Chapter 4

The prevalence of pervasive developmental disorders in children and adolescents with mental retardation

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Abstract

Insight into the prevalence of pervasive developmental disorders in children and adolescents with mental retardation is known to be of clinical importance. However, estimating this prevalence is complicated. The literature reports prevalence rates ranging from 3 through 50%. This variation seems to be related to the concepts of pervasive developmental disorders under study, the instruments used, and the studied populations. The present study aimed to estimate a reliable prevalence rate of pervasive developmental disorders. A total population based screening with the PDD-MRS and the ABC (n=825), was followed by further assessment of children and adolescents at high risk for pervasive developmental disorders according to these instruments, and for controls, with the ADI-R, ADOS and a DSM-IV-TR classification (n=188). The instruments lead to different prevalence rates. Based on the screening the estimated prevalence rates ranged from 7.8% to 15.9%. Further assessment with the ADI-R, ADOS and DSM-IV-TR classification, revealed higher estimated prevalence rates. The estimated prevalence rates will be related to the concept of pervasive developmental disorders they represent, to the instruments and to the investigated population.

4.1 Introduction

Insight into the prevalence of pervasive developmental disorders (APA, 2000) in children and adolescents with mental retardation is of clinical importance, since the co-occurrence of pervasive developmental disorders is known to cause many extra problems for the child and the environment (Volkmar et al., 1987; Tsai, 1996; Bryson, 1996, 1997; Kraijer, 1997; Carter et al., 1998). This insight will contribute to increasing the knowledge and awareness of health and educational professionals about pervasive developmental disorders, and will have far-reaching consequences for early identification and for providing services and interventions for those affected (Bryson, 1996; Fombonne, 1999).

However, establishing a reliable estimate of the prevalence of pervasive developmental disorders in children and adolescents with mental retardation is a complicated issue, since measuring pervasive developmental disorders in this population is not unequivocal. One reason for this is the behavioral overlap between children with mental retardation and children with pervasive developmental disorders, since low mental ages may account for less developed social and communicative behavior in itself (Wing, Gould, Yeates, & Brierley, 1977; DiLavore, Lord, & Rutter, 1995; Wing, 1997; Kraijer, 1997; Towbin, 1997). Another reason is the fact that the definition of pervasive developmental disorders includes a broad group of children. Especially children with a diagnosis in the category of PDD-NOS are difficult to describe as one homogeneous group (Bailey, Phillips, & Rutter, 1996; Buitelaar, Van der Gaag, Klin, & Volkmar, 1999; Luteijn et al., 2000). But even in the category of core autism, with more stringent criteria in both DSM-IV-TR (APA, 2000) and ICD-10 (WHO, 1992), not all children show the same behavior (Wing & Gould, 1979; Wing, 1997).

On the individual level, the final diagnosis should be based on thorough investigation, interviews with parents and teachers, combined with observations of the child. It is not surprising that classification of pervasive developmental disorders in large groups, to establish the prevalence, can never have the same thorough character of investigation. The reliability of the prevalence rate therefore heavily depends on the procedure used to estimate it.

In table 4.1 an overview is presented of 10 prevalence-studies in populations with mental retardation. For a review of studies on the overall epidemiology of autism we refer to Fombonne (1999).

Table 4.1 Overview of epidemiological research on PDD in populations with mental retardation (MR)

Authors	Year	Level of MR	Definition of studied PDD	Prevalence	Population	Age range	Instruments
Wing & Gould	*1979* *1981*	General population with MR	Triad of Wing & Gould	82.2% (prof) 47.4% (sev) 40.0% (mod) 1.8% (mild)	Camberwell, Great Britain, Mild n=700, Mod.+Sev.+Prof. n= 133	2-14 years	HBS, case notes
Gillberg	*1983*	Severe	Infantile autism, DSM-III Triad of Wing and Gould (1979) Marked problems in social interactions (Wing & Gould,1979)	8% 40% 50%	Swedish hostel, n=56	9-31 years	Highly structured interview by nurse who knew the subject, no systematic psychiatric assessments
Lund	*1985*	General population with MR	Early childhood autism, DSM-III Other psychosis since childhood, DSM-III	3.6% 7,6%	Denmark, n=302, Epidemiological	≥ 20 years	MRC-HBS
Gillberg et al.	*1986*	Severe (IQ≤50) Mild	Infantile autism, DSM-III Including triad of Wing and Gould (1979) and severe social impairments Infantile autism, DSM-III, and Asperger's syndrome (Wing, 1981) Including triad of Wing and Gould and severe social impairments	8% 49% 5% 13%	Representative 4-yr birth cohort from Göteborg, Sweden n=66 SMR n=83 MMR, Epidemiological	13-17 years	Individual assessment by a doctor in the field of child psychiatry, interview of parents, observation of child, review of existing files
King et al.	*1994*	Severe-Profound	Axis II diagnosis of pervasive developmental disorders, DSM-III-R	40%	Institution, Referred for initial psychiatric consultation, California, n=251	2-81 years	Examination/observation in residential, work, or schoolsettings, history, review of existing files, DSM-III-R
Kraijer	*1991*	General population with MR	Pervasive developmental disorders, DSM-III-R	34,9%	Observation clinic, Assen, Netherlands, n=393 7,9% PMR, 29,8 SMR, 34,6% Mod.MR, 27,7% MMR	0-14 years	Reviewing existing files, history, physical and psychological examination, observation, PDD-MRS
Deb & Prasad	*1994*	General population with MR	Autistic disorder, DSM-III-R	14.3%	Learning disabled, attending special schools, Grampian, Great Brittain n=634	5-19 years	Questionnaire for teachers, assessment by child psychiatrist
Steffenburg et al.	*1996*	General population with MR with epilepsy	Autistic disorder, DSM-III-R Autisticlike conditions, >6 crit for aut, DSM-IIIR Asperger's syndrome, Gillberg & Gillberg, 1989 Autistic traits, 3-5 crit for aut, DSM-III-R	27% 11% 3% 3%	Representative 11-year birthcohort from Göteborg, Sweden, N=90 Epidemiological	8-16 years	Clinical examination by child neuropsychiatrist, HBS, CARS, ABC, AS Diagnostic Checklist, GAFS, SOFAS
Kraijer	*1997*	General population with MR	Pervasive developmental disorders, DSM-III-R	38.3% (inst) 22.6% (day)	Institutions, n = 718 Day care facilities, n = 297 40% PMR, 40% SMR, 20% Mod MR, 17.3% MMR	3-80 years	Clinical Assessment, PDD-MRS
Morgan et al.	*2002*	General population with MR	Infantile Autism/Atypical Autism, ICD-10	30%	Learning disability, designated area, n=571	adults	PDD-MRS, clinical classification based on ICD-10

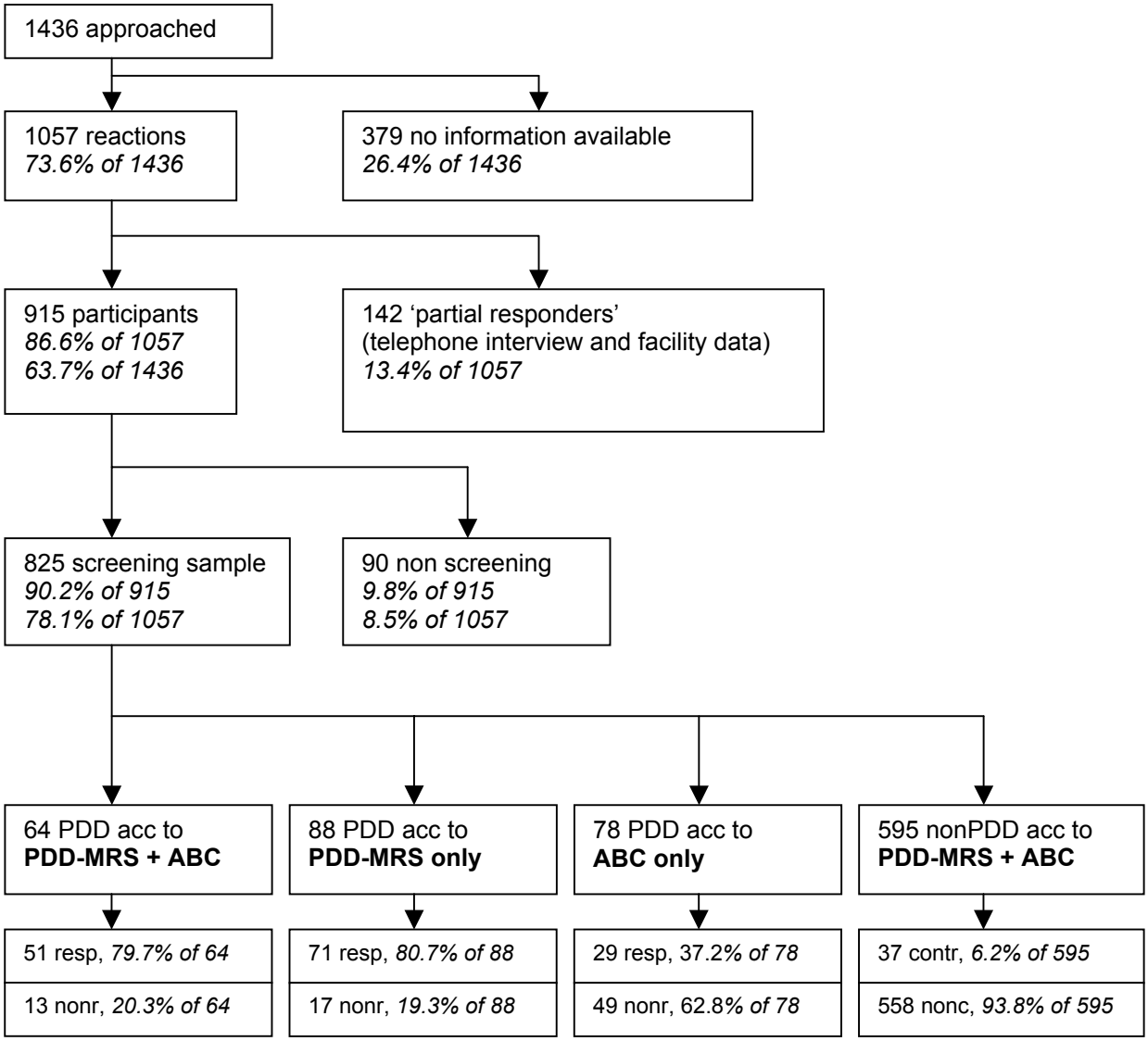
The studies in table 4.1 illustrate the difficulties in providing a reliable estimate of pervasive developmental disorders in individuals with mental retardation, since the procedures and therefore the prevalence rates vary widely over the studies. The concept of pervasive developmental disorders under study and the studied population seem to play the most important role therein. The concepts under study differ in narrowness or width in two ways. First, the width of the total concept of pervasive developmental disorders varies over the studies, e.g. the DSM-IV (APA, 1994) had a broader definition of PDD-NOS than the DSM-IV-TR (APA, 2000), including more children with less severe problems in the spectrum of pervasive developmental disorders. Second, the studies focus on different concepts within the PDD-spectrum, e.g. AD or the spectrum of pervasive developmental disorders, and both can be defined in a stricter or somewhat wider way. These differences in concept are related to the different theoretical frameworks, the different DSM-versions and, equally important, the different instruments that form the basis for the studies. It leads to the inclusion of different kinds or severity levels of problems in the area of pervasive developmental disorders in each study. The studied populations differ on many background variables, such as age range, level of mental retardation, or representativity (clinic, residential setting, school, total population, etc.).

Taking this into consideration, it is not surprising that the outcomes of the studies mentioned above are quite variable. To estimate a prevalence rate seems to be more straightforward than to interpret it and to generalize it to the total population with mental retardation. Nevertheless, as indicated above, a reliable and unambiguous estimate of the prevalence of pervasive developmental disorders is of major importance from a clinical perspective. Given this importance, further investigation of the prevalence is still needed.

The present study aims to establish an estimate of the prevalence of pervasive developmental disorders in the total population of children and adolescents with mental retardation, as reliable as possible. Therefore, two issues were considered to be important. First, the construct of pervasive developmental disorders, represented by a given prevalence rate should be specified. Second, the (representativity of the) studied population should be described precisely. With respect to the construct under study, we investigated the spectrum of pervasive developmental disorders, ranging from ‘core Autism’ to ‘broader PDD’. For each prevalence rate given, the concept represented will be specifically defined. With regard to the population, we decided to

investigate a total population of children and adolescents with mental retardation, in a designated area. The range of the population, including all levels of mental retardation, and the specific definitions of each concept will contribute to the reliability and the generalization of the estimated prevalence rate.

Figure 4.1 Procedure and participants



4.2 Method

Participants

The participants for this study were recruited from Friesland, a northern province of the Netherlands. All 1436 children and adolescents between 4 and 18 years, known to facilities for children and adolescents (suspected) with mental retardation (schools, day-care facilities and institutions), were approached. All levels

of mental retardation were included. No participants were excluded based on etiology of mental retardation, presence of sensory or motor impairments, or co-morbid psychiatric disorders or behavioral problems. Only children from families who did not speak the Dutch or Frisian language were excluded. Figure 4.1 gives an overview of the sampling.

The initial number of 1436 children, was the maximum estimate of children to include in our study. This number is probably too high, for in the ultimate response group (n=915), approximately 10% of the children did not fulfill all criteria and therefore had to be excluded from the analyses. A group of 1057 could be contacted personally. Due to privacy regulations, of 379 children and adolescents, no other information was available than type of facility. Comparing the response group of 1057 with the total 1436, showed that the responders were significantly deviant from the total population, concerning this one aspect (χ^2=34.4, df=3, p<.001). The response group contained more children from schools for severe learning problems, whereas the non-response group (n=379) largely consisted of children and adolescents from schools for mild learning problems. Table 4.2 presents the characteristics of the 1057 children and adolescents who were contacted personally.

Table 4.2 Characteristics of the population of children and adolescents with MR in Friesland (n=1057)

		Institution		**Day-care facility**		**School for severe learning problems**		**School for mild learning problems**	
		n	***%***	***n***	***%***	***n***	***%***	***n***	***%***
Sex	***Male***	31	75.6	60	55.6	320	64.8	255	61.6
	Female	10	24.4	48	44.4	174	35.2	159	38.4
Level of MR	***Profound***	29	70.7	62	57.4	4	0.8	1	0.2
	Severe	5	12.2	29	26.9	68	13.8	6	1.4
	Moderate	1	2.4	9	8.3	153	31.0	44	10.6
	Mild	6	14.6	5	4.6	237	48.0	328	79.2
	Non-MR	0	0.0	3	2.8	32	6.5	35	8.5
Age	***< 12***	9	22.0	87	80.6	211	42.7	190	45.9
	≥ 12	32	78.0	21	19.4	283	57.3	224	54.1
	Total	**41**		**108**		**494**		**414**	

In 86.6% (n=915) of the 1057 cases we received information from parents and from the school, day-care facility or institution. This group is referred to as

'participants'. In 13.4% (n=142) of the cases, we received information from the parents during a telephone interview, and information from school, day-care facility or institution. This group is referred to as 'partial responders'.

Comparing the partial responders with the participants revealed no significant differences between the two groups with respect to sex or age. However, a significant difference existed with respect to the facility of the child (χ^2=9.61, df=3, p=.019), and with respect to level of mental retardation (χ^2=19.62, df=4, p=.001). In the participants group, 47.7% of the children attended a school for severe learning problems, and 37.9% attended a school for mild learning problems. In the partial responders group, 38.7% attended a school for severe learning problems, and 47.9% attended a school for mild learning problems. The differences between the groups were much less explicit in institutions (partic. 3.4%, part-resp. 6.3%) and day-care facilities (partic. 11.0%, part-resp 7.0%). Additionally, the participants contained relatively many children from the severe and moderate levels of mental retardation, as opposed to partial responders, that for 70.4% consisted of children from the mild level of mental retardation. These factors are related to each other, since children from schools for mild learning problems are obviously more likely to have mild mental retardation than children from schools for severe learning problems (84.1% vs. 48.8%).

The differences between the two groups with respect to the distribution of facilities and the level of mental retardation, point into the same direction: children from lower levels of functioning are overrepresented in our sample, whereas higher functioning children are underrepresented, as compared to the total population.

The participants were assigned to the four levels of mental retardation as defined by the DSM-IV-TR (APA, 2000): profound (IQ 0-20), severe (IQ 21-35), moderate (IQ 36-50) or mild (IQ 51-70), and some were classified as non-mentally retarded (IQ 71 or above). In 80% of the cases, this classification was based on information from intelligence tests or developmental tests obtained by the facility. In most cases these were standardized tests, e.g. Dutch versions of the Wechsler Intelligence Scale for Children-Revised, WISC-R (Wechsler, 1974; Vander Steene et al., 1986), Wechsler Preschool and Primary Scale for Intelligence-Revised, WPPSI-R (Wechsler, 1989; Vander Steene & Bos, 1997), Snijders-Oomen Niet-verbale intelligentie test-Revised, SON-R (Snijders, Tellegen, Winkel, & Laros, 1996), and the Bayley scales of Infant Development (Bayley, 1969; Van der Meulen & Smrkovsky,

1983). In the other 20% of the cases, participants were assigned to one of these categories based on the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984), administered as part of the study, and on clinical review of functioning.

For this prevalence study, a division in two levels of mental retardation was made, i.e. mild mental retardation (IQ 51-70) and the combined levels of moderate/ severe/profound mental retardation (IQ ≤ 50). This division is mainly based on the character of the level of mental retardation. Children with mild mental retardation often show some more adaptive behavior development and often learn (at least some) reading and writing, whereas children with lower levels of mental retardation are considered to experience more severe consequences of their mental retardation, on a very basic level. This latter group is referred to as the combined lower levels of mental retardation. Additionally, the two groups contain more equal numbers of children (mild 460, combined lower levels 365).

For data analysis in this prevalence study, we only included children and adolescents who fulfilled the criteria of 1) mental retardation, 2) age 4 through 18 years when data collection started, 3) from Dutch or Frisian speaking families, and 4) information available from both parents and facility on behavior in the area of pervasive developmental disorders. A sample of 825 participants (age mean 11.4, sd 3.7) was included according to these criteria. This sample did not differ from the 915 participants, with respect to age, sex, or facility.

In the first step of the study, this sample was screened for pervasive developmental disorders, with the PDD-MRS (Kraijer, 1997, 1999; de Bildt et al., 2003b), and the ABC (Krug, Arick, & Almond, 1980; de Bildt et al., 2003b).

Based on the outcomes of the screening, 188 children and adolescents were further assessed, with the ADI-R (Lord, Rutter, & Le Couteur, 1994), the ADOS (Lord, Rutter, DiLavore, & Risi, 1998; Lord et al., 2000) and a DSM-IV-TR classification (APA, 2000). This sample contained 51 participants identified as PDD by both the PDD-MRS and the ABC, 71 by the PDD-MRS only, 29 by the ABC only, and 37 identified as non-PDD by both screeners. In figure 4.1 the response rate of each category is presented. In the PDD group according to the PDD-MRS, ABC or both, no significant differences were found for sex, age, facility or level of mental retardation. In the non-PDD group, the selected participants differed significantly from the non-selected with respect to age (p=.000), facility (χ^2=95.71, df=3, p=.000) and

level of mental retardation (χ^2=49.29, df=3, p=.000). The selected children were younger (mean 9.31, sd 3.46 vs. mean 12.22, sd 3.46), more often from day-care facilities (51.4% vs. 5.6%) and from the profound and severe levels of mental retardation (51.3% vs. 11.1%). The characteristics of the screening sample and the assessment sample are presented in table 4.3.

Table 4.3 Characteristics of the prevalence sample (n=825)

		Participants			
		Screening		Assessment	
		n	***%***	***n***	***%***
Sex	***Male***	519	62.9	133	70.7
	Female	306	37.1	55	29.3
Age	***< 12***	435	52.7	113	60.1
	≥ 12	390	47.3	75	39.9
Level of MR	***Profound***	78	9.5	39	20.7
	Severe	102	12.4	54	28.7
	Moderate	185	22.4	32	17.0
	Mild	460	55.8	63	33.5
Total		825		188	

Instruments

Screening

Scale of Pervasive Developmental Disorder in Mentally Retarded persons (PDD-MRS)

The PDD-MRS (Kraijer, 1997, 1999) is a Dutch instrument for the spectrum of pervasive developmental disorders, based on the DSM-III-R, including AD and PDD-NOS, but with no differentiation between them. The instrument is developed for children and adults with mental retardation, and is well studied and widely used in the Netherlands and Belgium (Kraijer, 1997; de Bildt et al., 2003b). It is a 12-item questionnaire completed by clinicians, with dichotomous items on the three aspects of pervasive developmental disorders: communication, social behavior and stereotyped behavior. Weighted factors 1, 2, or 3 are assigned to the item scores and the maximum score is 19. Psychometric qualities were reported to be good. Scores on the PDD-MRS are divided into three categories: a PDD category (scores of 10

and more), a doubtful PDD/non-PDD category (scores between 7 and 9) and a non-PDD category (scores of 6 or less). In this study, psychologists or teachers completed the PDD-MRS.

Autism Behavior Checklist (ABC)

At the time of this study, the ABC (Krug et al., 1980) was the only available standardized internationally applied instrument for autism, suitable for screening a large population. The ABC was included with the criteria as given by Oswald and Volkmar (1991). They proposed a cutoff of 58 and higher for AD and below 58 for non-AD, based on their study of sensitivity and specificity for the weighted sum scores, compared to a clinical DSM-III diagnosis of AD. Parents completed the ABC.

Assessment

Autism Diagnostic Interview-Revised (ADI-R)

Seven trained interviewers administered the ADI-R (Le Couteur et al., 1989; Lord et al., 1994) in the home of the parents/caregivers. All interviewers had reached 80% reliability in scoring the ADI-R as required. Data analysis involved the recommended algorithm, which provides information on the history and development of the individual (mostly information on 4-5 years of age, or 'ever'). The internal consistency of the ADI-R domains in our sample were slightly lower than described by Lord et al. (Lord et al., 1994), but followed the same pattern, with highest alpha's for social (.90) and communication (.76), and lowest for repetitive behavior (.59).

Autism Diagnostic Observation Schedule (ADOS)

Three trained examiners administered the ADOS (Lord et al., 1998, 2000) in the school, day-care facility or institution where the child was during the day. Each examiner had reached 80% reliability as required. All ADOS's were videotaped. Two trained raters coded the ADOS independently, immediately after administration. When raters disagreed on scoring, consensus was reached by discussing the item and reviewing the videotape.

Internal consistency of the domains varied between the different modules. For all modules the pattern of internal consistency corresponded with the pattern of internal consistencies described by Lord et al. (2000), with highest internal consistency for the social domain (.51-.82) and the combination of the social and

communication domain (.66-.87), lower for the communication domain (.58-.67) and lowest for stereotyped behavior (.07-.44). In general the values found for module 1 and 2 were higher than for module 3 and 4. This may be due to the number of individuals observed with each module.

Clinical classification

Clinical classifications were assigned by four experienced clinicians, two board certified child and adolescent psychiatrists, one clinical and developmental psychologist and one resident. When a case was considered difficult to assess, consensus classification was reached through reviewing and discussing the available information.

The clinical classification was made according to DSM-IV-TR criteria, based on parent information, collected with the ADI-R and observation of the child on video, during the ADOS. The clinicians were blind for the outcome on the algorithms of the ADI-R and the ADOS. Each combination of two clinicians classified ten children in common. They were unaware of which child, which other clinician or outcome of the other classification. In order to measure the level of agreement of the diagnostic classification (AD, PDD-NOS, non-PDD) between clinicians, a weighted kappa was calculated. The weights we used were 1 for exact agreement, .5 if one rater scored autism and the other PDD-NOS and 0 in all other cases. The percentage of agreement found was 81.2% and the weighted kappa coefficient was .66 (sd .13). Both the percentage of agreement and the weighted kappa values are considered good according to the criteria of Cicchetti (2001), that combine the criteria reported earlier by Cicchetti and Sparrow (1981) for weighted kappa values and the criteria reported by Cicchetti, Volkmar, Klin and Showalter (1995) for percentages of agreement.

4.3 Results

The screening resulted in three estimates of the prevalence rate of pervasive developmental disorders, in the total population of children with mental retardation. Table 4.4 presents 1) a prevalence rate based on classification by the PDD-MRS, 2) one based on the ABC, and 3) one based on the combination of both instruments. The prevalence rates based on one of the instruments include the participants

classified as PDD by that instrument and the participants classified as PDD with both instruments together.

According to both instruments (together and separately) the prevalence varies over the levels of mental retardation (PDD-MRS χ^2=56.99, df=1, p=.000; ABC χ^2=29.35, df=1, p=.000, both screeners χ^2=69.22, df=3, p=.000). Highest prevalences are found in the combined lower levels of mental retardation, and lowest rates in the mild level of mental retardation Specified for the lower levels of mental retardation, the results are as follows: Mod. PDD-MRS 16.8%, ABC 15.7%, Both 7.0%; Sev. PDD-MRS 43.18%, ABC 33.3%, Both 19.6%; Prof. PDD-MRS 43.6%, ABC 37.2%, Both 28.2%. The difference in prevalence rate between boys and girls is significant for the PDD-MRS (χ^2=12.75, df=1, p=.000) and for the combination of both screeners (χ^2=13.04, df=3, p=.005), with higher prevalences found in boys, especially in the mild level of mental retardation.

Table 4.4 Prevalence rates of PDD according to PDD-MRS, ABC and the combination of both

			PDD-MRS PDD	**ABC PDD**	**PDD-MRS + ABC PDD**
		n	***%***	***%***	***%***
Level of MR	***Mild***	460	9.3	10.9	2.0
	Comb. lower levels	365	29.9	25.2	15.1
	Total	825	18.4	17.2	7.8

These prevalence rates are applicable to the observed population of 825. To be generalized to the total population, a correction must be applied. First, a correction was needed with respect to the distribution over levels of mental retardation, since the observed sample was underrepresentative for the mild level of mental retardation. Additionally, we could simply follow the assumption that the prevalence rate is the same in the observed and the non-response population, and generalize the observed prevalence. This would result in an estimated prevalence of PDD on the PDD-MRS of 17.9% in the total population. However, from 128 children and adolescents of the non-response population, a PDD-MRS was available, indicating a lower prevalence rate in this population, i.e. 12.5%. Therefore, we applied a correction for non-response, based on the assumption that the prevalence rate in the non-response population is lower than in the observed population. This resulted in a corrected prevalence of PDD, according to the PDD-MRS, of 15.9% for the total population. For

the ABC, no data were available from the non-responders. However, we followed the same correction method and assumptions as for the PDD-MRS, leading to prevalence rates of 16.8% and 15.2%. An estimated prevalence rate based on classification by both screeners, would be 7.7%, assuming no difference between responders and non-responders with respect to prevalence of PDD based on both instruments. Correction, based on the assumption of a prevalence of 12.5% in the non-response population, does not seem meaningful for this group. The selection is much stricter here, and it is therefore supposed to narrow down the prevalence in the non-response population as well. The estimates are presented in table 4.5.

Table 4.5 Observed and corrected prevalence rates of PDD according to PDD-MRS and ABC

	PDD-MRS	**ABC**	**PDD-MRS + ABC**
	%	***%***	***%***
Observed	18.4	17.2	7.8
Assumption: No diff in prevalence between resp and non-resp	17.9	16.8	7.7
Assumption: Diff in prevalence between resp and non-resp	15.9	15.2	

Correction for non-response in the two levels of mental retardation did not result in major changes. First, since the distribution of the prevalence of PDD over the levels of mental retardation in the 128 non-responders was not known, correction based on that prevalence was impossible. Second, with respect to the mild group, correction based on the same prevalence rate in non-responders did not change the prevalence rate, due to the correction method, which was based on the four levels of mental retardation. For the combined moderate/severe/profound level, the prevalence rates changed only slightly after correction: PDD-MRS 30.4%, ABC 25.7%, and both 15.6%.

In table 4.6, the classifications of the ADI-R, ADOS and DSM-IV-TR are presented for children identified as PDD by the PDD-MRS, by the ABC, or by both. The first two groups (PDD-MRS PDD and ABC PDD) include all participants classified as PDD by that screener, the last group consists of participants who were classified as PDD by both screeners together.

The highest prevalence is found in the group identified as PDD by both the PDD-MRS and the ABC, according to the ADI-R (80.4% AD), the ADOS (72.5 + 19.6 = 92.1% PDD) and the DSM-IV-TR (56.9 + 25.5 = 82.4% PDD). The differences in

prevalence rate between the two levels of mental retardation are not significant for any of the three diagnostic instruments. When the PDD-MRS PDD group is considered, the prevalence rates vary over the two levels of mental retardation significantly for the DSM-IV-TR (χ^2=8.13, df=2, p=.017) and for the ADOS (χ^2=8.57, df=2, p=.014), with the highest prevalences in the combined lower levels of mental retardation. With respect to the ABC PDD group, the prevalence rates in the two levels of mental retardation differ significantly for the DSM-IV-TR (χ^2=8.20, df=2, p=.017), the ADOS (χ^2=18.53, df=2, p=.000), and the ADI-R (χ^2=5.78, df=1, p=.016), again with the highest prevalences in the combined lower levels group. Within the broader PDD classification of the ADOS and DSM-IV-TR, generally the AD:PDD-NOS ratio is approximately 3:1 for the ADOS, and 2.5:1 for the DSM-IV-TR, except for the PDD-MRS PDD subgroup with mild mental retardation (ADOS 1:1; DSM-IV-TR 1:2). This indicates that the proportions of children identified as AD by the ADOS and DSM-IV-TR resemble the proportion identified as AD by the ADI-R.

Table 4.6 Prevalence of PDD according to the ADI-R, ADOS and DSM-IV-TR in children identified as PDD by the PDD-MRS or the ABC or the combination

			ADI-R	**ADOS**	**DSM-IV-TR**
	Level of MR	***n***	***% AD***	***% PDD***	***% PDD***
PDD-MRS PDD	***Mild***	33	48.5	75.8	48.5
	Comb. Lower levels	89	56.2	87.1	67.4
	Total	**122**	**54.1**	**84.5**	**62.6**
ABC PDD	***Mild***	24	54.2	41.7	58.3
	Comb. lower levels	56	80.4	87.5	78.6
	Total	**80**	**72.5**	**73.8**	**72.5**
PDD-MRS +	***Mild***	6	83.3	66.7	83.3
ABC PDD	***Comb. lower levels***	45	80.0	95.6	82.2
	Total	**51**	**80.4**	**92.1**	**82.4**

To estimate meaningful prevalence rates in the total population, we generalized these numbers to the population of 825. Therefore, we applied a correction method based on the prevalence rates according to each diagnostic instrument, for those children in the observed sample of 188, who were identified as PDD by at least one of the two screeners. We assumed these numbers to be the most reliable to generalize to the population of 825. The prevalence rates observed in each group of

screen positives (e.g. PDD-MRS+/ABC+, PDD-MRS+/ABC- and PDD-MRS-/ABC+) were generalized to those children from the 825, who fell into one of these groups. Estimates were then established for the total population, and the subgroups mild and combined moderate/severe/profound mental retardation, as presented in table 4.7.

Table 4.7 Estimated prevalence of PDD based on a classification on the ADI-R, ADOS or DSM-IV-TR, in the total sample of 825, for the mild and combined moderate/severe/profound levels of mental retardation, and the total population

	DSM-IV-TR	**ADI-R**	**ADOS**
Level of MR	***% PDD***	***% AD***	***% PDD***
Mild	9.3	11.3	10.2
Comb. lower levels	26.1	21.5	32.1
Total	16.7	16.8	19.8

The estimated prevalence rates, based on PDD according to one of both screeners and the DSM-IV-TR or ADI-R, highly resemble each other in the total population. However, the ADI-R represents the prevalence of AD, whereas the DSM-IV-TR represents the prevalence of PDD, including AD (estimated prevalence 8.8%) and PDD-NOS (estimated prevalence 7.9%). The prevalence of PDD according to the ADOS, also including AD (14.5%) and PDD-NOS (5.3%), is somewhat higher. As expected, the estimated prevalence rates are higher in the combined lower levels of mental retardation, and the ratio of AD:PDD-NOS on the DSM-IV-TR and the ADOS is different in this group (estimated prevalence mild: DSM-IV-TR AD 3.0%, PDD-NOS 6.3%; ADOS AD 5.8%, PDD-NOS 4.4%; combined lower levels: DSM-IV-TR AD 16.1%, PDD-NOS 9.9%; ADOS AD 25.6%, PDD-NOS 6.6%).

4.4 Discussion

The objective of the present study was to establish a reliable estimate of the prevalence of pervasive developmental disorders in the total population of children and adolescents with mental retardation. Two issues were considered to be important for the reliability, and therefore for the clinical value of a given prevalence rate. First, the construct of pervasive developmental disorders, represented by a given prevalence rate, needs to be specified. Second, the (representativity of the) studied population should be described precisely. Regarding the construct of pervasive developmental disorders, this study focused on the spectrum of pervasive

developmental disorders, ranging from ‘core Autism’ to ‘broader PDD’. With respect to the population, a total population of children and adolescents with mental retardation, in a designated area, was approached for investigation. The range of the population, including all levels of mental retardation, and the specific definitions of each concept will contribute to the reliability and the generalization of the estimated prevalence rate.

The most reliable and well-founded estimate of the prevalence rate from this study possibly is the estimate based on the classification with the DSM-IV-TR, e.g. 16.7% for the total population, 9.3% in the mild, and 26.1% in the combined moderate/severe/profound levels of mental retardation. This prevalence represents the most recent DSM-definition of the PDD spectrum, including AD (estimated prevalence 8.8%) and PDD-NOS (estimated prevalence 7.9%). Additionally, it takes into account information from parents (as collected with the ADI-R) and from professionals who observed the child (by reviewing the videotape of the ADOS). This is in line with Fombonne (2003), who recently re-emphasized the importance of relying on multiple sources in epidemiological studies of pervasive developmental disorders. Furthermore, through this procedure, information about the current situation is combined with the developmental history of the child. The method we followed for the DSM-IV-TR classification most securely approaches the diagnostic process on an individual level.

To increase comparability with other studies, we also reported the prevalence rates, observed with each instrument. The estimated prevalence rate based on the ADI-R, e.g. 16.8% for all levels of mental retardation together, only represents AD as defined in the DSM-IV. This classification is mostly based on developmental history, and only takes into account parent information. It is known from various studies that the ADI-R seems to be over-inclusive in low functioning children and adolescents (Lord, Storoschuk, Rutter, & Pickles, 1993; Lord et al., 1994, 1997; Cox et al., 1999). Our own finding of a high interrelationship between the ADI-R and the DSM-IV-TR classification in low functioning children and adolescents (de Bildt et al., 2003a), was limited by the fact that the proportion of children and adolescents with PDD according to the DSM-IV-TR was high, and the variance within the group was very small. Hence, a cautious interpretation of the ADI-R prevalence rate of AD is needed, since it may overestimate the actual prevalence.

The same probably holds true for the estimated ADOS prevalence of PDD, e.g. 19.8%, including AD (estimated prevalence 14.5%) and PDD-NOS (estimated prevalence 5.3%) as defined in the DSM-IV. The ADOS prevalence is based on current information only, obtained by a professional. No developmental history or parent information is collected for this prevalence rate. Again, the literature is cautious when it comes to discriminating between low functioning children with and without PDD with the ADOS (Lord et al., 2000). Additionally, we found that the ADOS tended to be somewhat over inclusive in general when compared to the DSM-IV-TR classification (de Bildt et al., 2003a). Therefore, the ADOS prevalence rate of PDD is considered to overestimate the prevalence as well.

The estimated prevalence rates found with screening instruments only, are slightly lower than the ones estimated on the basis of more thorough assessment. Again, the PDD-MRS and the ABC only reflect the perspective of one informant. The prevalence of 15.9% based on the PDD-MRS most reliably represents the prevalence of the spectrum of pervasive developmental disorders (defined along the criteria of the DSM-III-R), according to professionals. The prevalence of 15.2% based on the ABC most reliably represents the prevalence of autism (no DSM or ICD definition), as viewed by parents. Both estimates are based on the observed prevalence in the participating population, and the hypothesized lower prevalence of 12.5% in the non-response population. This hypothesis can be partially corroborated for the PDD-MRS, since information on the prevalence based on the PDD-MRS, was available for a substantial group of non-responders. For the ABC, this hypothesis can be no more than an assumption that the prevalence of pervasive developmental disorders as found with the PDD-MRS, is also representative for the prevalence on the ABC in that group, which limits the correction for non-response to some extent. Nevertheless, the prevalence rates observed in the participants differ so little from each other, that the percentages of children identified in the non-response group are also assumed to resemble each other to a large extent. The prevalence of 15.2% based on the ABC is then a cautious estimate, compared to the one based on the assumption that there is no difference between participants and non-responders.

In comparison with the total population of children and adolescents with mental retardation in Friesland, our sample contained relatively many participants from the lower levels of mental retardation, and less from the mild level. Therefore, our sample was not totally representative, which may have influenced our results to some extent,

leading to overestimating the prevalence of pervasive developmental disorders in the total population of children and adolescents with mental retardation. However, with respect to the *screening*, we minimized this influence with our correction for non-response, in which we extrapolated our observed findings to the total population of 1436 based on the facilities of the children. Therefore, we consider the reported prevalence rates based on the screening, to be reliable and representative.

Correction for non-response was less well possible with respect to the *assessment procedure*. Children and adolescents who were identified as non-PDD by both screening instruments were strongly underrepresented in our assessment group. Therefore merely generalizing the results to the total population led to extremely high prevalence estimates (e.g. 38.8% for the DSM-IV-TR, 36.0% for the ADI-R and 60.1% for the ADOS) in the total population. Yet, we were able to generalize the observed findings during the assessment to the screening sample of 825, based on the identification of a child as PDD by either one of the screening instruments. This information was lacking in the other 611 children. The estimated prevalence rates for the separate levels of mental retardation are considered to be reliable and representative, for the main representativity issue (i.e. an underrepresentation of children from the mild level of mental retardation) does no longer influence the prevalence rates to such large extent.

In sum, the reported estimated prevalence rates based on the DSM-IV-TR, ADI-R and ADOS, are the most reliable, representative and well-founded estimates we can give. If the prevalence is assumed to be lower in the non-response population, these prevalence rates may be considered an overestimate of the actual prevalence. Additionally, we must emphasize the fact that we did not include an individual assessment of the participants, that would be needed for a final diagnosis. Although the children were clinically classified, the clinical perspective was less well integrated with the instrumental classification than would be the case in clinical practice.

Comparing our results to earlier prevalence studies from the literature is complicated, due to the differences in the concepts of PDD under study, and in the studied populations. None of the studies used the ADI-R or ADOS, the most recent DSM classifications were from the DSM-III-R (APA, 1987). Additionally, the sampling of the population complicates the comparison. Hence a precise and meaningful comparison cannot be made. Nevertheless, in general the prevalence rates from our study seem lower than those reported in some of the earlier studies. Besides the

variation in DSM classifications and instruments, the main explanation for this difference seems to be the specific populations studied in those studies (e.g. institutions/day-care facilities, where specifically uncomplicated children with mild mental retardation are underrepresented (Kraijer, 1997), mental retardation with epilepsy (Steffenburg, Gillberg, & Steffenburg, 1996), observation clinic (Kraijer, 1991), or severe/profound mental retardation (King, DeAntonio, McCracken, Forness, & Ackerland, 1994). Compared to our epidemiological sample, these populations could be considered as more or less 'high risk' populations, therefore leading to higher prevalence rates.

When we compare our results to prevalence rates that were recently reported in the total population (Baird et al., 2000; Chakrabarti & Fombonne, 2001; Bertrand et al., 2001; Yeargin-Allsopp et al., 2003), the prevalence of PDD is higher in our population. Fombonne (2003) considers a prevalence of autism spectrum disorders of approximately 60 per 10.000 to be representative. Compared to this prevalence of autism spectrum disorders of .6% in the total population, the DSM-IV-TR prevalence of pervasive developmental disorders, is 15.5 times higher in children and adolescents in our population with mild mental retardation, and even 43.5 times higher in the combined lower levels. This is not surprising, since over the years many studies have shown that mental retardation occurs very frequently in children with a pervasive developmental disorder, especially in autism or Autistic Disorder, with estimates between 70 and 90% (DeMyer et al., 1974; Wing & Gould, 1979; Wing, 1981; Rutter, 1983; Steffenburg & Gillberg, 1986; Wing, 1993; Bryson, 1996, 1997; APA, 2000). Due to the increasing attention for High Functioning Autism and Asperger's Syndrome, the percentage of individuals with a pervasive developmental disorder and mental retardation tends to decrease. Nevertheless, it is still substantial (Gillberg, 1999). However, the higher prevalence rate in children and adolescents with mental retardation implicates, that the recent interest in High Functioning Autism and Asperger's Syndrome, should not be at the expense of children and adolescents with pervasive developmental disorders and mental retardation.

From a clinical perspective, two main conclusions can be drawn from this study. First, on an individual level, it is important to consider the concept of pervasive developmental disorders when a child or adolescent shows a delay in his/her development that indicates mental retardation, for its relatively high prevalence. To identify whether a pervasive developmental disorder plays a role in the development

or behavior of the particular child, a thorough individual assessment is recommended, based on the current behavior and the developmental history, and combining information from parents and professionals. A pre-screening that identifies the individuals at high risk, would contribute to avoiding an unnecessary burden for the child and his/her family, and to work efficiently within the time limits of clinicians. In that case, it is important to involve multiple sources of information in the screening.

Second, on the more general level of the policy on planning services for children and adolescents with pervasive developmental disorders, governmental institutions and facilities for children with mental retardation should have a clear idea of the fact that pervasive developmental disorders are so wide spread. Services should be planned based on this high prevalence, to enhance early identification, and provision of preventing or intervening measures and services. This study gives an indication of the prevalence that may serve as a basis. Besides, facilities may wish to screen their own population to specify our indication for their particular populations.