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<th>Work package</th>
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<td>Task</td>
<td>T7.2 – Monitoring intellectual impairment</td>
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<th>Authors</th>
<th>Michelsen SI, Einarsson I, Boettcher L, vanBakel ME, Uldall P and Cans C</th>
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<tr>
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<td>External evaluator</td>
<td>Matt Muijen</td>
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| Internal Evaluation  |                                                                                             |
|----------------------|                                                                                             |
| Process indicators   | Recommendations to monitor severe intellectual impairment are produced page 27            |
| Output indicators    | Recommendations will be based on literature review and definitions used across Europe (reported from at least 1 Member state from each European region (N, E, S, W) page 39 |
| Outcome indicators   | Centres monitoring intellectual impairment will be able to provide data in a standardised way (at least 3 centres). page 42 |
Monitoring childhood intellectual disability
  - Studying the feasibility of a common European registration system

Michelsen SI, Einarsson I, Boettcher L, van Bakel ME, Uldall P and Cans C
with the collaboration of Craig S, Pildava S, Taube M, Marcelli M and Arnaud C
Preface

In the WHO background paper of 2010 "Better health, better lives: Children and young people with intellectual disabilities and their family" it is stated that "The effort to bring about meaningful change in this area is severely hampered by the general lack of easily available statistic information". The following European Declaration on this matter signed in Bucharest 26-27 Nov 2010 stated that one priority among the priorities for the European member states is "to collect essential information about the need and services and assure service quality". The present report sponsored by EU-DG-SANCO proposes and describes how to set up a network of data collection, some registers already existing in different European countries, with the further aim of sharing a European common database for children with ID. It is our hope that this report can be the background for such efforts to the benefit of people with intellectual disabilities.

The report has kindly been peer reviewed by Matt Muijen, regional advisor for mental health in WHO/Europe.
Summary ............................................................................................................................................................................................ 3

1. Introduction ................................................................................................................................................................................. 4
   1.1 Children with severe intellectual disability ............................................................................................................ 4
   1.2 Aim of monitoring children with severe intellectual disability ...................................................................... 5
   1.3 Aim of monitoring children with severe intellectual disability within the SCPE-NET project ............ 7

2. Definition and classification of children with severe intellectual disabilities .................................................. 7
   2.1 Literature on definition and classification ........................................................................................................... 10
   2.2 Workshop March 2011 ................................................................................................................................................. 14
   2.3 Suggestion of definition and classification for a monitoring system ......................................................... 15

3. Tools assessing severe intellectual disability in children ...................................................................................... 16
   3.1 The validity of IQ measures ...................................................................................................................................... 16
   3.2 Particular IQ measures (tests) ................................................................................................................................. 18
   3.3 Suggestion of tools for use in a future monitoring system ............................................................................ 20

4. Existing monitoring systems in Europe ........................................................................................................................ 21
   4.1 Literature on existing monitoring systems ........................................................................................................... 21
   4.2 Survey among SCPE centres ....................................................................................................................................... 22
   4.3 Contact with relevant institutions in Europe ...................................................................................................... 23
   4.4 Workshop September 2011 ........................................................................................................................................ 26
   4.5 Existing registers on ID in Europe ........................................................................................................................... 27

5. Suggestion of monitoring system in Europe ............................................................................................................... 35
   5.1 Aims of the future common database ..................................................................................................................... 35
   5.2 Recruitment sources of the future common database ............................................................................................ 35
   5.3 Inclusion criteria of the future common database ............................................................................................ 36
   5.4 Classification of intellectual functioning in future common database ...................................................... 36
   5.5 Variables other than intellectual disability in future common database ................................................. 37
   5.6 Denominators for estimation of prevalence ........................................................................................................ 39
   5.7 Current availability of suggested variables in existing registers of ID in Europe ................................... 39
   5.8 Implementation opportunities of a common database: strengths and weaknesses .................................. 41
   5.9 Potential participants in a new collaboration of registries on childhood ID ............................................. 42

6. Ethical considerations .......................................................................................................................................................... 46
Summary

Children with a severe intellectual disability have severe impairment in learning, difficulties with verbal communication and social adaptation in daily life. They need substantial support throughout life. Notwithstanding this, intellectual disability is a neglected area in public health with few reliable estimates of prevalence and few monitoring systems able to identify children with severe intellectual disabilities (1-4). Information on people with intellectual disabilities is lacking and comparable data on the prevalence and characteristics of children with intellectual disabilities is needed.

This report focuses on children with severe intellectual disability (ID) defined as IQ<50 and examines the feasibility of setting up a common European database for these children. The aims of such a database would be to describe characteristics of subgroups, monitor prevalence, trends and type of living and schooling of children with severe ID across Europe and to provide a cross-country analysis that can assist in improved understanding of the needs of children with severe ID.

On that basis we have drawn on the experience obtained from SCPE-net, an already existing network on cerebral palsy, SCPE-NET. SCPE-NET is a European project monitoring cerebral palsy in several European regions and countries. The monitoring system proposed builds on the work already under way in local registries. The network has 14 years of experience in harmonisation and collection of data and has included an aim of making recommendations for monitoring early childhood impairments at regional or national level.

Three key areas were examined before proposing a framework for a common database on children with severe ID and these areas are presented in the report as follows:

- the current definitions and classifications (section 2)
- available cognitive tools for measuring ID, compared and discussed (section 3)
- existing monitoring systems, that have been examined by way of literature review, through a survey among existing cerebral palsy registries and through contacts with relevant national institutions (section 4).

Existing registers are described and a future common database is proposed drawing from discussions held at cross-country workshops (section 4). On the basis of the findings of these activities, a proposal for a European network with a common database is presented (section 5).

We have adopted the American Association of Intellectual and Developmental Disabilities (AAIDD) term of intellectual disability defined as significant limitations in intellectual functioning, significant limitations in adaptive behaviour and onset before age 18.

We assume that description of ID will be derived from existing medical or psychological files. Tools used for assessing intellectual functioning are expected to vary between and within countries. We
suggest the inclusion of validated test instruments whenever possible, and reporting the name of the test, separate performance and verbal scores (WISC-III) or Cluster scores (WISC-IV) as well as a total score. In cases where clinical estimation is the only way of describing the child's intellectual functioning we suggest that this should preferably be multidisciplinary and supported by some test results.

We have restricted inclusion in the common database to children with a total IQ<50 (lowest value of the confidence interval) with an age of 6-8 years at diagnosis. The rationale for restricting the database to children with severe ID will, it is anticipated, result in fewer false diagnoses due to lack of information on adaptive behaviour. We suggest including results from a standardised test of adaptive behaviour whenever available and this should be aspired to in the future. We do not consider it likely that this information will be available at the present time for a substantial number of children. In addition it is proposed that information on comorbidity, schooling, living situation, birth as well as socio-demographic information on parents should be included.

Recruitment sources will differ between regions, depending on the organisation of health, social and educational sectors. Completeness of any register depends on the number and type of recruitment sources. Each register should specify and estimate type and number of recruitment sources needed to yield at least 80% completeness of the register. The estimation should be based on national/local validation studies where available or, if not, on references from national/local experts in the field.

Five registers (Iceland, Ireland, Latvia and two in France) that include children with severe ID exist in Europe and have potential to form part of a network charged with the task of establishing a common database. As they differ considerably in data collection practices and type of variables collected, harmonisation of data would be required to be able to contribute to a common database. Other regions familiar with registration of children with cerebral palsy in Norway, Italy and Denmark have indicated a willingness to join such a network in the future. Other registers or monitoring systems could be encouraged to follow.

1. Introduction

1.1 Children with severe intellectual disability
This report focuses on children with severe intellectual disability (ID) and examines the feasibility of a common European database for these children. Severe ID refers to children with an estimated IQ of 50 or below referring to both moderate (34<IQ<50), severe (19<IQ<35) and profound (IQ<20) intellectual disability categories. Ideally a common European database should also include children with mild intellectual disability but recruitment of children with mild intellectual disability is very difficult, includes a wider range of recruitment sources and so children with mild intellectual disability are known to have a significantly lower rate of identification (5). As full coverage is a key requirement for reliable data in a register network, we will focus on children with a severe intellectual disability only.
In western countries, the prevalence of severe ID (IQ<50) among school-aged children is around 3-4 per 1000 (6). This prevalence rate is clearly age-dependent, showing an increasing trend till the age of 15, probably reflecting how children are identified by service providers or schools (6,7). After the age of 15 there is a decreasing trend, probably due to higher mortality rates or differences in study populations or methods (6). Gender differences with an approximate 20% excess of males have consistently been reported (6).

Children with severe intellectual disability have severe impairment in learning, difficulties with verbal communication and social adaptation in daily life. They need substantial support throughout life. Notwithstanding this intellectual disability is a neglected area in public health with few reliable estimates of prevalence and only a small number of monitoring systems that are able to identify children with severe intellectual disabilities (1-3). Many children with intellectual disability are still not known to the health and public services in the respective countries of the EU(8).

1.2 Aim of monitoring children with severe intellectual disability

The establishment of a monitoring system of children with severe intellectual disability can address a number of aims including:

- profiling of children,
- greater identification of their needs and
- provision of data that can assist in a preventive way to address the likely causes of severe intellectual disability.

Overall there are two broad aims associated with registering children with severe intellectual disability: 1) Preventing severe intellectual disability and 2) Ensuring a high quality of life and participation in everyday life, as well as equal access to health care of children with severe intellectual disability. The second aim is supported by UN Conventions; for example, the Convention on Rights of Persons with Disabilities (article 23) states that “Children with disabilities should be able to participate on an equal basis with others in family life, health maintenance, education, public life, recreational, leisure and sporting activities” (2006).

Figure 1 shows ways of reaching these goals of prevention and support.
In a common monitoring system the following questions and hypotheses might be tested:
1) Do habilitation/rehabilitation strategies vary from region to region despite similar severity of disability?
2) Does prevalence of ID vary from region to region and is it dependent on socioeconomic conditions?
3) Would the introduction of the classification system ICF-CY improve achievements at school?
4) Does intensive care improve achievements at school?

Many variables are required to describe aetiology of intellectual disability and the life and needs of children with intellectual disability but too many variables might prevent full harmonisation and reduce the relevance of a European database. Completeness of information and accurate identification of children should have the highest priority at a common database level. A key consideration is the need for agreement on a small number of core variables – a minimum dataset – that would be common in all countries. A broad description of intellectual disability could - in addition to the monitoring purpose - also act as a basis for more specific research projects into severe intellectual disability in childhood.

A European monitoring system of children with severe intellectual disability could be based on representative local or regional registers in different countries delivering their data to a common database, in the same manner as the SCPE network is monitoring children with cerebral palsy. The absolute number of children with severe ID within each register each year is low and insufficient to study various etiological and clinical subgroups (for instance ID as a consequence of intra-uterine growth restriction or the co-existence of associated impairments) as well as service needs and provision for these subgroups.
1.3 Aim of monitoring children with severe intellectual disability within the SCPE-NET project

SCPE-NET is a European project monitoring cerebral palsy in 16 countries. The monitoring system is composed of 20 local registries providing data for a common database. The network has 13 years of experience in harmonisation and collection of data.

One specific objective of the SCPE-NET project is to make recommendations for monitoring early childhood impairments at regional or national level. The aim of task 7.2 within the SCPE-NET is to check the feasibility of applying, across Europe, the knowledge and experience gained from the network of cerebral palsy registers to other childhood impairments and chronic conditions such as intellectual impairment. The outcome of this task is to generate information to assist service planning, to better understand causes and improve clinical practice of children with severe intellectual disability.

For this, we need to be able to:

1) Describe characteristics of children with severe ID
2) Describe aetiology of severe ID in children
3) Monitor prevalence and trends in severe childhood ID and subgroups (according to various co-existing disabilities or etiological groups)
4) Monitor types of living and schooling in children with severe ID

2. Definition and classification of children with severe intellectual disabilities

An agreed definition and classification of childhood severe intellectual disability is needed to establish a common monitoring system and this may differ from approaches used for clinical purposes. Today the term intellectual disability has broadly replaced the term mental retardation in official documents and general use. However mental retardation is still used in some classifications such as ICD-10 (WHO) and DSM-IV (American Psychiatric Association), while the AAIDD (American Association on Intellectual and Developmental Disabilities) employs the term intellectual disability. The AAIDD states, however, that the term intellectual disability covers the same population as did the former term mental retardation. Definitions of mental retardation/intellectual disability are moving from descriptions of global incompetence to descriptions of abilities and participation. In addition, the majority of the recent definitions focuses on functioning taking the interaction between an individual and the context into account, which is in accordance with the International Classification of Functioning, Disability and Health (ICF-CY)(9).

Table 1 gives an overview of the most widely used definitions today.
Table 1 Formal definitions of mental retardation or intellectual disability

<table>
<thead>
<tr>
<th>Classification of Diseases (ICD-10 WHO 1990)</th>
<th>Mental retardation</th>
<th>Definition*</th>
<th>Classification</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td>1. Arrested or incomplete development of the mind</td>
<td>Mild 50-69</td>
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<td></td>
<td></td>
<td>2. Impairment of skills which contribute to the overall level of intelligence (i.e. cognitive, language, motor, and social abilities)</td>
<td>Moderate 35-49</td>
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<td></td>
<td>3. Manifested during the developmental period</td>
<td>Severe 20-34</td>
</tr>
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<td></td>
<td></td>
<td></td>
<td>Profound &lt;20</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Can be supplemented by scales assessing social adaptation in an environment (no or minimal vs. significant impairment of behaviour)</td>
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<thead>
<tr>
<th>Diagnostic and Statistical Manual of Mental Disorders (American Association of Psychiatrists DSM-IV 1994)</th>
<th>Mental retardation</th>
<th>Definition*</th>
<th>Classification</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>1. Significantly sub average intellectual functioning (IQ approx. 70 or below, for infants a clinical judgement)</td>
<td>Mild 50-55 to 70</td>
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<td></td>
<td></td>
<td>2. Concurrent deficits or impairments in adaptive functioning (in at least two of the following areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety)</td>
<td>Moderate 35-40 to 50-55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. Onset before age 18 years</td>
<td>Severe 20-25 to 35-40</td>
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<td></td>
<td></td>
<td>Profound &lt;20</td>
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<tr>
<td></td>
<td></td>
<td>1. Significant limitations in intellectual functioning (approx. 2SD below the mean with standardised measure)</td>
<td>Needed support to optimize functioning in five areas</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Significant limitations in adaptive behaviour as expressed in conceptual, social and practical adaptive skills (approx. 2SD below the mean in one of the three types or in overall adaptive behaviour measured with standardised measure)</td>
<td>I. Intellectual</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. Limitations should be apparent before age 18</td>
<td>II. Adaptive (conceptual, practical, social)</td>
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<td>III. Participation, interaction, social role</td>
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<td>IV. Health (physical, mental, aetiology)</td>
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<td>V. Social context (environment, culture, opportunities)</td>
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<tr>
<th>WHO definition (13)</th>
<th>Intellectual disability</th>
<th>Definition*</th>
<th>Dimensions</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td>1. Impaired intelligence (significantly reduced ability to understand new or complex information and to learn and apply new skills)</td>
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<td>2. Impaired social functioning (reduced ability to cope independently)</td>
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<tr>
<td></td>
<td></td>
<td>3. Begins before adulthood and with a lasting effect on development</td>
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*All points listed in each classification are necessary to fulfil the definitions
All definitions agree on onset in childhood. Both DSM-IV and AAIDD agree on deficits in adaptive behaviour as inclusion criteria, while ICD-10 suggests supplementing with a social adaptation assessment. Both ICD-10 and DSM-IV agree on a lower cut-off for mild mental retardation around IQ 50-55. The AAIDD no longer use statistical distribution of IQ scores to divide children with intellectual disability into subgroups; rather they recommend evaluation of five areas of functioning of the child (intellectual, adaptive, participation, health and social context). Nevertheless they still recommend use of SD for assessing intellectual functioning.

In 2013, DSM-V will be released which is expected to differ substantially from DSM-IV. The term “mental retardation” will be replaced by “intellectual developmental disorder” (14). Intellectual developmental disorder will be defined as deficits in general mental abilities with an intellectual deficit of at least 2 SD below the population mean for a person’s age and cultural group measured by an individualized, standardized, culturally appropriate and psychometrically sound test and with onset during the developmental period. The deficits relate to impaired functioning by limiting participation in one or more aspects of daily life resulting in the need for on-going support at school, work or independent life. Thus, intellectual developmental disorder requires a significant impairment in adaptive functioning, typically measured by an individualized, standardized, and culturally appropriate and psychometrically sound test.

An ICD-11 working group constituted of 30 experts from 13 countries very recently suggested a new framework for mental retardation/intellectual disability (15). The term “Intellectual developmental disorders” (IDD) is very similar to the term “Intellectual Disability (ID)” . IDD is considered as a clinical meta-syndrome, whilst ID refers to the functioning/disability counterpart. These two terms have replaced the term mental retardation. IDD is a health condition and not solely a constellation of disabilities. The proposed definition is as follows: “a group of developmental conditions characterized by significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behaviour and skills.”

Six main descriptors are proposed for this definition, of which two are of particular relevance to this report:

- Persons with IDD have difficulties with verbal comprehension, perceptual reasoning, working memory and processing speed.
- Persons with IDD typically manifest difficulties in adaptive behavior; that is, meeting the demands of daily life expected for one’s age peers, cultural and community environment. These difficulties include limitations in relevant conceptual, social, and practical skills.

The determination of IDD severity should rely on clinical description of the characteristics of each subcategory, IQ score should be considered as one clinical descriptor among others.

Among mental functions (b100-b199 codes), the ICF-CY defines intellectual function (b117) as “general mental functions, required to understand and constructively integrate the various mental functions, including all cognitive functions and their development over the life span” (9). Functions included in this area of b117 are functions of intellectual growth, intellectual retardation, mental retardation and dementia, while functions not included are memory functions (b144), thought functions (b160), basic cognitive functions (b163) and higher-level cognitive functions (b164). In addition, ICF-CY describes also other specific mental functions, each one with a specific code bxxx and
include attention, memory, psychomotor, emotional, perceptual, thought, basic cognitive, higher-level cognitive, mental functions of language and calculation functions (9).

In this report we will use the term “intellectual disability”.

2.1 Literature on definition and classification

Searches for scientific literature on definition and classification of children with severe intellectual disability were performed in PubMed from 1980 onwards.

The Mesh term “Intellectual disability” with subheading “Classification” was used and the search was limited to children age 0-18 years. In total 249 papers were identified and 16 papers with abstract or title mentioning definition or classification of ID were selected for full reading. A synthesis of seven papers on definition and classification is presented in Table 2 and described in this section.

Two papers from the 1980s focused on the importance of an organic aetiology of intellectual disability (16,17). They suggested a distinction between a “familial group” consisting of children whose intelligence is solely determined by a combination of genetic and environmental factors and “an organic group” which is further determined by an organic aberration (16). Most of the children with severe intellectual disability will be in the “organic group”. They state that research that does not take into account that aetiology will be confounded and will obscure much valuable information (17).

The importance of behavioural capacities is recognized, but it is recommended that social competences be described rather than used as inclusion criteria for the definition. This is so, because social competence is not sufficiently well-defined and also because social behaviour is age related while intelligence scores are more stable (16).

Instead of differentiating on the level of intellectual disability or aetiology, the AAIID definition of ID from 1992 focuses on the level of support and includes an assessment of adaptive behaviour. A review from 1994 summarizes the implications of this definition of ID (18). It is stressed that ID is no longer an absolute trait in a person but is a result of the functional impact from the interaction between personal capacities and the environment. The reviewers conclude that there is still a lack of reliable measures of adaptive behaviour and support needs. A number of other authors from the US also acknowledge that this deficit will increase the number of false positive and false negative classifications, particularly in the upper ranges of intellectual and adaptive behaviour continua (18). In addition, the decision not to distinguish subgroups or to enlarge the classification, but to focus on the need of support compromises how we specify services for different subgroups. However, these points should affect less the classification of the more severe forms of intellectual disability (18). When adaptive behaviour is taken into account, some of the children with cognitive impairment based on psychometric evaluation, would shift ID category (19,20). However children who would shift ID category between moderate, severe and profound categories would probably still require services, and therefore the misclassification is less problematic. Also the lack of suitable psychometric measures for the different adaptive domains is also an issue (21). It is suggested that adaptive behaviour develops during preschool years and consequently a precise measuring instrument is crucial (22). In practice,
adaptive behaviour is more likely to be ignored in epidemiological ID research. Both the American Psychological Association and the American Psychiatric Association (1994) have opted to retain descriptive levels of severity based on IQ for the time being.

An Australian review from 2002 discusses definition and methodology of studies on ID (23). As is evident, the ability to follow trends in prevalence as well as compare across populations depends on a stable definition, classification and exhaustive data sources in epidemiological studies. It is suggested that information about diagnosis (for example from ICD-10) supplemented with functioning (for example via ICF) provides a broader and more meaningful picture of ID in the population (23).

A comprehensive article on mental retardation from 2006 recommended the use of standardised measures of intelligence and adaptive behaviour, while considering cultural and environmental differences as well as associated impairments (24). Around school age these measures can be expected to correlate well with assessment later in life.
<table>
<thead>
<tr>
<th>Author, country, year and title of publication</th>
<th>Aim of paper</th>
<th>Definition and classification</th>
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<tr>
<td>Johnson CP et al. USA 2006 (24) &quot;Mental retardation: diagnosis, management, and family support&quot;</td>
<td>Describe diagnosis, search for aetiology, management and family support for ID.</td>
<td>Children with mild ID are likely to be identified not until they enter an academic setting, moderate at age 3-4 years and severe before age 1. Recommend to use standardised measure of intelligence and adaptive behaviour, while considering cultural and environmental differences as well as associated impairments and to define individual strength and needs across the five dimensions of AAIDD</td>
</tr>
<tr>
<td>Leonard H et al. Australia 2002 (23) &quot;The epidemiology of mental retardation: challenges and opportunities in the new millennium&quot;</td>
<td>Study prevalence data of ID according to age, gender, social class and ethnicity. Include issues of definition and methodology.</td>
<td>Information about diagnosis from ICD-10 supplemented with functioning (ICF) provides a broader and more meaningful picture of ID in the population. Epidemiological studies often focus solely on IQ for definition of ID, partly because no standardized measure of adaptive behaviour exists. Use of adaptive behaviour or nonverbal intellectual measures reduce false positive. Stable definition and classifications as well as type of data sources and purpose of data collection are critical considerations in epidemiological studies.</td>
</tr>
<tr>
<td>Vig et al. USA 1996 (22) &quot;Application of the 1992 AAMR definition: Issues for preschool children&quot;</td>
<td>Report special issues for preschool children of the new definition and classification of ID.</td>
<td>Researchers need level of ID to compare samples of children with varying degree of ID. Adaptive behaviour skills develop during preschool years for children with and without ID and consequently a measuring instrument is crucial. The Vineland Adaptive Behaviour Scales present four domains and only three of these resemble the ten areas in the new definition. In conclusion the new definition of ID without classification levels and limited relevance of adaptive behaviours is not useful for preschool children</td>
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<tr>
<td>Schalock RL et al. USA 1994 (18) &quot;The changing conception of mental retardation: Implications for the field&quot;</td>
<td>Summarize current thinking about the 1992 definition of ID and discuss major implications.</td>
<td>The new definition reflects the emerging trends in ID, but has implications: ID is no longer an absolute trait expressed solely by a person, but an expression of functional impact of an interaction between the capabilities of a person (expressed by intelligence and adaptive skills) and the person’s environment. There is a lack of standardized measures of adaptive behaviour and support needs. A change in definition potentially affects prevalence rates, but not necessarily upwards since the dual diagnosis demands reduced adaptive skills as well. Implications for research are increased costs in obtaining extended and more precise descriptions of individuals. This will result in better epidemiology and less emphasis on the individual level and more on the effect of the environment.</td>
</tr>
<tr>
<td>Author, country, year and title of publication</td>
<td>Aim of paper</td>
<td>Definition and classification</td>
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<tr>
<td>MacMillan D et al. USA 1993 (21) “Conceptual and psychometric concerns about the 1992 AAMR definition of mental retardation”</td>
<td>Comment on whether the “new” AAIDD definition of ID (1992) guides clinicians and researchers in classification.</td>
<td>New approach to adaptive skills introduces greater unreliability into the diagnostic process. No suitable psychometric measures are available for all the new domains. Not recognizing different subcategories results in a less precise and less reliable classification. Adopting IQ 75 cut off expands the number of people with ID.</td>
</tr>
<tr>
<td>Burack JA et al. USA 1988 (17) “Issues in the classification of mental retardation: differentiating among organic etiologies”</td>
<td>Demonstrate utility of differentiating among the various forms of organic retardation.</td>
<td>Two curves of intelligence exist. One comprises persons whose intelligence is determined by a combination of genetic and environmental factors and one of persons whose intellectual potential has been affected by organic insult. Research that does not specify aetiology will be confounded, obscuring much valuable information.</td>
</tr>
<tr>
<td>Zigler E et al. USA 1984 (16) On the definition and classification of mental retardation”</td>
<td>Proposal of classification system of ID based on etiology.</td>
<td>Suggest classification with three groups based on aetiology: organic damage (supplemented with specific diagnosis), familiar (result of interaction between hereditary and environment) and undifferentiated. Recommends the use of only IQ as a classificatory principle, but with additional description of correlates (for example health problems, physical stigmata and information on siblings and parents). Important behavioural consequences exist of having intellectual disability being mentally retarded due to organic causes. However social adaptation is not sufficiently well-defined to be a criterion for ID and behavioral capabilities should be used not only to define groups, but to help describe them. Using IQ plus social competence to define mental retardation makes it impossible to determine the true prevalence of ID in a society. The age aspect of classification is central. For the purpose of planning services we need to be able to predict future behaviour long after the initial assessment. However early intelligence scores of children with severe physiological damage do predict later intellectual ability relatively well. While chronological age has little effect on performance in intelligence tests, age probably affects less cognitively demanding social behavior and interests.</td>
</tr>
</tbody>
</table>
2.2 Workshop March 2011

A SCPE network workshop entitled “Definition, classification and measures of childhood intellectual impairment” was held in March 2011 in Grenoble, France. Participants came from a range of disciplines including epidemiology, psychology, special education, neuropaediatrics and neuropsychiatry and from several registers of the SCPE network with experience and interest in registering children with severe intellectual impairment (Denmark, Iceland, Latvia, Italy and France (Grenoble/ Toulouse). The aim of the workshop was to discuss harmonization of definition and classification criteria of severe childhood intellectual impairment and to agree on a common European definition for the purposes of registration.

Terminology

Terminology within the area of intellectual disability was discussed. Most SCPE partners use the term intellectual impairment when identifying children with cerebral palsy with an estimated intelligence quotient below 70. However, intellectual disability is the most widely used term internationally and all children with a severe intellectual impairment are expected to have an intellectual disability. Although mental retardation is still mainly used in the medical classifications, intellectual disability is agreed as the common term to be used in the future for those willing to register children with such impairment.

Focus on children with severe intellectual disability

Any future European monitoring system would ideally include all children with intellectual disability. However, we suggest the inclusion of only the group of children with severe intellectual disability with an estimated IQ < 50, as the majority of this group is likely to be in contact with a welfare system in order to receive special services. The rationale for the selection of this group is that it is easier to trace, more likely to be exhaustive with reduced ascertainment bias. Also, it represents the cut-off most widely used in epidemiological studies (23) and conforms to the various definitions. At a later stage it may be possible to examine the inclusion of children with mild ID (50-70 IQ).

Definition

With the aim of pooling data from several registers in a common database on severe childhood intellectual disability, there is a need for an ID definition suitable for the purposes of registration. Three major classification systems exist. ICD-10 classifies according to assessment of IQ, DSM-IV according to a combination of assessment of IQ and adaptive behaviour, while the definition from AAIDD is mainly based on adaptive behaviour and a non-quantified decreased IQ, based on the likelihood of benefit from educational services.

Although the classifications might be good for identifying ID, they are often less useful for goals assessment and for the determination of supports. The ICF (conceptually very different from other classifications) is complementary by focussing on the functional aspects of the disorder.

Classification

Classifying ID is an important aspect of the work overall. IQ measurement might be useful for research, while information on adaptive behaviour as well as activities and participation as set out in the ICF
could be essential for policy and service planning. Also levels of adaptive behaviour could be important to avoid false negatives in research as well as the appropriate clinical setting especially for the group of children with mild intellectual disability.

Most classification systems are based on statistical distribution of IQ scores. The number of standard deviations from the mean delimit the mild (2SD) and severe ID (3SD) categories. However classifications today increasingly include a measure of adaptive behaviour. As noted earlier, the AAIDD and DSM-IV directly includes deficit in adaptive behaviour in their definition of ID while ICD-10 (WHO) suggests an evaluation of adaptive functioning as supplementary information to the intellectual disability diagnosis.

In each regional register data are obtained from sources such as medical or psychological files or reports from service providers. For some of the children a cognitive evaluation will be present, while for others the assessment will rely on a clinical estimation (mainly for the most severe cases). However, at present the evaluation of adaptive behaviour is not common for the majority of children.

**Age at assessment/inclusion**
Most children with severe intellectual disability are identified and diagnosed early in life in the health care system and evaluations of intellectual functioning correlate well with later assessments from around school age. To be able to publish prevalence data and trends using a common database on children with severe intellectual disability it is important to restrict the age for inclusion. Also although intellectual functioning might not change considerably over time, other collected variables included in the database (e.g. epilepsy and type of schooling) might do so and a narrow age range for inclusion in the database is preferred.

**2.3 Suggestion of definition and classification for a monitoring system**
We propose to use the term intellectual disability and our definition includes:

1) Significant limitations in intellectual functioning
2) Significant limitations in adaptive behaviour
3) Onset before age 18

As feasible inclusion criteria for the common database we also suggest:

1) Total IQ<=50 (lowest value of the confidence interval)
2) Age 6-8 years at IQ estimation

At present a measure of adaptive behaviour is not part of our suggested criteria, but we recommend including additional information on adaptive behaviour whenever available.
Classification according to level of ID is as follows:
1) IQ < 20
2) IQ 20 - 34
3) IQ 35 - 50

Among children with information on adaptive behaviour, an additional categorization should be considered based on results of a standardised test of adaptive behaviour.

3. Tools assessing severe intellectual disability in children

We know from the literature that there are many challenges in acquiring IQ measures of young children with intellectual disability and below we describe these challenges.

3.1 The validity of IQ measures

In classical test theory validity denotes the agreement between the test score (e.g. IQ) and the characteristic it is supposed to measure (level of intellectual functioning). The validity coefficient is supposed to guide us in deciding the extent to which the outcome/measure represents or reflects the true ability of the child.

The different tests help us to estimate the IQ of the child. Based on the IQ estimate of a test, the child may be categorized according to one of the ID categories (Table 3). It is desirable to avoid both false positives and false negatives.

Table 3 Validity of tools to measure intellectual disability

<table>
<thead>
<tr>
<th>Test score shows low IQ</th>
<th>Test score shows normal IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child with intellectual disability</td>
<td>True positive. Valid result</td>
</tr>
<tr>
<td>Child with normal intellectual abilities</td>
<td>False positive</td>
</tr>
</tbody>
</table>
The test theory approach can be supplemented with the decision theory approach introduced by Cronbach (25). From this point of view, validity is concerned with the relationship between the information generated by the test (e.g. IQ) and a criterion variable (identification of individuals with intellectual disability for registration purposes). The focus is on how well one or more tests help in the decision about whether a particular child belongs to the category 'intellectual disabled' or a particular level of intellectual impairment?

It is crucial to know, therefore, the validity of different IQ measures and the extent to which IQ obtained with different measures have high correlates with each other (e.g. Bayley Scales of Infant Development second or third version with Wechsler Intelligence Scale for Children third or fourth version (WISC-III or IV)).

The problem of reliability and suggestions of how to use IQ measures to guide reliable decision-making about intellectual functioning

Reliability concerns the extent to which measurements are accurate. Any test result will include measurement error, but it is important to ensure that measurement error is as small as possible. The validity of a measurement requires that the measurement is reliable. However, the assumption that error is random might be violated in relation to children with intellectual disability. First of all, children with ID often show more variability in their performance than children without ID. Secondly, many children with intellectual disability have additional impairments; motor impairments, sensory impairments, perceptual impairments, communicational impairments that interfere with their performance on most tests. This may weaken both reliability and validity of IQ measures of children with intellectual disability. However, the tests might still inform the examiner about the cognitive functional level of the child by the use of qualitative interpretations of the child's performance and thus their categorisation.

We suggest the following use of test scores in deciding the child's level of intellectual functioning:

- Tests completed without adaptation or with minor adaptations only -> IQ score including confidence interval.
- Tests completed with significant adaptations -> IQ score might not be reliable or valid as an exact measure, but might still be a valid guide in the clinical estimation of the child's level of intellectual functioning.
- Tests only partially completed -> Child's performance used to guide clinical estimation of the child's level of intellectual functioning.
3.2 Particular IQ measures (tests)

WISC-IV
The Wechsler Intelligence Scale for Children, fourth version (WISC-IV) is aimed at children in the age range of 6 to 16.11 with IQs from 40 and upwards. The psychometric properties of the test are very good, including the fact that the floors and ceilings of the different subtests are good (the test has good discrimination in the lower range too) and the test can be used confidently for the identification of individuals with intellectual disability (26). For children with IQ below 40, subtests from the WISC-IV can be used to guide clinical estimation. For children with severe IQ, WISC-IV will often need to be substituted with other measure or clinical estimation as formal assessment will often not be feasible.

WISC-IV provides Full Scale IQ (Population mean 100, standard deviation 15) based on 10 subtests and four index scores; verbal comprehension, perceptual reasoning, working memory and processing speed. Interpretations of the child level of cognitive functioning based on one subtest only has a low level of validity as the base rate of getting one low subtest is quite high in the general population (64.5% in a standard population with one subtest score 1 standard deviation below the mean(27). Index scores can be used to estimate the level of cognitive functioning of a child even though they do not provide an IQ. This may be relevant if a child has a very varied performance as the full scale IQ may be invalid if the child’s scores in different subtests differ between each other with 15 points or more.

The main difference between WISC-III and WISC-IV is the discarding of the verbal/performance IQ from the third to the forth version. The correlation between WISC-III and WISC-IV is high, although not perfect (Table 4). One particular problem that is worthy of attention is the Flynn effect. Since the introduction of standardised IQ measures, there has been a systematic rise in IQ, at least in the western part of the world (28,29). To counter this rise in IQ measures, the difficulty of the different versions of Wechsler Intelligence Scales for Children has been adjusted accordingly, each time the test has been re-normed, to keep the population mean at 100. Typically, this has happened in relation to the introduction of a new version. This means that an individual that received a score of 105 on the WISC-R (the WISC before WISC III) would on average receive a score of 100 on the new WISC-III. Similarly, clinicians have reported the fourth version to be more difficult for the individuals who are tested than the previous one. This decline in IQ in association with the introduction of a new version of the Wechsler Intelligence Scales for Children has been reported in children with ID (30). We therefore recommend that the particular version of the Wechsler Intelligence Sales for Children is reported along with the test result.

A short form of the WISC-IV has been developed, consisting of seven subtests. It shows excellent correlation with the full version of the test (r=.99 (31)). The employment of the short form in clinical use is unknown.

The authors of the WISC-IV have mainly correlated WISC-IV with other measures they have published. Correlations between WISC-III and WISC-IV and other measures have been found in different studies in journal articles and book chapters (26) (32-35). In table 4, those most frequently used IQ tests for
children with ID have been listed along with the relevant age range and their correlations with WISC-III and IV.

**WPPSI-R, WPPSI-III**
The Wechsler Preschool and Primary Scale of intelligence- revised and third version (WPPSI-R and WPPSI-III) are downward extensions of the WISC tests to younger age groups.

**Bayley-II and Bayley-III**
Both Bayley scores (II and III) contain a full test and a screening test. It provides an age-standardized composite score for cognitive scale (mean 100; SD 15; range 55-155). The Bayley tests are aimed at children between 1 month and 3.5 years but are sometimes used for older children with intellectual disability, for whom the WPPSI or WISC tests are too difficult or inappropriate.

**Griffiths’ Mental Development Scales (GMDS-ER 2-8)**
Griffiths’ Mental Development Scales was developed to measure development in infants and small children. The current version is from 2006 and has been extended upwards to cover the age range from birth to eight years. It consists of six subscales that are combined in a General Quotient. The application of GMDS-ER in children with intellectual disability is similar to the Bayley Scales. However, the GMDS-ER has been found to result in higher age equivalents than those obtained with the Bayley Scales both in typical children (36) and in children with Down syndrome (37). The correlation between older versions of the Griffiths’ scales and the Bayley Scales has been found to vary between .73 and .97 (36,38). The literature recommends that scores from the Griffiths and the Bayley Scales are not used interchangeably (36,38).

**Leiter-R**
Leiter- R is a revised version of the original Leiter test, comprising both a full and a short version. The test has been developed as a thoroughly non-verbal test. The Leiter-R Brief and Full IQ have been found to correlate well with each other and this indicates that the Brief IQ can be used as a proxy for a Full IQ (39).

**SON-R (Snijders-Oomen non-verbal intelligence test)**
The SON-R test was developed for deaf children and contains guidelines for the interpretation of this test for children with intellectual disability, autism, language disorders, deaf or hear-of-hearing and immigrant children.

**The Differential Ability Scale (DAS)**
The DAS is considered especially useful for preschool children and for low-functioning children since it allows for determination of ability structure and calculation of IQ scores below 45. The test has been translated into several languages.
### Table 4: Correlations between IQ measures in use

<table>
<thead>
<tr>
<th>Measure</th>
<th>Outcome</th>
<th>Age range</th>
<th>Correlation with WISC-IV</th>
<th>Correlation with WISC-III</th>
</tr>
</thead>
<tbody>
<tr>
<td>WISC-IV</td>
<td>IQ</td>
<td>6-16.11 years</td>
<td>1.0</td>
<td></td>
</tr>
<tr>
<td>WISC-III</td>
<td>IQ</td>
<td>6-16 years</td>
<td>.89</td>
<td>1.0</td>
</tr>
<tr>
<td>WPPSI-R</td>
<td>IQ</td>
<td>2.11-7.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WPPSI-III</td>
<td>IQ</td>
<td>2.6-7.3 years</td>
<td>.89</td>
<td></td>
</tr>
<tr>
<td>Bayley-II</td>
<td>MDI or MA</td>
<td>1 month to 3.5 year</td>
<td>.73 with WPPSI-R</td>
<td></td>
</tr>
<tr>
<td>Bayley-III</td>
<td>IQ</td>
<td>2-20 years</td>
<td>.86</td>
<td></td>
</tr>
<tr>
<td>Leiter-R</td>
<td>IQ</td>
<td>4-11.11 years</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Raven (CPM)</td>
<td>IQ</td>
<td>2.6-7 year</td>
<td>.81</td>
<td></td>
</tr>
<tr>
<td>SON-R</td>
<td>IQ</td>
<td>2.6-7.11 years</td>
<td>.61</td>
<td></td>
</tr>
<tr>
<td>Stanford-Binet IV</td>
<td>IQ</td>
<td>2.6-12.5 years</td>
<td>.61</td>
<td></td>
</tr>
<tr>
<td>K-ABC</td>
<td>IQ</td>
<td>3.0-18.11 years</td>
<td>.76-.81 with WPPSI-III</td>
<td></td>
</tr>
<tr>
<td>KABC-II</td>
<td>IQ</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DAS</td>
<td>GCA</td>
<td>2.6-17.11 years</td>
<td>.89 with WPPSI-R</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: MDI: Mental development index, MA: Mental age equivalent, CPM; Coloured Progressive Matrices, K-ABC: Kaufman Assessment Battery for Children, KABC-II: Kaufman Assessment Battery for Children second version, DAS: the Differential Ability Scale, GCA, General Conceptual Ability.

Often, low correlations are found between early and later measures of intellectual functioning (e.g. Bayley scales and WISC-tests, not in table) (32). The main reason is presumed to lie in the fact that the abilities measured by tests for very young children are different from those measured in tests aimed at school children.

### 3.3 Suggestion of tools for use in a future monitoring system

As mentioned earlier we assume that description of intellectual functioning (including test results) will be derived from existing medical or psychological files. Tools used for assessing intellectual functioning are expected to show some variation between and within countries, mainly between IQ measures obtained with the Wechsler tests (WPPSI-R, WPPSI-III, WISC-III and WISC-IV) and the...
Bayley scales (version II or III) as these tools tend to dominate in most countries. As a consequence recommendations on use of tools need to be flexible. We suggest the inclusion of validated test instruments whenever possible, reporting the name of the test and reporting separately performance and verbal scores (WISC-III) or Cluster scores (WISC-IV) as well as the total score. In cases where clinical estimation is the only way of determining the child’s intellectual functioning we suggest this should preferably be multidisciplinary and supported by some test results.

4. Existing monitoring systems in Europe

4.1 Literature on existing monitoring systems

Searches for scientific literature on monitoring systems were performed in PubMed. The searches were limited to publications later than 1980 and written in English.

The Mesh term Intellectual Disability with subheading “Classification” in combination with data collection, survey, monitoring, register, registry, registries or epidemiology were searched. In total 128 references were identified and titles or abstracts were reviewed. Five papers surveying childhood intellectual disability and discussing monitoring systems were identified. Another search was performed with the terms intellectual disability in combination with monitoring and epidemiology. In total 116 references were identified within this second search and only one new paper was selected after review of titles or abstracts. Nine additional scientific papers reporting on these issues were found. All selected papers (n=15) are listed in the Appendix.

A recent review concludes that although data on the overall prevalence of disability exist, data on people with intellectual disabilities are lacking in spite of the presence of a good statistical infrastructure and the richness of data in the Western world (2). The importance of well-defined goals for surveillance is emphasized. The existing National Intellectual Disability Database in Ireland is well-recognised as an impressive monitoring system of ID indicators (2,40).

A substantial Australian review stated that it is crucial in terms of valid prevalence data that case definition, ascertainment sources and methods are kept constant over time (23). Epidemiological studies needs to be population based (23).

It is commonly recognised that the prevalence of mild ID varies greatly in the literature, while the prevalence of severe ID is largely agreed to be around 0.4 (41).

A recent US paper shows the number of children with ID reported by their parents to be stable around 0.7% from 1997 to 2008 (42), while a US disability survey program reports a higher prevalence of ID (1.55% in 1996 and 1.20% in 2000) (43). The lower parent reported prevalence is mainly explained by parents not being aware of their children being tested in an education context. This is in accordance with an Australian database of intellectual disabilities finding an average prevalence of ID=1.52% with 50% recruited only through the education system (44). Children below the age of 9 were seldom recruited from the education system.
Data from a National Health Survey can be useful to describe characteristics and needs of persons with ID (45), although the definition of ID is not straightforward and thus the number of persons identified with ID might depend on the number and specificity of the questions in the survey (46).

A Canadian population based study on ID recruited most children from the educational sector (47). They found a low prevalence of children with mild ID and suspected that this might be due to a low identification of these children due to high educational integration (47). This might result in unrecognized children with unmet needs (47).

In an older Norwegian study 87% of children with ID were referred from the educational sector (48). In total 48% of the identified children had no previous psychological tests and needed further psychometric evaluation.

An older review from the Netherlands concludes that the prevalence of severe ID is not complete until the age of 5 and increases until the age of 15 (49). The prevalence of severe ID varied little between regions indicating little influence of exogenous factors (49). Prevalence of children with ID of unknown cause are around 0.24%, only 20% of these had severe ID (50).

A French register on severe childhood disability exists, and includes severe ID (51). Prevalence in 7 year old children was stable during the period 1980-91. Parental refusal to participate in the register was about 2% (51).

An on-going American monitoring system of developmental disabilities found that 2 out of 3 of children with severe ID have additional neurological conditions (52). These results support the idea of two distinct types of ID: isolated ID and ID combined with other neurological conditions (52).

A study in Finland found the prevalence of severe ID to be 0.63%, the high prevalence was explained by a high completeness and high number of children with Down Syndrome again explained by a high maternal age (53).

When no routine system exists, prevalence may still be determined, e.g. a recent school survey in Italy (54) showing a prevalence rate of 0.9% of children with ID or an older US survey showing a prevalence rate of 0.78% of children with ID (45).

A recent Norwegian study finds a higher administrative prevalence of intellectual disabilities in less populated municipalities as compared to urban areas and suggested that better access to support for children with mild ID in smaller communities, is leading to more accurate diagnosis (55). The authors recommend inclusion of data regarding urban-rural living as well as practices in diagnosis in future studies (55).

### 4.2 Survey among SCPE centres

All registers of SCPE were contacted by mail in order to get information on existing monitoring systems of childhood intellectual disability and on the possibilities of finding these children in different European countries. Four registers were already known to have register children with ID:
two in France (one in Grenoble covering the Isere, Savoie and Haute-Savoie counties and Toulouse covering the Haute-Garonne county) and one in Iceland, Ireland and Latvia. These registers are described in 4.5.

In total 11 other registers representing 9 countries (Croatia, Denmark, Germany, Italy, Lithuania, Norway, Portugal, Slovenia, Spain and UK) responded to the survey. The respondents were typically either epidemiologists or child neurologists.

Only one register – Croatia - reported that some kind of register existed but this was not further described.

Denmark reported having an administrative database from the Ministry of Education which shows a rising yearly % of children receiving special education on the basis of general intellectual disabilities (that to some extent, represents children with an IQ less than 70); 3% were reported in 2009 (Report in Danish).

Two registers reported that a recent survey had been carried out in their country. One in Lithuania not further described and one from Denmark found a prevalence of children with IQ under 50 of 4 per 1000 and 8.9 per 1000 for children below IQ 70. 25% of these children were unknown to the health system and only found by the social/education system (paper in Danish (56)).

Most countries identified the educational and/or social system as well as the health system as the best sources of identifying children with IQ less than 50. In Eastern Europe the emphasis is on the health system whereas, in Italy and Norway the emphasis is on the educational system. The definition of the health system differed and included some or all of the following: paediatric departments, child development clinics, mental hospitals, child mental clinics and neuropsychiatric clinics. In Germany it was mentioned that, by law, it is forbidden to register people with intellectual handicap.

In conclusion: Few registers and recent surveys on intellectual disability exist in Europe. Identifying these children cannot be done without searching in the social/educational system as well as the health system depending on the country.

4.3 Contact with relevant institutions in Europe

1. Centre for educational development, appraisal and research, University of Warwick, UK.

This center has written a report in 2010 called; Procedures used to diagnose a disability and to assess special educational needs: International review. The report has been written by Dr. Martin Desforges and Professor Geoff Lindsay and was commissioned by the National Council for Special Education in Ireland.

The report was commissioned because the policy regarding special educational needs (SEN) in Ireland was under review. The context for the report was the growing resistance to diagnostic labeling as a mean of securing resources for SEN. It was questioned whether it was necessary to have a diagnosis in order to provide SEN.
This issue is of interest when establishing monitoring systems for intellectual disabilities since it may be possible to identify children through the education system.

The review is very extensive including internet interviews with professional working with SEN system in 8 different countries around the world. The key findings were that there was a substantial variation in both policy and practice across different countries. Half of the countries did not require a diagnosis of disability when assessing SEN. Terminology and categorization of disabilities varied both within and between countries. They found fundamental problems with reliability in classifying disabilities. The recommendations for Ireland were that a diagnosis should not be a prerequisite or determinant for the allocation of additional resources for a child with SEN. They reviewed WHO international classification of functioning, disability and health (ICF, WHO 2002) and found that this instrument did not provide reliable diagnosis although it might have potential, but they were not convinced that appropriate tools for good psychometric probabilities would be developed in the near future.

This conclusion does not support the argument that the educational system should be used to identify children with ID.

http://www2.warwick.ac.uk/fac/soc/cedar/projects/completed2010/intreview

2. World Health Organization – regional office for Europe


A background paper on the health of children and young people with intellectual disabilities entitled: “The case for change” was published in 2010 (57). The purpose of the declaration was:

1) To promote and support good physical and mental health and well-being
2) To eliminate health and other inequalities and preventing other forms of discriminating, neglect and abuse
3) To provide support that prevents family separation and allows parents to care for and protect children and young people.
4) To support children and young people in the development of their potential and the successful transitions through life.

The report stresses that the focus should be on the children not on their disabilities. The authors of the report acknowledge that institutionalization is an active source of harm (dehumanisation) and is inadequately serving the health needs of children with ID. Importantly, from the perspective of this report, there is a lack of data on the number of children with intellectual disabilities. The prevalence is expected to rise. The uncertainty of prevalence was seen especially in Eastern Europe but also in the UK.

In the report, the main emphasis was put on the importance of deinstitutionalization by taking action in order that children and young people with ID among other things could: be allowed to grow up in
the family environment, be transferred from institutions to home, the treatment should be coordinated, be identified by collecting essential information about the needs and services for these children. It recommends that European countries should legislate according to the declaration and the WHO regional director was invited to take action to support research initiatives, monitor the health status of children with ID and assess progress towards implementation of the declaration.

The declaration explicitly supports directly the establishment of a monitoring system of children with ID.


3) Improving Health and Lives: Learning Disabilities Observatory, UK
This Institution has been established to provide better data on children with ID/LD. The institution publishes yearly reports on children with learning disabilities. Since no monitoring system covering the whole country exists, they rely on regular surveys. It is noted that numbers from different parts of the country cannot be compared because some local authorities are more likely than others to judge children with ID to have special needs. It is estimated that 3.6% of all children have mild learning disability and 0.47% have severe learning disability. A census of children getting SEN resources is undertaken regularly.

http://www.improvinghealthandlives.org.uk/

4) National Institute of Public Health, Croatia.
Tomislav Benjak, PhD from the Institute in Croatia noted that the definition of intellectual disability relies on ICD-10 as reported from medical doctors. The national prevalence is 7 per 1000 children. It was not possible get national figures through the social or education systems.

5) National Institute for Health and Welfare, Finland
From the above website and professor Matti Kuorelahti, PhD, professor in special education, University of Oulu, we were informed that the register on individuals with intellectual disabilities was shut down in the early 1990’s. A register following the use of certain services for ID does exist, but this would be expected to vary from region to region. All children with severe ID get social services and are registered locally. In the school years 30% of all school pupils receive special education of some kind.


6) POMONA
PONOMA was a European Commission Public Health funded project, that produced two sequential projects POMONA I (2002-2004) and POMONA II (2005-2008), that aimed to identify health indicators for people with intellectual disability (58). By developing health indicators specifically for people with
intellectual disability POMONA contributed to the advancement of the future understanding of health of people with intellectual disabilities across the European Union. Using a set of 18 health indicators previously developed there was an opportunity to gather standardized European-wide health data on behalf of this population. Data were collected on a sample of 1269 adults with intellectual disability from thirteen Member States. Analysis of this data showed the disparity in health status and access to health care services between people with intellectual disabilities and their age related peers, and provided evidence that people with intellectual disabilities are typically excluded from health surveys and are rarely considered in major public health campaigns.

4.4 Workshop September 2011
A second workshop entitled “Suggestion of a monitoring system for severe intellectual disability” was held September 1-2, 2011 in Copenhagen, Denmark. Participants were representatives from SCPE registers with experience in registering children with severe intellectual impairment (Iceland, Latvia, France and Ireland) as well as a psychologist experienced in measuring intellectual disability. The aim of the workshop was to discuss use of already existing registers and to work on the potential for establishing new registers. The four existing databases on childhood ID are described in detail in 4.5, while the final suggestion of a future common database is described in 5.

Recruitment sources
The existing registers on ID are based on different recruitment sources. In France and Iceland the identification of children is linked to the social service system, while in Ireland and Latvia this is linked to the health sector. At present Denmark does not monitor childhood severe intellectual disability, but it is anticipated that children can be identified through the health sector and the municipal department responsible for allocating children to special education.

Suggested variables in a future common database

Potential variables that were discussed:

Demographics
Age, gender
Ethnicity, nationality
Parental age, parental ethnicity and nationality

Pre- and perinatal factors
Conditions during pregnancy, at birth, birth weight, gestational age, neonatal complications

Etiological factors
Associated medical conditions (for example Down syndrome, brain malformation etc.)
Relatives with ID or psychiatric disorders (type of disorder and which family members)
Family morbidity

1) Developmental or psychiatric disorders in the family
   a. No developmental or psychiatric disorder
   b. Not known
   c. Yes
      i. Type of disorder
         1. Name/description:
         2. ID
         3. Autism
         4. Schizophrenia/psychotic disorders
         5. Mood disorder, bipolar, major depression
         6. Not known
      ii. Extent of disorder
         1. Father
         2. Mother
         3. Siblings (number)
         4. Grandparents
         5. Siblings of parents
         6. Not known

Socio-economic data
Parents' education, employment, finances
Living (home, institution)
Schooling

Support
Obtained and required

Co-morbidity
Physical/motor disability
Sensory
Developmental
Psychiatric
Gastro-intestinal
Sleep disorder
Epilepsy

4.5 Existing registers on ID in Europe
Five of the centres participating in the SCPE-NET have on-going registration of children with severe ID and are described in detail below. The registers in Ireland, Iceland and Latvia cover the whole country,
while the two registers in France are exhaustive population based registers on a geographically restricted area.

The National Intellectual Disability Database (NIDD) in Ireland

The database is funded and owned by the Ministry of Health and has been a national database on ID since 1995. The database is administered regionally by the health service providers the Health Service Executive and locally by agencies that provide services to people with ID. The Health Research Board manages the national system and collates and publishes annual statistics from anonymized data received from the Health Service Executive. Information is collected on all of those who are known to have an intellectual disability, both adults and children. The database adheres to existing data protection legislation in Ireland.

The aims of the database are to 1) improve accuracy of service planning, 2) guide future service development and 3) monitor trends in ID and facilitate research on subgroups of ID.

Ireland has another national database on physical and sensory disabilities. A person is not registered on both databases but in rare cases children may appear on both where there is no diagnosis yet available. If an individual has more than one disability he/she is registered on the database that best reflects his/her needs.

Registration on the NIDD is voluntary and consent is sought. Very few refuse consent, but no data regarding this are available. Refusal to register on the NIDD does not affect the individual’s access to services. Service provider agencies have a key role to play in the data collection and update of the database. These agencies are often non-statutory, traditionally religious-run organisations and are funded by the state to provide a range of services. The NIDD was developed in collaboration with the service providers who can use the web-based database to monitor and track their own service users.

The inclusion criteria are having an ID and using or requiring specialized intellectual disability services. ID is measured with ICD-10 cut-offs and in addition the database uses a category of “not verified” which can apply particularly to children who haven’t had an assessment of their IQ level. The focus of the database is needs-based intervention and the day, residential and respite services that individuals need. The database does not record specific diagnosis. However this is continuously evaluated and it is planned that ICF-based measures captured on the physical and sensory disability database be extended and adapted to the ID database.

Persons of all ages are recruited. The persons in the ID database are referred from neonatal and paediatric departments, other health professionals, disability services and voluntary organizations who provide services. The form for the ID database is completed by service providers, based on face-to-face interview with participants and/or their families.

The database contains information on level of ID, physical/sensory disabilities, current day/residential/respite service, multidisciplinary support (like, for example physiotherapy, occupational therapy, and speech and language therapy) and future services needed.
The database currently has information on about 8000 children aged 0 to 17 years. All levels are registered, however people with mild ID are under-represented as they are more likely to be in mainstream settings rather than require specialised disability specific services. An annual review of all completed forms ensures frequent updating of the database.

The prevalence of severe ID (IQ<50) is estimated to be 3 per 1000 (Figure 2).

**Figure 2 Prevalence of ID in Ireland**

![Graph showing prevalence of ID in Ireland from 1998/9 to 2010.](image)

**The register of child Intellectual Disability in Latvia**

The register was started in 1997 and is owned and administered by the Latvian government. The aim of the register is to develop a uniform data information system to ensure statistical information for the EU and others.

The PREDA system includes nine diagnostic groups and one of these is mental disorders which includes a subgroup of intellectual disability. Great efforts are made regarding data protection.

Persons are recruited via the health sector. The public sector reports more completely than the private sector, but most children with ID are in the public system. Parents are not asked for consent, but they can refuse to give it.

Psychiatrists (the only profession allowed to enter data on mental disorders) either submit data by post or computer (at the moment 50% each).

ID diagnosis is based on ICD-10 F.
Another Latvian governmental database registers information on school and to be allocated to a special school a child needs a diagnosis from the Health sector. It is possible to link the two databases with an identification number of the child.

The completeness of the register is considered good after comparing with an official governmental system that registers all people receiving treatment. Prevalence for severe ID is estimated to be around 3 per 1000.

**The intellectual disability database of Iceland**

*Greiningar- og rødgjørðstøð ríkisins* is named the „State Diagnostic and Counselling Centre“ (SDCC) in English run by the Ministry of Welfare. The intellectual database of Iceland is located at the SDCC. The SDCC offers interdisciplinary services, which serves children and adolescents with a disability wherever they live in Iceland. The Centre operates according to an Act (83/2003) which is intended to ensure that children and adolescents can avail of diagnosis, counselling and have access to services for serious developmental disorders which may lead to handicap. The Diagnostic Centre also has a duty to educate parents and professional bodies about children’s disabilities and principal treatment methods. It has an important role in supporting disabled children and their families. The main disorders which lead to referrals are autistic spectrum disorders, intellectual disabilities and motor disorders. The SDCC also handles other projects, such as research, registration and education, and has a duty to safeguard knowledge in the field. It is involved with teaching and research institutions within Iceland and abroad. The SDCC served for many years under the Ministry of Social Affairs and has therefore always been closely linked to the service needs of children with disabilities. Since the beginning of the year 2011 the Centre has been run by a new Ministry of Welfare. The database is run as part of the daily activities at the Centre and owned by the State.

The database was established as an administrative tool for the centre at its establishment (1986) and covers all individuals assessed at the SDCC from the start.

The database has been administered by the director’s office at the SDCC of the centre. As the centre is obliged to serve the 0-18 years old population with disabilities of the whole country the database covers the entire population.

The data is recorded at a production of a discharge summary after an assessment. An update is done after each additional assessment.

The number of variables has increased over the years according to the needs/purposes of the dataset. The database includes demographic information about individuals, information about the referrals, the tools used for assessments and most importantly information on clinical diagnoses (disability/developmental-, co-morbid-, etiologic- and relevant previous and social diagnoses) according to ICD-10 classification.

The database has not only been used as an administrative tool for daily activities, but also for current and future planning of services. The database has been useful for municipalities, schools and social services to plan for transition periods, both for each individual and the group as a whole.
In general

The population of Iceland is very small (~320,000), compared to the size of the country, and the inhabitants are quite widely spread around the country (mainly at the coasts). The database has been a significant source of information for the institutions, municipalities and other service providers that prepare and implement services. This information has been of value for both state run services and also for locally based services. For example small municipalities usually need some extra funding if they have individuals needing more support during their life. The database has been used to distribute funding appropriately.

The database has been used to follow prevalence trends in disabilities for the whole population and also at different areas of the country. Total number of births in Iceland is around 4500 per year. The data have been useful to observe the changing trends in practice of referrals to the Centre and been useful for the centre to plan its approach to meet increasing demand for service from SDCC. There has been a significant increase in the number of referrals to the centre for the last decade. The changing trends can be observed in all age groups and is mainly explained be increased awareness of the autistic spectrum.

The database is also used to produce annual reports for the SDCC, which monitor trends in prevalence of disabilities, which actually reflects the change in referrals to the centre from the whole country. The main increase in the workload of the centre has again been around assessments and diagnoses of autistic spectrum disorders.

The database has also been of value in the area of research on childhood disabilities. For every research-related project, permission is required from the director of SDCC. The Data Protection Authority in Iceland and the National Bioethics Committee control access to individualised data in the database. This is also true if the SDCC is involved in submitting data for other projects, like the SCPE project.

The centre gets a written informed consent from every parent of a child referred to the centre to gather information about their child and to use the information gathered to support the work of the centre.

Information about individuals with ID in the Icelandic SDCC database

On the 18th of March 2011 there were 3981 individuals, born in the years of 1980 to 2006 (26 years), registered in the database. Of those there were 1136 individuals diagnosed with intellectual disability. Of the 1136 persons there were 657 diagnosed with mild and 479 with “severe” ID (IQ under 50). During those same 26 years there were 111,982 individuals born in Iceland. These numbers give an administrative prevalence of 1% with intellectual disability in Iceland for those 26 years and 0.43% for “severe” ID (Figure 3).
Figure 3: Prevalence of mild (50≤IQ≤70) and severe ID (IQ<50) in Iceland

Prevalence of ID in Iceland increased from 1980 to 1999 according to diagnoses and registration at SDCC. Before 1996 the SDCC did not accept referrals for mild ID at school age, therefore the prevalence might be lower during the early period. During that period there may have been under ascertainment of severe ID as well as of borderline cases. Overall, the prevalence of severe ID has been stable between 1990 and 2006. This could indicate that it would be appropriate to support the proposed age range for inclusion (7 years) in suggested common European database.

Registers of severe childhood disabilities in France (RHEOP, RHE31)
In France, two registers on severe neurosensory childhood (and developmental) disabilities exist: 1) the register of severe disabilities and perinatal observatory (RHEOP) in Grenoble which started in 1991 and covers the counties of Isère, Savoy and High-Savoy where approximately 30,000 live births are recorded each year, and 2) the register of severe disabilities in Toulouse, called RHE31, created in 1999, which covers the county of Haute-Garonne, and counts around 14,000 live births each year. Both registers collect data on children presenting severe motor disabilities (requiring at least permanent re-education or physical aid and including all cerebral palsy cases), severe intellectual disabilities (IQ below 50 in RHEOP and equal or below 50 in RHE31), Down syndrome, severe auditory impairment (having a bilateral loss of more than 70 decibel before correction), severe visual impairment (visual acuity of the best eye < 3/10 after correction) and pervasive developmental disorders, including autism. The inclusion criteria, data collection and procedures are essentially the same for the two registers. The main objectives for both registers are 1) to monitor prevalence rates and describe characteristics (associated disabilities, risk factors, management of care) of the severe deficiencies mentioned above, 2) to assist in service planning according to the presence or absence of associated severe impairments, and they also play a role in 3) the evaluation of practices and 4) research related
to risk factors. Apart from severe deficiencies, the register of Grenoble also monitors mortality rates, through the registration of intrauterine fetal death or death per partum (≥ 22 weeks of gestation or birth weight ≥ 500g), and all therapeutic interruptions independent of gestational age.

Data are collected from medical files of children living during their 8th year of life (Haute Garonne has in addition data on children in their 5th year of life) in the geographical area covered by each register, once parents are informed. In about 3% of the cases, parents object. Data collection is usually performed by medical or paramedical investigators who actively collect information from medical records at medical-social establishments, psychiatric institutions and institutions that decide on special education and financial allowances. The latter source is the main place of ascertainment. RHEOP has data available for the generations 1980 to 2002 for Isere, and from 1997 till 2002 in the Savoy Counties. Haute-Garonne has data available from 1986 till 2002.

ID is based upon the result of a validated IQ test or assessment by a clinician. Cases are then classified according to the ICD-10 classification. As well as data on diagnosis, information is collected on other co-morbidities and deficiencies (severe or not); the aetiology, medical and educational care and demographic variables is done, whenever available.

The average annual prevalence rate of severe intellectual disabilities is 3.0 per 1000 children [95% CI: 2.9-3.2], being in their 8th year of life for the County of Isere (average of 23 years) and 3.0 per 1000 [95% CI: 2.8-3.2] in Haute-Garonne County (average of 17 years).

In nearly half of the cases, severe ID is associated with another severe disorder, e.g. psychiatric or severe motor disorder and, in such groups, a higher proportion of children have severe or profound mental retardation. One third of children with severe ID have a severe or profound mental retardation (IQ below 35). The aetiology of severe ID is difficult to identify, but a prenatal origin is most frequent in cases without any other associated disorder. About 40% of children are in mainstream schooling, but only 15% of them are in regular units, often in nursery schools and/or with the support of a student assistant. Schooling highly depends on the type of associated disorder. The time trend analysis shows that, in the case of children with no associated disorder, the prevalence rate of severe ID has been stable for twenty years, although the prevalence rate of children with Down syndrome has been decreasing.

The prevalence rates of severe intellectual deficiency (IQ<50) remained stable between 1980 and 2002 in Isere County, France (Figure 4a), and between 1986 and 2002 in Haute-Garonne county (Figure 4b).
Figure 4a Trends in prevalence rates of severe intellectual deficiency (IQ<50) in children residing in their eight year of life in Isere County, France

![Graph showing trends in prevalence rates of severe intellectual deficiency (IQ<50) in children residing in their eight year of life in Isere County, France.]

Figure 4b Prevalence rates in RHE31 (Haute-Garonne County), ID defined by IQ ≤50, children aged 8y. Birth cohorts 1986-2002.

![Graph showing prevalence rates in RHE31 (Haute-Garonne County), ID defined by IQ ≤50, children aged 8y. Birth cohorts 1986-2002.]

5. Suggestion of monitoring system in Europe

5.1 Aims of the future common database
We suggest a common database to achieve the following aims:

1) Describe prevalence of severe ID in childhood
2) Describe children with severe ID from a clinical perspective according to subgroups defined by aetiology and associated impairment
3) Describe type of schooling and living arrangements of children with severe ID
4) Create a database from which further studies on aetiology, service needs and provision of services can be based

This will enable a description of trends over time in relation to the above and in relation to prevalence of ID in children age 6-8 years. Public services for children with ID aged 6-8 years can potentially be guided directly by the data on prevalence, type of ID, school and living arrangement. Aetiology, services, quality of life and participation can be studied by collection of additional data on subgroups of individuals with ID identified in the common database.

5.2 Recruitment sources of the future common database
The common database should receive regularly data from participating local registers. The chosen covered area of each register should be representative of the country or cover a specific geographical area in this country, with a sufficient number of live births per year (see 5.6).

Recruitment sources will differ between regions, depending on the organisation of health, social and educational sectors. Completeness of any register depends on the number and type of recruitment sources.

Each register should specify and estimate type and number (preferable more than two) of recruitment sources needed to yield at least 80% completeness of the register. The estimation should be based on national/local validation studies whenever possible or, if not, on references from national/local experts in the field. The types of relevant recruitment sources needed for at least 80% completeness include:

a. Health sector (specify whether active or passive recruitment is needed)
b. Educational (specify whether active or passive recruitment are needed)
c. Institution allocating finances or education/social services (specify whether active or passive recruitment is needed)

Each register should then describe their current recruitment sources and compare them to those recommended above. Detailed information should be provided regarding the type of Health sector sources (paediatric, genetic, psychiatric or other departments), and the type of Educational source (teachers, special educational needs sector or others).
Registers not recruiting from all relevant sources should note whether this is due to practical, financial or other reasons.

Registers participating in a common database should aim to conduct a validation study of completeness of the register, and this should be repeated regularly.

5.3 Inclusion criteria of the future common database

It is proposed to use 50 as the IQ cut-off value. This value is in accordance with the ICF and DSM classifications that classify children with an IQ below 50 as having moderate, severe or profound intellectual disability. Adaptive behaviour is not included in the inclusion criteria since this information is not likely to be present for a substantial number of children. The children should not be included earlier than age 6, where evaluation according to school placement should have been made. To be able to estimate prevalence and compare data from different regions and time periods no child assessed later than the age of 8 years should be included.

1. IQ ≤ 50 (lower limit of 95% CI 50 or less)
   Assessed by
   a. Test (preferable validated)
   b. Clinical assessment (preferable multidisciplinary and supported by test)

2. Age
   Preferred minimal age at assessment = 6 years
   However if the child is only tested at a younger age and with a result of IQ ≤ 50, the child should be included at age 6.
   Maximum age at assessment = 8 years

5.4 Classification of intellectual functioning in future common database

Level of intellectual functioning will follow the most common categories. IQ is preferable assessed by a validated test combined with a clinical assessment. If testing is not possibly the reason for this should be explained and if possible the clinical assessment should be multidisciplinary and supported by incomplete results of tests.
1) Level of ID (<20, 20-35, 35-50)
2) Mode of assessment ID
   a. Test of IQ
      i. Name of test:
      ii. Result of test (overall IQ, Performance and Verbal score/index):
      iii. Test of IQ not available
   b. Test of adaptive behaviour
      i. Name of test:
      ii. Result of test (score):
      iii. Test of adaptive behaviour not available
   c. Clinical
      i. Untestable
      ii. Test not available
3) Date of assessment for database

5.5 Variables other than intellectual disability in future common database

Following the suggested aims in 5.1 we need to include information about possible aetiology and risk factors as well as clinical signs and type of schooling. We do not include service needs and received support in the common database; however, study populations for research projects in this field could be identified in the database.

A sound demographic and socio-economic description of the children included is crucial to future epidemiological studies based on the database. Ethnicity is difficult to measure and countries have different traditions and ethical policies on how to do this. We suggest collecting data on country of birth of child and parent.

Below are the suggested core variables (c) and possible supplementary variables are listed:

**Demographic**

1) Date of birth (c)
2) Gender (c)
3) Country of birth
4) Mother’s date of birth (c)
5) Father’s date of birth
6) Mother’s country of birth
7) Father’s country of birth
8) Consanguinity between biological mother and biological father
Socio-economic
1) Mother’s occupation
2) Father’s occupation
3) Mother’s education (c)
4) Father’s education (c)
5) Cohabitation of child (mother and father, mother, father, institution) (c)
6) Living area (urban, rural) (c)

Schooling
Type of school c
a. Mainstream
   i. Mainstream class
   ii. Special class
   iii. Time in both special and mainstream class
b. Special
   i. Daytime
   ii. Residential

Information on birth
1) Birth weight (c)
2) Gestational age
3) Neonatal unit care

Comorbidity (for each category text and if possible ICD-10 code)
1) Motor (c)
2) Psychiatric
3) Epilepsy (c)
4) Pervasive Developmental Disorder (autism spectrum disorders) (c)
5) Hearing (c)
6) Vision (c)
7) Rare Diseases
8) Congenital malformation
9) Syndrome
10) Metabolic diseases and genetic disease

Diagnose responsible for severe ID, if known
1) Text (c)
2) ICD-10 code (c)
3) Age of diagnosis (c)
5.6 Denominators for estimation of prevalence

Estimation of population in each region represented in the common database will be needed for the calculation of prevalence. We suggest that this be based on the number of live births from each birth cohort, living in the area at age 6-8. In places where migration is important before 6-8 years old, it should rather be based on the number of children whose family is living in the area when they are 6-8 years old. The ideal size for coverage in relation to a register on childhood ID is around 30 000 live births per year, and a minimal size is 10 000 live births per year.

5.7 Current availability of suggested variables in existing registers of ID in Europe

As described in 4.5, five registers on childhood intellectual disability currently exist. None of these have all of the variables suggested for the future common database. This section presents an overview of the possibility of building a common database from these registers.
In table 5, the variables proposed for the future database and their availability in each of the five existing registers have been listed.
Table 5  Common data to be included in the European database on childhood intellectual disabilities and availability in each of the five registers

<table>
<thead>
<tr>
<th>Variables</th>
<th>France-RHEOP</th>
<th>France-RHE31</th>
<th>Iceland</th>
<th>Ireland</th>
<th>Latvia</th>
</tr>
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<tbody>
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<td><strong>Inclusion</strong></td>
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</tr>
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<td>Moderate intellectual disability (IQ=35-49)</td>
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<tr>
<td>combined with profound ID</td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>combined with severe ID</td>
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</tr>
<tr>
<td><strong>Demographic</strong></td>
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<td>dd/mm/yy</td>
<td>dd/mm/yy</td>
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</tr>
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<tr>
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</tr>
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</tr>
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<tr>
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</tr>
<tr>
<td>Occupation father</td>
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<td></td>
</tr>
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<td>Mainstream (mainstream or special class) or special school (residential or daytime)</td>
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</tr>
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<td>Time spent in mainstream school</td>
<td>&lt; 1/2 of the time, 1/2 time, &gt; 1/2 of the time</td>
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<td><strong>Comorbidities (text and ICD-10 code)</strong></td>
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<tr>
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<td>Vision disability</td>
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<tr>
<td>Text</td>
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<td>yes</td>
<td>yes</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>ICD-10 code</td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>Age of diagnosis</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
</tr>
</tbody>
</table>

1 classification based on cognitive tests (Intelligent Quotient) in all registers except Iceland that includes also the adaptive behaviour
Age, gender, description of ID, comorbidities, schooling and living arrangements are covered well by all four registries, except for information on diagnosis of ID and comorbidities in Ireland. Socio-demographic data regarding parents as well as information on birth is not consistently collected in the four registries.

Consequently a common database on these four registries would probably be able to provide prevalence data on severe ID, additional data on motor disability, epilepsy, visual or hearing disability in children aged 6-8 years. Type of schooling and living arrangements would be possible to report as well. To report in more detail on these children would require further work on harmonisation of data.

In table 7, the 5 year average prevalence rate of severe, moderate and profound intellectual disability for each register is listed. In Table 8 the population figures (residents in each of the regions covered by the register) for the same age groups are listed.

5.8 Implementation opportunities of a common database: strengths and weaknesses

A common database offers several advantages: it will force a harmonisation of definitions and inclusion criteria necessary to compare prevalence and practices (e.g. education and health services) across countries. Also, it will increase the number of cases and subsequently the power for analyses on risk factors and consequences, particularly when looking at subgroups.

A comparison of existing registers, showed several similarities which offer strengths to the initiative of a common database (Table 6). Four of the five registers have already local epidemiological aims of monitoring trends in prevalence, and one has future plans to use the data for epidemiological purposes. Each of the registers is using the WHO definition for severe intellectual disability apart from the register of Toulouse, France RHE31, that includes IQ=50 as well. In relation to subgroups, Latvia is the only country unable to provide separate data on severe and profound intellectual disability. Furthermore, in each register 6-8 year olds are captured which is the age range selected for the common database. Finally, all of them code according to the ICD-10 classification.

However, on some other points the databases/registers differ, and therefore data may not be fully comparable. First, unlike other countries examined, the registers in France are population based registers covering a specific geographical area. Thus, these French registers do not necessarily represent the whole country.
Second, cases are recruited differently across registers. The two registers in France recruit their patients actively by sending medical surveyors to different sources, while recruitment by the other databases is done passively by health professionals when a case is consulting. However, apart from Latvia, in all other centres there is a financial or educational incentive to seek contact with a health professional or in case of the French registers an administrative requirement. Therefore, we assume that the proportion of children with an IQ<50 that do not seek contact with a health professional should be very low. In Latvia registration depends on the willingness of the psychiatrist to refer cases to the central database. In Ireland and Iceland, on the other hand, data are sent to a central database in order to obtain financial help/services but there is no financial incentive in Latvia, which may make the psychiatrist more reluctant to refer. This may partly compromise the completeness of the Latvian database. However, this only concerns private psychiatrists as the information from public psychiatrists who are funded by the government can be cross checked. As probably only a few patients consult private psychiatrists, the number of patients not included in the database, seem to be negligible. Finally, the number of cases with cognitive and adaptive test results, is nearly complete only in Iceland and consequently it will not be possible to include this information for the time being. With regard to cognitive test results, this should not affect the number of inclusions as long as there is a clinical assessment. Neither should the adaptive behaviour test results affect the prevalence significantly since children with severe ID are being studied.

5.9 Potential participants in a new collaboration of registries on childhood ID

Five registers (Iceland, Ireland, Latvia and two in France) that include children with severe ID exist in Europe and potentially could form the basis of a network to establish a common database. As they differ considerably in data collection practices and type of variables collected, harmonisation of data would be required to be able to contribute to a common database. Also other regions familiar with registration of children with cerebral palsy in Norway, Italy and Denmark would be willing to join such a network. In the future other registers or monitoring systems could be encouraged to follow. Data security has to be carefully specified and respected.
<table>
<thead>
<tr>
<th>Register</th>
<th>Type and definition</th>
<th>Aims</th>
<th>Collection of data</th>
<th>Sources</th>
<th>Tests</th>
<th>Classification</th>
<th>Age of registration</th>
<th>Update</th>
<th>Exhaustivity</th>
<th>Refusal rate</th>
<th>Nr of sources per patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iceland</td>
<td>mild, moderate, severe, profound ID</td>
<td>1) administrative tool for daily activities 2) current and future planning of services 3) surveillance of prevalence 4) research purposes</td>
<td>Passively when cases consult a psychiatrist or psychologist</td>
<td>Mainly psychiatrists, school psychologists and psychologists</td>
<td>Cognitive tests: ≥95% Adaptive behaviour tests: 80-90%</td>
<td>ICD-10</td>
<td>All ages</td>
<td>Continuous</td>
<td>Estimated to be very high but exact numbers not known.</td>
<td>Written informed consent of all cases</td>
<td>Cases are mostly reported by one health center, grouping several health professionals. Exact nr of sources per patient are not known.</td>
</tr>
<tr>
<td>Ireland</td>
<td>mild, moderate, severe, profound ID</td>
<td>1) to provide data to assist with service planning; 2) to monitor trends in prevalence rates; 3) to assist with research</td>
<td>Passively: By organisations involved in provision of services, referral through health services</td>
<td>Psychiatrists, pediatricians and other professional working in the health services</td>
<td>Level of Intellectual disability generally determined by cognitive assessment 85% of those registered have an assessment</td>
<td>ICD-10</td>
<td>All ages</td>
<td>Yearly</td>
<td>95%</td>
<td>Only a few deny consent</td>
<td></td>
</tr>
<tr>
<td>France (RHEOP)</td>
<td>moderate, severe, profound ID</td>
<td>1) to monitor trends in prevalence rates 2) to describe characteristics of children with severe ID 3) to provide data to assist with service planning; 4) to assist with research on risk factors</td>
<td>Actively by medical surveyors</td>
<td>Medical files at administrative institutes that decide for special education and financial allowances, psychiatric centers, medical-social institutions</td>
<td>Cognitive tests: ≥57% Clinical appreciation: 43% No adaptive behavior test results</td>
<td>ICD-10</td>
<td>7 year old</td>
<td>1 point in time</td>
<td>no method of formal assessment but probably ≥95%</td>
<td>Passive consent. About 3% of all parents having children with disabilities (also those not included upon our criteria) registered at administrative institute for special education and financial resources deny consent</td>
<td>1 (generations 1998-2002)</td>
</tr>
<tr>
<td>France (RHE31)</td>
<td>moderate, severe, profound ID</td>
<td>1) to monitor trends in prevalence rates 2) to describe characteristics of children with severe ID 3) to provide data to assist with service planning; 4) to assist with research on risk factors</td>
<td>Actively by medical surveyors</td>
<td>Medical files at administrative institutes that decide for special education and financial allowances, Psychiatric services for children through coding of hospital stays</td>
<td>Cognitive tests: 35% of children have been tested, numerical results available for 62% of them Clinical appreciation: 68% No adaptive behavior test results</td>
<td>ICD-10</td>
<td>8 year old</td>
<td>1 point in time</td>
<td>no method of formal assessment</td>
<td>Passive consent. About 4.9% of all parents with children having severe disabilities registered at administrative institute for special education and financial resources deny consent</td>
<td>1 source for 88% of cases 2 sources for 12% of cases (mean 1,1)</td>
</tr>
<tr>
<td>Latvia</td>
<td>mild, moderate, severe, profound ID</td>
<td>1) To develop unified data information system 2) To ensure the implementation of State statistical program 3) To supply international institutions with statistical information (Eurostat, WHO, EU IDE, EMCE2DA, IARC etc.) 4) In near future it will satisfy also epidemiological purposes</td>
<td>Passively when cases consult a psychiatrist</td>
<td>Psychiatric centers [private and public sectors]</td>
<td>Based on clinical appreciation No information on % of cognitive and adaptive behavior tests</td>
<td>ICD-10</td>
<td>All ages</td>
<td>Updated when diagnosis changes</td>
<td>≤ 95%</td>
<td>Passive consent. No exact data available on refusal</td>
<td></td>
</tr>
</tbody>
</table>

mild ID=50≤IQ<69; moderate ID=35≤IQ<50 except RHE31 (France) where moderate ID=35≤IQ≤50; severe ID=IQ<35; profound ID=IQ<20
Table 7  Average prevalence over 5 generations of moderate, severe and profound intellectual disability in the five registers participating in the future ID network

<table>
<thead>
<tr>
<th>ID category</th>
<th>Total (n)</th>
<th>Male (n)</th>
<th>Female (n)</th>
<th>Prevalence</th>
<th>Total (n)</th>
<th>Male (n)</th>
<th>Female (n)</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moderate (F71)</td>
<td>605</td>
<td>395</td>
<td>210</td>
<td>2.1</td>
<td>231</td>
<td>151</td>
<td>80</td>
<td>1.7</td>
</tr>
<tr>
<td>Severe (F72)</td>
<td>260</td>
<td>152</td>
<td>108</td>
<td>0.9</td>
<td>159</td>
<td>96</td>
<td>63</td>
<td>0.2</td>
</tr>
<tr>
<td>Profound (F73)</td>
<td>50</td>
<td>25</td>
<td>25</td>
<td>0.2</td>
<td>19</td>
<td>16</td>
<td>3</td>
<td>0.2</td>
</tr>
<tr>
<td>Severe/profound (F72-73)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other ID (F78)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>n.s. (F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other ID or n.s. (F78-F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not verified</td>
<td>802</td>
<td>572</td>
<td>230</td>
<td>2.8</td>
<td>409</td>
<td>263</td>
<td>146</td>
<td>3.2</td>
</tr>
<tr>
<td>Total</td>
<td>1717</td>
<td>1144</td>
<td>573</td>
<td>6.0</td>
<td>492</td>
<td>318</td>
<td>174</td>
<td>4.1</td>
</tr>
<tr>
<td>95% CI</td>
<td>[5.7-6.2]</td>
<td>[7.3-8.2]</td>
<td>[3.8-4.4]</td>
<td></td>
<td>[3.9-4.8]</td>
<td>[4.8-6.1]</td>
<td>[2.7-3.7]</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>ID category</th>
<th>Total (n)</th>
<th>Male (n)</th>
<th>Female (n)</th>
<th>Prevalence</th>
<th>Total (n)</th>
<th>Male (n)</th>
<th>Female (n)</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moderate (F71)</td>
<td>131</td>
<td>69</td>
<td>59</td>
<td>1.7</td>
<td>90</td>
<td>63</td>
<td>27</td>
<td>0.8</td>
</tr>
<tr>
<td>Severe (F72)</td>
<td>34</td>
<td>21</td>
<td>13</td>
<td>0.4</td>
<td>37</td>
<td>20</td>
<td>17</td>
<td>0.5</td>
</tr>
<tr>
<td>Profound (F73)</td>
<td>12</td>
<td>7</td>
<td>5</td>
<td>0.2</td>
<td>23</td>
<td>16</td>
<td>7</td>
<td>0.2</td>
</tr>
<tr>
<td>Severe/profound (F72-73)</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0.02</td>
<td>52</td>
<td>33</td>
<td>19</td>
<td>0.6</td>
</tr>
<tr>
<td>Other ID (F78)</td>
<td>43</td>
<td>25</td>
<td>18</td>
<td>0.6</td>
<td>202</td>
<td>132</td>
<td>70</td>
<td>2.0</td>
</tr>
<tr>
<td>n.s. (F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other ID or n.s. (F78-F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not verified</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>222</td>
<td>123</td>
<td>96</td>
<td>2.9</td>
<td>202</td>
<td>132</td>
<td>70</td>
<td>2.0</td>
</tr>
<tr>
<td>95% CI</td>
<td>[2.5-3.3]</td>
<td>[2.6-3.7]</td>
<td>[2.0-3.1]</td>
<td></td>
<td>[2.5-3.3]</td>
<td>[3.0-4.3]</td>
<td>[1.6-2.6]</td>
<td></td>
</tr>
</tbody>
</table>

1 aged 5-9 years in 2007, ID diagnosis may have been updated after this age
2 aged 8 years between 2006 and 2010, ID diagnosis may have been updated after this age
3 these children may also have mild ID
Table 7 continued

<table>
<thead>
<tr>
<th>ID category</th>
<th>Iceland (generations 1998-2002, aged 8 years)²</th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td>Male</td>
<td>Female</td>
<td>Total</td>
<td>Male</td>
</tr>
<tr>
<td></td>
<td>n</td>
<td>n</td>
<td>n</td>
<td>n</td>
<td>n</td>
</tr>
<tr>
<td>Moderate (F71)</td>
<td>48</td>
<td>36</td>
<td>12</td>
<td>2.3</td>
<td>3.4</td>
</tr>
<tr>
<td>Severe (F72)</td>
<td>17</td>
<td>10</td>
<td>7</td>
<td>0.8</td>
<td>1.0</td>
</tr>
<tr>
<td>Profound (F73)</td>
<td>6</td>
<td>1</td>
<td>5</td>
<td>0.3</td>
<td>0.1</td>
</tr>
<tr>
<td>Severe/profound (F72-73)</td>
<td>0</td>
<td>1</td>
<td>5</td>
<td>0.3</td>
<td>0.1</td>
</tr>
<tr>
<td>Other ID (F78)</td>
<td>40</td>
<td>24</td>
<td>16</td>
<td>1.9</td>
<td>2.3</td>
</tr>
<tr>
<td>n.s. (F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other ID or n.s. (F78-F79)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not verified</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>111</td>
<td>71</td>
<td>40</td>
<td>5.4</td>
<td>6.8</td>
</tr>
<tr>
<td></td>
<td>[4.4-6.4]</td>
<td>[5.3-8.5]</td>
<td>[2.8-5.3]</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Table 8  Children resident in each of the areas covering the registers for the same ages at which children with intellectual disabilities are registered

<table>
<thead>
<tr>
<th></th>
<th>All children between 5 and 9 resident in Ireland in 2007</th>
<th>All children of 8 year living in Latvia between 2006 and 2010</th>
<th>All children of 7 year living in Isère County, France, between 2005 and 2009</th>
<th>All children of 8 year living in Haute-Garonne county, France, between 2006 and 2010</th>
<th>All children of 8 year living in Iceland between 2006 and 2010</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>288325</td>
<td>94588</td>
<td>77543</td>
<td>70603</td>
<td>20733</td>
</tr>
<tr>
<td>Male</td>
<td>147 984</td>
<td>48373</td>
<td>39484</td>
<td>36363</td>
<td>10514</td>
</tr>
<tr>
<td>Female</td>
<td>140 341</td>
<td>46216</td>
<td>38059</td>
<td>34240</td>
<td>10219</td>
</tr>
</tbody>
</table>

6. Ethical considerations

It is obvious that a cross-country register on people with intellectual disabilities will raise concerns about data security and sensitivity. In Denmark 40 years ago a nationwide register on people with intellectual disabilities was closed down because of the risk of misuse of data. Today such data in Denmark are partially available through new registers like the hospital discharge register. We are also aware that a register probably will not be permitted in Germany, for example, because of their present laws and regulations. It is possible, then, that some countries will not be able to contribute to the register.

The WHO European Declaration of Bucharest 2010, however, has as a priority to "collect essential information about needs and services and assure service quality" of people with intellectual disabilities and this will be difficult without some kind of registration.

It is important to stress that data security on the information held in any future register on intellectual disability has to be carefully specified and respected. This will be a core part of any further planning for such a register.
### Appendix I  Articles on monitoring childhood severe intellectual disability (n=15)

<table>
<thead>
<tr>
<th>Author, country, year and title of publication</th>
<th>Background</th>
<th>Aim and method</th>
<th>Conclusion on monitoring ID</th>
</tr>
</thead>
</table>
| Boyle et al. USA 2011 (42)  
“Trends in the prevalence of developmental disabilities in US children, 1997-2008” | Data on parent reported prevalence of developmental disabilities have been used to assess educational, medical and social needs for support. During the last 10-15 years a number of factors may have influenced the prevalence. Improved survival and diagnosis as well as increased awareness might have increased prevalence, while medical practices might have decreased prevalence. | To examine prevalence (and changes over time) of developmental disabilities according to demographic and socioeconomic characteristics, by analyzing data from population-based surveys with parent reported diagnose of disability | The study reported developmental disabilities in general increased over the last decade, but number of children with parent reported ID is stable around 0.71%. This rather low prevalence might be due to some parents never being told of their child’s functioning in the intellectual disabilities range. Prevalence of children with ID was highest in the oldest children (11-17 years), black children and children of parents with short education and poor financial situation. |
| Fujiura GT et al. USA 2010 (2)  
“Make measurable what is not so: national monitoring of the status of persons with intellectual disability.” | Focus on human rights has changed the dialogue over disability policy from attenuation of anomalies to access and equity of opportunity. But life prospects lack behind the rhetoric. Cross-nationally comparable indicators are needed to monitor the status of people with ID | A literature review to establish the role of cross-nationally comparable indicators as a tool for monitoring the status of persons with ID. | Monitoring ID is an essential step in implementing disability policies. Aim of monitoring needs to be set. Currently a statistical infrastructure exists, but disability and ID are seldom identified. Statistical data is limited to North America, few European countries and parts of the Pacific Rim for example Australia and China. Surveys demands many resources and are not replicated or occur to infrequently. Dedicated cross-national ID monitoring initiatives are extremely rare. No optimal form of ID data exists. Ireland probably the best. Standard convention of capturing severity is lacking. This is critical in data utility and informing policy. |
| Sondenaa E et al. Norway 2010 (55)  
“Prevalence of intellectual disabilities in Norway: Domestic variance.” | People who receive services often labeled “administrative ID”. Epidemiologists distinguish between administrative and true prevalence of ID. Administrative prevalence in Norway: 0.42-0.48%. (International true prevalence 3% of schoolchildren. WHO suggest 2-3% mild ID). Norwegian municipalities receive funds according to number people with ID. | To investigate administrative prevalence of ID in Norway, by comparing Norwegian regional data on inhabitants receiving funds. | Administrative ID prevalence= 0.44% (Norway population=4.77 mill), including only inhabitants receiving funds. Varied across municipalities and regions. The smaller municipality the higher prevalence, maybe due to small communities supporting mild ID more often or socio-economic-cultural differences. Recommends to monitor urban-rural changes over time as well as practice in diagnosis and service. |
<table>
<thead>
<tr>
<th>Source</th>
<th>Title</th>
<th>Summary</th>
<th>Objective</th>
<th>Additional Information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Faravelli C et al.</td>
<td>Italy 2009 (54)</td>
<td>Most adult mental disorders origin in early life and are associated with substantial difficulties later in life. Epidemiology of childhood psychiatric disorders are sparse.</td>
<td>To report on prevalence of mental disorders in children in Italy based on a representative sample of children attending primary school (N=999, age 6-11 years)</td>
<td>Prevalence of ID=0.9% (CI 0.31-1.48).</td>
</tr>
</tbody>
</table>
| Bhasin TK et al. | USA 2006 (43) | The disability surveillance program MADDSP serve as a model for 17 states and provides regular, systematic monitoring of prevalence of selected disabilities according to various demographic characteristics of children and their mothers. | To provide prevalence estimates for five disabilities including ID for 1996 and 2000 according to specific demographic and severity characteristics, by analyzing data from a cross-sectional population-based study on children recruited from multiple sources for example public schools, hospitals and public service agencies (N=568 (1996) and 522 (2000), age 8 years) | Prev. ID in 1996=1.55% (mild ID= 1.00, severe ID=0.43)  
Prev. ID in 2000=1.20% (mild ID=0.73, severe ID=0.33).  
Decrease in mild ID, but not severe ID, is significant. The prevalence estimates of 2000 is in accordance with prevalence’s before 1996 and reasons for the higher prevalence in 1996 are not known and needs to be further explored.  
A surveillance system like MADDSP is used to identify various perinatal, postnatal and socio-demographic risk factors of developmental disabilities. |
<p>| Petterson B et al. | Australia 2003 (44) | ID places heavy demands on families, communities and health and education systems, but is a much-neglected area of public health. Causes are heterogeneous and are known in less than 50% of cases. Ongoing population-based ascertainment of ID facilitates the planning of services and provides a tool for studying causal pathways. | To report on a monitoring system for ID. A database of ID has been maintained in Western Australia since 1953 with recruitment based on support services, but from 1999 children are in addition recruited from public and private education sectors. | Average prevalence of ID=1.52% with 50% recruited only through the education system. Below the age of 9 relatively fewer children are recruited from the educational system. |</p>
<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Country</th>
<th>Title</th>
<th>Summary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Larson et al.</td>
<td>US 2001 (45)</td>
<td></td>
<td>“Prevalence of mental retardation and developmental disabilities: Estimates from the 1994/1995 National Health Interview Survey Disability Supplements”</td>
<td>Accurate estimates of prevalence of intellectual disability and developmental are crucial to estimate number of people needing assistance. Prevalence estimates provide an important statistical context to efforts within a society to plan and provide for groups of interest. To describe how national health interview surveys can be used to study ID. In an initial visit persons with disability who had an additional visit. 20% were interviewed by proxy. ID defined if yes to a question about ID, yes to ID as cause of general activity limitation or if reason for use of health care system was ID. Prevalence of ID in non-institutionalized people=0.78%</td>
</tr>
<tr>
<td>Cans C et al.</td>
<td>France 2003 (51)</td>
<td></td>
<td>“Disabilities and trends over time in a French county 1980-9.”</td>
<td>Improved obstetric and neonatal care has resulted in a decrease in infant mortality, but this has not been followed by a decrease in infant morbidity. Very few surveys study trends in childhood disability. To describe type and prevalence trends over time for severe childhood disabilities (at age 7) in a French county 1980-91, by data from a French register on children with disabilities mainly recruited from institutions allocating support for special schools. Prevalence of severe ID stable from 1980-1991 with a mean of 0.28%. Parental permission of inclusion in register was denied in 2% of children.</td>
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<td>Bradley EA et al.</td>
<td>Canada 2002 (47)</td>
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<td>“Mental retardation in teenagers: prevalence data from the Niagara region, Ontario.”</td>
<td>Research on ID prevalence permit more rational planning of services. True ID prevalence: WHO= 3%, US= 1-3%, Scandinavia=1% Severe ID (age 5-19 years) are stable across time and countries =0.38 % and administrative equals true prevalence. Mild ID vary considerably: Low rates explained by underestimation, but also increased mean IQ or improved environment. Mild ID is associated with class, race and parental occupation. Previous Canadian studies find severe ID (8-10 years)=0.37/0.38% and all ID (15-29 years)=min 0.77 % To provide Canadian population based prevalence data for ID in young people, by identifying and assessing children in community agencies and educational, social and residential institutions (N=175, age 14-20) Overall prevalence ID = 0.718 % Mild ID (IQ 50-75) = 0.354 % (lower than in US studies, same as in some Scandinavian). Severe ID (IQ&lt;50) = 0.364 % (as in literature). Mild ID, but not severe ID decreased with age (this might be due to persons outside an academic setting not being identified as often). Mild ID more frequently in less advantaged social strata. These Canadian results with low mild ID are similar to earlier Swedish results and this is explained by policies of integration with educational integration avoiding stigmatization, although some young persons with mild ID might by unrecognised and have unmet educational and other needs.</td>
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<td>Leonard H et al. Australia 2002 (23)</td>
<td>&quot;The epidemiology of mental retardation: challenges and opportunities in the new millennium.&quot;</td>
<td>The definition, classification and measurement of ID have involved considerably controversy over time. To be able to apply epidemiological principles we need consensus in this area.</td>
<td>To provide an overview of opportunities and challenges in epidemiological studies of ID, by examination of ID data, according to age, gender, social class and ethnicity including issues of definition and methodology, by a review of literature. Traditionally ID has been classified by statistical distribution of IQ, but AAIDD increasingly focus on adaptive behavior in ten applicable skill areas. Epidemiological studies often focus solely on IQ for definition of ID, partly because no standardized measure of adaptive behavior exists. Use of adaptive behavior or nonverbal intellectual measures help to reduce false positive. Type of data sources and purpose of data collection are critical considerations in epidemiological studies. Most children are ascertained from educational data sources. Socio-demographics affect mild ID, but maybe not severe ID. Case definition, ascertainment sources and methods kept constant will allow prevalence to be measured over time.</td>
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<td>Croen LA et al. USA 2001 (50)</td>
<td>&quot;The epidemiology of mental retardation of unknown cause.&quot;</td>
<td>Cause of ID is only known in 30-50% of children with ID. The existing literature most often distinguish between mild and severe ID and assume the same risk factors apply to children with known and unknown cause of ID. An understanding of the epidemiology of ID of unknown cause might add to the description of a causal pathway.</td>
<td>To report infant and maternal characteristics of children with ID of unknown cause, by analyzing data from life birth certificates of children identified with ID by a statewide service agency (N=11114, age 4-12 years). Prevalence of children with ID of unknown cause =0.24%. Of these 64% has mild ID, 20% severe ID and 16% ID of unknown severity. Risk factors of mild and severe ID of unknown cause were male gender and low birth weight as well as black race, increasing maternal age and decreasing level of education. This is contrast to the previous understanding of socioeconomic position having little or no association with severe ID. Additional risk factors for mild ID were multiple births, second or later-born children.</td>
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<td>Stromme P et al. Norway 1998 (42)</td>
<td>&quot;Mental retardation in Norway: prevalence and sub-classification in a cohort of 30037 children born between 1980 and 1985.&quot;</td>
<td>ID is one of the most frequent neurological handicaps among children. True prevalence is difficult to measure. Administrative data are used to identify severe ID. Prevalence rates have decreased in industrialized Western countries and corresponding mean IQ has increased. Reliable baseline data are needed to observe new trends.</td>
<td>Estimate the true prevalence of ID in a large community with a representative sample, by recruiting children from educational and health institutions responsible of diagnosing ID and subsequently assessing their IQ (N=213, age 7-16 years). Nearly all cases with IQ&lt;50 had been neurobiological investigated prior to referral (compared to less than half of IQ 50-70).</td>
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Roeleveld N et al.
Netherlands 1997
(6)
“The prevalence of mental retardation: a critical review of recent literature.”
Mild ID prevalence’s are more difficult to estimate than severe ID. Valid severe ID prevalence’s: register-based case ascertainment followed by re-evaluation of IQ levels. Valid mild ID prevalence’s: register-based case ascertainment supplemented by additional research or a population-based survey including extended psychometric evaluation.
To establish valid estimates of true prevalence rates of severe and mild ID in children (age 5-19 years) and analyze variation in prevalence rates, by a review of literature.
Prevalence of severe ID= 0.38%. Increases until age 15 and younger than 5 is not complete. Decrease after age 15 is probably due to high mortality. Little variation between populations, indicating the aetiology is not influenced greatly by exogenous factors.
Prevalence of mild ID varies greatly between populations and complete assessment is not achieved before maturity. It is difficult to estimate, maybe around 2.98%. Striking association with social class, race and parental occupation. But this might also be due to specific aetiologic factors in lower social classes (endemics, intoxication, parental occupation with chemicals etc.). Strong need for standardisation of definition and research methods in this area. Possibilities for prevention of severe ID are marginal but the prevalence of mild ID could be reduced by improving the biological and psychosocial environment.

Murphy C et al.
USA 1995 (41)
“The administrative prevalence of mental retardation in 10-year-old children in Metropolitan Atlanta, 1985 through 1987.”
Prevalence rates of ID are important to determine educational and health care needs of these children. Prevalence rates vary between 0.3-0.97 (age 10) due to real differences over time and populations, but also due to differences in case definition, identification method and the demographic, social and cultural characteristics of the studied populations.
To estimate prevalence of ID and associated disabilities in children, by analyzing data from a cross-sectional population-based study on children recruited from multiple sources for example public schools, hospitals and public service agencies (N=1074, age 10 years)
Administrative prevalence of mild ID=0.84%, probably underestimated. Administrative prevalence of severe ID=0.36% probably close to true prevalence.
Black race, male gender (mainly mild ID), In total 12.1% of children with mild ID and 44.9% of children with severe ID had coexisting disabilities, mainly cerebral palsy and epilepsy. Children with severe ID more often had multiple neurological conditions.

Rantakallio et al.
Finland 1986 (53)
“Mental retardation and subnormality in a birth cohort of 12000 children in Northern Finland.”
Perimortaly figures have dropped considerably since 1950s, but no clear evidence of a drop in incidence of mental handicap, perhaps even an increase is expected. Relevant figures for incidences of various handicaps are needed, but difficult to find.
To investigate incidence and prevalence of ID (IQ<70) and subnormality (IQ=71-85) (N=12058 born in Northern Finland in 1966)
Prevalence of severe ID=0.63% at age 14 years. The incidence is high possibly due to excess of children with Down syndrome (percentage of mothers above 35 years was high) and to a high completeness in a longitudinal and multisource case finding.
Reference List


11. Diagnostic and Statistical Manual of Mental Disorders DSM-IV. American Association of Psychiatrists. 1994;


