Disabilities and trends over time in a French county, 1980–91

C Cans, P Guillem, J Fauconnier, P Rambaud, P S Jouk

Aim: To describe trends over time and types of disability for children born in a French county from 1980 to 1991.

Methods: Data were collected from medical records of a morbidity register; disabled children with at least one severe deficiency have been included. Prevalence rates are given per 1000 resident children, over four three-year periods. Results: Overall, 7.73 per 1000 children (that is, 1360 children), had a severe childhood disability, and the prevalence rate had increased significantly since 1980. This increase was mainly owing to an increase in cerebral palsy and psychiatric disorder prevalence rates. Conclusion: Future research aimed to explain these trends over time can be based on such data. The present knowledge is useful for planning purposes.

Concurrent changes in obstetric and neonatal care over the past decades have resulted in a decrease in infant mortality. However, several authors have noted that this decrease in infant mortality has not been followed by a decrease in infant morbidity. In some places, the cerebral palsy prevalence rate has remained stable, while in others it has increased; a decrease in some subgroups has recently been reported.

The educational and medical care to be given to disabled children has been laid down by law in France since 1975. Since then, very few surveys have described the trends in childhood disability. However, knowledge about types of childhood disabilities and trends over time might prove to be very useful for those specialised schools and care institutions that deliver care to these children, and for those making health policy decisions. Thus, in this paper, we describe types of childhood disabilities and their prevalence trends over time for children born in one French county between 1980 and 1991.

POPULATION AND METHODS

Population

The morbidity register, “Registre des Handicaps de l’Enfant et Observatoire Périnatal de l’Isère” (RHEOP), was established in 1992 in France, with the aim of monitoring trends over time for severe childhood disabilities. Isère county has 1 000 000 inhabitants, and 14 000 live births per year. All disabled children living in Isère county, aged 7 years, with a clinical feature of a severe deficiency are registered.

Only children with a severe neurosensory deficiency were recorded. The criteria used to determine the severity of the deficiency were those usually used (table 1). Children with two or three mild or moderate deficiencies were not included. With respect to population migration in and out of the county, it was decided to use results for prevalence rates per 1000 resident children rather than those per 1000 live births, as a previous survey showed that the magnitude of migration into this county was the same for healthy children and disabled children. Analysis of prevalence rates over time was carried out for three year periods, and the Armitage trend test was used when checking for trends over time.

RESULTS

Types of disability

A total of 1360 children born between 1980 and 1991 (797 boys and 563 girls), satisfied the inclusion criteria for “at least one severe deficiency”—that is, around 120 children for each birth year cohort, with a significant overall male/female sex ratio of 1.42 (p < 0.001). Table 1 shows the different types of severe disabilities present in these 1360 children. As a child might present more than one disability, the total number of disabilities is greater than the number of disabled children. The mean number of ascertainment sources was 1.7 per child.

Trends over time

Table 2 shows the prevalence rates for the overall period, and for consecutive three year periods. The overall prevalence rate of 7.73 per 1000 has increased significantly since 1980 (p < 0.001). The prevalence rate of motor disability has increased slightly (p = 0.17); this increase mainly concerned the cerebral palsy prevalence rate (p = 0.01), while the prevalence rate of other motor disabilities has remained stable (p = 0.31).
(severe mental retardation) has remained stable over time (p = 0.94), even when looking at subgroups—that is, Down’s syndrome (p = 0.68), or other intellectual disabilities (p = 0.70). Overall, the prevalence rate has increased for psychiatric disorders (p < 0.001). However, for psychosis with severe mental retardation, the prevalence rate has remained stable (p = 0.41), while for autism and other psychoses, the rates have actually increased (p = 0.001 and p < 0.001, respectively). The prevalence rate for sensory disability has not changed since 1980 (p = 0.80).

**Characteristics**

The socioeconomic status, determined by the occupation of the father and/or mother of the disabled child, was compared with the distribution figures for parental occupation within the Isere population in 1990. Mothers of the disabled children were often without occupation, more often than other women of the same age in the whole population (55% versus 31%, p < 0.001). Parental occupation statistics were different, with more fathers of low economic status (working in factories) and fewer fathers of high economic status (businessmen, scientists, etc.).

**Table 1** Severe disabilities at 7 years of age in Isere county 1980–91 (1360 children, 1643 disabilities)

<table>
<thead>
<tr>
<th>Disability (1)</th>
<th>Birth years 1980–91 [n]</th>
<th>% within each type of disability</th>
<th>Proportion among all severely impaired children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Motor disability (1)</td>
<td>558</td>
<td>38.7</td>
<td></td>
</tr>
<tr>
<td>&quot;IMC&quot; among cerebral palsy</td>
<td>272</td>
<td>48.7</td>
<td></td>
</tr>
<tr>
<td>Other cerebral palsy</td>
<td>88</td>
<td>15.8</td>
<td></td>
</tr>
<tr>
<td>Nervous system congenital anomalies</td>
<td>37</td>
<td>6.6</td>
<td></td>
</tr>
<tr>
<td>Other congenital anomalies</td>
<td>66</td>
<td>11.8</td>
<td></td>
</tr>
<tr>
<td>Degenerative disease (hereditary or not)</td>
<td>52</td>
<td>9.3</td>
<td></td>
</tr>
<tr>
<td>Other motor disability</td>
<td>43</td>
<td>7.8</td>
<td></td>
</tr>
<tr>
<td>Psychiatric disorders (2)</td>
<td>353</td>
<td>24.5</td>
<td></td>
</tr>
<tr>
<td>Kanner autism</td>
<td>31</td>
<td>8.8</td>
<td></td>
</tr>
<tr>
<td>Other autisms</td>
<td>58</td>
<td>16.4</td>
<td></td>
</tr>
<tr>
<td>Psychosis with severe mental retardation</td>
<td>58</td>
<td>16.4</td>
<td></td>
</tr>
<tr>
<td>Other psychoses</td>
<td>206</td>
<td>58.4</td>
<td></td>
</tr>
<tr>
<td>Intellectual disability (3)</td>
<td>484</td>
<td>33.6</td>
<td></td>
</tr>
<tr>
<td>Down’s syndrome</td>
<td>167</td>
<td>34.5</td>
<td></td>
</tr>
<tr>
<td>Other chromosomal anomalies</td>
<td>20</td>
<td>4.1</td>
<td></td>
</tr>
<tr>
<td>Other congenital anomalies</td>
<td>63</td>
<td>13.0</td>
<td></td>
</tr>
<tr>
<td>Other intellectual disability</td>
<td>234</td>
<td>48.4</td>
<td></td>
</tr>
<tr>
<td>Hearing disability (4)</td>
<td>146</td>
<td>10.1</td>
<td></td>
</tr>
<tr>
<td>Without any other disability</td>
<td>85</td>
<td>58.2</td>
<td></td>
</tr>
<tr>
<td>With at least one associated disability</td>
<td>61</td>
<td>41.8</td>
<td></td>
</tr>
<tr>
<td>Visual disability (5)</td>
<td>102</td>
<td>7.1</td>
<td></td>
</tr>
<tr>
<td>Without any other disability</td>
<td>40</td>
<td>39.2</td>
<td></td>
</tr>
<tr>
<td>With at least one associated disability</td>
<td>62</td>
<td>60.8</td>
<td></td>
</tr>
</tbody>
</table>

**Severity criteria**

(1) None for Infirmité Motrice Cérébrale (IMC) which is part of the whole group entitled Cerebral Palsy (CP), defined as a clinical association of posture and movement disorders resulting from a non-progressive lesion on an immature brain. Need for specific help and/or continuous physiotherapy care was the severity criterion for the other motor deficiencies.

(2) ICD codes F84.0 to F84.5.

(3) IQ <50 or mental retardation classified as profound, severe, or moderate.

(4) Loss of >70 dB in each ear before correction.

(5) Visual acuity <3/10 in each eye after correction.

**Table 2** Prevalence rates of severe disabilities in children born 1980–91

| Disability                                   | Birth years 80–82 83–85 86–88 89–91 Total 95% CI |
|----------------------------------------------|-------------------|-------------------|-------------------|-------------------|
| Children with at least one severe disability | 314 308 351 387 1360 | 314 308 351 387 1360 | 314 308 351 387 1360 | 314 308 351 387 1360 |
| Of whom cerebral palsy                       | 134 132 144 148 558 | 134 132 144 148 558 | 134 132 144 148 558 | 134 132 144 148 558 |
| Psychiatric disorders                        | 65 65 105 118 353 | 65 65 105 118 353 | 65 65 105 118 353 | 65 65 105 118 353 |
| Of whom autism                               | 1.42 1.51 2.42 2.70 2.01 | 1.42 1.51 2.42 2.70 2.01 | 1.42 1.51 2.42 2.70 2.01 | 1.42 1.51 2.42 2.70 2.01 |
| Severe mental retardation                    | 0.31 0.28 0.81 0.64 0.51 | 0.31 0.28 0.81 0.64 0.51 | 0.31 0.28 0.81 0.64 0.51 | 0.31 0.28 0.81 0.64 0.51 |
| Hearing or visual disability                | 128 122 104 130 484 | 128 122 104 130 484 | 128 122 104 130 484 | 128 122 104 130 484 |

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professors, etc) among families with disabled children (46% versus 38%, and 8% versus 13% respectively, p < 0.001).

One hundred and nine disabled children (8.0%, range 6.6–9.4%) had a clear recognised postneonatal aetiology. Among the other severely disabled children with available information on pre/perinatal characteristics, 34.5% had a congenital anomaly, 6% had a very low birth weight (<1500 g), and 42% had been hospitalised in a neonatal care unit. Although there was a slight increase for the proportion of severely disabled children with a very low birth weight (from 5.9% to 8.1%, p = 0.12), the proportions with congenital anomaly or hospitalised in a neonatal care unit were remarkably stable during the studied period (p = 0.87 and p = 0.93 respectively).

One third of the severely disabled children (33.1%, range 30.1–36.1%), were born with a normal birth weight, without congenital anomaly, and had not been hospitalised during their first month of life. This proportion varied according to the type of deficiency: 25.1% for motor deficiencies, 22.0% for intellectual disability, 53.1% for psychiatric disorders, and 42.9% for sensory deficiencies.

DISCUSSION
According to our inclusion criteria, 7.73 per 1000 children born 1980–91 were registered with a severe disability at 7 years of age.

Data collection was conducted slightly differently before and after the 1984 birth cohort, with a retrospective data collection before 1984. This might account for the observed increase after 1984, as it is usually more difficult to find cases retrospectively than prospectively. However, we do not believe that this is the case, for two reasons:

- With the capture recapture method we were able to obtain a regular follow up of the degree of ascertainment, which remained stable over time, from 96% in the first period to 93% in the last period.
- We registered only severely disabled children, and it seems unlikely that children with such severe disability would have escaped retrospective data collection.

The increase in the overall prevalence rate for severely disabled children between 1980 and 1990 is mainly a result of the increase in the rates of cerebral palsy and severe psychiatric disorders. The cerebral palsy rate observed in our study is 2.04 per thousand, similar to the rates and trends observed elsewhere in Europe. The rate of autism observed over the whole period, 0.51 per 1000, is close to the rates published in other studies, and doubled during the study period from 0.31 to 0.64. The recent increase in the autism prevalence rate observed in different places has led to much discussion, without any clear and/or complete explanations being put forward. Gillberg et al., in Sweden, have suggested that the most probable reasons for this increase are classification bias and migratory movement into the area of study. Certainly it is necessary to obtain a more consistent definition of autism, and to have routine monitoring of autism trends, in order to avoid the problems of differences in definitions.

The rate of severe mental retardation remained stable during the 1980–90 period, 2.75 per 1000, while elsewhere in France a decreasing trend was observed, Down’s syndrome excluded, between 1976 and 1985. In England, the prevalence rate of severe mental retardation was higher in the 1970s, 3.0–4.1 per 1000 for children aged 5–9 years. Recent studies in other countries have shown a rate of around 3.0 per 1000 children.

The rate of severe visual deficiency was 0.57 per 1000, which is lower than the rate of 0.70 per 1000 observed in the Northern Europe register, but very close to the rate observed in Italy, 0.50 per 1000. This cannot be explained by differences in definitions, as the threshold for defining severity was the same in these studies. In France, Arnaud et al. observed great variations between counties, according to the number of specialised care places in each area.

Social class differences in the prevalence rates for neurodevelopmental disabilities have been described, and it has been suggested that the excess of disability in lower social classes could be the result of pregnancy complications, which are more frequent in deprived mothers.

In future studies about disabled children, the existence of a large group of children without any adverse pre/perinatal characteristics must be kept in mind. Knowledge of the high proportion of severely disabled children with congenital anomaly is useful for planning purposes, and also for early screening and care strategies in order to provide optimal support as soon as possible.

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We acknowledge the Conseil Général de l’Isère

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Images in Paediatrics

Dumbbell tumour of the mediastinum

A 6 month old male infant presented with fever and cough of one month duration. The chest radiograph showed bilateral radio-opacities occupying the medial aspect of each hemithorax (fig 1). Computerised tomography (CT) revealed a dumbbell shaped tumour occupying both hemithoraces (fig 2). The tumour was continuous across the midline by an isthmus passing posterior to the descending aorta and displacing the oesophagus anteriorly. The tumour had fat and soft tissue attenuation and did not enhance significantly. It was completely excised by bilateral posterolateral thoracotomy. Postoperative recovery was uneventful. Lipoblastoma was diagnosed on histopathology.

Lipoblastoma is a rare tumour of embryonal fat that occurs almost exclusively in children under the age of 3 years.1 The tumour presents as a localised, well circumscribed lesion known as lipoblastoma or a diffuse/multicentric form known as lipoblastomatosis.2 Most lesions present as a subcutaneous soft tissue mass on the extremities or head and neck. Truncal lesions are less common.3 Focal tumours behave in a benign manner and diffuse lesions may recur if incompletely excised.6 To our knowledge, only five patients with mediastinal lipoblastoma have been reported in the literature.4 This is the only instance of a dumbbell shaped mediastinal tumour occupying both hemithoraces.

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Arch Dis Child 2003 88: 117
doi: 10.1136/adc.88.2.117

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