratory movements which they observed took place when anaesthetics or analgesics were administered. Pentobarbital sodium, ether, nitrous oxide and cyclopropane were administered to animals in these circumstances. Only in the case of cyclopropane did anaesthesia of the mother result before respiratory depression of the foetuses occurred. As a result of their studies they concluded that all volatile anaesthetics and barbiturates were liable to result in respiratory depression of the foetus, but that cyclopropane appeared to be the least liable to do so in anaesthetic concentrations.

A great many reports have appeared comparing the effects of one anaesthetic agent with another and different combinations of premedication with anaesthetic, (Smith, 1939; Smith and Barker, 1942; Cole and Kimball, 1934; Cole, Kimball and Daniels, 1939; /
TABLE 224

Rate of foetal loss, (Neonatal deaths and stillbirths) for deliveries under caudal compared to other anaesthesias. Hingson et al, 1946

Rate of foetal loss per 1000 live births for the following study.

<table>
<thead>
<tr>
<th>City</th>
<th>No. of births</th>
<th>Caudal</th>
<th>Spinal</th>
<th>General</th>
<th>None</th>
</tr>
</thead>
<tbody>
<tr>
<td>New York</td>
<td>1685</td>
<td>16.2</td>
<td>28.3</td>
<td>162.2</td>
<td></td>
</tr>
<tr>
<td>Philadelphia</td>
<td>3580</td>
<td>10.6</td>
<td>-</td>
<td>45.6</td>
<td></td>
</tr>
<tr>
<td>Memphis</td>
<td>2626</td>
<td>36.6</td>
<td>74.2</td>
<td>73.2</td>
<td>144.0</td>
</tr>
</tbody>
</table>
1939; Lund, 1942). The consensus of opinion appears to be that the method of delivery and the nature of the previous labour, together with the premedication, are more important determinants of the occurrence of neonatal apnoea than is the choice of the anaesthetic agent. In many cases it is possible to deliver the infant before the full concentration of the anaesthetic has crossed the placental barrier to reach the foetus. Prolonged administration of large amounts of anaesthetic are especially dangerous for the foetus, (Lund, 1942; Smith, 1948). To some extent any form of general anaesthesia is a potential contributory factor to foetal hypoxia, but its importance is clearly likely to be much greater in cases in which the foetus is already hypoxic or has suffered trauma before the anaesthetic takes effect. This appears to have been the situation in a large proportion of the children in the present series.

**Analgesic drugs used during labour.** Numerous observations have been made on the effect of various analgesic drugs and their combinations on the pain experienced by the mother during labour and delivery and their effect on the foetus. With the exception of three agents it has been demonstrated that all result in a degree of respiratory depression in a proportion of the foetuses in analgesic doses, (Smith, 1948; Irving et al, 1934; Mengert, 1942).
TABLE 225

Comparative effects of various analgesics on respiration at birth.

Irving et al, 1934

<table>
<thead>
<tr>
<th>Pre-anaesthetic medication</th>
<th>Delivery anaesthesia</th>
<th>Infants breathing spontaneously %</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>none</td>
<td>98</td>
</tr>
<tr>
<td>None</td>
<td>none</td>
<td>80</td>
</tr>
<tr>
<td>Pentobarbital and rectal ether.</td>
<td></td>
<td>65</td>
</tr>
<tr>
<td>Pentobarbital and scopolamine.</td>
<td></td>
<td>63</td>
</tr>
<tr>
<td>Sodium amytal and scopolamine.</td>
<td></td>
<td>61</td>
</tr>
<tr>
<td>Sodium amytal and rectal ether.</td>
<td>Nitrous oxide, oxygen and/or ether.</td>
<td>59</td>
</tr>
<tr>
<td>Pantopon (opiate) and rectal ether.</td>
<td></td>
<td>53</td>
</tr>
<tr>
<td>Pernocton (barbiturate).</td>
<td></td>
<td>53</td>
</tr>
<tr>
<td>Pentobarbital and paraldehyde.</td>
<td></td>
<td>50</td>
</tr>
<tr>
<td>Pantopon and scopolamine.</td>
<td></td>
<td>33</td>
</tr>
<tr>
<td></td>
<td>No. of cases</td>
<td>10-30 secs.</td>
</tr>
<tr>
<td>-----------------------</td>
<td>--------------</td>
<td>-------------</td>
</tr>
<tr>
<td>None</td>
<td>666</td>
<td>96.7</td>
</tr>
<tr>
<td>Sodium pentobarbital.</td>
<td>127</td>
<td>90.4</td>
</tr>
<tr>
<td>Morphine</td>
<td>172</td>
<td>84.9</td>
</tr>
</tbody>
</table>
The action of morphine in causing foetal hypoxia has been found to be indirect in animal experiments performed by Snyder, (1949, 1953). He attributes the hypoxia to the effect of the drug in inhibiting uterine contractions, and prolonging labour with consequent diminished oxygenation of the foetal blood rather than to a direct toxic effect on the foetal respiratory centre. He brings forward the great tolerance of the foetus to morphia during pregnancy as favouring this theory. The particular dangers of morphia as a cause of respiratory depression have been stressed by Shute and Davis, (1933).

Though the dangers from neonatal apnoea appear to be somewhat less with heroin and pethidine than with morphia itself, they are still considerable. (Lund and Harris, 1943; Smith, 1948).

The barbiturates, essentially hypnotic drugs, have been found to be dangerous when given in analgesic dosage by a number of workers, and they have been shown to act chiefly as toxic agents to the foetal respiratory centre. (Moore, 1933; Dreisbach and Snyder, 1943). The combination of barbiturates with scopolamine appeared to be associated with little extra foetal risk in the survey of Irving et al, (1934). Intravenous anaesthesia with sodium pentothal was found to be excessively /
excessively dangerous for mother and child in a survey by Hellman, Shettles, Manahan and Eastman, (1944). A foetal and neonatal mortality of 4.9% in 1415 cases was obtained.

The three agents about which there appears to be conflicting evidence of their effects on the foetus is analgesic doses are paraldehyde, chloroform and nitrous oxide. The tolerance of the foetus during pregnancy and for the first weeks of life to chloroform is markedly increased in dogs and the toxic effects found in adults do not occur. In analgesic doses to the mother, chloroform does not appear to have been shown to be a cause of foetal apnoea. Paraldehyde was found in one investigation to have no anoxic effect on the foetus and others have reported that the blood levels in the foetus are much below that of the mother's when it is given in analgesic doses, (Colvin and Bartholomew, 1935; Gardner, Levine and Bodansky, 1940). Its main disadvantages appear to be the difficulties of its administration, its unpleasantness for the patient and its relatively poor analgesic effect. Nitrous oxide has not been shown to be a cause of foetal hypoxia. When given in combination with less than 15% of oxygen, however, a number of reports indicate that foetal hypoxia may occur, (Eastman, 1936).
In this series the classification of which drugs may be regarded as harmful has been rather arbitrary. Morphia, large doses of heroin or barbiturates have been regarded as possible causes of foetal hypoxia if given repeatedly during labour or if administered in large doses within four hours of delivery. General anaesthesia or analgesia with ether, chloroform, nitrous oxide or cyclopropane, spinal and local anaesthesia have not been considered as causes of foetal hypoxia. Pentothal anaesthesia has been classified as a possible cause of foetal hypoxia.

Neonatal apnoea as a cause of neonatal hypoxia. There appears to be rather less controversy about the causes of failure to breathe after birth than there is about the causes of spontaneous respiration commencing, (Barcroft, 1946; Smith, 1948; Snyder, 1949). Whereas the mechanisms involved in causing the infant to take its first breath are still only partly understood, numerous studies have been published which deal with the causes and effects of neonatal apnoea, (Lund, 1941; Schreiber, 1943; Snyder, 1939, 1950; Courville, 1945, 1952; Cole et al, 1939, 1943). It seems to be established that immaturity of the respiratory centre is never the sole cause of neonatal apnoea, for it is well developed before the lungs have matured sufficiently to /
to be functional, (Gairdner, 1954). On the other hand, a functional respiratory centre is necessary before respiration can commence. The most frequent causes of it not functioning appear to be intracranial trauma resulting in structural damage, pre-existing hypoxia damage, or poisoning by analgesic or anaesthetic agents.

Even when the centre is functional, however, the infant may still not begin to breathe. The lungs may be incapable of aeration, because of extreme structural immaturity, because they are full of amniotic fluid or oedema, or because of congenital malformation. They may be capable of aeration, but the foetus may be unable to expand them because it is too weak to do so on account of prematurity or other abnormality.

The clinical applications of these rather theoretical considerations are of interest for a number of studies have shown that the common causes of neonatal apnoea are as follows —

1. Prematurity
2. Hypoxia of pregnancy or delivery resulting in —
   (a) poisoning of the respiratory centre or structural damage to it.
   (b) the air passages being obstructed as a result of the infant’s premature efforts to breathe before delivery.
   (c) Intracranial trauma resulting from labour and abnormal forms of delivery.
   (d) Poisoning by anaesthetic agents.

Other /
Other aetiological factors of some importance are foetal malformations, especially congenital heart disease, direct injury to the chest wall or to the spinal cord and failure to breathe of unknown aetiology following Caesarean section, (Potter, 1951; Smith, 1948).

Since neonatal apnoea can be caused by a variety of agents it cannot be regarded as a manifestation of any particular one of them in the living subject. In particular, so far as the present investigation is concerned it cannot be regarded merely as an indication of pre-existing hypoxia or trauma.

Neonatal apnoea has a double significance. It is not only the end result of many different forms of foetal injury, but it is also an important cause of further hypoxia damage in the newborn infant. It is necessary, therefore, to consider in what circumstances it is most likely to cause damage to the child, for this is likely to vary considerably in different circumstances in different infants.

In general it has been shown by animal experiments that the tolerance to hypoxia of the newborn infant is much greater than that of the adult and that the premature infant has an increased intolerance compared to the mature, (Barcroft, 1946; Snyder, 1949; Glass, Snyder and Webster, 1944).
This is in accordance with what is found in the human being. Much greater tolerance to hypoxia has been demonstrated to be present in the premature as compared to the mature infant, (Himwich, Alexander and Fazekas, 1941). In clinical conditions, however, the most important factor in determining whether an infant survives anoxia or suffers irreparable damage is the presence or absence of previous hypoxic episodes. Clearly the child which has been hypoxic throughout late pregnancy and labour is likely to sustain irreparable damage as a result of a relatively short period of apnoea. On the other hand, an infant whose apnoea is due to the respiratory centre being depressed by anaesthetic and who has a normal blood oxygen saturation at the time of delivery is likely to show a high tolerance to the hypoxia of apnoea, (Smith, 1948). In a great many infants in the present series there had been previous hypoxic episodes during pregnancy and delivery. Their tolerance is, therefore, likely to have been low.

In mature infants who have not previously been anoxic the incidence of cerebral damage is thought to be high after about ten minutes of apnoea and to rise progressively rapidly after this time. Between twenty and forty minutes irreversible changes are observed in the cortex which are usually lethal, (Windle, /
For the purposes of this investigation apnoea of any duration is regarded as being a possible hypoxic insult and apnoea of a longer duration than ten minutes is regarded as a probable cause of hypoxic damage.

**Hypoxia due to other neonatal complications.**

Many of the causes of apnoea in the period immediately following birth are also causes of hypoxia after that time. The lungs may be persistently atelectatic because of immaturity, bronchial obstruction or failure of the foetus to expand them. Pulmonary respiratory exchange may be handicapped by hyaline membrane formation especially in the premature infant, by pulmonary oedema, intrauterine pneumonia, or to inadequate ventilation due to feebleness of respiratory efforts, (Meyer, 1953; McGregor, 1953). Some indication of the importance of persistent atelectasis as a cause of anoxic death in the neonatal period is given by the statistics of Gibberd, (1953).
To these causes of pulmonary dysfunction, capable of producing hypoxic cerebral damage must be added the dangers of aspiration of food contents, especially in the premature, and respiratory infections, especially pneumonias which result from this and other causes.

To assess the degree of hypoxia which these disorders are likely to cause is extremely difficult; and to determine the amount of cerebral damage is impossible. For the purposes of the present investigation respiratory disorders accompanied by cyanosis lasting for longer than one hour have been accounted possible causes of hypoxic brain damage in the newborn infant. (somewhat arbitrarily).

Conditions resulting in cyanosis which persists for longer than six hours have been classified as probable causes.

Trauma/
Chapter 2e.

Trauma in Labour and Delivery.

It is now necessary to define the conditions in labour and delivery which are considered to be traumatic in the present investigation.

Thirty years ago the criteria of what constituted traumatic labour and delivery seemed well established. Intracranial haemorrhage resulting from instrumental or breech delivery was thought to be the commonest form of lethal cerebral damage in the infant (Holland, 1922, 1926; Ehrenfest, 1931). The importance of prematurity
prematurity as a contributory cause of the mortality resulting from stillbirth and neonatal death was attributed largely to its effect in predisposing the foetus to trauma.

In recent years the tendency has been to discount the importance of trauma in birth injury and to emphasise the dangers of hypoxia. Neuropathological conditions such as lobar sclerosis, porencephaly and partial atrophies which used to be attributed to the effects of birth trauma, have been produced in experimental foetal animals by rendering them hypoxic, (Windle, 1950; Courville, 1953).

Whereas intracranial haemorrhage used to be regarded as an almost certain indication of traumatic birth injury, it is now thought that hypoxia is a far more important aetiological factor than trauma in subarachnoid, intraventricular and intracerebral haemorrhage, (Craig, 1938; Drillien, 1947; Balf, 1953; Macgregor, 1948). On the other hand, there is general agreement that subdural haemorrhage is usually traumatic in origin, (Craig, 1938; Balf, 1948; Macgregor, 1948; Potter, 1953; Ingraham and Matson, 1944, 1949), whilst in his survey of perinatal deaths due to intracranial haemorrhage, Craig found that only nine of his forty-two
forty-two cases of subdural haemorrhage with dural tears were premature. Whereas 56% of the mothers with infants who died from subarachnoid, intraventricular or intracerebral haemorrhage had complications of pregnancy liable to produce hypoxia, only eleven of the sixty-two pregnancies, (approximately 15%) in the subdural group were similarly complicated. On the other hand, 65% of those with intracranial, subarachnoid or intraventricular haemorrhages were delivered spontaneously by the vertex, and only 27% of the 64 cases with fatal subdural haemorrhages were born by spontaneous vertex delivery. In eight, delivery was by the breech, thirty-four were forceps deliveries and in fourteen of the latter, manual or instrumental rotation of the head was performed before extraction. The author concluded, "In general, bleeding into the subarachnoid space and into the ventricular system was associated with illness of the mother during pregnancy, and haemorrhage into the brain substance and into the subdural space with instrumental delivery, and with delivery at or after term".

The forms of abnormal delivery which were particularly liable to result in subdural haemorrhage were similar to those described by Beneke, 1910, 1920; Holland, 1922, 1926; Greenwood, 1924; Ehrenfest, 1931). Subdural haemorrhage was most often the result of tears of the dural ligaments which /
which involved either the tributaries of the sagittal/sinus or the great cerebral vein itself. Tears were especially liable to occur in full-time infants when excessive head moulding, rapid distortion of the skull or abnormal pressure on it was such that undue or oblique stress was placed on the tentorial cerebelli or the falx. Though hypoxia may increase the risk of rupture of venous sinuses and their tributaries through causing them to be distended and congested, their actual tearing appears to be the result of trauma in the majority of cases, (Ingraham and Matson, 1949; Capon, 1922).

Of 73 cases with tentorial tears in Holland's series, only three were delivered spontaneously by the vertex, twenty-four were delivered by forceps and thirty-two by the breech, seventeen of the latter having been subjected to version prior to extraction.

In a contemporary survey to that of Holland into the causes of intracranial haemorrhages in the newborn, Capon also emphasised the association of traumatic birth with subdural haemorrhage. He considered that the following forms of abnormal labour and delivery in ascending order of their danger were especially liable to result in subdural haemorrhage -

Prolonged /
Prolonged labour due to disproportion or malpresentation.
Precipitate delivery.
Forceps delivery.
Breech extraction.
Version and extraction.

Since there appears to be quite adequate evidence to suggest that subdural haemorrhage is predominantly the result of birth trauma, and that the forms of labour and delivery listed above are those which are most often associated with subdural haemorrhage, they will be regarded as causes of possible or probable trauma in patients in the present series.

Recent Studies.

A number of recent studies have attempted to assess the relative dangers of traumatic injury to the foetus in various types of complicated labour and abnormal modes of delivery. In two reports of the causes of stillbirth and neonatal death in Chicago Lying-in Hospital, cases were classified according to the mode of delivery, birth weight and pathological cause of death, Potter and Adair, 1939, 1943). Table 227. By excluding cases in which death did not appear to be the result of abnormalities of labour or delivery, the authors were able to make some assessment of the relative dangers of the
the various modes of delivery in causing traumatic damage to the child. Any form of instrumental delivery was associated with an increase of the stillbirth and neonatal death rates. Caesarean section was the form of delivery least often associated with traumatic damage, whilst mid or high forceps delivery, version and extraction and breech delivery were the most dangerous. Unfortunately their figures do not distinguish between the effects on the child of assisted delivery and the effects of the abnormalities of labour which made the assisted deliveries necessary. Rather similar conclusions to those of Potter and Adair were reached by Drillien (1947), and d'esopo and Marchetti (1942). The latter authors attempted to take account of "predisposing factors" in the cases of birth injury which occurred amongst 25823 births in hospital. Tables 227 and 228. Cephalopelvic disproportion, uterine inertia, malpresentations and rapid labour were the commonest abnormalities of labour which appeared to cause or predispose to foetal trauma.

A controlled investigation of the effects of prolonged labour, forceps delivery, precipitate delivery and natural labour and delivery on the foetus in 28,698 hospital vertex deliveries was carried out by Tucker and Benaron (1953). They thought /
thought that the incidence of neonatal apnoea and perinatal death was proportional to the degree of trauma which the different forms of labour and delivery caused. 1.07% of infants died perinatally when delivered spontaneously by the vertex, whereas comparable figures were 1.55% in precipitate births, 1.54% in low forceps deliveries, 7.2% in mid cavity forceps deliveries and 29.41% in high forceps deliveries. Only very occasional babies born after natural labour showed evidence of traumatic intracranial lesions, though these were relatively common in those delivered with the aid of instruments.

As a result of a study of 7,599 births in the Simpson Memorial Maternity Pavilion it was concluded that instrumental delivery was as safe as spontaneous delivery for the premature infant, but that it carried an increased risk in the mature. The greatest perinatal mortality by mode of delivery was found in breech extraction in both premature and mature infants. A high proportion of these deaths were ascribed to trauma, (Drillien, 1947).
TABLE 227

Percentage mortality by mode of delivery and cause of death in 25823 deliveries.
d'Esopo and Marchetti, 1942

<table>
<thead>
<tr>
<th>Method of delivery</th>
<th>Total cases</th>
<th>Birth injury</th>
<th>Asphyxia</th>
<th>Congenital pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spont. vertex</td>
<td>19916</td>
<td>0.01</td>
<td>0.04</td>
<td>0.01</td>
</tr>
<tr>
<td>Low forceps</td>
<td>2672</td>
<td>0.04</td>
<td>0.01</td>
<td>0.03</td>
</tr>
<tr>
<td>Mid forceps</td>
<td>1011</td>
<td>2.9</td>
<td>0.01</td>
<td>1.3</td>
</tr>
<tr>
<td>Breech delivery</td>
<td>1084</td>
<td>1.7</td>
<td>2.5</td>
<td>0.07</td>
</tr>
<tr>
<td>Version and extraction</td>
<td>182</td>
<td>6.6</td>
<td>9.3</td>
<td>0.06</td>
</tr>
<tr>
<td>Craniotomy</td>
<td>38</td>
<td>18.4</td>
<td>7.9</td>
<td>28.95</td>
</tr>
<tr>
<td>Caesarean section</td>
<td>782</td>
<td>0.04*</td>
<td>2.9</td>
<td>0.01</td>
</tr>
<tr>
<td>High forceps</td>
<td>29</td>
<td>20.7</td>
<td>10.4</td>
<td>0.0</td>
</tr>
</tbody>
</table>

* due to damage sustained during labour, prior to section

The authors proceeded to analyse the predisposing and precipitating aetiological factors in the major pathological causes of death, Table 228
<table>
<thead>
<tr>
<th>Factors</th>
<th>No. of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mature</td>
</tr>
<tr>
<td>Cephalopelvic disproportion</td>
<td>22</td>
</tr>
<tr>
<td>Uterine Inertia</td>
<td>16</td>
</tr>
<tr>
<td>(some due to narcosis)</td>
<td></td>
</tr>
<tr>
<td>Not known</td>
<td>11</td>
</tr>
<tr>
<td>Malpresentation</td>
<td>9</td>
</tr>
<tr>
<td>Breech</td>
<td>4</td>
</tr>
<tr>
<td>Brow</td>
<td>1</td>
</tr>
<tr>
<td>Forceps trauma or bad mechanics</td>
<td>8</td>
</tr>
<tr>
<td>Rapid hard labour</td>
<td>6</td>
</tr>
<tr>
<td>Rapid labour after medical induction</td>
<td>3</td>
</tr>
<tr>
<td>Trauma from version and extraction (no other apparent cause)</td>
<td>1</td>
</tr>
<tr>
<td>Asphyxia with rapid labour</td>
<td>3</td>
</tr>
<tr>
<td>Prolonged second stage</td>
<td>3</td>
</tr>
<tr>
<td>Other</td>
<td>4</td>
</tr>
</tbody>
</table>
A combined mortality due to stillbirth and neonatal death of approximately 10% in 1977 mature cases of breech delivery was reported by Goethals (1939). Unfortunately the pathological diagnosis of the cause of death was not stated but the author considered that the majority were due to asphyxia and intracranial trauma. In cases presenting by the breech in which caesarean section was performed the mortality from all causes was very much lower than when delivery had been per vaginam.

**TABLE 229**

The proportion of stillbirths and neonatal deaths following breech delivery due to various causes.

*d'Esopo and Marchetti, 1942*

<table>
<thead>
<tr>
<th>Cause of death</th>
<th>% of 152 stillbirths and neonatal deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prematurity</td>
<td>23</td>
</tr>
<tr>
<td>Asphyxia</td>
<td>18.4</td>
</tr>
<tr>
<td>Congenital anomalies</td>
<td>17.1</td>
</tr>
<tr>
<td>Maceration</td>
<td>17.1</td>
</tr>
<tr>
<td>Birth injury</td>
<td>11.8</td>
</tr>
<tr>
<td>Congenital pneumonia</td>
<td>5.9</td>
</tr>
<tr>
<td>Congenital syphilis</td>
<td>1.3</td>
</tr>
<tr>
<td>Erythroblastosis</td>
<td>1.3</td>
</tr>
<tr>
<td>Other</td>
<td>4.1</td>
</tr>
<tr>
<td>All causes</td>
<td>100.0</td>
</tr>
</tbody>
</table>

It is apparent from these studies, that breech birth is commonly associated with a high rate of stillbirth.
stillbirth and neonatal death and that the commonest pathological causes of death attributable to the form of delivery are anoxia and intracranial trauma. The latter probably causes the deaths of between 2 and 5% of most large series of breech deliveries. Delivery by the breech is considered to be a probable cause of trauma in the present series.

**Version and extraction.** Without exception the general surveys of the causes of stillbirth and neonatal deaths report a high mortality in infants following version and extraction. A total mortality of 29.3% was reported in 246 cases studied by Potter and Adair (1943). The mortality possibly related to delivery was 22.0%. 48.3% of the stillbirths and neonatal deaths resulted from trauma and 37.9% from anoxia. In a series of 182 cases of version and extraction studied, a combined mortality of 23.1% was found by d'Esopo and Marchetti (1942). Death from trauma occurred in 6.6% and death from asphyxia in 7.9% of all cases delivered by version and extraction. In a series of 186 cases delivered by internal version and extraction in hospital a mortality by stillbirth and neonatal death of 24.7% was reported by Rosenholm (1954). A series of 1146 cases delivered by version and extraction was reported by Erving and Kenwick (1954). These authors found a relatively low total foetal mortality.
mortality of 5.5% resulting from stillbirth and neonatal death. In 16 of 39 cases of stillbirth and neonatal death attributable to delivery, intracranial haemorrhage was the cause.

Version and extraction is a method of delivery which has been used by most obstetricians in recent years only when faced with very dangerous abnormalities of labour.

Clearly the risks to the foetus in version and extraction are those of breech delivery with the additional effects of version and comparatively rapid delivery and the probability of rapid changes in the direction and severity of the forces acting upon the foetal skull. The ratios of deaths due to trauma and deaths due to anoxia were almost identical for infants delivered by the breech and infants delivered by version and extraction in the series of d'Esopo and Marchetti (1942). Version and extraction is considered a probable cause of trauma in the present series.

**Forceps delivery.** The general studies quoted above show a higher percentage of foetal mortality in forceps deliveries than in spontaneous vertex deliveries, and also that the danger to the foetus is increased in high forceps deliveries as compared to mid cavity or low forceps deliveries. Thus in the /
the series of Potter and Adair (Table 219), low forceps delivery was associated with a comparable mortality to that found in spontaneous vertex delivery and this was also the finding of Harer (1932). On the other hand, both these series reported that mid cavity or high forceps delivery resulted in a stillbirth and neonatal mortality four or five times as great. Similar differences were found by Drillien (1947).

Whereas only 1% of the stillbirths and neonatal deaths associated with spontaneous vertex delivery were traumatic, 34% of low forceps deliveries, and 66% of mid and high forceps deliveries showed evidence of trauma at autopsy in the series of Potter and Adair (1943). In a series of hospital and domiciliary births it was reported that traumatic intracranial haemorrhage accounted for 22.4% of stillbirths and neonatal deaths following low forceps delivery, and 54.7% of those following mid cavity or high forceps as compared to 13.9% after spontaneous vertex delivery, (Bundesen et al, 1951) Bundesen, H.N., Potter, E.L., Fishbein, W.L., Bauer, F.C., and Plotke, F. "Progress in the Prevention of Needless Deaths". Chicago Health Dept. Annual Report, 1951). The pathological examination of a series of 65 cases of stillbirth and neonatal death following mid cavity forceps delivery resulted in 46.1% being classified as being due to trauma by d'Esopo /
d'Esopo and Marchetti (1942). When cases in which the pathological causes of death which could not be attributable to the method of delivery, (including congenital malformation and extreme prematurity) the percentage of cases dying as a result of birth trauma becomes 55. In their analysis of the causes of the high incidence of foetal trauma in forceps delivery the authors emphasise the importance of extraction in the presence of dispropoition and unrecognised abnormalities of pelvic configuration. In a proportion of cases forceps were applied too late to vulnerable anoxic infants and traction was too rapid. Infants were killed by forceps trauma in an attempt to save them from the perils of hypoxia.

It is a matter of considerable difficulty to distinguish traumatic effects of abnormal labour from the traumatic effects of instrumental delivery. Some indications of the dangers of mid cavity and high forceps delivery can be obtained, however, when roughly comparable series of cases delivered by mid-cavity and high forceps and by caesarean section are compared. It is found consistently that the mature infants born by caesarean section after abnormal forms of labour show a lower mortality than do those of comparable birth weight born by instruments and that deaths from trauma are significantly less frequent, (Drillien, 1947; Potter and Adair, 1943;
of Jeffcoate, approximately 17% of cases delivered instrumentally after prolonged labour died, whereas the combined foetal and neonatal mortality in cases delivered by caesarean section after similar labours was approximately 5%.

Thus, there is good evidence that high or mid cavity forceps delivery increases the risk to the child, and that the majority of deaths attributable to this method of delivery are traumatic. On the other hand, it has been demonstrated by a number of authors that methods of delivery which are probably harmless to the foetus in good condition, may have the most serious results in infants who are anoxic or have already suffered from birth trauma. Low forceps delivery may probably be considered a prophylactic measure in the healthy full-time foetus, preventing intracranial trauma as a result of obstruction at the perineum, (Aldridge and Meredith, 1941; Potter and Adair, 1939, 1943; Harer, 1932). In the premature foetus, however, or the mature which has already suffered from the effects of prolonged labour or is anoxic, low forceps delivery results in a considerably raised mortality compared to that found after spontaneous vertex delivery (Clifford, 1934; Dana, 1946; Peckham, 1932, 1938). Even in the /
the healthy, mature infant the trauma of mid cavity or high forceps delivery appears to be a factor of importance in causing stillbirth and neonatal death, as has recently been stressed by Douglas and Kaltreider (1953). For the purposes of the present survey, therefore, low and mid cavity forceps are considered to be possible causes of foetal trauma and high forceps delivery a probable cause.
Abnormalities of labour.

There is much work reported in which the effects of the duration of labour on stillbirths and neonatal death rates have been studied. Both very rapid labour and very prolonged labour have been shown to be associated with an increased foetal and neonatal mortality, (Peckham, 1932; Bourne and Bell, 1933; Douglas and Stander, 1943; Potter and Adair, 1939, 1943; Drillien, 1947; Jeffcoate, 1948; Baird, 1955; Tucker and Benaron, 1953).

Prolonged labour. Yet there are many abnormal clinical states which result in prolonged labour, for example, labour may be unduly long because there is failure of the uterine powers, because there is mechanical obstruction to the passage of the foetus through the pelvis, or a combination of these factors may be present. The further classifications of prolonged labour which have been attempted from the obstetric point of view are not of great help to the paediatrician whose interest is primarily on the effects of the abnormalities of labour on the child. Thus the distinction between hypotonic and hypertonic states of the uterine muscle and the different types of dystonia suggested by Jeffcoate are of limited value for the effects on the child of the disorders he includes in each category.
category are largely unknown, (Jeffcoate, 1948, 1949).

The few studies of the effect of uterine inertia on the child which have been made indicate a rather high combined foetal and infant mortality. Thus, in 49 cases the mortality rate was 42% in the series of Bourne and Bell (1933). In a series of 631 cases in which pelvic contraction was absent and in which labour lasted over 48 hours a combined foetal and neonatal mortality of 21.8% was reported by MacRae (1939). Sixteen of 91 stillbirths whose deaths appeared to be due to trauma studied by d'Esopo and Marchetti (1942) were attributed to the effects of primary or secondary uterine inertia.

A little more evidence about the dangers to the child of delivery through contracted pelvis is available, though large recent series have been spoilt from an academic point of view by the frequency with which Caesarean section has been performed. The ways in which asymmetrical and tearing stresses on the foetal head resulted in tearing of the tributaries of the longitudinal sinus or the great cerebral vein were described by Beneke (1910, 1920), Holland (1922, 1926), Greenwood (1924), Ehrenfest (1922). Stillbirth and neonatal death rates were found to be considerably higher when there was cephalopelvic /
cephalopelvic disproportion by Caldwell, Moloy, and d'Esopo (1934), Daichman and Pomerance (1948), d'Esopo (1947). As might be expected the dangers to the foetus are accentuated when the infant is large and a particularly high mortality is found in large postmature infants, (Rathbun, 1943; Potter and Koff, 1939; McKiddie, 1939). Apparently the actual capacity of the pelvis (its size) is considerably more important in determining the course of labour and the fate of the child than its actual shape, (Ince and Young, 1940).

In a series of 742 deliveries to mothers with "borderline" pelves, it was found that the stillbirth rate was approximately 1.5% when Caesarean section was used freely, (Jacobs, 1942). On the other hand, a very high mortality as a result of foetal trauma has been reported after labour and instrumental delivery though even a moderately contracted pelvis, (Bill, 1954). A number of authors have described methods of measuring the pelvis accurately so that the risk to the foetus may be assessed, (Thoma, 1937; Mengert, 1954; Moir, 1947; Caldwell et al, 1939). Mengert found that the combined foetal and mortality was 5.3% when the inlet capacity was 79% or less, compared to 2.3% when it was 90% or more. When the midplane capacity /
capacity was 90% or more the mortality was 2.6%,
but when it was 79% the mortality was 3% irrespective of the method of delivery.

Cephalopelvic disproportion due to large size of the foetal head exposes the foetus to the same risks of trauma as does pelvic contracture. In a study of 195 last postmature infants with an average gestation of 288 days a combined foetal and neonatal mortality of 10% was found by Potter and Koff (1939). The average duration of labour was 29 hours 43 minutes. Many of the deaths were due to hypoxia, the increased risk of which in the postmature infant has been described. In four of fifteen cases in which autopsy was performed, however, traumatic intracranial haemorrhage was found.

Unfortunately the evidence about the effects on the foetus of these abnormalities of labour which result in prolonged labour is not sufficient for their relative dangers to be assessed with any accuracy. On the other hand, the dangers of prolonged labour, irrespective of cause have been studied frequently. The majority show a steadily increasing mortality in labours of over thirty hours duration, (Peckham, 1932; Jeffcoate, 1943). The combined stillbirth and neonatal mortality was found to be more than doubled in labours lasting over 30 hours /
hours compared to a control series by d'Esopo and Marchetti (1942), Drillien (1947). More detailed figures are given by Baird (1955). Table 23A. It seems justifiable therefore to classify labour of thirty hours or more as a possible cause of foetal trauma and labour of 48 hours or more as a probable cause, rather than to try, for example, to assess the effect on the foetus of uterine inertia or cephalopelvic disproportion.

Precipitate labour. Precipitate labour has been noted to result in a much higher mortality than labour of average duration. In both white and coloured patients a much increased risk of stillbirth and neonatal death as a result of asphyxia or traumatic intracranial haemorrhage was noted by Peckham, (1932). The combined stillbirth and neonatal mortality was found to be more than twice as great in cases in which labour lasted less than three hours than when it lasted between 18 and 23 hours (d'Esopo and Marchetti, 1942). In labours lasting less than two hours an increase in the number of stillbirths of over 50% compared to cases with labour of average duration was noted by Tucker and Benaron, (1953). That the chief dangers to the foetus in precipitate delivery are the production of intracranial haemorrhage due to unduly rapid and forceful change in the shape of the foetal skull and hypoxia, possibly caused by persistent contraction/
<table>
<thead>
<tr>
<th>Labour</th>
<th>Less than 38 wks.</th>
<th>38-39 wks.</th>
<th>40-41 wks.</th>
<th>42 wks. &amp; over</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 24 hours</td>
<td>24.5</td>
<td>17</td>
<td>4.2</td>
<td>7.5</td>
</tr>
<tr>
<td>24 - 48 hours</td>
<td>25.0</td>
<td>10.6</td>
<td>8.2</td>
<td>10.7</td>
</tr>
<tr>
<td>48 hours plus</td>
<td>12.5</td>
<td>10.0</td>
<td>10.8</td>
<td>27.2</td>
</tr>
<tr>
<td>All lengths of labour</td>
<td>23.4</td>
<td>13.1</td>
<td>7.09</td>
<td>13.7</td>
</tr>
</tbody>
</table>
contraction of the placental site, has been stressed in a number of studies (Holland, Lane-Claypon, 1926; Peckham, 1932; Hughes, 1932; Craig, 1938; Jeffcoate, 1955).

From the point of the present study it is necessary to take account of the fact that the rate of stillbirth and neonatal death following labours of under 2 hours is not much more than twice as great as that after labours of average duration. Precipitate labour is, therefore, considered a possible and not a probable cause of foetal trauma.
Malrotation of the foetal head as a cause of prolonged labour and foetal trauma. The abnormal direction of the stresses exerted upon the foetal skull during labour when the vertex has not adapted to the pelvic shape have already been described. When the head fails to rotate into the occipito anterior position it is usually because of minor degrees of mid pelvic contraction, (d'Esopo, 1941; Thomas, 1933). Nevertheless, this failure to rotate in the upper and mid pelvis is liable to place undue strain upon the dural ligaments when the head enters the lower pelvis and tearing may result, (Holland, 1922; Capon, 1922). A series of 333 cases of persistent occipito posterior, 98 occipito transverse and 1432 occipito anterior positions was studied by Harer (1932). It was found that the two former positions were associated with a higher stillbirth and neonatal death rate than the latter, (Table 231). It will be seen that the foetal mortality, irrespective of the method of delivery is approximately four times as high in persistently occipito posterior and lateral positions than it is in occipito anterior. The author stressed that the majority of the deaths associated with these positions occurred in mature infants and were due to trauma and should be preventable. Unfortunately, pathological findings were not correlated with the position /
position during labour in this study. Nevertheless, the majority of authors appear to agree with Harer and d'Esopo (1941) and Steele and Javert (1942).

This series also indicates the high rate of instrumental delivery which is required in these positions of the vertex and which add to the already considerable mortality.

**Other presentations.** A series of 93 mature infants who presented transversely was published by Eastman (1932). The combined foetal and neonatal mortality was 42.3% in patients delivered vaginally. In 17 cases in which labour lasted between 12 and 24 hours, it was 64.7% and in patients with labours lasting more than 24 hours it was 100%. The chief dangers to the child appeared to lie in premature rupture of the membranes, prolapse of the umbilical cord (in 15.7% of cases) and hour glass or tetanic contraction of the uterus. In the series, twenty-one patients were delivered by caesarean section without a foetal death. The author emphasised that a considerable proportion of the deaths could be attributed to the trauma of prolonged labour though a protracted pelvis and that of the necessary manipulations to correct the bad presentation.

The /
<table>
<thead>
<tr>
<th>Character of delivery</th>
<th>No. of cases</th>
<th>% mortality</th>
<th>No. of cases</th>
<th>% mortality</th>
<th>No. of cases</th>
<th>% mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spontaneous delivery with rotation in post and lateral cases.</td>
<td>893</td>
<td>2.51</td>
<td>39</td>
<td>8.51</td>
<td>6</td>
<td>50</td>
</tr>
<tr>
<td>Forceps extraction with manual rotation</td>
<td>72</td>
<td>2.77</td>
<td>11</td>
<td>0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Forceps extraction</td>
<td>488</td>
<td>1.65</td>
<td>36</td>
<td>2.77</td>
<td>3</td>
<td>33.3</td>
</tr>
<tr>
<td>Scanloni manœuvre</td>
<td>131</td>
<td>15.38</td>
<td>60</td>
<td>5.17</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Version</td>
<td>46</td>
<td>20.45</td>
<td>37</td>
<td>19.44</td>
<td>8</td>
<td>14.28</td>
</tr>
<tr>
<td>Caesarean section</td>
<td>5</td>
<td>0.0</td>
<td>14</td>
<td>30.77</td>
<td>5</td>
<td>20.0</td>
</tr>
<tr>
<td>Delivered as occipito post</td>
<td>4</td>
<td>0.0</td>
<td>1</td>
<td>0.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Craniotomy</td>
<td>0</td>
<td>0.0</td>
<td>4</td>
<td>100.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>1432</strong></td>
<td><strong>2.77</strong></td>
<td><strong>333</strong></td>
<td><strong>11.35</strong></td>
<td><strong>98</strong></td>
<td><strong>13.68</strong></td>
</tr>
</tbody>
</table>
The latter included a high proportion of versions and extractions.

The dangers of face and brow presentations in predisposing to strain upon and tearing of the dural ligaments have been stressed by Holland (1922), Capon (1922) and Potter (1953). Unfortunately there appear to be no large series of cases with controls on which any estimate of the incidence of intracranial trauma associated with these presentations may be based. In a series of 27 face and brow presentations in 3260 deliveries studied, six stillbirths attributed to trauma were described by Lennie (1949).

Abnormal positions of the vertex are considered to be a possible cause of trauma in the present series.

The traumatic effects of different methods of delivery.

**Breech birth.** A detailed investigation into the causes of the relatively high foetal and neonatal mortality associated with breech delivery was made by Wilcox (1949). He found that the combined foetal and neonatal mortality in 1660 cases was 1\%\%\%. Unfortunately, autopsies were not carried out in all the cases and his figure of 3\% of deaths being due to intracranial haemorrhage may be /
be too low. Even so, when deaths due to congenital malformations and "prematurity" are excluded it is found that 16% of deaths were associated with bleeding within the skull. The figures of d'Esopo and Marchetti (1942) may be treated in the same way. Birth injury is then found to be responsible for more than one third of the deaths which can in any way be attributed to the method of delivery, a figure comparable to that of Potter and Adair (1943).
CHAPTER 2d

Toxic agents in parturition.

Under this heading are considered the more important causes of foetal damage which act by poisoning the foetus. They may be the result of disturbances in the child's or mother's metabolism, as in uraemia or rhesus incompatibility, or the result of infections or drugs. For present purposes those which act in early pregnancy are regarded as possible causes of foetal malformation and are discussed later. Only toxic agents acting in the last trimester of pregnancy and during labour and the neonatal period will be considered here.

The distinction between which agents are toxic and which hypoxic in their effect on the foetus is an arbitrary one. However, it is possible to make such a distinction in broad terms. For example, while pituitrin appears to affect the foetus by virtue of its interference with placental circulation and thus cause hypoxia, the toxins of diphtheria have more direct action on the child, and may be regarded as being "toxic", rather than "hypoxic".

The literature gives little information about the effect of maternal illness in causing cerebral palsy. Little thought that maternal ill-health was chiefly of importance as a predisposing cause of /
of prematurity. Osler was impressed by "the vigorous and healthy looking condition of the mothers" especially those of his hemiplegic patients. However, he described a case of diplegia born to a mother who suffered from tuberculosis and a patient with hemiplegia whose mother suffered from rheumatic chorea during pregnancy, (Osler, 1889). Freud listed maternal infectious diseases as an occasional contributory cause of cerebral palsy, (Freud, 1897).

The infectious disease which attracted most attention was syphilis, though assessment of the published cases is handicapped by the fact that there was no serological diagnosis. Children suffering from diplegia and hemiplegia with syphilitic parents were described in case reports (Giles de la Tourette, Erlenmeyer, Rosenberg, Friedman and Fournier.) Freud pointed out that the syphilis was not necessary causal in these cases though he acknowledged that it could be an important contributory aetiological factor in cerebral palsy, (Freud, 1897).

Most recent surveys hardly mention ill-health of the mother as an aetiological factor in cerebral palsy, (Willie, 1951; Evans, 1948; Phelps, 1941; Gustafson and Carceau, 1941). Two cases of congenital hemiplegia whose mothers suffered respectively /
Ively from influenza and pneumonia at the time of delivery were described by Byers (1941). Three children with cerebral palsy whose mothers were diabetic were mentioned by Asher and Schonell (1950). Andersen found that 37 of 173 mothers had some illness during pregnancy. Diabetes was present in three, syphilis in two, thyrotoxicosis in one and toxoplasmosis in one. Greenspan and Deaver reported two diabetic mothers amongst 94 of those children who attended a clinic for cerebral palsy. Six of 99 mothers of children with cerebral atrophy demonstrated by aircencephalography had infectious diseases during pregnancy, (Faber, 1947).

The effect of toxic agents in late pregnancy on the foetus.

For the purposes of description, toxic agents which operate during late pregnancy, labour and delivery have been classified as follows.

1. Conditions which act by causing the production of abnormal, toxic amounts of normally occurring substances, e.g. Rhesus incompatibility producing an excess of bilirubin and ammonia in the foetus, maternal uraemia causing an excess of nitrogenous waste products to accumulate, diabetes in which an excess of a variety of hormonal substances may be produced which are damaging to the foetus.

2. Conditions in which abnormal toxins are produced /
produced which cause foetal damage. In some of these maternal infections there may be no invasion of the foetus by micro-organisms, e.g. pylitis, diphtheria, meningococcaemia. In others, secondary infection of the foetus is relatively common, e.g. syphilis, toxoplasmosis.

3. Artificially administered toxins such as quinine.

Conditions which act by causing toxic accumulations of naturally occurring substances.

Rhesus Incompatibility. The classical example of a condition of this type is erythroblastosis foetalis due to rhesus incompatibility in which the normally occurring products of haemoglobin breakdown occur in such excess as to cause cerebral damage to the foetus or newborn infant. The relevant literature on this subject and on the kernicterus of prematurity is reviewed in the section on Dyskinesia.

The toxins which cause kernicterus directly or indirectly remain partly unknown. It seems certain that a high serum bilirubin level is not the only toxic agent and that other naturally produced poisons probably play a part, hypoglycaemia, hypoxia, tissue immaturity and abnormalities of ammonia metabolism have all been suggested as contributory causes of kernicterus, (Gerrard, 1957; Leikin, 1957).

In the present series the demonstration of significant blood incompatibility between the mother and /
and child together with evidence that the child has been affected as a result is regarded as a probable toxic factor. Other clinical signs which suggest the presence of kernicterus or the presence of a serum bilirubin titre of above 20 milligrams per cent are regarded as indicating that the child has probably been exposed to damaging toxins, (Aidin et al, 1950).

**Diabetes mellitus and endocrine disorders.**

It has long been recognised that maternal diabetes or a maternal pre-diabetic state is associated with a much increased foetal and neonatal mortality, (White and Hunt, 1940; Miller, Hurwitz and Kuder, 1944; Barns and Morgans, 1949; White, 1946; Sisson, 1940; Farquhar, 1959). There are multiple causes of this high mortality. The incidence of congenital malformations in the foetus is increased compared to that found in the offspring of unaffected mothers, (Laurence and Oakley, 1942; Miller, Hurwitz and Kuder, 1944). But other more complex aetiological factors appear to be responsible for the majority of perinatal deaths, (Barns and Morgans, 1949; White, 1946).

Unfortunately it is difficult to obtain full descriptions of the cerebral abnormalities encountered in autopsy in the children of diabetic mothers. The two fullest descriptions both comment on the relatively low brain weight observed in these cases, White /
(White and Hunt, 1943; Cardell, 1953). But no evidence of focal cerebral damage is reported by them, or in other descriptions of pathological findings.

Obviously the frequency with which apnoeic episodes occur in the infants of diabetic mothers and their liability to pulmonary atelectasis might be expected to predispose to hypoxic cerebral damage in the neonatal period. On the other hand, many of the foetal deaths could not be explained on this basis even after pathological examination, (Cardell, 1953; Warren and Le Compte, 1952). There is at least suggestive evidence that other toxic factors attributable to the mother's diabetes may be playing a part, probably in contributing to the liability to cerebral damage from trauma and hypoxia.

Combining the figures of three recent authors it is found that 8 of 606 mothers giving birth to children who showed congenital cerebral palsy had diabetes, (Asher and Schonell, 1950; Andersen, 1957). Unfortunately the type and origin of the cerebral palsy in these cases is not clearly stated. It would obviously be of interest to know whether there were indications of the cerebral palsy being due to congenital malformation in view of the increased /
increased incidence of congenital malformations in the children of diabetic mothers, (Miller, Hurwitz and Kuder, 1944). Nevertheless, this apparent excess of diabetic mothers compared to those of normal children suggests that diabetes or a demonstrated pre-diabetic state of the mother may have aetiological significance. Maternal diabetes is classified as a possible toxic factor to the foetus for the purposes of the present series.

**Hyperthyroidism.** There is some argument as to whether maternal hyperthyroidism should be regarded as a cause of foetal or neonatal death. This is hardly surprising when the comparative rarity of the condition is realised. It may be rare, in part, because of diminished fertility in the thyrotoxic woman, (Keynes, 1952). An incidence of toxaemia of pregnancy of 76% in thyrotoxic mothers was reported by Javert (1940). Mussey reported a combined foetal and neonatal mortality (excluding abortions) of one child in eighteen fully documented pregnancies and similar statistics were presented by Keynes (1948), Mussey, Haines and Ward (1948). No foetal mortality in children weighing over 1,500 grams was found by Javert (1940). There appears to be some justification for the statement that "there is no evidence that exophthalmic goitre unfavourably affects the course of pregnancy", (Browne, 1955). In the absence of statistics suggesting /
suggesting a frequent association of thyrotoxicosis in pregnancy and cerebral palsy, and in the absence of evidence that it causes brain damage, thyrotoxicosis is not considered a possible or probable toxic agent. This is in contrast to the opinion which has been expressed occasionally that thyrotoxicosis should be regarded as a contributory cause of the condition.

Infectious diseases of the mother during pregnancy. As pointed out by Wesselhoeft (1955) "I. The infection may pass off without injury (to the foetus) and allow full term delivery of a normal child.

2. The infection may induce spontaneous abortion or premature labour on the part of the mother.

3. The infection may (a) cause the child to be born with a congenital form of the disease (b) cause the death of the foetus in utero, or (c) bring about anomalies of foetal development resulting in major or minor congenital deformedies”.

It is predominantly with the third category of effects of infections with which the present study is concerned, though the effect of causing premature labour is also important.

For the purposes of description, infections likely to result in direct foetal invasion will be discussed first and then infections in which toxic effects /
effects are more important.

**Syphilis.** There is no shortage of cases of children with cerebral palsy whose disabilities are attributed to syphilis in the literature, (Freud, 1897; Ford, 1926, 1953; Ferguson and Critchley, 1930; Merritt, 1946).

It is possible to define a number of clinical neurological syndromes of congenital origin in which syphilis appears to be of prime aetiological importance. Of these, congenital paretic neurosyphilis appears to be the commonest. Menninger and Jelfiffe (1936) were able to collect 426 cases of this condition from the literature and 23 cases were summarised from their own practice by Merritt et al, (1946). Congenital cerebral vascular syphilis appears to be less common, partly, possibly because it is symptomatic at an earlier age and therefore more difficult to diagnose (Merritt et al, 1946; Ford, 1953). Rarer still is hydrocephalus due to chronic syphilitic meningitis though most large series of cases of hydrocephalus contain one or two cases, (Russell, 1949). In 100 cases of hydrocephalus described by Ford, three were due to syphilitic meningitis (Ford, 1926).

It appears to be generally agreed that these "congenital neurosyphilitic syndromes" have become progressively /
progressively uncommon in the antibiotic era and the majority of cases in the United States now occur amongst negroes, neurosyphilis being rare amongst whites.

The aetiological importance of syphilis in congenital hemiplegia, diplegia, dyskinesia and ataxia is much more difficult to define, and has been a subject of debate for many years, (Freud, 1897; Sachs and Hausmann, 1926; Ferguson and Critchley, 1930; Ford, 1926).

Quite a large number of cases of hemiplegia, apparently of congenital origin, diplegia and ataxia (usually with hydrocephalus), in association with congenital syphilis have been described in the quite recent literature by authors to whom the Wassermann reaction was available, (Babboniex, 1930; Nasso, 1924; Vaglia, 1920; Ferguson and Critchley, 1930; Merritt et al, 1946). It is difficult to assess the aetiological significance of the syphilis in these cases. In many cases the appearance of neurological signs and symptoms after some years makes it impossible to regard them as being (strictly speaking) of congenital origin, (Kinnier Wilson, 1954). Nevertheless, there remain a number of cases in which no other cause of typical cerebral palsy syndromes than congenital syphilis is apparent, (Ford, 1952). In the four modern series /
series which have been reported the prevalence of
maternal syphilis in cases of cerebral palsy does
not appear to be significantly higher than amongst
mothers in the general population, (Andersen, 1954;
Asher and Schonell, 1950; Whipple and Dunham, 1938).
This is in contrast to the impression of a number
of authors that maternal syphilis may be rather
underestimated as an aetiological factor in \( \text{A} \) and

\( \text{c} \) ebral palsy, (Wyllie and Shrubshull, 1931.)

On the other hand, there is much evidence that
syphilis causes foetal death and injury in a high
proportion of cases in which the mother is infected,
(Carr, 1935; Turner, 1934; Moseley et al, 1940).

Whilst preserving an open mind about the ways
in which syphilis may act in predisposing the child
to cerebral palsy, it seems justifiable to regard
the condition as a probable contributory aetiological
factor. In the present series, therefore,
evidence of active syphilis in the mother or child
will be regarded as being a probable toxic factor.

**Toxoplasmosis.** The recognition of "A small
number of cases of non-luetic encephalitis of
obscure aetiology in young infants" which appeared
to be due to true intrauterine infection led to the
recognition of toxoplasmosis as a cause of permanent
neurological sequelae in a high proportion of
affected /
affected children, (Wolf and Cowen, 1937). The infecting organism was soon found to be a protozoan parasite which was often a commensal organism on the affected mother, (Sabin, 1941).

Babies who were affected might be stillborn. Frequently they were abnormal at the time of birth, developed epileptic fits within a matter of days or weeks, showed jaundice in the neonatal period, and often had feeding difficulties. All milestones were retarded and later mental defect, a variety of types of cerebral palsy and epilepsy became apparent. Frequently, microcephaly, or hydrocephalus, a variety of eye defects, most commonly microphthalmia and chorioretinitis, and patchy intracranial calcification. Visceral lesions were also found. The course of the condition varies. Progressively severe epilepsy, retardation of development and secondary infections might result in death in a matter of years or months. In other cases though epilepsy persisted and mental development ceased, a fatal outcome did not occur and the condition appeared to become arrested. Surviving children were likely to show persistent cerebral palsy which might be of hemiplegic or diplegic distribution, (Callahan, Russell and Smith, 1946; Fisher and Wilson, 1948; Riley and Arneil, 1950; Farquhar, 1950; Hall, Moss and Ryan, 1953). A representative case and a survey /
survey of the literature were presented by Wyllie and Fisher, (1950).

The occasional association of toxoplasmosis with congenital malformations originating in early pregnancy has been recorded. For example, toxoplasmosis associated with situs inversus totalis was recorded by Van Creveld, Arus and de Bruyne, (1948). The association is probably coincidental.

Toxoplasmosis is probably a rare cause of cerebral palsy. In Sweden only two of 23,260 unselected infants were found to be suffering from toxoplasmosis by Svante (1953). Nevertheless, when its existence can be demonstrated in the child suffering from cerebral palsy it must be regarded as a probable aetiological factor.

Pyelitis. The danger to the foetus when the mother suffered from pyelitis of pregnancy has been realised from the beginning of the century. Cathala stated, "The prognosis for the child is grave as delivery may be premature and infection be transmitted to it". (Cathala, 1904). The early literature has been fully reviewed by Athenstaedt (1933).

A foetal mortality of 10% was reported by Haselhorst (1931). Traut (1937) found a combined foetal and neonatal mortality of 12% largely due to /
to premature delivery. A combined foetal and neonatal mortality of 15.8% was given by McLane (1939). In a survey of 252 pregnancies complicated by urinary infections which occurred between 1927 and 1944, a combined foetal and neonatal mortality of 8.3% (excluding abortions) was found by Dodds (1945). Twelve of the twenty-one stillbirths and neonatal deaths were prematurely born. 1.4% of 272 premature deliveries were attributed to pyelitis by Mauzey (1940). It seems probable, however, that whilst premature by weight, some of these babies were, in fact, delivered at or near term with small birth weights due to poor nutrition during late pregnancy. Unfortunately, most of the statistics which have been quoted relate entirely to the pre-antibiotic era and it is extremely difficult to find out to what extent the danger to the foetus has been lessened in recent years. Even in papers discussing "Chemotherapy in Obstetrics and Gynaecology", no reference of changes in foetal mortality is made (Douglas, 1940). Pratter (1941), however, on rather inadequate evidence felt that chemotherapy improved the foetal prognosis.

It has been stated "children born of pyelitic mothers are not abnormal unless prematurity results from the severity of the infection". On the other hand, the feebleness, liability to apnoea following delivery/
delivery and poor temperature control of the offspring of mothers with pyelitis are well recognised clinical findings. Unfortunately no statistics are available which give any idea of the relative frequency of the association between pyelitis and cerebral palsy, nor are the pathological findings in any large series of children of mothers with pyelitis reported. It seems likely that pyelitis in pregnancy may damage the foetus by its contributory toxic effects when in combination with other agents, especially hypoxia. On the other hand it is felt justifiable to consider it only as a possible, not as a probable toxic factor in the present study.

_Pneumonia_. Most of the studies of the effect on the foetus of pneumonia of the mother during pregnancy were made before chemotherapy came into widespread use. There is no doubt that is used to be extremely dangerous to the foetus. In 1614 cases in which pneumonia complicated the last three months of pregnancy it was found that 40% of children of viable weight (above 4 pounds) were stillborn or died in the neonatal period (Ransdell, 1905). In a series of 40 pregnancies complicated by pneumonia after seven months gestation, the offspring was stillborn or died in the neonatal period in 43% of cases (Bullowa, 1937). In this study it was found that the presence of pneumococcal septicaemia in the foetus /
foetus was of little prognostic help. The author laid stress on the importance of foetal hypoxia and toxic factors in causing damage, but emphasised that premature onset of labour was the main factor. Similar results and conclusions were presented by Finland and Dublin (1939), and Oppel (1939). In the majority of autopsies of offspring dying in the neonatal period, or born dead, pathological examination revealed changes compatible with hypoxic death.

It is extremely difficult to estimate the current danger of maternal pneumonia in the last trimester of pregnancy to the foetus, but it seems justifiable to consider pneumonia as a possible toxic factor injuring the foetus. Certainly in pregnancy pneumonia was recorded twice amongst the mothers of 349 cases of congenital cerebral palsy by Asher and Schonell, (1950).

**Bronchitis and bronchiectasis.** It is impossible to assess the significance of these conditions in causing death or damage to the foetus in the absence of studies of the requisite large series of cases. They are rather reluctantly not classified as possible or probable causes of toxic damage to the foetus in the present study.

**Other infectious diseases.** The effect of diphtheria in causing foetal death and damage and neonatal /
neonatal death certainly appears to have been marked in the pre-antibiotic era (Ollier, 1904, 1904; and de Lee and Greenhill, 1943). It is interesting, incidentally to note that the consistently quoted abortion rate of 33% appears to be derived from some experiments on pregnant rabbits made by Anderodias at the turn of the century (Holban and Seitz, 1929). In a more recent study of five cases, all the children were delivered alive and well, and it was considered that the placenta was probably impermeable to diphtheria toxins (Robinson et al, 1947). Fortunately no cases requiring classification of maternal diphtheria occurred in the present series.

Other diseases which appear to be of importance in causing toxic damage and foetal and neonatal death are recorded in association with cerebral palsy in the earlier literature and have been studied in the past twenty years, include Smallpox, (Rolleston and Ronaldson 1940), Meningococcal septicaemia, (Brandelberg and Vergan 1951), Typhoid fever, (Curshmann 1923) and (Rolleston and Ronaldson 1940), Ulcerative Colitis (Burgen et al 1939) and (Feren and Wolersky 1948), and Subacute Bacterial Endocarditis (Pedowitz and Hellman 1953). None of these conditions complicated the pregnancies of mothers in the present series. Therefore they have not been classified as toxic to the foetus for the purposes of this study.
CHAPTER 3

The dangers of parturition in the patients suffering from cerebral palsy in the present series.

In Tables 232 to 236 are shown the possible and probable hypoxic, traumatic and toxic insults to which the patients may have been subjected as a result of abnormalities of parturition. Cases of acquired cerebral palsy are omitted except for those with acquired hemiplegia, who form a rough control group, for purposes of comparison. Congenital cases without apparent abnormalities of parturition are also omitted though the total number of congenital cases is stated for each category. The cases are presented in the same order as when their birth histories were given in detail to facilitate ease of reference. To avoid arithmetical complications, each "possible insult" is counted as "one insult" and each "probable insult" as two insults, when the total potential stresses to which the child has been subjected are considered.
The number of hypoxic, toxic and traumatic insults suffered by patients suffering from congenital cerebral palsy.

In Table 233 are shown the numbers of hypoxic, toxic and traumatic insults thought to have been suffered by patients suffering from acquired hemiplegia, hemiplegia of unknown origin and congenital cerebral palsy in the present series. In Table 232 these are expressed as total insults of each type per 100 patients. Since the same criteria were used to interpret the case histories in each category, the numbers of insults per 100 patients may be directly compared, though the small number of patients in some of the categories makes some of the comparisons of doubtful value.

It will be observed that the greatest number of insults per 100 patients is found in congenital hemiplegia, then in dyskinesia, premature diplegia, ataxia, ataxic diplegia, mature diplegia and least in bilateral hemiplegia. A smaller number of insults is found in acquired hemiplegia and hemiplegia of unknown origin. There is, in fact, a statistically significant difference between the number of insults in acquired hemiplegia and in the number in bilateral hemiplegia, the form of congenital cerebral palsy with least insults.

In general, these results are what might be expected from a review of the literature on the aetiology.
aetiology of congenital cerebral palsy, and from the clinical impression of the case histories. There is general agreement that birth injury is of predominant importance in the aetiology of congenital hemiplegia. Birth injury (including the effects of rhesus incompatibility in this term) is also recognised to be present in the vast majority of patients suffering from dyskinesia. Abnormalities of parturition must be expected to be prevalent in any large series of premature infants and it is hardly surprising to find that there are more insults in the group of premature than in the group of mature diplegic patients. But the suggestions which have been made that birth injury is less important in diplegia than in congenital hemiplegia or dyskinesia gains some support from this study (Collier, 1899). A proportion of cases of ataxic diplegia, ataxia and bilateral hemiplegia have been thought to be the result of developmental malformations, often genetically determined, by a number of authors. The relatively low number of insults found in these categories is compatible with this opinion.

The types of insults and their timing.

Apart from a rather marked excess of toxic insults in dyskinesia, due entirely to rhesus incompatibility being considered a toxic insult, there are no significant differences in the proportions /
proportions of hypoxic, toxic and traumatic insults in the various categories, Table 232. Excepting dyskinesia from consideration, there is a tendency for there to be a rather higher ratio of hypoxic to traumatic insults in those categories in which there are most insults. Thus the ratio of hypoxic to traumatic insults in congenital hemiplegia is about 2.6:1, in premature diplegia about 2.3:1, in mature diplegia about 1.8:1, and in bilateral hemiplegia 1.5:1.

One might assume that the reason for the higher ratio of hypoxic to traumatic insult in the "birth injury categories" is due to a greater prevalence of abnormalities of pregnancy, which are predominantly hypoxic in their effects. This does not appear to be the case, however, for in Table 234, is shown the timing of the various types of insults. The proportion of insults sustained during pregnancy is remarkably constant at between 28 and 37% except in the case of ataxic diplegia in which relatively few insults occur during pregnancy and a rather high proportion during labour and delivery and in the neonatal period. Similarly, the proportion of insults occurring during labour and delivery and during the neonatal period vary little except in ataxic diplegia.

Thus, /
<table>
<thead>
<tr>
<th>Condition</th>
<th>No. of patients</th>
<th>No. with insults</th>
<th>No. of insults per 100 patients</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acquired hemiplegia</td>
<td>33</td>
<td>8</td>
<td>27 12 9</td>
<td>48</td>
</tr>
<tr>
<td>Unknown hemiplegia</td>
<td>11</td>
<td>3</td>
<td>18 0 9</td>
<td>27</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>30</td>
<td>30</td>
<td>297 20 113</td>
<td>430</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>7</td>
<td>5</td>
<td>129 0 86</td>
<td>217</td>
</tr>
<tr>
<td>Mature diplegia</td>
<td>43</td>
<td>31</td>
<td>131 12 75</td>
<td>218</td>
</tr>
<tr>
<td>Prem. diplegia</td>
<td>34</td>
<td>30</td>
<td>213 12 96</td>
<td>321</td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>10</td>
<td>7</td>
<td>140 0 90</td>
<td>230</td>
</tr>
<tr>
<td>Ataxia</td>
<td>9</td>
<td>8</td>
<td>176 22 77</td>
<td>275</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>16</td>
<td>16</td>
<td>144 87 100</td>
<td>331</td>
</tr>
</tbody>
</table>
Thus, it may be said that apart from there being an excess of toxic factors in dyskinesia, and a relative poverty of hypoxic insults during pregnancy in ataxic diplegia, the categories show little difference in either the type or timing of insults suffered during parturition. On the other hand, insults appear to be significantly more numerous in some categories of congenital cerebral palsy than in others.

This is not to say that there is a greater chance of a given baby suffering from hemiplegia than diplegia if insults are particularly numerous. It is possible to visualise all kinds of predisposing factors which might make him more susceptible to one condition or the other, quite apart from any abnormalities of parturition. Moreover, the presence or absence of abnormalities of parturition does not prove that birth injury has caused cerebral palsy in an individual child. All that has been shown is that there are more potential dangers to the child in the birth histories of those who suffer from hemiplegia or dyskinesia, than for example, those with bilateral hemiplegia or ataxic diplegia.

The numbers of insults per patient in the categories of cerebral palsy compared.

The significance of the number of insults found /
Types of insults by category of cerebral palsy.

<table>
<thead>
<tr>
<th>Category</th>
<th>Total patients</th>
<th>Total without insults</th>
<th>Total with insults</th>
<th>No. of hypoxic insults</th>
<th>No. of toxic insults</th>
<th>Traumatic insults</th>
<th>Total insults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acquired hemiplegia</td>
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<td>25</td>
<td>8</td>
<td>9</td>
<td>4</td>
<td>3</td>
<td>16</td>
</tr>
<tr>
<td>Unknown hemiplegia</td>
<td>11</td>
<td>8</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
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<td>30</td>
<td>89</td>
<td>6</td>
<td>34</td>
<td>129</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
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<td>2</td>
<td>5</td>
<td>9</td>
<td>0</td>
<td>6</td>
<td>15</td>
</tr>
<tr>
<td>Mat. diplegia</td>
<td>143</td>
<td>12</td>
<td>31</td>
<td>56</td>
<td>6</td>
<td>32</td>
<td>94</td>
</tr>
<tr>
<td>Prem. diplegia</td>
<td>34</td>
<td>4</td>
<td>30</td>
<td>71</td>
<td>4</td>
<td>32</td>
<td>107</td>
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<td>Ataxic diplegia</td>
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<td>23</td>
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<tr>
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<td>23</td>
<td>14</td>
<td>16</td>
<td>53</td>
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</table>
### The timing of insults

<table>
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<tr>
<th>Category</th>
<th>Total insults</th>
<th>Pregnancy</th>
<th>Labour and delivery</th>
<th>Neonatal</th>
<th>No. of cases</th>
</tr>
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<tr>
<td>Congenital hemiplegia</td>
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<td>54</td>
<td>38</td>
<td>30</td>
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<tr>
<td>Bilateral hemiplegia</td>
<td>15</td>
<td>5</td>
<td>6</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>Mat. hemiplegia</td>
<td>94</td>
<td>31</td>
<td>44</td>
<td>19</td>
<td>43</td>
</tr>
<tr>
<td>Prem. hemiplegia</td>
<td>106</td>
<td>33</td>
<td>43</td>
<td>30</td>
<td>34</td>
</tr>
<tr>
<td>Ataxic hemiplegia</td>
<td>23</td>
<td>4</td>
<td>11</td>
<td>8</td>
<td>10</td>
</tr>
<tr>
<td>Ataxia</td>
<td>27</td>
<td>9</td>
<td>11</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>53</td>
<td>19</td>
<td>22</td>
<td>12</td>
<td>16</td>
</tr>
</tbody>
</table>

### Approx. % of insults at each time

<table>
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<th>Category</th>
<th>Total</th>
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</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>28</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>33</td>
</tr>
<tr>
<td>Mat. diplegia</td>
<td>33</td>
</tr>
<tr>
<td>Prem. diplegia</td>
<td>31</td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>17</td>
</tr>
<tr>
<td>Ataxia</td>
<td>34</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>37</td>
</tr>
<tr>
<td>Type of palsy</td>
<td>Numbers</td>
</tr>
<tr>
<td>--------------</td>
<td>---------</td>
</tr>
<tr>
<td>Number of insults</td>
<td>Acquired hemiplegia</td>
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<td>0</td>
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</tr>
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<tr>
<td>7</td>
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</tr>
<tr>
<td>8</td>
<td>0</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>30</strong></td>
</tr>
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</table>
### TABLE 236

<table>
<thead>
<tr>
<th>Acquired</th>
<th>Congenital</th>
<th>Bilateral</th>
<th>Mat. Prem.</th>
<th>Ataxic Hemiplegia</th>
<th>Ataxic Diplegia</th>
<th>Hemiplegia</th>
<th>Diplegia</th>
<th>Total</th>
<th>Percentage of Patients by Category of Cerebral Palsy and Number of Insults</th>
</tr>
</thead>
<tbody>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>100</td>
</tr>
</tbody>
</table>
found in a given category of cerebral palsy will depend largely on the numbers within it with very few, or no abnormalities of parturition. For example, it is possible to conceive of one series of 100 patients with 430 insults which contains 10 patients with six insults, each on average, twenty patients with five insults, sixty patients with four insults, and ten with three insults. In such a series there would appear to be a constant factor of abnormal parturition and one would be justified in suspecting (however cautiously) that abnormal parturition was of aetiological importance in the majority of cases. But the 430 insults might also be due, in another series, to there being 50 patients without any abnormality of parturition, 30 patients with 9 insults, and 20 with 8 insults. In this series, abnormal parturition could be held to be of aetiological importance in only a proportion of patients. One would be tempted, however unwillingly, to postulate that "birth injury" was unlikely to be significant aetio logically in the others.

The numbers of patients by the number of insults they may have suffered during and immediately after parturition is shown in Table 235. A high /
high proportion of patients with acquired hemiplegia had no insults and only three per cent. had more than two. On the other hand, the majority of patients suffering from congenital cerebral palsy had more than two insults, and 19% had more than 5. The difference in these figures is significant statistically, (p being less than .01, using the X2 test).

There are some interesting differences in the categories of congenital cerebral palsy when they are analysed in this way. Statistically there is a significantly higher proportion of hemiplegic and dyskinetic patients than mature and premature diplegic patients with more than three insults (p less than .01).

It will be noted, for example, that 18 of the 43 mature, and 5 of the 134 prematurely born diplegic patients had one or no insults.

The figures in Table 236 are graphed in Fig. 13 (percentage of patients by number of insults). It will be seen that, broadly speaking, there are three types of curve. Firstly there is that of acquired hemiplegia. Relatively few patients in this category have suffered many insults during parturition and the curve is a linear descending one. Secondly are the curves of congenital hemiplegia and dyskinesia, which are very similar. The majority of patients
patients have had many insults in the course of pregnancy, labour and delivery. The curves are ascending and linear. Thirdly are the other congenital categories, patients suffering from bilateral hemiplegia, diplegia, ataxic diplegia and ataxia. These curves are biphasic. They share characteristics of the curve of acquired hemiplegia and those of congenital hemiplegia and dyskinesia. As indicated in the Table 237, a relatively high proportion of patients have few or no insults, but a proportion have a history of numerous hypoxic, toxic and traumatic episodes during pregnancy, labour and delivery.

Further study of the distribution of patients in the various categories by the number of insults sustained.

For the purposes of further study, three composite categories will be considered. In the first are included patients suffering from acquired hemiplegia in whom birth injury does not appear to be important. These form a rough control group. The second category comprises patients suffering from dyskinesia and congenital hemiplegia. A high proportion of patients in both these categories show many insults, the percentages by number of insults being statistically similar. The curve on the graph Fig. 23 has been described as being of "birth injury type". In the third category are included
those cases of congenital cerebral palsy in the categories of diplegia, ataxic diplegia, ataxia and bilateral hemiplegia. The percentages of patients in these forms of cerebral palsy by the number of insults are not significantly different statistically, and in the graph they all tend to show the biphasic curve which has been described. Fewer patients have no insults than in acquired hemiplegia; fewer have many insults than in diplegia, ataxic diplegia, ataxia or bilateral hemiplegia.

In Table 237 are shown the distributions of patients in these three composite categories by numbers of insults. The differences in these distributions between any two categories are highly significant, (using the X2 test, p is less than .01). Thus, 97% of patients with acquired hemiplegia had less than two insults during parturition, but only 17% in composite category two, and 53% in composite category three. In contrast, only 3% of patients with acquired hemiplegia had four or more insults, but there were 53% in composite category two and 33% in composite category three.

When patients without any apparent abnormalities of parturition are excluded from consideration it is found that the difference in the distribution of the remainder by number of insults is still statistically/
statistically significant, (using the X2 test, p is less than .01), in the three categories. Thus the difference in the apparent distribution of patients by the number of insults in the composite categories is not only the result of there being more patients with no insults in some than in others. Patients who have been subjected to abnormal parturition tend to suffer fewer insults in composite category one than these in composite category three (comprising diplegia, ataxic diplegia, ataxia and bilateral hemiplegia), and these in composite category two (comprising congenital hemiplegia and dyskinesia) relatively many.

It is impossible to draw firm conclusions about the relative importance of birth injury in the different categories of cerebral palsy on the basis of these findings. One reason for this is that it cannot be shown that the number of insults suffered by the child during parturition is necessarily proportional to the danger to which he is exposed. Assuming that the number of insults does give some indication of the likelihood of his being damaged, certain tentative conclusions about the aetiological importance of birth injury in the various categories of cerebral palsy may be offered.

The fact that a minority of patients suffering from /
<table>
<thead>
<tr>
<th>No. of insults</th>
<th>Acquired hemiplegia</th>
<th>Congenital hemiplegia and dyskinesia</th>
<th>Diplegia and Ataxic diplegia, Ataxia, Bilateral hemiplegia</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>25</td>
<td>0</td>
<td>22</td>
</tr>
<tr>
<td>1</td>
<td>4</td>
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</tr>
<tr>
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</tr>
<tr>
<td></td>
<td>33</td>
<td>45</td>
<td>103</td>
</tr>
</tbody>
</table>
**TABLE 238**

Numbers and percentages of patients in previous grouped categories by number of insults

<table>
<thead>
<tr>
<th>Type of hemiplegia</th>
<th>Acquired hemiplegia</th>
<th>Congen. hemiplegia and dyskinesia</th>
<th>Diplegia, Ataxic diplegia, Ataxia, Bilateral hemiplegia</th>
<th>Approx. %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. of cases</td>
<td>No. of insults</td>
<td>No. of cases</td>
<td></td>
</tr>
<tr>
<td></td>
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<table>
<thead>
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<th></th>
<th>Number</th>
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</table>
from congenital hemiplegia and dyskinesia have few insults, and the majority have many, suggests that birth injury may be of considerable aetiological importance in these conditions. The fact that it is uncommon to find patients suffering from congenital hemiplegia or dyskinesia in whom there is not a history suggestive of hypoxic, toxic or traumatic damage being sustained by the child supports this conclusion. The possibility of a minority of cases being due to developmental malformations of the brain associated with abnormalities of parturition cannot be excluded.

In contrast to patients suffering from congenital hemiplegia and dyskinesia, a quite considerable proportion of those with diplegia, ataxic diplegia, ataxia or bilateral hemiplegia have no insults, and fewer have many insults. The fact that so many have no insults (21% of the composite category) suggests that aetiological factors other than birth injury may be important in these conditions. At the same time, in as many as one third of cases there are four or more insults (Table 238). If these postulated insults are considered to give some indication of the danger to the foetus than it seems likely that birth injury may have been important in a proportion of the patients. The biphasic nature of the curve on the graph of number of /
Figure 3.

Percentages of Patients by number of Insults.

Acquired Hemiplegia

Congenital Hemiplegia and Dyskinesia

Congenital Diplegia

Ataxia -- Ataxic Diplegia -- and Bilateral Hemiplegia

Number of Insults

Percentages of Patients by number of Insults.

Acquired hemiplegia

Congenital hemiplegia & Dyskinesia

Congenital Diplegia, ataxic diplegia, ataxia & bilateral hemiplegia

Number of Insults
of patients by number of insults may then be explained in two ways. The composite category may be considered as containing two groups of patients from the aetiological point of view. A proportion of patients appear to suffer from congenital cerebral palsy which is not the result of birth injury. A proportion of these, like patients with acquired hemiplegia, are likely to have been born after abnormal pregnancies, labours or deliveries, and some will have been exposed to apparent insults, usually small in number. The second group of patients in the composite category appear to have many insults and their cerebral palsy, like that of patients suffering from congenital hemiplegia and dyskinesia may be regarded as being due to birth injury.

A more complicated explanation of the findings is that there is a continuum of patients between the two groups described above. One may postulate that in a minority of patients, aetiological factors other than birth injury operate to cause cerebral palsy, and that in another minority, birth injury is the only cause of the cerebral palsy. Between these minorities there may be a series of patients in whom birth injury, and aetiological factors other than birth injury are both important in varying degrees. Thus, a child may be predisposed to suffer /
suffer irreparable cerebral damage as a result of very slight birth trauma by the presence of congenital malformations or metabolic abnormalities, possibly genetically determined. On present evidence it is impossible to guess at the relative importance of birth injury and other etiological factors in these cases which appear to lie between the birth injury, and the other than birth injury groups.
SECTION 6

CONGENITAL MALFORMATIONS AS A CAUSE OF CEREBRAL PALSY.
CHAPTER 1.

Congenital malformation as a cause of cerebral palsy.

Since the earliest medical interest in cerebral palsy there has been speculation about the importance of malformation of the brain as a cause of the condition. Yet the precise aetiological significance of malformation remains unknown. In this section the various phases of interest in congenital malformation as a cause of cerebral palsy will be described. Evidence will be presented which suggests that in a proportion of patients in the present series there have been developmental malformations of the brain.

Historical. Pathologists made the first attempts to distinguish between the effects of birth injury and developmental malformations on the brain. Casauvielh tried to define criteria by which the pathological findings characteristic of damage sustained by the brain as a result of trauma or disease could be distinguished from those which were the result of "vices primitives" - the failure of the brain to develop normally from the time of its earliest stage of development. (Casauvielh, 1327).

Contemporary pathologists argued about the significance of various pathological appearances for years. For example, some considered that cerebral atrophy was the result of "Vice primitive", whilst others attributed it to the end result of disease or damage to the brain during late pregnancy or birth. (Breschet, 1831; von Lellemand, 1834; Cruvehlier, 1862).

Cruvehlier pointed out that the appearances of the brains of patients who had suffered from cerebral palsy were due not only /
only to the effects of the damaging agent, but also to the
processes of repair. In many cases the repair processes might
so alter appearances that the nature and time of onset of the
original damaging agent were impossible to determine retrospectively. He thought that it was almost impossible to determine
whether some pathological appearances were due to "vices
primitives", or were the result of disease occurring at a very
early stage of prenatal cerebral development, for in both cases
arrest of growth was likely to be the most striking neuropathological finding.

As more became known about the appearances of brains at
various stages of prenatal development it became possible to
assess with greater accuracy the time at which brain growth had
been interfered with, though to distinguish between "vices
primitives" and disturbance of growth due to acquired disease
in the earliest stages remained impossible. Kundrat, for
example, was able to date approximately the time at which the
disease processes responsible for true porencephaly had been
present by examining the nature and patterns of convolutions
surrounding the lesion. He showed that the disturbance of
growth had occurred in the majority of patients before the
fourth or fifth month of prenatal life. (Kundrat, 1882).

There continued to be considerable argument about the
nature of the prenatal disease processes which were responsible
for the pathological changes observed in the brains of patients
suffering from cerebral palsy. Some authors, including
Abercrombie,
Abercrombie maintained that prenatal cerebral embolism was the cause in many cases, others including Kundrat postulated transient defects in cerebral blood supply during prenatal life. Strumpell considered that prenatal encephalitis was the most important cause in a majority of cases of cerebral palsy. The controversy about which disease processes were important tended to confirm Cruvehlé's opinion that it was often difficult to make a firm retrospective pathological diagnosis. (Cotard, 1868; Henoch, 1869; Kundrat, 1882; Strumpell, 1884; Abercrombie, 1888).

Until the end of the nineteenth century clinical studies were little help in determining the aetiological importance of developmental malformations of the nervous system in cerebral palsy, largely because so little was known about the causes or effects of malformations until this time. Moreover, the work of Little and McNutt, which was so enthusiastically and uncritically accepted, appeared to demonstrate a definite causal relationship between birth injury and cerebral palsy. (Gowers, 1888; Sachs and Peterson, 1890). A few authors had speculated as to whether patients might be predisposed to develop cerebral palsy by hereditary factors. Richardiere thought that heredity was the most important single determining factor in hemiplegia acquired before the age of three years and in congenital cases. Wallenberg described "neurological disease" in the parents or siblings of 14 of 160 cases of hemiplegia of whom 141 had acquired /
acquired the condition postnatally, and Marie instanced the case of a boy with congenital hemiplegia whose three siblings had all died with convulsions. (Richardiere, 1835; Wallenberg, 1836; Marie, 1833). On the other hand, hereditary factors were not considered to be important in congenital or acquired hemiplegia by Wuillamier, (1832); Osler (1839); or Freud and Rie, (1891).

Opinions also differed about the importance of hereditary factors in diplegia. It was generally agreed that a high proportion of patients (34% Freud, 42.7% Ganghofner), had no obvious history of birth injury or other likely known cause of cerebral damage. Freud thought that the importance of "Little's factors" had been over-emphasised, and he criticised McNutt's assumptions that the majority of cases were due to traumatic subdural haemorrhages very severely. He postulated that abnormal parturition and an abnormal foetus might be joint manifestations of some underlying maternal abnormality, rather than cause and effect. He recognised that reports of "generalised rigidity" or "paraplegia" occurring in siblings were not infrequent, and analysed those of Feer, (1890); Ganghofner, (1895); and Schultz, (1886) in detail. He speculated as to whether some of these cases might not be transitional between the two recently described familial syndromes of Tay-Sachs disease (which he regarded as a progressive form of diplegia), Friedreich's disease, and true Little's disease. (Freud, 1893).

Collier extended and elaborated the theories of Freud about the aetiology of diplegia. Unfortunately he included within this category, cases of Tay Sachs disease, which is hereditary, /
hereditary, and probably other forms of progressive cerebral degeneration, and it is difficult to be sure to what extent his cases may be regarded as representative of "diplegia" as defined in this thesis. He thought that in only a relatively small proportion of patients was diplegia the result of birth injury. In the majority of cases arrest of neuronal formation and maturation had occurred in prenatal life. The reasons for the arrest were not very specifically described, but he appears to have believed that genetic factors were responsible in some cases (for example, in amaurotic family idiocy), and that in others reproductive failure of the mother, often associated with ill-health in pregnancy, might produce an uterine environment in which full neuronal development could not occur. In fact, he postulated two inter-related causes for the majority of cases: firstly, in some patients there were inborn factors dating from conception which resulted in nerve cells being formed which were predestined to degenerate on account of their intrinsic defects. Secondly, in other patients a brain undergoing normal maturation would be "frosted" because of an unfavourable uterine environment, often dependent upon maternal disease, (Collier, 1899, 1926). Clearly these theories added little to those put forward by Freud, but in Britain and America, Collier's articles on cerebral palsy were certainly more widely read than those of Freud.

In his descriptions of patients suffering from congenital cerebellar ataxia, Batten made a distinction between those cases which
which he attributed to the effects of birth damage and those which he thought were the result of partial or complete cerebellar agenesis. He did not speculate as to the relative importance of malformation and birth injury as a cause of the condition, and there was considerable argument amongst subsequent authors about this, (Batten, 1903; 1905; Foerster, 1910, and Clark, 1913).

More Recent Work.

With the turn of the century the results of research in other fields of medical interest were applied increasingly to the problems of determining the aetiology of cerebral palsy. Advances in experimental teratology, developmental neuroanatomy and pathology, human genetics and improved methods of neurological diagnosis all contributed to increased understanding of developmental malformations as a cause of cerebral palsy.

The contribution of experimental teratology. Though experimental interference with normal growth processes had been made as early as 1822 by St. Hillaire, (the elder) using the chick embryo, it was not until the early nineteenthundreds that the results of such experiments on the central nervous system were fully analysed and became widely known, (Ballantyne, 1893, 1904; Schwalbe, 1906 (in difficult German), Dareste, 1891). von Monakov, (1926) described their relevance to malformations in the human nervous system and emphasised the general principle that the timing of an insult directed towards the developing central nervous system was more important than its nature. The severity with which tissues were damaged appeared to be related more /
more to their rate of growth at the time of interference than to the type of insult used.

With subsequent refinement in research methods it has been possible to prove the truth of von Monakov's concept more rigorously. Different malformations of the brain have been produced by employing the same teratogenic agent at different stages of prenatal development in experimental animals. For example, different degrees of malformation varying from small areas of microgyria, to complete anencephaly, as well as microcephaly, hydrocephalus, true porencephaly and partial agenesis of the cerebrum and cerebellum have been produced by using prenatal irradiation. It is even possible to predict which abnormalities will result from a given dose of irradiation at different gestational ages. Many of the malformations which can be produced in this way are similar to those encountered in the brains of patients who have suffered from cerebral palsy, (Warkany, 1947; Stockard, 1920; von Monskov, 1901; Minkowski, 1952).

In addition to providing information about the types of malformation to be expected if growth of the central nervous system is interfered with at different stages of its development, animal experiments have also given information about the agents which are likely to be damaging. Thus it was the early experiments of von Hippel and Pagenstecher reported in 1907 which first attracted the attention of clinicians to the dangers of maternal irradiation, (Zappert, 1926; Goldstein and Murphy, 1929). On the other hand many agents which may be shown to be teratogenic /
teratogenic in animals, such as gross vitamin deprivation, lithium and selenium intoxication, or massive corticosteroid overdosage, do not appear to be of great clinical importance. One dietary deficit which may be rarely significant is severe maternal iodine lack, for there are a number of reports of endemic cretins in which diplegia was associated with prenatal arrest of cerebral development, (Lotmar, 1933; Warkany, 1927). There is still remarkably little evidence about the effect of severe maternal hypoxia in the first trimester of pregnancy on the foetus; it is still impossible to decide whether the hypoxia of threatened abortion may cause subsequent aberration of foetal development in the human being, though in animals such an effect does not appear to have been demonstrated.

The contribution of embryology and neuropathology. At the same time as experimental malformations in the brains of animals were being studied, the early stages of the development of human brains were also being investigated. Very detailed studies were made of large series of human brains showing developmental malformations. Full descriptions of many microcephalic brains were presented by Ballantyne, (1902, 1904); Vogt, (1905); Schwalbe, (1906 and Schob, (1930), (the previous authors' work being massively and enthusiastically summarised by him). These and similar studies showed that a number of pathological appearances hitherto thought to be the result of birth injury might equally well, or better, be explained on the basis of developmental malformation. In the light of increased embryological /
embryological knowledge and the experience of animal experiment
findings such as partial aplasia of the cerebellum, corpus
callosum and cerebral hemispheres, areas of micro or macrogyria,
heterotopic islands of grey matter and obstructive and compens-
satory hydrocephalus could often be explained on the basis of
developmental aberrations. At the same time, experimental
work on the effects of birth injury in animals and more detailed
study of human birth injuries clarified the neuropathological
pictures to be expected as a result of paranatal insults.
Status marmoratus and cystic encephalomalacia, for example, are
now generally accepted to be the result of paranatal injury,
rather than developmental malformation, (Benda, 1952; Minowski,
1952).

There remain, however, a number of neuropathological
findings which cannot be interpreted as being entirely due,
either to developmental malformation or birth injury with any
certainty. For example, there is a marked tendency for mal-
formed brains to be damaged by trauma during birth. Haemorr-
hage may occur into preformed porencephalic defects, or sub-
dural haemorrhage over areas of lobar sclerosis, Griepentrog,
1954). Even in very early prenatal cerebral damage there
tends to be gliosis as well as tissue loss and the process of
repair may obscure the nature of the original cerebral damage
very greatly.

A further difficulty in the interpretation of pathological
appearances is that even when they can be ascribed to develop-
mental malformation with some certainty, there is commonly
little /
little indication as to whether they are the result of genetic factors, or of disease or trauma acting in early prenatal life. It has been shown, for example, that there is commonly a genetic basis for anencephaly, as there is for, what Penrose calls, "true microcephaly", yet both these conditions may be caused experimentally by interfering with the development of the central nervous system at an early stage of prenatal life. (Penrose, 1933).

Clinical and pathological studies. Close study of large series of diplegic patients from the clinical and pathological points of view led both Patten and Stewart to believe that developmental malformation of the brain was a more important aetiological factor than had hitherto been believed. Patten was impressed by the remarkable consistency from patient to patient of the clinical findings in diplegia, and by the fact that 77.7% of patients without a history suggestive of birth injury showed symmetrical cerebral palsy. He argued that such symmetry of pathological findings and clinical effects would be much more easily produced by developmental malformations during the course of brain development than by birth injury with its greater tendency to cause focal and therefore asymmetrical effects, (Patten, 1931). Stewart studied the pathological findings in fifty mentally defective patients who suffered from "Diplegia". Unfortunately at least seven of the patients he classifies as suffering from diplegia would not be placed in this category in the present work, and there must be doubts about some of the others. He was impressed by the diversity of /
of the neuropathological findings in his cases, and suggested that this made it likely that different aetiological factors might be responsible for them. He felt that the very small size and weight of the brains in some of the cases, many of whom had no history indicating birth injury was in favour of a disturbance in the early stages of brain development and, "afforded clear proof of the prenatal origin of the condition". He thought that "Collier's conception of a primary degeneration of the cerebral neurones — best explained many of the lesions encountered", (Stewart, 1942-3.)

Though Freud had collected a number of cases of diplegia occurring in siblings and cousins from the literature and his own experience, fuller elucidation of the genetic background of diplegia required the detailed study of a large number of patients and their families. This was undertaken by Penrose in 1938. He made a clinical and genetic study of 1280 mentally defective patients of whom "66 suffered from bilateral and asymmetrical cerebral lesions which dated from birth or early life .... pyramidal lesions predominated in 55 cases, and in 11 the lesion was wholly or mainly extrapyramidal". The latter group included some cases of progressive cerebral paralysis and is obviously more diverse in content than patients classified as suffering from diplegia in this work. Nevertheless, the majority of his patients certainly come within this category.

In at least seven cases "bilateral pyramidal disease" occurred in two siblings and in other cases there was a significant excess of mentally retarded siblings over that expected in...
in the general population. Consanguinity of parents was present in a total of six of the sixty-six cases. Parents were less often defective than were those of children suffering from other types of mental defect. Penrose considered that there was evidence to suggest that congenital diplegia was inherited as a mendial recessive trait, another manifestation of which might be mental defect. No such genetic background was found in a much smaller group of patients suffering from congenital hemiplegia.

A rather similar series of 36 mentally defective patients suffering from cerebral palsy was studied by Yannet, (1944). He classified them into three categories; firstly those suffering from diplegia, secondly those suffering from "asymmetrical cerebral palsy" (predominantly hemiplegia) and thirdly those with ataxia. Unfortunately he grouped "patients with striatal symptoms in their respective diplegic or cerebellar groups - thus avoiding re-duplication of data", instead of making a separate category for them, but since there were only 14 with striatal symptoms his main conclusions are probably valid. Yannet found that in all three groups the mean maternal age was higher than expected, that later born children in the birth order tended to be affected (especially by diplegia), and that there was an excess of similarly affected children and siblings who were mentally retarded. The frequency of abnormal parturition was impressive only in the hemiplegic group of patients, but there was an excess of prematurely born patients in both the hemiplegic and diplegic groups. The small head size of many of the diplegic patients was notable and there was a high prevalence /
prevalence of congenital abnormalities of the eyes. He compared these findings to those of other workers in large series of patients suffering from undoubted developmental malformations. He was impressed by the fact that his diplegic patients had a similar high mean maternal age, high ordinal birth rank, a high proportion of similarly affected and mentally retarded siblings, a high incidence of premature birth, and by the fact that associated physical anomalies were frequently encountered in his findings. He felt that this suggested that developmental malformations were probably important aetiologically in a relatively high proportion of diplegic patients. He thought that the very small head size of many of his patients was in favour of there having been a disturbance in brain growth at an early stage of development, and that this also was in favour of developmental malformation, rather than birth injury being the damaging agent in many of his diplegic patients.

In a later paper, Yannet studied 42 cases of "asymmetric spastic infantile cerebral palsy" (the majority of whom suffered from hemiplegia) in more detail. (McGovern and Yannet, 1947). They found that 13% of patients had a convincing history of birth injury, and 16% of postnatally acquired disease. They then compared these patients in whom some presumptive cause of the hemiplegia was present to the remainder in whom no cause was evident. They found that the group with unknown aetiology showed a higher maternal age, had a smaller average head size and a greater prevalence of prematurity. The right side was more often affected than the left. They concluded that the group /
group with unknown aetiology "had many factors in common with the spastic diplegias, and probably represents the result of prenatal conditions leading to developmental cerebral defects". Other authors, before and since, however, have found a high proportion of hemiplegic patients with a history suggestive of birth injury, (Stewart, 1943; Woods, 1957). It must also be remembered that Yannet's patients were all severely mentally retarded, and therefore not a true sample of the population of cerebral palsied people suffering from hemiplegia.

**The contribution of human genetics.** The greatest contribution of human genetics to the increased knowledge of the aetiology of cerebral palsy was the elucidation of the human rhesus genotypes, first theoretically by Fisher, and later clinically by other workers, (Mollison, Mourant and Race, 1952). But study of patients from the genetic point of view has also led to the recognition of a number of rare hereditary syndromes of cerebral palsy, and the realisation that the commoner types of cerebral palsy may be the result of genetic factors in a proportion of patients. The findings of Penrose in diplegia and Batten in ataxia have been described.

Of hereditary conditions which might cause cerebral palsy, tuberous sclerosis was probably the first to be studied thoroughly from a genetic standpoint. It was shown that though the mode of inheritance varied, the majority of cases were the result either of genetic mutation or mendelian dominant inheritance, (Penrose, 1935; Penrose and Haldane, 1935). The frequency /
frequency with which cerebral palsy of non-progressive type occurs in tuberous sclerosis is difficult to assess. It is probably significant, however, that the most closely studied series, one of 29 patients, contains the highest proportion, one case of hemiplegia, one of ataxia, and two of "atonic diplegia", (Gritchley and Earl, 1932). Another condition which is associated with cerebral palsy in a significant proportion of cases is phenylketonuria, first described by Fajing in 1934. The metabolic effect, primarily an inability to metabolise phenylalanine completely, is inherited as a Mendelian recessive trait. It is almost invariably associated with mental defect, though whether this can be attributed to the defect of amino acid metabolism is not certain, (Penrose, 1935, Tredgold, 1952). Cerebral palsy most commonly diplegia, occurs in approximately 4% of patients, though many others show minor abnormalities on neurological examination, (Jervis, 1954).

Other clinical syndromes have also been recognised. "Hereditary congenital spinocerebellar ataxia accompanied by congenital cataract and oligophrenia" was reported in 14 patients in two families by Sjögren, (1950). Their findings were similar to those observed in another family by Marinesco et al, (1931). These were congenital in origin and from the neurological point of view considered of a mixture of atactic and pyramidal disorders, partly of spinal and partly of cerebral origin. Though there was a tendency to neurological deterioration in adult life, many of the symptoms remained fairly static in /
in childhood and the condition may thus be regarded as a form of cerebral palsy. Inheritance is Mendelian recessive in type.

A further syndrome of oligophrenia in association with congenital ichthyosis and spastic diplegia was later described in 28 patients by Sjögren and Larsson, (1957). Again inheritance was by a monohybrid autosomal recessive gene.

Another rather rare form of cerebral palsy which is familial and genetically determined is hereditary congenital spastic paraplegia. In some of the families in which this condition has occurred there have also been cases of hereditary progressive spinocerebellar ataxia, and others of pes cavus only (for example in those reported by Phillips, (1949), and Sutherland, (1957). In other families, hereditary congenital paraplegia appears to have occurred in relatively pure culture, as in those reported by Ginsburg, (1939).
A study of the importance of developmental malformation amongst patients suffering from congenital cerebral palsy in the present series.

It is not possible to study the likelihood of there being developmental malformation of the nervous system amongst patients suffering from cerebral palsy in the same way as the likelihood of birth injury may be studied. The clinical manifestations of developmental malformation are much less specific and definite and more difficult to assess on a quantitative basis. For example, it has been shown in a number of studies that children suffering from many malformations are more often born prematurely after abnormal parturition, and to older mothers, than are normal children. But gross abnormalities of parturition must obviously mean that there is at least a possibility, and sometimes even a probability of birth injury. Whether the abnormalities of parturition are severe or slight makes developmental malformation no more or less likely.

In a few cases there may be clinical indications that developmental malformations are likely to have been responsible for cerebral palsy. There may be a family history of malformations, or the patient himself may show malformations outwith the central nervous system. These relatively positive criteria are likely to apply in only a minority of living patients, however. The most that can be done in the majority is to compare the histories and clinical findings in a series of conditions known to be malformations (like anencephaly and cleft palate), with those of series of patients suffering from cerebral palsy. In fact it will be shown that the histories and /
and clinical findings in the present series suggest that malformations tend to be more frequent in some categories of cerebral palsy than in others.

**Direct causes of developmental malformation in the human.**

As a result of experimental work and clinical observations, a number of agents have been recognised which are liable to provoke developmental abnormalities in the unborn child.

**X-Irradiation.** Following the isolated case report of Aschenheim, (1920), more than 20 instances of microcephalic infants being delivered after the mothers had received therapeutic doses of X-irradiation, usually in the first three months of pregnancy, were collected by Zappert, (1926). He coined the term, "Roentgenogenic foetal microcephaly".

The connection between X-irradiation and congenital foetal defects was further studied by Goldstein and Murphy (1929) and Murphy, (1947). A total of 106 women who received therapeutic doses of x-ray, (or radium treatment) during pregnancy. Of the 75 children born at term, 33 were abnormal. In 10 of these factors other than radiation might possibly have caused or contributed to the foetal abnormality, (two of the mothers were syphilitic and one tuberculous). In the remaining 28, the foetal irradiation sustained during pregnancy was considered to be the cause of the abnormalities. In 23 of the women, irradiation had occurred before the fifth month of pregnancy, and before the third in at least eighteen of them. Of the 28 malformed /
malformed infants, sixteen were microcephalic, and one hydrocephalic. Unfortunately the clinical descriptions are not full enough to determine what proportion actually suffered from any of the classical syndromes of cerebral palsy. All the microcephalic children who survived appeared to have been mentally impaired, the majority being described as idiots. From other reports it seems that diplegic syndromes, and ataxia, often complicated by epilepsy, are the most frequent neurological manifestations, (Ford, 1952).

Rubella Encephalopathy. The observation by Gregg that mothers who had suffered from rubella in the early months of pregnancy tended to produce offspring with congenital cataracts and other eye deformities, was soon followed by reports of associated congenital abnormalities in other systems of the body, (Gregg, 1941, 1942, 1943; Evans, 1944; Albaugh, 1945; Carruthers, 1945; Pendargast, 1946).

By 1947, Murphy was able to collect 295 cases from the literature of congenital abnormalities which had been attributed to maternal rubella in early pregnancy. Abnormalities of the eyes were present in 161, cardiac lesions in 117, deaf mutism in 83, dental defects in 24, microcephaly in 16. In the Australian epidemic, first described by Gregg, the incidence of congenital abnormalities in the offspring was thought to be as high as 100% if the mother suffered rubella in the first two months of pregnancy, and sixty per cent if she was attacked during /
during the third month, (Swan, 1944). It soon became apparent, however, that the incidence of malformations was very much less than this in most other epidemics. Nevertheless, it is high enough for severe epidemics of rubella to be reflected by a rise in the incidence of congenital malformations, (Stevenson and Worcester, 1950).

It has been stated that "In rubella embryopathy, cerebral alterations, sensu strictiori seem to be much less frequent and pronounced than anomalies of the eyes, of the internal ear, the heart and the teeth", (Minkowski, 1952). Nevertheless, as Murphy's statistics show, the brain is affected in a significantly high proportion of cases. In the majority, mental defect, rather than cerebral palsy appears to be the presenting clinical feature, but "athetosis" and spastic paraplegia have been recorded by Beswick, Warner and Warkany, (1949), and Ford has observed cases suffering from diplegia of which he gives no details, (Ford, 1952).

Though the recognition of rubella encephalopathy led to an intensive search for other congenital malformations in the offspring of mothers attacked by other virus infections during pregnancy, there is no convincing evidence that they occur significantly often, (Haia et al, 1952; Greenley et al, 1949; Campbell, 1953; Swan, 1949).

In a sense it might be argued that congenital syphilis and congenital toxoplasmosis should be considered as being maternal infections giving rise to congenital malformations in the offspring. /
offspring. But as Minkowski points out, these conditions attack the brain at a much later stage than agents which cause the majority of major congenital cerebral malformations. They have, therefore, been included under the heading of "Paranatal toxic or infective causes of brain damage", (Minkowski, 1952).

Known causes of developmental malformations in patients suffering from congenital cerebral palsy. The only mother to have therapeutic X-irradiation during her pregnancy was in Case 15. In this patient, x-ray therapy was given for a rapid advancing carcinoma of the breast. The child appeared to be quite normal until the age of ten months, when an acute hemiplegia developed in the course of a febrile illness. In these circumstances it is difficult to believe that the x-ray therapy to the mother was of any aetiological importance in the child's hemiplegia.

Rubella is known to have occurred during pregnancy in only one mother of children with cerebral palsy in the present series, Case 17. It occurred at 4 months gestation. This child suffered from acute staphylococcal mastoiditis and complicating acute hemiplegia in infancy, after the mother had developed a staphylococcal breast abscess. It seems unlikely that the rubella was of any aetiological importance.

Influenza occurred in early pregnancy in Case 203, in which a prematurely born boy suffered from diplegia and retrolental fibroplasia.
Social factors in developmental malformations and in the patients of the present series. The evidence about the social background in children suffering from developmental malformations is conflicting. Details of the social circumstances of 494 malformed children who had been stillborn, or died in the neonatal period were studied by Murphy (1947). Twenty-two of these were born to destitute women who were illegitimately pregnant, and "married women of the same class" who came from a hospital which catered for "individuals of this type". This hospital gave a rate of 113 malformed individuals per 10,000 live and stillbirths, more than twice that in the general population studied by Murphy. Murphy considered that malformations occurred with unusual frequency in families of poor economic and social status. He thought that 40% of the mothers with malformed children had taken an unsatisfactory diet during pregnancy and anaemia was commoner than in a control group. Baird reached a similar general conclusion as a result of comparing the stillbirth and neonatal death rates attributable to malformations in a Nursing Home (Social Classes I. and II. of the Registrar General) and hospital cases (Social Classes III. IV. and V.) in Aberdeen, (Baird, 1945).

In contrast to these findings, the stillbirth and neonatal mortality attributable to congenital malformations was found to be remarkably similar in the different social classes by Woolf, (1946). This author emphasised that in this way the mortality rate from malformation differed from other causes of stillbirth and /
and neonatal death and he pointed out that this was in accordance with the finding of the Registrar General (Decennial Suppl. 1921).

Record and McKeown criticised the validity of the conclusions reached by Murphy. They studied 930 cases of stillbirth or neonatal death attributable to malformations of the nervous system in Birmingham. They failed to discover any significant difference in the stillbirth or neonatal mortality rates by Social Class or by living standards assessed by other criteria, (Record and McKeown, 1949-50).

Stillbirths and infant deaths attributed to foetal defects of all parts of the body were relatively more frequent amongst children whose fathers were in Social Class III, than those in Social Classes I. or II. and more frequent amongst those in Social Classes IV. and V. than those in Social Class III. in Scotland in 1951, (Registrar General for Scotland). Table 159 Rather less marked differences by social class in the rates of stillbirth attributed to congenital malformations were observed in England and Wales in 1950 by Heady and Heasman, (1959). The rates in social classes I. and II. was 34 per 10,000 live and stillbirths, in social class III. 43, and in IV. and V. 44. The trend, however, was similar. In spite of a relatively higher maternal age in social classes I. and II. which might be expected to result in an increase in the incidence of malformed offspring, the rate is lower than that in Classes III, IV. and V. in both the Scottish and English statistics.

Neither Murphy, nor Record and McKeown were able to find any/
any significant difference in the prevalence of developmental malformations amongst children of employed and unemployed fathers or mothers, though Woolf thought that fatal congenital malformations might be more frequent in the offspring of "Occupational groups where women do prolonged or heavy work during pregnancy", (Murphy, 1947; Record and McKeown, 1949-50; Woolf, 1945).

There is still no confirmation of Murphy's finding that developmental malformations may be frequent when the maternal diet is poor during pregnancy. Only a small, not statistically significant increase in the incidence of human developmental malformations was reported during the period of severe dietary deprivation which occurred in Holland at the end of the second World War, (Smith, 1949).

The prevalence of fatal congenital malformations does not appear to be different in illegitimate and legitimate children, (Murphy, 1947; Record and McKeown, 1949-50; Registrar General for Scotland, 1951).

Social conditions in patients suffering from cerebral palsy. The distribution by social class of patients suffering from congenital and acquired cerebral palsy is shown in Tables 160 - 161. It will be observed that there is a relative excess of congenital patients in Social Classes I. and II. and IV. and V. and an excess of acquired cases in Social Classes IV. and V. compared to all live births in counties of cities in Scotland in 1951. The differences are statistically significant. (p" being less than .01).
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</tr>
</thead>
<tbody>
<tr>
<td>Stillbirth rates</td>
<td>33</td>
<td>50</td>
<td>59</td>
</tr>
<tr>
<td>Infant deaths</td>
<td>45</td>
<td>47</td>
<td>60</td>
</tr>
<tr>
<td>Combined rates of</td>
<td>78</td>
<td>97</td>
<td>119</td>
</tr>
<tr>
<td>stillbirths and</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>infant deaths</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
TABLE 160

The distribution and approximate percentage distribution by social class of fathers of patients suffering from congenital cerebral palsy compared to that for all live and stillbirths in counties of cities.

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Congenital Hemiplegia</th>
<th>Bilateral Hemiplegia</th>
<th>Ataxic Diplegia</th>
<th>Ataxic Hemiplegia</th>
<th>Dyskinesia</th>
<th>Others</th>
<th>All cong. cases</th>
<th>All live and stillbirths in counties &amp; cities</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
<td>No.  %</td>
</tr>
<tr>
<td>I.</td>
<td>1  3</td>
<td>1  9</td>
<td>1 14</td>
<td>5 7</td>
<td>0 0</td>
<td>0 0</td>
<td>3 19</td>
<td>0 0</td>
</tr>
<tr>
<td>II.</td>
<td>3 10</td>
<td>2 18</td>
<td>0 0</td>
<td>13 17</td>
<td>0 0</td>
<td>1 11</td>
<td>4 25</td>
<td>0 0</td>
</tr>
<tr>
<td>III.</td>
<td>12 42</td>
<td>3 27</td>
<td>6 86</td>
<td>28 35</td>
<td>4 53</td>
<td>5 56</td>
<td>5 32</td>
<td>1 100</td>
</tr>
<tr>
<td>IV.</td>
<td>6 21</td>
<td>2 18</td>
<td>0 0</td>
<td>15 20</td>
<td>2 28</td>
<td>2 22</td>
<td>2 12</td>
<td>0 0</td>
</tr>
<tr>
<td>V.</td>
<td>7 23</td>
<td>3 27</td>
<td>0 0</td>
<td>17 21</td>
<td>1 14</td>
<td>1 11</td>
<td>2 12</td>
<td>0 0</td>
</tr>
<tr>
<td>Unknown</td>
<td>1 -</td>
<td>1 -</td>
<td>0 -</td>
<td>0 -</td>
<td>0 -</td>
<td>0 -</td>
<td>0 -</td>
<td>0 -</td>
</tr>
<tr>
<td>Total</td>
<td>30 100</td>
<td>12 99</td>
<td>7 100</td>
<td>73 -</td>
<td>7 100</td>
<td>9 100</td>
<td>16 100</td>
<td>1 100</td>
</tr>
</tbody>
</table>
TABLE 161

The distribution and percentage distribution of patients suffering from acquired cerebral palsy by social class

<table>
<thead>
<tr>
<th>Social Class</th>
<th>Acquired hemiplegia</th>
<th>Acquired ataxic diplegia</th>
<th>Acquired ataxia</th>
<th>Other</th>
<th>All acquired cerebral palsy</th>
<th>All live and stillbirths in counties of cities, Registrar General, 1951</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
<td>%</td>
<td>No.</td>
<td>%</td>
</tr>
<tr>
<td>I.</td>
<td>1</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>II.</td>
<td>2</td>
<td>6</td>
<td>1</td>
<td>20</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>III.</td>
<td>14</td>
<td>43</td>
<td>2</td>
<td>40</td>
<td>3</td>
<td>60</td>
</tr>
<tr>
<td>IV.</td>
<td>11</td>
<td>33</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>40</td>
</tr>
<tr>
<td>V.</td>
<td>5</td>
<td>15</td>
<td>2</td>
<td>40</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Unknown</td>
<td>0</td>
<td>-</td>
<td>0</td>
<td>-</td>
<td>1</td>
<td>-</td>
</tr>
</tbody>
</table>

Total 33 100 5 100 6 0 47 100 100.0
Unfortunately the numbers of cases in the individual categories are too small for statistically significant differences to be demonstrable except in those of congenital and acquired hemiplegia and diplegia. For acquired hemiplegia the excess of patients in social classes IV. and V. is statistically significant. For congenital hemiplegia and diplegia the excess of patients in social classes I. and II. and IV. and V. is statistically significant.

As might be expected with the relatively high proportion of fathers in social classes IV. and V. paternal unemployment and maternal employment, whole or part-time, occurred commonly, Table 162. Unfortunately, statistics for comparison giving the proportions of unemployed father and employed mothers by social class in the general population of Edinburgh could not be found. The numbers of mothers of patients in the present series doing hard manual work until late in pregnancy was striking.

Unfortunately it was not possible in the circumstances of the investigation to make special studies of maternal nutrition during pregnancy, but except in a few cases where the mothers were mentally retarded, psychotic or alcoholic, diets were probably adequate.

The proportion of illegitimate patients suffering from cerebral palsy, 14 in 208, and for congenital cerebral palsy, 11 in 160 (approx. 7%) was not dissimilar to that amongst all live /
live births in Scotland 1951. It is not significantly different from the illegitimacy rate amongst children with fatal congenital malformation of the nervous system.

<table>
<thead>
<tr>
<th></th>
<th>Father unemployed</th>
<th>Mother employed</th>
<th>Total Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Completely</td>
<td>Often</td>
<td>Part-time</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>5</td>
<td>10</td>
<td>14</td>
</tr>
<tr>
<td>Bilateral</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Diplegia</td>
<td>8</td>
<td>5</td>
<td>11</td>
</tr>
<tr>
<td>Ataxic Diplegia</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Ataxia</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
<td>17</td>
<td>28</td>
</tr>
<tr>
<td>Approx. %</td>
<td>7</td>
<td>8</td>
<td>13</td>
</tr>
</tbody>
</table>

The conclusions which can be drawn from this study are limited. The most significant finding is the marked excess of patients suffering from congenital cerebral palsy in social classes I. and II. compared to the proportion of malformed children who die during infancy or are stillborn. Table 163
in social classes IV. and V. is not strikingly different, but there is a smaller proportion of patients than of malformed children in social class III. The excess of patients in social class I. and II. is most marked in dyskinesia, hemiplegia of unknown origin and diplegia, which are the three congenital categories with the highest maternal age.

Whilst the higher proportion of elderly mothers in social classes I. and II. compared to the other classes might be considered to favour their producing malformed children, this cannot be regarded as a strong argument in favour of these forms of cerebral palsy being due to malformation. In fact, a rather greater percentage of stillbirths attributed to birth injury (12.6%) occur in social classes I. and II. in Scotland, than do stillbirths attributed to congenital malformation (9.3%) Table 163. Thus, the excess of patients in social classes I. and II. might equally well be interpreted as being due to the older mothers in these classes being more liable to produce birth injury.

The distribution of patients suffering from congenital cerebral palsy by social class is different from that of children suffering from fatal congenital malformations in Scotland 1951, or fatal malformations of the nervous system, (Record and McKeown, 1949-50). This suggests either that children with congenital malformations of the nervous system who survive to show cerebral palsy are differently distributed by social class from those who die, or that the majority of patients suffering cerebral /
cerebral palsy do not in fact suffer from developmental malformations. Obviously the latter is the more likely possibility, but it is not an argument against there being developmental malformations in a significant minority of patients.
TABLE 163

A comparison of the percentage distributions by social class of fathers of all live and stillbirths in Scotland, 1951, live and stillbirths in counties of cities, 1951, stillbirths due to congenital malformations and birth injury, and children in Edinburgh born between 1938 and 1952 who suffer from cerebral palsy. (Figures for stillbirths and births from Registrar General, 1951)

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>I. &amp; II.</td>
<td>12.5</td>
<td>21.0</td>
<td>9.3</td>
<td>12.6</td>
<td>14.2</td>
</tr>
<tr>
<td>III.</td>
<td>58.3</td>
<td>40.0</td>
<td>54.9</td>
<td>52.2</td>
<td>54.7</td>
</tr>
<tr>
<td>IV. &amp; V.</td>
<td>29.2</td>
<td>39.0</td>
<td>35.7</td>
<td>35.2</td>
<td>31.1</td>
</tr>
<tr>
<td>Total all classes</td>
<td>100.0</td>
<td>100.0</td>
<td>100.0</td>
<td>100.0</td>
<td>100.0</td>
</tr>
</tbody>
</table>

* Including deaths due to "toxaemia", "antepartum haemorrhage" "other placental and cord conditions" and "difficult labour"
The family history in cases of congenital malformation.

Since many congenital malformations are known to be inherited, the family histories of cases of congenital malformation are of interest.

Parents. It is impossible to assess the proportion of parents suffering from congenital defects who have given birth to children with malformations on the basis of published reports. Neither Murphy, nor Record and McKeown (loc. cit.) note the presence of congenital malformations in the parents of their cases. Since both papers deal with fatal malformations, however, it seems unlikely that many of the parents suffered from malformations similar to those in their dead children.

Other relatives. The difficulties in obtaining reliable information about congenital malformations in distant relatives and in obtaining a control group of cases have been discussed by Record and McKeown, (1949-50). It is impossible to present trustworthy evidence of the frequency with which they occur. Murphy found a history of malformation in other relatives in 39 of his 390 cases. In approximately 41% of them the child’s defect was duplicated in the relative. Duplication occurred three times more often in the mother’s family than in the father’s.

Eighty of 742 cases of fatal malformations of the nervous systems had a family history of other congenital malformations and of these thirty-seven involved the nervous system. The figures /
figures in a control group were respectively twenty-eight and ten, (Record and McKeown, 1949-50). The incidence of familial abnormality was higher for anencephalus and spina bifida than for hydrocephalus.

**Siblings.** A tendency for the siblings of malformed children to be malformed, often in a similar manner to the propositus has been noted by a number of authors, (Macklin, 1936). Penrose studied 1041 siblings of malformed children of whom 454 were the brothers or sisters of propositi with abnormalities of the central nervous system. Of them, 10, or approximately 2.3% were malformed, and 7 had defects of the central nervous system, (approx. 1.5%). In five cases the original malformation was reproduced.

In 294 fraternities which contained at least one malformed child, an incidence of recurrent deformity (not necessarily the same as the original deformity) amongst the siblings was found in 2.1% of cases by Malpas (1937). For anencephalus the incidence of malformations in other siblings was 2.6%.

Murphy found that when one malformed child had been born in a family, the chances of further malformations occurring were about one in every eight subsequent pregnancies. In 57.5% of his forty cases of fraternal malformations, the abnormalities were repeated. He concluded that there was a "very strong tendency for identical congenital defects to affect brothers and sisters. This appears to be the case regardless /
regardless of the type or seriousness of the malformation”.

The findings of Record and McKeown are similar in their series of neurological malformations. They found that 1.89% of the siblings of malformed propositi showed abnormalities of the nervous system compared to 0.29% in a control group. There was also a significantly increased risk of other types of malformations, including cardiac defects and pyloric stenosis) in the siblings after the birth of a child with abnormalities of the central nervous system, (Record and McKeown, 1949-50).

**Consanguinity.** Malpas received 109 replies relating to consanguinity from the parents of 294 malformed children. In none was a history of cousin marriage obtained, (Malpas, 1937). Murphy obtained a history of consanguinous marriage in 1.3% of 553 families of children with congenital malformations. Amongst 742 families in which congenital malformation of the nervous system had occurred, four, 0.54% parents were consanguinous, compared to 0.13% in a control group, (Record and McKeown 1949-50). As has been pointed out by Hogben, however, the incidence of consanguinity is of little importance when the gene frequencies being considered are relatively high. They are high in many congenital malformations of the nervous system (Roberts 1959).

**Consideration of the family histories of patients suffering from congenital cerebral palsy.**

**Family history of cerebral palsy.** A reliable history of /
of cerebral palsy occurring in parents, siblings, grandparents, uncles, aunts, or cousins was obtained in 16 cases, or 10% of the patients suffering from congenital cerebral palsy (including patients with hemiplegia of unknown origin). In two instances, two children who were siblings were included in the series, (Cases 98 and 199 and Cases 158 and 159. Tables 240 - 244).

Since the number of relatives other than parents and siblings was unknown the prevalence of cerebral palsy amongst them could not be calculated. A history of cerebral palsy amongst them was given in 5% of the cases. Three parents suffered from cerebral palsy. The father of a diplegia child was hemiplegic, and two mothers of daughters with ataxic diplegia showed the same condition themselves. Thus, approximately 1.8% of cases had a parent who suffered from cerebral palsy, and 0.9% of parents were known to be affected. Approximately 4.4% of the siblings of patients suffered from cerebral palsy, or if the four patients who each had a sibling in the series are counted as one case, 3.2% of siblings were affected. This is about fifteen times the prevalence of cerebral palsy amongst the children of Edinburgh, as determined during the survey, and almost twenty times the prevalence of congenital cerebral palsy.

Cerebral palsy of the type shown by the patient was present in a relative in six cases (including cases 158 and 159). It was of different type in eight (including cases 98 and 199). The precise type of cerebral palsy was unknown in two instances.
All four patients suffering from hemiplegia who had a positive family history had relatives with diplegia. Two of the seven diplegic patients with a positive family history had relatives with hemiplegia, but in the other five the cerebral palsy in relatives was diplegic in type. Of the three cases of ataxic diplegic with a positive family history, one had a hemiplegic sibling, and in the other two there was a family history of ataxic diplegia.

The relationship of the affected relatives to the patients is important. Of the four hemiplegic patients with a family history of cerebral palsy, siblings or a cousin were diplegic in three. Of the seven diplegic patients, four had a sibling or a cousin who was diplegic, and one was hemiplegic. The father of a diplegic child in case 47 was hemiplegic, but it is apparent from the tables that in hemiplegia and diplegia, cerebral palsy tends to affect the family tree in a horizontal, rather than a vertical direction, when there is a positive family history. That this is so, immediately suggests that diplegia and hemiplegia may be the result of a Mendelian recessive gene in a proportion of cases, but conclusive evidence of this cannot be shown as a result of the present investigation. Environmental factors shared by siblings and to a lesser extent, cousins, have also to be taken into consideration, though the findings are much more easily interpreted on a genetic basis.

The findings in ataxic diplegia are strikingly different, for
### TABLE 240

**Familial Cerebral Palsy (Parents and Siblings)**

<table>
<thead>
<tr>
<th>Parent</th>
<th>Sibling</th>
<th>Other relatives</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 29</td>
<td>Cong. Hemiplegia. (Father epileptic)</td>
<td>Maternal grandmother diplegic.</td>
</tr>
<tr>
<td>&quot; 201 &quot;</td>
<td>&quot; &quot;</td>
<td>Mother's sister's child diplegic.</td>
</tr>
<tr>
<td>&quot; 39 &quot;</td>
<td>&quot; &quot;</td>
<td>1 diplegia</td>
</tr>
<tr>
<td>&quot; 199 &quot;</td>
<td>Unknown hemiplegia. (Mother feeble-minded)</td>
<td>F's uncle cerebral palsy unknown type.</td>
</tr>
<tr>
<td>&quot; 1 &quot;</td>
<td>Bilateral &quot;</td>
<td>F's sister's boy hemiplegic.</td>
</tr>
<tr>
<td>&quot; 10 &quot;</td>
<td>Diplegia</td>
<td>M's sister's son diplegic.</td>
</tr>
<tr>
<td>&quot; 177 &quot;</td>
<td>Diplegia</td>
<td>M's brother's son diplegic.</td>
</tr>
<tr>
<td>&quot; 163 &quot;</td>
<td>Diplegia</td>
<td>F's cousin probably diplegic.</td>
</tr>
<tr>
<td>&quot; 47 &quot;</td>
<td>Diplegia</td>
<td>M's brother mentally defective.</td>
</tr>
<tr>
<td>&quot; 98 &quot;</td>
<td>Diplegia (both feeble-minded)</td>
<td>Maternal uncle mentally defective.</td>
</tr>
<tr>
<td>&quot; 158 ) Diplegic twins (mother feeble-minded)</td>
<td>1 half-sister hemiplegic</td>
<td></td>
</tr>
<tr>
<td>&quot; 159 )</td>
<td></td>
<td>Mother's two cousins cerebral palsy.</td>
</tr>
<tr>
<td>&quot; 14 &quot;</td>
<td>Ataxic diplegia.</td>
<td></td>
</tr>
<tr>
<td>&quot; 101 &quot;</td>
<td>Ataxic diplegia.</td>
<td>Mother's two cousins cerebral palsy.</td>
</tr>
<tr>
<td>&quot; 82 &quot;</td>
<td>Ataxia.</td>
<td></td>
</tr>
</tbody>
</table>

*In the series*
for in two of the three cases with a positive family history, the mother was affected, and in one of these a sibling showed the same condition. The evidence is strongly suggestive of a Mendelian dominant form of inheritance in at least a proportion of patients with ataxic diplegia. This is perhaps hardly surprising when the rather close relationship between this syndrome and some forms of progressive spinocerebellar degeneration is recalled.

The fact that in this series, patients with hemiplegia have relatives with diplegia, diplegic patients relatives with hemiplegia, and patients with bilateral hemiplegia and ataxic diplegia relatives with hemiplegia, required comment.

It seems unlikely that the association of these conditions in families is accidental in view of the high prevalence of cerebral palsy shown. That the clinical pictures in relatives are so different is less disturbing if the underlying neuropathology is considered. It is extremely diverse in type in both diplegia and hemiplegia, and yet many of the appearances are remarkably similar in both conditions. For example, atrophic lobar sclerosis, porencephaly, hemisphere atrophy may be found quite commonly in both conditions, (Stewart, 1942-43; 1943). It seems at least conceivable that differences in gene penetrance, intraterne environment, the effects of birth trauma and possibly other unknown factors may modify considerably both the extent and severity of brain damage and the resultant clinical picture. /
**TABLE 241**

**Familial Cerebral Palsy, Congenital cases**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Total cases</th>
<th>Number with positive family history</th>
<th>Parent</th>
<th>Sibling</th>
<th>Distant relatives</th>
<th>Cousin</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia.</td>
<td>30</td>
<td>3</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Hemiplegia unknown origin.</td>
<td>11</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>7</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Diplegia</td>
<td>76*</td>
<td>6*</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>10</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ataxia</td>
<td>9</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>16</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Others</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Totals</strong></td>
<td><strong>160</strong></td>
<td><strong>15</strong></td>
<td><strong>3</strong></td>
<td><strong>5</strong></td>
<td><strong>4</strong></td>
<td><strong>9</strong></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Number of cases</th>
<th>160</th>
<th>318</th>
<th>242</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>% with cerebral palsy</td>
<td>.94</td>
<td>2.07</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>% of cases</td>
<td>100</td>
<td>1.88</td>
<td>3.10</td>
<td>5.00</td>
</tr>
</tbody>
</table>

* including diplegic twins 153/159 as one case
resultant clinical picture.

Other neurological and mental abnormalities. A rather high proportion of siblings, parents, uncles, aunts, cousins and grandparents suffered from epilepsy, neurological diseases, or were mentally retarded. The abnormalities encountered in the present series are shown in Tables 240-244.

Epilepsy. Epilepsy was present in 1.86% of parents and in 2.06% of siblings. 4.31% of patients had more distant relations who were stated by their parents to be epileptic. The frequency of epilepsy amongst siblings and parents appears to be in excess of that expected in the community at large, though figures for Edinburgh are not available for comparison. Assuming that the frequency of adults with recurrent convulsions in adults is in the region of one in 200, (Chao, Druckman and Kellaway, 1958), parents of children with cerebral palsy have epilepsy about four times as frequently as one would expect. Assuming that children with febrile convulsions (not idiopathic epilepsy) have been efficiently excluded from the present series, the excess of siblings with recurrent convulsions is probably of the same order. The numbers of parents and siblings with epilepsy in each category are too small for comparison of its prevalence to be of value.

Mental retardation. Mental retardation was generally assessed on the basis of the individual's inability to keep his /
### TABLE 242

<table>
<thead>
<tr>
<th>Other relatives</th>
<th>Congenital Hemiplegia</th>
<th>Unknown Hemiplegia</th>
<th>Bilateral Hemiplegia</th>
<th>Diplegia</th>
<th>Ataxia 10</th>
<th>Ataxia 9</th>
<th>Dyskinesia</th>
<th>Others</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total cases</td>
<td>30</td>
<td>11</td>
<td>7</td>
<td>76</td>
<td>10</td>
<td>9</td>
<td>16</td>
<td>1</td>
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<tr>
<td>Cerebral palsy</td>
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<td>0</td>
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<td>2</td>
<td>1</td>
<td>2</td>
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<td>0</td>
<td>8</td>
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<td>Epilepsy</td>
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<td>0</td>
<td>1</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Mental retardation</td>
<td>2</td>
<td>0</td>
<td>3</td>
<td>5</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>12</td>
</tr>
<tr>
<td>Congenital malformation</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Other neurological disorders</td>
<td>0</td>
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<td>0</td>
<td>2</td>
<td>0</td>
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<td>1</td>
<td>0</td>
<td>3</td>
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<tr>
<td>Total of patients with abnormal relatives</td>
<td>4</td>
<td>0</td>
<td>3</td>
<td>13</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
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</tr>
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</table>

### % of cases with affected relatives

<table>
<thead>
<tr>
<th>Other relatives</th>
<th>% of cases with affected relatives</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral palsy</td>
<td>6.66</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>3.33</td>
</tr>
<tr>
<td>Mental retardation</td>
<td>6.66</td>
</tr>
<tr>
<td>Congenital malformation</td>
<td>0</td>
</tr>
<tr>
<td>Other neurological disorders</td>
<td>0</td>
</tr>
</tbody>
</table>

### % of patients with abnormal relatives

<table>
<thead>
<tr>
<th>Other relatives</th>
<th>% of patients with abnormal relatives</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cerebral palsy</td>
<td>13.32</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>0</td>
</tr>
<tr>
<td>Mental retardation</td>
<td>42.6</td>
</tr>
<tr>
<td>Congenital malformation</td>
<td>17.03</td>
</tr>
<tr>
<td>Other neurological disorders</td>
<td>30.0</td>
</tr>
<tr>
<td>Others</td>
<td>22.0</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>6.25</td>
</tr>
<tr>
<td>Total</td>
<td>17.01</td>
</tr>
</tbody>
</table>
### Table 243

<table>
<thead>
<tr>
<th></th>
<th>Congenital Hemiplegia</th>
<th>Unknown Hemiplegia</th>
<th>Bilateral Hemiplegia</th>
<th>Diplegia</th>
<th>Ataxia</th>
<th>Ataxia</th>
<th>Dyskinesia</th>
<th>Others</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Number of siblings</strong></td>
<td>43</td>
<td>26</td>
<td>5</td>
<td>109</td>
<td>21</td>
<td>13</td>
<td>18</td>
<td>7</td>
<td>242</td>
</tr>
<tr>
<td><strong>Cerebral Palsy</strong></td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td><strong>Epilepsy</strong></td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td><strong>Mental retardation</strong></td>
<td>7</td>
<td>1</td>
<td>0</td>
<td>11</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<td>19</td>
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<td><strong>Congenital malformation</strong></td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td><strong>Other Congen. malform.</strong></td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td><strong>Total siblings abnormal.</strong></td>
<td>8</td>
<td>4</td>
<td>1</td>
<td>17</td>
<td>3</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>36</td>
</tr>
<tr>
<td><strong>Total cases with abnormal siblings.</strong></td>
<td>6</td>
<td>3</td>
<td>1</td>
<td>12</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>26</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>% of siblings abnormal</th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cerebral Palsy</strong></td>
<td>2.32</td>
<td>3.84</td>
<td>0</td>
<td>1.83</td>
<td>9.15</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>2.48</td>
</tr>
<tr>
<td><strong>Epilepsy</strong></td>
<td>0</td>
<td>3.84</td>
<td>0</td>
<td>1.83</td>
<td>4.75</td>
<td>7.69</td>
<td>0</td>
<td>0</td>
<td>2.06</td>
</tr>
<tr>
<td><strong>Mental defect or retardation</strong></td>
<td>16.24</td>
<td>3.84</td>
<td>0</td>
<td>10.03</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7.35</td>
</tr>
<tr>
<td><strong>Congenital malformation</strong></td>
<td>0</td>
<td>3.84</td>
<td>0</td>
<td>0</td>
<td>7.59</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1.65</td>
</tr>
<tr>
<td><strong>CNS</strong></td>
<td>0</td>
<td>0</td>
<td>20</td>
<td>1.83</td>
<td>0</td>
<td>7.69</td>
<td>0</td>
<td>0</td>
<td>1.65</td>
</tr>
<tr>
<td><strong>Other Congen. malform.</strong></td>
<td>0</td>
<td>0</td>
<td>20</td>
<td>1.83</td>
<td>0</td>
<td>7.69</td>
<td>0</td>
<td>0</td>
<td>1.65</td>
</tr>
<tr>
<td><strong>% of abnormal siblings.</strong></td>
<td>18.56</td>
<td>15.36</td>
<td>20</td>
<td>15.57</td>
<td>14.25</td>
<td>23.07</td>
<td>0</td>
<td>0</td>
<td>14.36</td>
</tr>
</tbody>
</table>
### Table 26

<table>
<thead>
<tr>
<th>Family History</th>
<th>Cerebral Palsy</th>
<th>Epilepsy</th>
<th>Mental Retardation</th>
<th>Other Neurological Disorders</th>
<th>Congenital Malformations</th>
<th>Total Parents</th>
<th>% of Parents Abnormal</th>
<th>Total Cases with Abnormal Parents</th>
<th>% of Cases with Abnormal Parents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>60</td>
<td>3</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>96</td>
<td>5.01</td>
<td>5</td>
<td>15.03</td>
</tr>
<tr>
<td>History</td>
<td>11</td>
<td>14</td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>23</td>
<td>6.92</td>
<td>0</td>
<td>0.93</td>
</tr>
<tr>
<td>Hemiplasia</td>
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<td>18</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>32</td>
<td>6.25</td>
<td>0</td>
<td>0.93</td>
</tr>
<tr>
<td>Total</td>
<td>160</td>
<td>318</td>
<td>3</td>
<td>5</td>
<td>0</td>
<td>36</td>
<td>10.00</td>
<td>0</td>
<td>1.36</td>
</tr>
</tbody>
</table>

*Note: The table shows the distribution of various neurological disorders and their associated parents with abnormal cases.*
his place in a normal school on account of mental handicap. It occurred amongst 10.24% of parents, 3.04% of siblings and more distant relatives were affected in 7.64% of cases in the present series. Feeble mindedness was very much more frequent than frank mental defect. Though the estimate of the number of siblings affected is probably artificially low because of the difficulties of detecting anything but marked mental defect in preschool children, the percentage affected is still almost five times that to be expected in children of school age in the general population. Only 1 in 35 Edinburgh children require special educational provision on account of mental handicaps. (Edinburgh Education Committee Report, 1956)

It is noteworthy that the parents suffering from mental retardation were almost exclusively in Social Classes IV and V. and that the great majority of regarded siblings also occurred in these groups. Possibly the fact that they were of low intelligence determined their economic state. It is, however, impossible to determine whether the parents were liable to have children suffering from cerebral palsy because of their genetic constitution, their social background, or whether both genetic and environmental factors were involved.

An excess of individuals with some degree of mental retardation is to be expected in any population group consisting largely of Social Classes IV. and V. and it is very difficult to determine when mental retardation should be regarded merely as a mild variant of the norm, or as definitely /
definitely pathological. The fact that a rather small proportion of children than parents were mentally retarded suggests that there may have been a swing-back towards the norm of intelligence amongst the offspring of these parents, as would be expected in any group of parents of rather below average intelligence. Certainly this finding is against there being a Mendelian recessive form of inheritance of mental retardation in any large proportion of cases. This is important in diplegia for Penrose suggested that in some cases mental defect and diplegia might be different expressions of the same Mendelian recessive gene, (Penrose, 1937).

In view of the probability that the majority of those classified as mentally retarded represent deviation from the normal average of intelligence, rather than being children suffering from gross neuropathological abnormality, mental retardation has not been considered a neurological abnormality in what follows.

Other neurological abnormalities. These were recorded in more distant relatives of two diplegic patients, (Case 40 in which neurofibromatosis occurred, and Case 118 in which there was a history of hydrocephalus). Thus, 1.89% of cases of congenital cerebral palsy had a history of more distant relatives with neurological abnormality.

Two siblings, 0.83% had other neurological abnormalities. One brother of a child with hemiplegia died of hydrocephalus, and /
and a sibling of an ataxic patient had a short weak arm.

Parents had other neurological abnormalities in five cases, a prevalence amongst them of 1.5%. The defects were tuberous sclerosis in the father of a hemiplegic child (Case 162), otosclerosis, disseminated sclerosis and acquired hemiplegia in the parents of diplegic patients. The mother of a child with dyskinesia suffered from Bell's palsy, with residual paresis. Apart from the cases of tuberous sclerosis these disorders do not appear to be of great aetiological importance.

Other congenital anomalies not affecting the nervous system and which did not result in death before the age of one year were recorded amongst distant relatives in one case in which two maternal uncles had died with multiple congenital malformations (Case 174). No parents were recorded as having significant congenital malformations.

Four siblings (1.6%) had congenital malformations. One sibling of a hemiplegic child had bilateral dislocations of the hips (Case 137). Two patients with diplegia had siblings with fragilitas ossium (Case 22) and congenital heart disease (Case 128) respectively. In Case 28, a sibling of a child with ataxia died of multiple congenital malformations.

The frequency of apparent congenital malformations amongst siblings who survived more than one year is high, especially when it is probable that a large number of minor malformations such /
Buch as partial syndactyly, hypomanibulosis, hypertelorism were unaccounted. These could only have been accurately diagnosed by the routine detailed examination of every sibling. This would have been impossible in the conditions of the survey. Abortions, stillbirths, and infant deaths. Any attempt to assess the prevalence of cerebral palsy, neurological abnormalities and developmental malformations amongst the relatives of patients must take account of those who were aborted, stillborn or died in infancy, for it is probable that a proportion meeting these fates were affected. In the present investigation it was possible to study only the stillbirths and infant deaths amongst siblings. The prevalence of abnormalities amongst the pregnancies of mothers of children suffering from cerebral palsy was 7.6%, probably not very dissimilar from that in pregnancies of women in the general population. The prevalence of abortions amongst the pregnant women is unknown. The abortions have already been discussed from the obstetric point of view, and since hardly any of the aborted embryos were examined pathologically, the nature of their defects if any remain unknown. The prevalence of abnormalities amongst the stillbirths and infant deaths amongst siblings, if any, is unknown. Since not all stillbirths or infants who died came to autopsy, it is difficult to determine the frequency with which the stillbirth and infant deaths were due to developmental malformations, or damage to the central nervous system. Without autopsy, malformations tend to be missed, and deaths attributed to "convulsions" or "prematurity".
"prematurity", (Drillien, 1947). However, the histories of the stillbirths and neonatal deaths are summarised in Table 245. It will be observed that in addition to the seven children who died in convulsions, 4 of the 29 children (about 14%) who were stillborn, or died in infancy were considered to have abnormalities of the central nervous system. This figure is similar to that found by Worcester, Stevenson and Rice, (1950). But in the present series it is almost certainly a considerable underestimate in view of the fact that so few autopsies were performed.

Consideration of all live and stillborn siblings.

Siblings with evidence indicative of, or suggesting the presence of abnormalities in the nervous system or congenital anomalies elsewhere are indicated in Table 243. It is possible that some of the children who were stillborn or died in infancy may have been suffering from cerebral palsy similar to that shown by the patients, but in other cases the abnormalities appear to be very dissimilar.

It will be observed (Table 245) that the proportions of abnormal siblings is relatively high (6.5%), even when those with mental retardation only are excluded from consideration. Moreover, though the numbers of abnormal siblings in the individual categories are too small for statistical comparison to be possible, it is interesting to observe that there are considerable differences in the proportions which are abnormal.
<table>
<thead>
<tr>
<th>Diagnosis of patient</th>
<th>Case No.</th>
<th>Classification</th>
<th>Cause of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia</td>
<td>5</td>
<td>Stillbirth.</td>
<td>Prematurity.</td>
</tr>
<tr>
<td></td>
<td>70</td>
<td>Stillbirth.</td>
<td>&quot;Birth injury.&quot;</td>
</tr>
<tr>
<td></td>
<td>80</td>
<td>Stillbirth.</td>
<td>&quot;Birth injury.&quot;</td>
</tr>
<tr>
<td></td>
<td>109</td>
<td>Stillbirth.</td>
<td>Prematurity.</td>
</tr>
<tr>
<td></td>
<td>171</td>
<td>Stillbirth.</td>
<td>Erythroblastosis.</td>
</tr>
<tr>
<td></td>
<td>171</td>
<td>Stillbirth.</td>
<td>Erythroblastosis.</td>
</tr>
<tr>
<td></td>
<td>34</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td>Hemiplegia of unknown origin</td>
<td>134</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>134</td>
<td>Postneonatal death.</td>
<td>Endocardial fibrolastosis.</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Diplegia</td>
<td>34</td>
<td>Stillbirth.</td>
<td>Birth injury. (Placenta previa)</td>
</tr>
<tr>
<td></td>
<td>41</td>
<td>Neonatal death.</td>
<td>Spina bifida.</td>
</tr>
<tr>
<td></td>
<td>41</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>41</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>42</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>174</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>105</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>76</td>
<td>Postneonatal death.</td>
<td>&quot;Diphtheria&quot; Convulsions.</td>
</tr>
<tr>
<td></td>
<td>157</td>
<td>Postneonatal death.</td>
<td>Bronchopneumonia, convulsion</td>
</tr>
<tr>
<td></td>
<td>157</td>
<td>Postneonatal death.</td>
<td>Bronchopneumonia, convulsion</td>
</tr>
<tr>
<td></td>
<td>161</td>
<td>Postneonatal death.</td>
<td>Post vaccinal encephalitis, convulsions.</td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>57</td>
<td>Stillbirth.</td>
<td>Unknown.</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Neonatal death.</td>
<td>Erythroblastosis.</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>Neonatal death.</td>
<td>Erythroblastosis.</td>
</tr>
<tr>
<td></td>
<td>57</td>
<td>Postneonatal death.</td>
<td>Tuberculous meningitis.</td>
</tr>
<tr>
<td>Ataxia</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>54</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>54</td>
<td>Neonatal death.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>145</td>
<td>Postneonatal death.</td>
<td>Respiratory infection.</td>
</tr>
<tr>
<td></td>
<td>197</td>
<td>Postneonatal death.</td>
<td>Respiratory infection.</td>
</tr>
<tr>
<td>Other</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>
## TABLE 245a

<table>
<thead>
<tr>
<th>Congenital malformations and neurological disorders in the siblings of patients suffering from congenital cerebral palsy.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Congenital hemiplegia</strong></td>
</tr>
<tr>
<td>No. of cases</td>
</tr>
<tr>
<td>Total no. of conceptions</td>
</tr>
<tr>
<td>Abortions</td>
</tr>
<tr>
<td>Live and stillborn babies other than patients.</td>
</tr>
</tbody>
</table>

Stillbirths and infant deaths attrib. to malformations.
- 1            | 0             | 0             | 2            | 0            | 0            | 0            | 0            | 0          | 0.3        |
- 1            | 0             | 0             | 2            | 0            | 0            | 0            | 0            | 0          | 0.7        |

Infant deaths due to convulsions.
- 0            | 1             | 0             | 6            | 0            | 0            | 0            | 0            | 0          | 0.7        |

Siblings congen. cerebral palsy.
- 1            | 1             | 0             | 2            | 2            | 0            | 2            | 0            | 0          | 0.6        |

Sibs. with malformed nervous systems.
- 0            | 1             | 0             | 0            | 0            | 1            | 0            | 0            | 0          | 0.6        |

Sibs. with other serious malform.
- 0            | 0             | 1             | 2            | 0            | 1            | 0            | 0            | 0          | 0.4        |

Epileptic siblings.
- 0            | 1             | 0             | 2            | 1            | 1            | 0            | 0            | 0          | 0.5        |

Total with malform or neurological disease.
- 2            | 4             | 1             | 14           | 3            | 3            | 0            | 0            | 27         | 0.6%       |

Approx. % of other live and stillbirths with malform. or neurol. disease of live and stillbirths.
- 4            | 14.2          | 40            | 7.1          | 28.6         | 23           | 0            | 0            | 0          | 6.5%       |
The two categories with the lowest proportion of abnormal siblings, dyskinesia and congenital hemiplegia, are those in which the strongest evidence of birth injury being important was found. The categories of bilateral hemiplegia, ataxic diplegia, diplegia in which birth injury was thought to be less important, showed higher proportions of abnormal siblings. The highest proportion of abnormal siblings was found in ataxia, in which birth injury was felt to be of intermediate importance, though in view of the very small number of cases too much importance cannot be attached to this finding.

That the siblings are not abnormal as a result of birth injury is suggested by the finding that abnormal parturition occurs with equal frequency in normal and abnormal siblings. Whether they tend to be abnormal because of genetic traits inherited by their parents, because of a high mutation rate (associated with high maternal age in some cases), or because of environmental influences at an early stage of intrauterine life cannot be determined on the basis of the present study. Nevertheless, the finding that there is an excess of siblings with malformations in these categories does favour the postulate that a proportion of the patients themselves may also be suffering from malformations rather than birth injuries.

Consanguinity of parents. The cases in which consanguinity was known, and in which it was suspected are shown in Table 246. Unfortunately the findings are of no real significance.
<table>
<thead>
<tr>
<th>Consanguinity</th>
<th>Known</th>
<th>Case</th>
<th>Hemiplegia</th>
<th>Bilateral hemiplegia</th>
<th>Double hemiplegia</th>
<th>Congenital hemiplegia of unknown origin</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>0</td>
<td>16</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>0</td>
<td>9</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>0</td>
<td>10</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>2</td>
<td>7</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>0</td>
<td>7</td>
<td></td>
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<tr>
<td>1</td>
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</tr>
</tbody>
</table>

Consanguinity in cerebral palsy

**TABLE 246**
significance. The types of cerebral palsy in which there are enough cases available for study are those with a relatively high prevalence in the community. Assuming that diplegia is a condition inherited as a result of an autosomal Mendelian recessive gene, as suggested by Penrose and that 1 in 1000 children in the community suffer from it, then approximately one of every 15 individuals in the population must carry the gene heterozygously, (Penrose, 1938; Roberts 1959). For congenital hemiplegia, about 1 in every 25 of the population would be heterozygous, assuming that the condition is due to an autosomal Mendelian recessive gene and that it occurred once in every 2,500 children. In these circumstances it would require a huge excess of consanguineous parents in either of these conditions for the findings to be of aetiological significance. In fact the excess is relatively small. In the other types of cerebral palsy the numbers of cases are too small for statistical analysis.
A number of studies have shown that mothers producing offspring suffering from congenital malformations tend to show certain characteristics of age and reproductive history. These will be described briefly and then the mothers of patients in the series will be studied to see to what extent they conform to the pattern of those with offspring known to be malformed.

**Maternal age in congenital malformations.** In most of the studied series of mothers who have produced children suffering from developmental malformations it has been found that the maternal age at the time of the birth of the offspring has been relatively high, (Landtmann, 1948). The incidence of anencephalus amongst offspring was found to be ten times higher amongst women in the maternal age group 46-50 than in the age group 16-20 by Malpas, (1937). The same author also found that there was an increased frequency of hydrocephalus amongst the children of elderly mothers.

In a study of the maternal age in 570 cases of fatal congenital malformations in Philadelphia, Murphy found that the percentage of malformed children born at any given maternal age over 30 increased significantly in each subsequent five-year period. The proportion of malformed offspring born to mothers in the age group 40-50 was three times as that observed amongst those whose mothers were under thirty years.
years of age. The age of fathers did not appear to have any relationship to the frequency of malformations, (Murphy, 1957).

Record and McKeown were at pains to separate the effects of high parity from those of maternal age in their study of fatal developmental malformations of the nervous system, for other studies had shown that as well as being more frequent amongst the children of older mothers, congenital malformations were also more often encountered in children of high birth rank. They found that maternal age appeared to be of much greater importance in some malformations than in others. (Record and McKeown, 1949-50).

Their findings tended to confirm the observations made by the Registrar General for Scotland on anencephaly and the more detailed studies of Penrose on the significance of high maternal age in Mongolism, (Penrose, 1934). In the latter paper the relative importance of high maternal age and hereditary genetic factors in the causation of mongolism are discussed in a way which still provides something of a revaluation of the way in which genetic theory can help to elucidate clinical problems. Penrose noted that a positive family history was more often encountered when mongols were born to younger mothers than when maternal age was high. His subsequent postulate that the offspring of older mothers might show a higher incidence of gene mutation, has been confirmed by subsequent chromosome studies.

Maternal parity and developmental malformations. In most /
most series of cases of developmental malformations the high maternal parity has been thought to be of less aetiological importance than relatively high maternal age, but both factors are associated with an increase in the incidence of malformed offspring. In small series of cases it may be impossible to separate the effects of the two factors, and in some larger series the authors have not attempted to assess their relative importance, (Malpas, 1937; Murphy, 1947).

Penrose found that there was a greater tendency for first born and later born children to be affected by mongolism and also for later born children to suffer from anencephaly, spina bifida and congenital hydrocephalus, even allowing for the effect of maternal age, (Penrose, 1934, 1946).

In a study of 160 cases of spina bifida the first three birth ranks were noted to contain fewer cases than later birth ranks even when the factors of maternal was taken into consideration by Hindes-Nielsen, (1933).

In the study of Record and McKeown it was observed that there was increased risk of fatal congenital malformation of the nervous system at first parity, and thereafter above the sixth parity. (These authors took abortions into account when calculating parity).

Reproductive efficiency of mothers with malformed children. Murphy reviewed 405 families in which there had been one child with fatal developmental malformations and one or /
or more other children. He found that of the total of 1732 conceptions, only 1094 (63.2%) resulted in the birth of living children who later showed no abnormality. There were 405 malformed offspring and 233 conceptions which ended in abortion, premature birth or stillbirth. The disturbances of pregnancy which occurred were noted significantly more frequently in positions close to that which resulted in the birth of the malformed child, and were especially liable to occur immediately before it. Of the intervals between pregnancies he found that the one immediately preceding the birth of the malformed child was longest, (Murphy, 1947).

A high abortion rate in mothers of mongols was also reported by Smith and Record (1955).

Record and McKeown found that mothers giving birth to children with fatal developmental malformations of the nervous system had a higher abortion rate than mothers in a control series. Mothers with children suffering from anencephaly and hydrocephalus had a significantly higher stillbirth rate though their infant mortality rates of their offspring were not significantly different from that of the controls. They also observed that mothers appeared to be slightly less fertile before the birth of the affected child and that the immediately preceding pregnancy had a greater chance of being lost as a result of abortion, stillbirth or infant death than did other conceptions. This was especially likely to be so if/
if the developmental malformation was hydrocephalus. They were highly critical of Murphy's interpretations of his data, however, and suggested that the apparent period of relative infertility before the birth of the affected offspring had not the significance which Murphy thought, (Record and McKeown, 1949-50).

Maternal health before the birth of the malformed child. No correlation between the occurrence of chronic maternal diseases, including syphilis, and malformations amongst offspring could be found by Worcester, (1950), Murphy, (1947), Record and McKeown, (1949-50). No significant increase in the incidence of menstrual abnormalities was found in a series of over 600 mothers giving birth to malformed children by Murphy, nor in their series of mothers with offspring showing fatal malformations of the nervous system by Record and McKeown, (1949-50).

Disorders of parturition and developmental malformations of the offspring. There have been a number of studies of the pregnancies, labours and deliveries resulting in the births of series of children suffering from fatal and non-fatal malformations of the nervous system and other parts of the body.

42.5% of 73 mothers of malformed children were found to have suffered from "morbid states" during pregnancy by Landtmann, (1943). 12% suffered from acute infections in early pregnancy, and 15% from chronic diseases of various types /
types. Diets were thought to have been adequate in all cases, but Murphy thought as a result of his study that maternal diets had been inadequate in approximately 40% of the mothers with children who died as a result of malformations. Diets were thought to have been adequate in the mothers whose children suffered from fatal malformations of the nervous system studied by Record and McKeown, (1949-50). These authors criticised Murphy's conclusions. No statistically significant increase in the incidence of congenital malformations was noted in war-time Holland at a time when maternal diets during pregnancy were demonstrably inadequate by Smith (1949).

Polyhydramnios was noted in 10.4% of mothers whose offspring suffered from fatal and non-fatal malformations by Worcester et al (1950). 51% of mothers giving birth to anencephalic infants had shown polyhydramnios in 119 of 526 pregnancies resulting in the birth of malformed children were similarly complicated in the series studied by Murphy (1947).

Threatened abortion occurred in 13% of the pregnancies studied by Worcester et al. Unfortunately they had no adequate control series with which to compare this finding. Threatened abortion occurred slightly more commonly in pregnancies resulting in the birth of anencephalic infants than in a control series. But threatened abortion was not more frequent in pregnancies resulting in the births of children with /
with other fatal congenital malformations of the nervous system in the cases studied by Record and McKeown (1949–50). Most authors do not distinguish between threatened abortion and antepartum haemorrhage in presenting their findings. Vaginal haemorrhage occurred at some time during pregnancy in 47 (approximately 9%) of the 526 pregnancies studied by Murphy, and in 10.5% of cases (three times the expected prevalence) in which the offspring suffered from malformations studied by Drillien (1947). Antepartum haemorrhage was found significantly more frequently amongst pregnancies resulting in malformed infants than amongst controls by Landtmann (1943).

Greenhill analysed the records of 4446 cases of placenta previa and noted that 2.5% of the offspring showed developmental malformations. He postulated that there was an association between placenta previa and the occurrence of malformations in the child, (Greenhill, 1923, 1939). A similar association was thought to be present by Potter and Adair (1943). Murphy was unable to find any evidence that this was so in his series. There was a slight excess of mothers with placenta previa who gave birth to anencephalic infants in the series of Record and McKeown, but mothers with children dying from other malformations suffered from placenta previa no more frequently than expected.

Women with hypertension and albuminuria were not found to have children with severe congenital malformations any more frequently /
frequently than those without by Murphy (1947). No excess of toxaemic mothers giving birth to malformed children was found by Drillien (1947), Landtmann (1948), or Record and McKeown (1949-50). A "slight excess" of toxaemia amongst hospitalised mothers giving birth to live and stillborn children suffering from congenital malformations, over that expected in other hospitalised mothers was reported by Worcester et al. (1950). No matched control series was available for comparison. Drillien observed that the prevalence of abnormalities of pregnancy was only slightly greater in pregnancies which resulted in the birth of malformed infants than in those which resulted in the births of healthy infants. This was in contrast to the much greater proportion of complicated pregnancies found when infants died as a result of "asphyxia", "prematurity" and "infections", (Drillien, 1947).

All authors agree that malformed infants tend to be prematurely born by weight more frequently than normal infants. 21.1% of malformed infants had birth weights of 5 pounds, eight ounces or less in the series of Drillien. 26.5% in the series of Worcester et al, and 36.3%, compared to 3.1% in a control series in that of Record and McKeown.

Abnormalities of labour and delivery occur more frequently when the infant is malformed than when it is normal. Of the infants who suffered from congenital malformations studied by Potter and Adair, natural cephalic delivery occurred in 64%,
64%, forceps delivery in 15%, breech delivery in 11%, version and extraction in 7% and caesarean section was employed in 4%. (Percentages approximate). Instruments were used no more frequently than in a control group in the series studied by Drillien, but delivery was by the breech in 19.2% of cases, four times as often as expected, and by caesarean section twice as often, (Potter and Adair, 1939; Drillien, 1947).

It is apparent from these studies that in any large series of children suffering from developmental malformations, disorders of pregnancy, especially hydramnios, vaginal haemorrhage may be expected to occur rather more frequently than expected, though not as often as in a series of children dying from some other causes. Abnormalities of labour and delivery, especially breech birth, and prematurity may be expected much more frequently than when the offspring is normal.

Maternal age in cerebral palsy. Patients in each category were grouped according to the mother's age at the time of their birth in Table 208. and the average maternal age is shown in Table 208. Compared to the distribution by maternal age of all live legitimate births in the Counties of Cities in Scotland, there is no statistically significant difference in the distributions of the whole group, though the excess of mothers over the age of forty is noteworthy. (Registrar General for Scotland 1951).

Analysis /
Analysis of the individual categories shows that compared to the general population there are statistically significant differences in congenital hemiplegia and in diplegia. (Using the X2 test, p is .01 and .02 respectively). In both these categories there is an excess of older mothers, and in diplegia there are also more mothers under the age of 24 than would be expected. The distribution of diplegic patients by age of mothers is, in fact, "biphasic", though more markedly so than that found in mongols by Penrose (1934). The relatively large proportion of young mothers in diplegia makes the high average maternal age in this category the more striking.

The excess of older mothers in diplegia and congenital hemiplegia is certainly compatible with some of the cases in these categories being due to developmental malformation, but is no real indication that they are. The finding can also be interpreted as being the result of the higher prevalence of birth injury amongst the offspring of older mothers.

Parity of mothers with children suffering from congenital cerebral palsy. The distribution of patients by the number of previous pregnancies is compared to figures for births in Counties of Cities given by the Registrar General in Table (Registrar General for Scotland 1951). Whereas there is an apparent excess of children of high birth rank in acquired cerebral palsy (of doubtful statistical significance, p being .05), there is an excess of children of low birth rank amongst those
### TABLE 247

**Fertility of mothers of patients with congenital cerebral palsy, hemiplegia of unknown origin and acquired hemiplegia**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number of mothers</th>
<th>Number of pregnancies (including patients)</th>
<th>Average number of pregnancies per mother</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia.</td>
<td>30</td>
<td>84</td>
<td>2.8</td>
</tr>
<tr>
<td>Hemiplegia unknown origin.</td>
<td>11</td>
<td>42</td>
<td>3.8</td>
</tr>
<tr>
<td>Acquired hemiplegia.</td>
<td>33</td>
<td>134</td>
<td>4</td>
</tr>
<tr>
<td>Congenital bilateral hemiplegia.</td>
<td>7</td>
<td>13</td>
<td>1.9</td>
</tr>
<tr>
<td>Congenital diplegia.</td>
<td>76</td>
<td>214</td>
<td>2.8</td>
</tr>
<tr>
<td>Ataxic diplegia.</td>
<td>9</td>
<td>29</td>
<td>2.9</td>
</tr>
<tr>
<td>Congenital ataxia.</td>
<td>10</td>
<td>24</td>
<td>2.7</td>
</tr>
<tr>
<td>Congenital dyskinesia.</td>
<td>16</td>
<td>40</td>
<td>2.5</td>
</tr>
</tbody>
</table>
those suffering from congenital cerebral palsy (of high statistical significance, p being less than .01). 52.5% of the 160 cases are in fact first born, and over half of these were only children at the time of examination.

**Number and spacing of pregnancies of the mothers with offspring suffering from congenital hemiplegia.** The numbers of pregnancies to mothers of patients suffering from congenital cerebral palsy, at the time of examination are shown in Table 247, where they are compared to the number of pregnancies of mothers of patients with acquired hemiplegia. It will be observed that mothers of children suffering from bilateral hemiplegia have remarkably few other pregnancies and that the only category in which the fertility of mothers appears to approach that of those in the category of acquired hemiplegia is ataxic diplegia. Thus, in spite of their rather higher average age, mothers of patients with congenital cerebral palsy appear to be less fertile than mothers of children with acquired hemiplegia. A more detailed study of the two largest categories of congenital cerebral palsy, namely congenital hemiplegia and diplegia, is possible. This suggests that mothers under the age of 24 with children suffering from congenital hemiplegia or diplegia are as fertile or more fertile than those in the general population, but that there is a much steeper decline in fertility thereafter.

A study of the spacing of pregnancies in those categories
in which there were enough for analysis revealed that there appeared to be a period of relative sterility immediately before and immediately after the birth of the child with congenital cerebral palsy. The clinical impression that this was not the result of contraceptive measures being employed received some support from the observation that the sterile period was greater immediately before the birth of the affected child than was the subsequent one. Table 247.

The finding of a disproportionately large lapse of time between prior and subsequent pregnancies to those which resulted in the birth of the affected child is similar to that of Murphy in his study of fatal developmental malformations. Another similarity between his series and the mothers of patients suffering from congenital hemiplegia and diplegia is that in both there is a marked tendency for miscarriages and perinatal deaths to occur in the conceptions continuous to those which produced the abnormal children. (Table 5.)

As well as conceiving less frequently than mothers in the general population, the mothers of some of the patients suffering from congenital hemiplegia and diplegia appear to have a period of relatively greater sterility at about the time the child with cerebral palsy is produced. When they do conceive in the years immediately prior and subsequent to the birth of the affected child, they seem particularly liable to abort or lose the offspring that result.

These /
These findings at least suggest that in a proportion of mothers of patients with congenital hemiplegia and diplegia there is some fundamental abnormality in the ability to conceive and carry children normally which is most marked at the time the patients are conceived. Whether the patients are abnormal in these circumstances, only because the mothers are more likely to have complications of parturition which result in birth injury of the offspring, or whether there are primary developmental abnormalities in the offspring is impossible to determine. However, the similarities of the findings to those in the series of developmental malformations studied by Murphy are striking.

The significance of a history of abnormal parturition in congenital cerebral palsy. The numbers of patients in each category of congenital cerebral palsy who were born after disorders of pregnancy, labour or delivery are shown in Table 247. It will be observed that 28 patients (excluding 3 with hemiplegia of unknown origin) or approximately 19% had a history of clinically normal parturition. The highest proportion was in bilateral hemiplegia, in which 2 out of seven patients, approximately 29% had no abnormalities of parturition. In diplegia, 26% had no such history; in ataxic diplegia approximately 14%; in dyskinesia 12.5%; and in ataxia 11%. Only one out of the thirty patients with congenital hemiplegia gave no history of abnormal pregnancy. labour /
labour or delivery. The highest incidence of prematurity was in diplegia, in which category 44% of patients were prematurely born by weight, compared to 33% of patients with ataxia, 10% of dyskinetic children, 14% of those with ataxic diplegia, and 13% of those with congenital hemiplegia. No patients suffering from bilateral hemiplegia were premature by weight.

These findings have already been discussed from the point of view of attempting to determine the likelihood of birth injury being important in the various categories. The information they give about the relative aetiological importance of developmental malformations in each category is more limited.

If developmental malformation was the only cause of a particular form of cerebral palsy, the histories of parturition might be expected to resemble those found amongst large series of children with other types of developmental malformations. A slight increase in the number of abnormal pregnancies compared to normal, abnormalities of labour and delivery in up to 40% of cases and premature birth in between 15 and 25%.

In fact, taking all the cases of congenital cerebral palsy together, less than 50% of cases have a history of uncomplicated pregnancy and less than 50% a history of apparently uncomplicated labour and delivery. This would appear to indicate that the majority of cases do not behave as one would /
would expect them to if congenital cerebral palsy were always the result of developmental malformation. On the other hand, it does not exclude the possibility that a minority of patients behave as if they suffered from developmental malformation.

When the individual categories are reviewed from this point of view it is observed that those containing the highest proportion of patients with a history of abnormal parturition are dyskinesia, ataxia and hemiplegia, the lowest proportions are found in bilateral hemiplegia, diplegia, and ataxic diplegia. Four out of eight patients with bilateral hemiplegia were born after apparently normal pregnancy, three out of seven with ataxic diplegia, and 43% of those with diplegia. The high proportion of prematurely born diplegic patients (44%) is greater than expected in most congenital malformations. The proportion is rather low in ataxic diplegia and there were no prematurely born patients amongst those suffering from bilateral hemiplegia.
### TABLE 248 a. & b.

**Birth histories of patients with congenital cerebral palsy**

<table>
<thead>
<tr>
<th></th>
<th>Congenital hemiplegia</th>
<th>Unknown hemiplegia</th>
<th>Bilateral hemiplegia</th>
<th>Diplegia</th>
<th>Ataxic diplegia</th>
<th>Ataxia</th>
<th>Dyskinesia</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Numbers</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pregnancy, labour and delivery uncomplicated.</td>
<td>1</td>
<td>8</td>
<td>2</td>
<td>20</td>
<td>2</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Pregnancy abnormal. Labour and delivery uncomplicated.</td>
<td>6</td>
<td>4</td>
<td>2</td>
<td>15</td>
<td>1</td>
<td>4</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Pregnancy normal. Labour and delivery complicated.</td>
<td>7</td>
<td>0</td>
<td>2</td>
<td>17</td>
<td>3</td>
<td>1</td>
<td>7</td>
<td>0</td>
</tr>
<tr>
<td>Pregnancy and labour and/or delivery abnormal.</td>
<td>16</td>
<td>0</td>
<td>1</td>
<td>26</td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>30</strong></td>
<td><strong>12</strong></td>
<td><strong>7</strong></td>
<td><strong>78</strong></td>
<td><strong>9</strong></td>
<td><strong>9</strong></td>
<td><strong>16</strong></td>
<td><strong>1</strong></td>
</tr>
</tbody>
</table>

|                  |                      |                    |                      |          |                 |        |            |       |
| B. Approximate percentages |                   |                    |                      |          |                 |        |            |       |
| Pregnancy, labour and delivery uncomplicated. | 3         |       | 26                 | 29       | 22.5           |        |            |       |
| Pregnancy abnormal. Labour and delivery uncomplicated. | 21        | 19      | 30                 | 23.1    |                |        |            |       |
| Pregnancy normal. Labour and delivery complicated. | 23        | 22      | 24                 | 22.5    |                |        |            |       |
| Pregnancy and labour and/or delivery abnormal. | 53        | 33      | 17                 | 31.9    |                |        |            |       |
| **Total**        | **100**             | **100**           | **100**              | **100.0**|                |        |            |       |

### Birth histories of patients with acquired cerebral palsy.

<table>
<thead>
<tr>
<th></th>
<th>Acquired hemiplegia</th>
<th>Other</th>
<th>Total</th>
<th>Approx. %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pregnancy, labour and delivery normal</td>
<td>25</td>
<td>10</td>
<td>35</td>
<td>74.3</td>
</tr>
<tr>
<td>Pregnancy abnormal. Labour and delivery normal</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>8.6</td>
</tr>
<tr>
<td>Pregnancy normal. Labour and delivery abnormal.</td>
<td>3</td>
<td>6</td>
<td>9</td>
<td>12.3</td>
</tr>
<tr>
<td>Pregnancy, labour and/or delivery abnormal.</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4.3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>33</strong></td>
<td><strong>14</strong></td>
<td><strong>47</strong></td>
<td><strong>100.0</strong></td>
</tr>
</tbody>
</table>
The child with congenital malformations.

A number of studies have been made of the characteristics of children suffering from developmental malformations which merit summary.

Sex. The ratios of males to females are significantly different in different malformations. Murphy found a male to female ratio of 170.2/100 for hydrocephalus without spina bifida and a male to female ratio of 36.5/100 for anencephalus. For all malformations in his series the ratio was 115/100 compared to 103.2/100 for all live births in Philadelphia, the town from which his series was drawn, (Murphy, 1947).

For fatal malformations of the nervous system the male to female ratio amongst patients was 63.8/100 in the series of Record and McKeown, (1949-50).

<table>
<thead>
<tr>
<th>Table 249</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex incidence of malformations of the central nervous system resulting in stillbirth or infant death. Birmingham 1940-47. Record and McKeown 1947</td>
</tr>
<tr>
<td>Number of cases</td>
</tr>
<tr>
<td>-----------------</td>
</tr>
<tr>
<td>Anencephalus</td>
</tr>
<tr>
<td>Spina bifida</td>
</tr>
<tr>
<td>Hydrocephalus</td>
</tr>
<tr>
<td>Other</td>
</tr>
<tr>
<td>All</td>
</tr>
</tbody>
</table>

In /
In England and Wales 1949-50, the male to female ratios of neonatal and post neonatal deaths attributable to malformations of the nervous system were approximately 69/100 and 71/100 respectively, (Heady and Heasman, 1959). These ratios are greatly influenced by the high proportion of infants suffering from anencephalus an almost universally fatal condition which is much more frequent in the female than the male. Possibly conditions compatible with survival in a higher proportion of patients might show a less female biased ratio.

**Multiple births and congenital malformations.** The evidence is conflicting as to whether there is a significant excess of multiple births amongst children with developmental malformations. Twins were reported to occur about twice as frequently as expected amongst the 677 children with congenital malformations studied by Worcester et al (1950). A high incidence of twins amongst the siblings of malformed children was reported by Murphy (1947). No greater tendency for twins than singleton offspring to suffer fatal malformations of the nervous system was reported by Record and McKeown (1951). Moreover, the chances of the twin of a malformed child being similarly affected were no greater than that of a later born sibling.

**Multiple malformations.** In most series of malformed children a significant proportion of patients are found to have abnormalities of more than one part or system of the body.
Thus, of the 955 malformed individuals studied by Murphy, 77.2% had malformations "involving one portion of the body", whereas the remaining ones (22.8%) had defects affecting more than one part, (Murphy 1947).

**The child with congenital cerebral palsy.**

**Sex.** In Table 250 are shown the numbers and percentages of male and female patients in the various categories of congenital cerebral palsy. In all categories except dyskinesia and bilateral hemiplegia, in which the numbers of patients were small, there is an excess of males. When hemiplegia is considered, the male to female ratio is 200.8/100, and this figure is similar to that found in acquired hemiplegia. In diplegia the ratio is 360/100 which may indicate a rather lower liability of the male to suffer from this condition than from hemiplegia.

The only category showing a very high female preponderance which might suggest sex linked inheritance of a determining gene is the small one of bilateral hemiplegia in which all seven patients were girls. Though it is interesting that these patients seemed likely to be suffering from congenital malformations for other reasons, the sample is not large enough for the sex ratio to be of statistical significance. Taking all the patients in the different categories of cerebral palsy together, the male to female ratio (155.4/100) is less like that for neonatal deaths due to malformation of
The sex ratios of patients suffering from congenital cerebral palsy

<table>
<thead>
<tr>
<th>Condition</th>
<th>Male</th>
<th>Female</th>
<th>Approximate ratio</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia</td>
<td>20</td>
<td>10</td>
<td>200</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>9</td>
<td>3</td>
<td>300</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Diplegia</td>
<td>48</td>
<td>30</td>
<td>160</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Ataxic diplegia</td>
<td>5</td>
<td>2</td>
<td>250</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Ataxia</td>
<td>4</td>
<td>5</td>
<td>80</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>7</td>
<td>9</td>
<td>77</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td>0</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total congenital cases</td>
<td>84</td>
<td>64</td>
<td>155</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>23</td>
<td>10</td>
<td>221</td>
<td>100</td>
<td></td>
</tr>
<tr>
<td>Hemiplegia of unknown origin</td>
<td>9</td>
<td>3</td>
<td>300</td>
<td>100</td>
<td></td>
</tr>
</tbody>
</table>
the nervous system (approximately 69/100) or postneonatal deaths (71/100) than the ratio for intracranial birth injury which was fatal in the neonatal period, (176/100) in England and Wales 1949–50. Heady and Heasman 1959. Social and Biological Factors in Infant Mortality, No. 15 of Studies on Medical and Population Subjects. H.M.S.O.). On the other hand, the findings are perfectly compatible with a minority of the patients in the present series suffering from cerebral palsy as a result of developmental malformations.

Multiple births and congenital cerebral palsy. In the series of 160 patients with cerebral palsy of congenital origin, there were six who were twins, Table 251. This gives a prevalence of twins of 3.3% compared to 2.5% amongst all live and stillbirths in Scotland in 1951 not a difference of statistical significance.

It will be observed that five of the twins suffered from diplegia and that all were premature by weight. The significance of this finding is that there is a greater tendency for premature children to suffer from diplegia than any other form of cerebral palsy. Thus, the apparent excess of twins amongst diplegic patients may be related to the fact that they were premature rather than that they were the result of multiple births.
Figure 24

Identical twins with diplegia, both patients being functionally paraplegic and similarly mentally retarded. (Cases I58-I59)
<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Other twin</th>
<th>Sex</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>162</td>
<td>F.</td>
<td>Congenital hemiplegia.</td>
<td>Healthy.</td>
<td>M.</td>
<td>Both premature by weight.</td>
</tr>
<tr>
<td>41</td>
<td>M.</td>
<td>Diplegia.</td>
<td>Healthy.</td>
<td>M.</td>
<td>Both premature by weight.</td>
</tr>
<tr>
<td>63</td>
<td>F.</td>
<td>Diplegia.</td>
<td>Healthy.</td>
<td>M.</td>
<td>Both premature by weight.</td>
</tr>
<tr>
<td>159)</td>
<td>M.</td>
<td>Diplegia.</td>
<td></td>
<td>M.</td>
<td></td>
</tr>
<tr>
<td>181</td>
<td>F.</td>
<td>Diplegia.</td>
<td>Foetus papyraceous.</td>
<td>Unknown</td>
<td>Premature by weight.</td>
</tr>
<tr>
<td>193</td>
<td>F.</td>
<td>Diplegia.</td>
<td>Healthy.</td>
<td>M.</td>
<td>Premature by weight. (but no other details)</td>
</tr>
</tbody>
</table>
Other congenital abnormalities amongst patients with congenital cerebral palsy. The congenital malformations which did not appear to be directly related to the abnormalities of the nervous system found in patients with congenital cerebral palsy are shown in Table 251. The need to exclude conditions which are secondary to abnormalities of the nervous system means that a number of minor abnormalities of possible significance has to be left out of consideration. These included hyperextensibility of the digits, strabismus, dislocation of the hip even when apparently congenital and mild cases of talipes equino deformity. Abnormalities which be regarded as normal variants when mild have only been listed when quite extreme. For example, the diagnosis of partial syndactyly has only been made when the interdigital cleft extends at least up to the second interphalangeal joint, hypertelorism only when associated with marked epicanthic folds.

In Table 252, is shown the frequency with which congenital malformations occur in the various types of congenital cerebral palsy. It will be observed that they occur with least frequency in congenital hemiplegia and dyskinesia in which birth injury appeared to be of greatest etiological importance. They occur most frequently in cases of bilateral hemiplegia and ataxic diplegia in which there was other evidence to suggest that at least a high proportion of cases were not the result of birth injury. It is interesting that the proportion of patients with acquired hemiplegia with congenital abnormalities was smaller than in any of the congenital categories except dyskinesia.
<table>
<thead>
<tr>
<th>Case</th>
<th>Diagnosis</th>
<th>Congenital Malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>29</td>
<td>Congenital hemiplegia.</td>
<td>Very high arched palate (irregular dentition later)</td>
</tr>
<tr>
<td>71</td>
<td>Congenital hemiplegia.</td>
<td>Very high arched palate (irregular dentition)</td>
</tr>
<tr>
<td>192</td>
<td>Congenital hemiplegia.</td>
<td>Partial syndactyly of 2nd &amp; 3rd toes.</td>
</tr>
<tr>
<td>58</td>
<td>Hemiplegia of unknown origin.</td>
<td>Hypertelorism.</td>
</tr>
<tr>
<td>113</td>
<td>Hemiplegia of unknown origin.</td>
<td>Hypomandibulosis.</td>
</tr>
<tr>
<td>3</td>
<td>Diplegia.</td>
<td>Multiple nasi.</td>
</tr>
<tr>
<td>22</td>
<td>Diplegia.</td>
<td>Severe talipes equino varus deformities. (7 arthrogryposis).</td>
</tr>
<tr>
<td>40</td>
<td>Diplegia.</td>
<td>Hypertelorism.</td>
</tr>
<tr>
<td>62</td>
<td>Diplegia.</td>
<td>Multiple nasi.</td>
</tr>
<tr>
<td>81</td>
<td>Diplegia.</td>
<td>Congenital heart disease, ventricular septal defect.</td>
</tr>
<tr>
<td>98</td>
<td>Diplegia.</td>
<td>Partial syndactyly of 2nd &amp; 3rd toes.</td>
</tr>
<tr>
<td>120</td>
<td>Diplegia.</td>
<td>Severe bilateral talipes equino varus.</td>
</tr>
<tr>
<td>131</td>
<td>Diplegia.</td>
<td>Hypertelorism.</td>
</tr>
<tr>
<td>140</td>
<td>Diplegia.</td>
<td>Severe bilateral hypertelorism.</td>
</tr>
<tr>
<td>154</td>
<td>Diplegia.</td>
<td>Hypertelorism.</td>
</tr>
<tr>
<td>158</td>
<td>Diplegia.</td>
<td>Accessory auricles.</td>
</tr>
<tr>
<td>177</td>
<td>Diplegia.</td>
<td>Hypomandibulosis.</td>
</tr>
<tr>
<td>183</td>
<td>Diplegia.</td>
<td>Partial syndactyly of 2nd &amp; 3rd toes bilaterally.</td>
</tr>
<tr>
<td>205</td>
<td>Diplegia.</td>
<td>Hypospadias.</td>
</tr>
<tr>
<td>8</td>
<td>Ataxic diplegia.</td>
<td>Partial syndactyly of 2nd &amp; 3rd toes.</td>
</tr>
<tr>
<td>14</td>
<td>Ataxic diplegia.</td>
<td>Hypomandibulosis.</td>
</tr>
<tr>
<td>150</td>
<td>Ataxic diplegia.</td>
<td>Multiple nasi.</td>
</tr>
<tr>
<td>182</td>
<td>Ataxic diplegia.</td>
<td>Hypertelorism.</td>
</tr>
<tr>
<td>73</td>
<td>Ataxia.</td>
<td>Congenital hydrocephalus.</td>
</tr>
<tr>
<td>78</td>
<td>Ataxia.</td>
<td>Congenital hydrocephalus.</td>
</tr>
<tr>
<td>23</td>
<td>Dysskinesia.</td>
<td>Hypospadias.</td>
</tr>
<tr>
<td>202</td>
<td>Dysskinesia.</td>
<td>Cleft palate.</td>
</tr>
<tr>
<td>35</td>
<td>Bilateral hemiplegia.</td>
<td>Low hair line.</td>
</tr>
<tr>
<td>102</td>
<td>Other forms of cerebral palsy.</td>
<td>Arthrogryposis.</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>Number of cases</td>
<td>Number with malformations</td>
</tr>
<tr>
<td>-----------------------------------</td>
<td>-----------------</td>
<td>---------------------------</td>
</tr>
<tr>
<td>Congenital hemiplegia</td>
<td>30</td>
<td>5</td>
</tr>
<tr>
<td>Hemiplegia of unknown origin</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>Bilateral hemiplegia</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>Diplegia</td>
<td>76</td>
<td>18</td>
</tr>
<tr>
<td>Ataxic Diplegia</td>
<td>10</td>
<td>4</td>
</tr>
<tr>
<td>Ataxia</td>
<td>9</td>
<td>2</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>16</td>
<td>2</td>
</tr>
<tr>
<td>Other</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>160</td>
<td>38</td>
</tr>
<tr>
<td>Acquired hemiplegia</td>
<td>33</td>
<td>4</td>
</tr>
</tbody>
</table>
Though the numbers of patients are too small for statistical analysis the proportions of patients in the various categories with congenital malformations are in inverse proportion to the apparent importance attached to birth injury. They provide a little more evidence in favour of congenital malformation being an important aetiological factor in congenital cerebral palsy.

Head size of patients with cerebral palsy as an indication of congenital malformation rather than birth injury.

The suggestion that small head size in groups of patients suffering from congenital cerebral palsy might be a point in favour of congenital malformation, rather than birth injury, has been mentioned, (Yannet, 1944). The head circumferences of selected groups of patients were compared with the expected normal in the following graphs. In the first the cases with bilateral hemiplegia are compared to those with congenital hemiplegia and hemiplegia of unknown origin. It will be observed that there is a marked tendency for the heads of children with bilateral hemiplegia to be smaller. On the other hand, a comparison between patients with congenital diplegia who had and had not a history of abnormal parturition was unrewarding. No significant difference in the distribution of these cases by their head sizes being apparent. Possibly reduction in head size compared to normal is only very marked when congenital anomalies of the brain are gross, as tend to be in bilateral hemiplegia. (Figs. 25)
The head size of children suffering from cerebral palsy.
Figure 26:

The head size of patients suffering from cerebral palsy.

| Comparison of Head size of patients with Edinburgh norms (Ellis 1955, Thomson 1956) |
|----------------------------------|----------------------------------|
| Ataxic diplegia                  | Ataxia                           |
| Dyskinesia                       |                                  |
CHAPTER 3

CONCLUSIONS

It has been shown that it is even more difficult to define satisfactory positive diagnostic criteria for "Developmental Malformation" as a cause of cerebral palsy than it is for "Birth Injury". If malformations causing cerebral palsy were always the result of Mendelian dominant or recessive inheritance it would be difficult to assess the relative importance of birth injury and genetic factors without a large series of closely studied patients with full family histories. Since developmental malformations in cerebral palsy may also be the result of genetic mutation and deviations from the normal patterns of development in early foetal life, the problem of aetiological diagnosis is yet more complex.

There is relatively strong evidence that developmental malformations were responsible for a significant proportion of patients in two categories of cerebral palsy in the present series, bilateral hemiplegia and ataxic diplegia. All seven congenital cases of bilateral hemiplegia were girls. Four had gross congenital abnormalities affecting other parts of the body than the nervous system. One patient without associated malformations had a family history of cerebral palsy and mental defect. The mothers tended to be very infertile. The pregnancies, labours and deliveries which resulted in the births of the patients were not thought to have been likely to /
to cause gross hypoxia or trauma. All the patients had extremely small heads, only one having an occipitofrontal circumference of more than 19 inches at the age of six years.

In three of the ten patients suffering from congenital ataxic diplegia, there was a family history of cerebral palsy. In one case, a half sister was hemiplegic, but in the other two, both girls, the mothers also suffered from ataxic diplegia and in one of them a sister was similarly affected. A rather high proportion of patients siblings and relatives suffered from other neurological disorders. Mothers were relatively infertile, and their ages at the time of birth tended to be high. Four patients had associated minor developmental malformations. It seems probable that at least a proportion of patients suffering from ataxic diplegia have developmental malformations as a result of Mendelian dominant inheritance. It is possible that there may be other malformations of different aetiology in this group of patients. If there are, the series is too small to demonstrate them.

The evidence in favour of malformations of genetic or later developmental origin being important in other categories of cerebral palsy is still more circumstantial. That they may be important is suggested by various pieces of evidence which have been presented. For example, the presence of familial genetic factors in congenital hemiplegia may be postulated since 10 per cent. of cases had a family history of /
of cerebral palsy. The fact that four patients with congenital diplegia (counting the pair of diplegic twins as one case) had relatives with diplegia, including a sibling and two first cousins, indicates that diplegia may be the result of Mendelian recessive inheritance in some cases. But it is difficult to weigh the significance of these various pieces of evidence when one wishes to assess the importance of malformations in the various categories. For example, is it more important that 75% of patients with hemiplegia of unknown origin had no history of abnormal parturition, or that a rather high proportion of patients suffering from diplegia and ataxia were premature by birth weight?

With such a small series of patients, the most than can be attempted is a comparison of the various categories of cerebral palsy by the factors which are believed to suggest developmental malformation. The factors chosen for this comparison were as follows:

1. Known family history of cerebral palsy in siblings, parents, uncles, aunts or cousins.
2. Abnormal siblings suffering from congenital malformations or neurological disease, excluding mental retardation.
3. Birth after apparently uncomplicated pregnancy, labour and delivery.
4. Premature birth by weight (five pounds, eight ounces or less).
5. Associated congenital malformations of patients, excluding those possibly secondary to cerebral palsy.
6. /
<table>
<thead>
<tr>
<th>Type of cerebral palsy</th>
<th>Family history of tableed</th>
<th>Congenitally abnormal siblings</th>
<th>Normal pregnancy and birth</th>
<th>Prematurity</th>
<th>Associated malformations</th>
<th>Occipito fronto circumference below first centile</th>
<th>Total ratings</th>
<th>Total ratings less than those for normal pregnancy and birth</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hemiplegia.</td>
<td>5</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>3</td>
<td>5</td>
<td>19</td>
<td>18</td>
</tr>
<tr>
<td>Hemiplegia of unknown origin.</td>
<td>6</td>
<td>5</td>
<td>7</td>
<td>1</td>
<td>2</td>
<td>6</td>
<td>27</td>
<td>20</td>
</tr>
<tr>
<td>Bilateral hemiplegia.</td>
<td>3</td>
<td>4</td>
<td>6</td>
<td>1</td>
<td>7</td>
<td>7</td>
<td>28</td>
<td>22</td>
</tr>
<tr>
<td>Diplegia.</td>
<td>7</td>
<td>3</td>
<td>5</td>
<td>7</td>
<td>4</td>
<td>4</td>
<td>30</td>
<td>25</td>
</tr>
<tr>
<td>Ataxic diplegia.</td>
<td>2</td>
<td>6</td>
<td>4</td>
<td>4</td>
<td>6</td>
<td>1</td>
<td>23</td>
<td>19</td>
</tr>
<tr>
<td>Ataxia.</td>
<td>4</td>
<td>7</td>
<td>3</td>
<td>6</td>
<td>4</td>
<td>3</td>
<td>27</td>
<td>24</td>
</tr>
<tr>
<td>Dyskinesia</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>5</td>
<td>1</td>
<td>3</td>
<td>13</td>
<td>11</td>
</tr>
</tbody>
</table>
6. Occipito frontal circumference of less than one percentile for age.

Patients in each of the seven categories were rated according to the proportion of patients in which these factors were positive. Thus, the category containing the smallest proportion of patients with a family history of cerebral palsy would be rated one, whilst that with the largest proportion would be rated seven. The results of this rating are shown in Table 253.

It will be observed that there are considerable variations in the ratings for various factors in each category. For example, congenital hemiplegia and hemiplegia of unknown origin score 5 and 6 respectively for "Family History of Cerebral Palsy", but only 1 and 3 for "Prematurity". The low score for ataxic diplegia in which a significant proportion of patients are almost certainly affected because of hereditary transmission of a genetic trait is also noteworthy. These facts emphasise the need for caution in interpreting the results of such a crude method of investigation in which the aetiological factors compared are arbitrarily chosen. Nevertheless, there are interesting differences in the total scores of the different categories.

Dyskinesia and congenital hemiplegia are the categories with the lowest scores. Bilateral hemiplegia, and diplegia are those with the highest, closely followed by ataxia and hemiplegia.
Comparison of ratings of categories of congenital cerebral palsy (and hemiplegia of unknown origin) by "Birth injury" insults and by factors suggesting the presence of developmental malformation.

<table>
<thead>
<tr>
<th>Order of categories by &quot;birth injury&quot; insults per 100 patients</th>
<th>Order of categories by &quot;malformation&quot; ratings</th>
</tr>
</thead>
<tbody>
<tr>
<td>2. Bilateral hemiplegia.</td>
<td>2. Congenital hemiplegia.</td>
</tr>
<tr>
<td>3. Ataxic diplegia.</td>
<td>3. Ataxic diplegia.</td>
</tr>
<tr>
<td>5. Ataxia.</td>
<td>5. Bilateral hemiplegia.</td>
</tr>
</tbody>
</table>
hemiplegia of unknown origin. These findings are compatible with developmental malformation being of greater importance in diplegia and bilateral hemiplegia, and possibly in ataxia and hemiplegia of unknown origin, than in dyskinesia and congenital hemiplegia.

It is interesting to compare ratings of the categories by these factors which might be taken as suggesting developmental malformation with the ratings according to the number of "Birth injury" insults per 100 patients. In order to do this the scores for "Normal pregnancy and birth" are removed from the total scores in the "Malformation" ratings, since normality or abnormality of birth is scored in the "Birth injury" rating also.

It will be observed in Table 254, that the two categories with the lowest "Malformation" score are those with the highest "Birth injury" score. This may indicate that birth injury is a more important aetiological factor in these categories than the others, and that fewer patients suffering from these conditions have developmental malformations than in the other categories of cerebral palsy.

The two categories with the least indication of "Birth injury" being important aetio logically are hemiplegia of unknown origin, and bilateral hemiplegia and these come fifth and fourth respectively in the "Malformation" ratings. This suggests /
suggests that developmental malformation may be more important than birth injury in the etiology of these conditions.

Ataxia and diplegia come relatively high in both the birth injury and the malformation ratings. There are several possible theoretical explanations of this finding. The categories may be heterogenous etiologically; some patients being diplegic or ataxic because of developmental malformations, and others because of birth injury. Alternatively, both malformation, possibly acting as a predisposing factor, and birth injury may be required to make the great majority of the patients ataxic or diplegic. In fact, there is some evidence within the group of diplegic patients that both theoretical explanations may be correct. It was shown in the section discussing the etiology of diplegia that patients with family histories of malformed siblings and relatives suffering from neurological diseases, or who themselves had associated malformation, had fewer birth injury insults than those with negative family histories and no associated developmental malformations.

It is possible to think in terms of malformation, (whether the result of heredity, genetic mutation, or teratogenic deviation during later development), as being the only cause of diplegia or ataxia in a proportion of cases. Similarly, "Birth injury" may be the only cause in others. In a third group of cases, both etiological factors may be present.
Some, in whom malformation is a relatively mild predisposing factor, may require severe birth injury to produce ataxia or diplegia; others, in whom malformation is a strong predisposing factor need many fewer hypoxic traumatic or toxic insults to produce these disorders. If the majority of patients required both the predisposition of "malformation" and the effects of "birth injury", it would explain the high rating on both scales in both ataxia and diplegia.

It is much more difficult to explain the comparatively low rating of ataxic diplegia in both scales. A tendency for patients in this condition to suffer either from birth injury without any predisposition by "Malformation" factors, or from malformation with any birth injury would tend to make the rating lower than it would otherwise be on both scales, but this can only be a hypothetical explanation. A higher rating on the "Malformation" scale might have been expected in view of the fact that a significant proportion of cases are almost certainly hereditarily determined.
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