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Epidemiologic data on Asperger disorder

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In 1944, Asperger described a syndrome that has subsequently been given his name, although there is evidence from the earlier European literature that clinical descriptions matching this disorder were available in the 1920s [1]. Asperger's work, however, was largely ignored up to the seminal article by Wing [2], which led to a resurgence of interest in this diagnostic concept. Asperger disorder (AD) was only introduced as a separate diagnostic category in the latest revisions of ICD-10 [3] and DSM-IV [4]. Epidemiologic research on this disorder has only started in recent years and therefore data are still scarce on the prevalence of this syndrome. The goal of this article is to review available epidemiologic surveys that have shed light on the prevalence of AD.

Study selection

Previous reviews of epidemiologic studies of autism and pervasive developmental disorders (PDD) were identified [5–7], and studies providing relevant information for the epidemiology of AD were selected. Medline searches were also performed to identify any other survey not included in the prior reviews. Seven studies that fell into two groups were selected at the end of this process: studies investigating only AD and no other pervasive developmental disorder (one survey), and generally larger studies providing specific estimates for the prevalence of AD alongside estimates for other subtypes of pervasive developmental disorders (six surveys).

Survey of Asperger disorder only

One survey conducted in Sweden investigated the prevalence of AD in a small sample of children attending five mainstream schools in a Göteborg middle-class

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Table 1
Asperger syndrome (AS) in recent autism surveys: study characteristics

Reference	Country	Area	Size of target population	Median age of sample
Sponheim and Skjeldal, 1998	Norway	Akershus County	65,688	5
Taylor et al, 1999	United Kingdom	North Thames	490,000	7
Kadesjö et al, 1999	Sweden (Central)	Karlstad	826	8
Powell et al, 2000	United Kingdom	West Midlands	25,377	8
Baird et al, 2000	United Kingdom	South-East Thames	16,235	7
Chakrabarti and Fombonne, 2001	United Kingdom Midlands	Staffordshire, Midlands	15,500	5

	Screening		Intensive assessment		
Reference	Instruments	Informants	Informants	Instruments	Diagnostic Criteria
Sponheim and Skjeldal, 1998	10 items screening Schedule	Pediatricians + maternal-child health clinics + child psychiatrists	Parent Child	Parental Interview + direct observation CARS, ABC	ICD-10
Taylor et al, 1999	Computerized special needs/disability registers at CDC, and records from special schools	All data on files	Record	Rating of all data available in child record	ICD-10

Kadesjö et al, 1999	Letter to professionals to identify children with autistic conditions + direct assessment of 1 child in 2 in target population together with parent/teacher interviews.	Well-baby clinics, autism assessment teams	Child Parent Professional	ADI_R, Griffiths Scale or WISC Asperger Syndrome Screening Questionnaire	DSM-III-R /ICD-10 Gillberg's criteria (Asperger syndrome)
Powell et al, 2000	Medical records	Pediatricians Child psychiatrists	Records	ADI-R Available data	DSM-III-R DSM-IV ICD-10
Baird et al, 2000	CHAT Checklist for referrals PDD questionnaire Record search	Parents, GP	Parents Child Other data	ADI-R psychometry	ICD-10 DSM-IV
Chakrabarti and Fombonne, 2001	All cases referred to Child Development Center, with multiple or severe developmental impairments.	Health visitors, speech and language therapists, GPs, pediatricians	Child Parent Professional	ADI-R, 2 wks multidisciplinary assessment, Merrill-Palmer, WPPSI	ICD-10 DSM-IV

Abbreviations: ABC, Autism Behavior Checklist; ADI-R, Autism-Diagnostic Interview-Revised; CARS, Child Autism Rating Scale; CHAT, Checklist for Autism in Toddlers; DSM-III-R, Diagnostic and Statistical Manual, 3rd edition revised; DSM-IV, Diagnostic and Statistical Manual, 4th revision; ICD-10, International Classification of Diseases, 10th revision; GP, general practioner; PDD, pervasive developmental disorder; WISC, Wechsler Intelligence Scale for Children; WPPSI, Wechsler Preschool and Primary Scales of Intelligence.

borough [8]. In this study, 1519 children aged 7–16 years were included in the screening stage. Screening involved lecturing and dissemination of brochures to teachers and other school personnel, followed by the completion of a 27-item screening questionnaire by teachers on each individual pupil. Questionnaires were subsequently reviewed for patterns of scores and 18 subjects were selected for further investigation. The second phase of investigation involved a mixture of direct assessments of the child, parental interviews, direct observations, and teacher interviews. Fourteen subjects participated to this stage, and of these, four definite cases of AD according to ICD-10 were identified, yielding a prevalence rate of 28.5 per 10,000 (95% CI: 0.6–56.5/10,000 calculated by the authors). Different sets of diagnostic criteria were applied, with the number of subjects meeting criteria for AD ranging from 3–7, leading to huge variations in prevalence estimates. This study was meritorious as it was the first systematic epidemiologic inquiry on AD. Several methodologic weaknesses should be noted in its design and conduct, however: (1) the target population was small and, as illustrated by the wide confidence intervals for prevalence estimates, not much confidence can be attached to the various estimates provided by the authors, (2) no rationale was given for studying the prevalence of AD in this particular borough and details were generally lacking about school selection and sampling procedures, (3) limited evidence was produced only on the psychometric properties of the screening questionnaires, and the procedures followed to select subjects for further assessments were not entirely clear, (4) reliance on teachers' reports as sole informants for screening was not adequately justified; in fact, parental contribution to the final diagnosis was much more significant than teacher reports and interviews, suggesting that parents should be included as primary informants at the initial screening stage, and (5) prevalence estimates varied enormously according to specific diagnostic criteria, raising questions about the overall validity of the case determination; in particular, it was apparent that some cases meeting a set of diagnostic criteria for AD also could be classified as autism cases according to another set of diagnostic criteria.

Surveys of Asperger disorder and other pervasive developmental disorders

The ambiguous boundaries that persist between AD and other autism spectrum disorder subtypes call for epidemiologic surveys that focus on the prevalence of all subtypes of PDD within the same sample of children.

Six epidemiologic surveys [9–14] have *simultaneously* assessed the presence of autistic disorder and Asperger disorder in epidemiologic samples. The study characteristics are summarized in Table 1. These studies were all conducted in Europe, in countries that have a robust tradition of performing population-based surveys of child health. In five of six surveys, reasonable sample sizes for the target population (> 15,000) were achieved. In one study [11], the sample size was extremely small and included only 826 children. Surveys were usually designed in two phases, one of screening and one of diagnostic confirmation.

The instruments used to screen and to confirm the diagnoses were not specifically devised to assess AD. Rather, they were tools used primarily to assess clinical features of classic autism. Even though several studies relied on standardized measures such as the Autism Diagnostic Interview, these measures generally do not provide specific algorithms or cut-offs for the diagnosis of AD.

The prevalence rates for AD range from 0.3–48.4 per 10,000, a huge variation that reflects methodologic differences across studies. The highest estimate derived from the study with the smallest sample size [11], in which 4 out of 826 7-year-old children met criteria for AD, leading to a prevalence estimate of 48.4 per 10,000 (95% CI: 1.1–95.8/10,000). As in the Swedish study [8], the point prevalence estimate is high but the wide confidence interval indicates the extreme lack of precision of this study.

In these six studies, it was therefore possible to compare directly *within surveys* the prevalence of AD relative to that of autistic disorder. In all studies, the rate of AD was consistently lower than that of autistic disorder as indicated by the prevalence rate ratio of autism to AD that is uniformly greater than 1 (Table 2, right-hand column). How much lower is more difficult to establish. When the authors pooled together the data from these six surveys, however, the number of children with autism was on average five times higher than that for AD. Thus, with a conservative prevalence estimate of 10 per 10,000 children with autistic disorder [7], these figures suggest that the prevalence of AD might be in the neighborhood of 2 per 10,000.

Few surveys of AD have been performed to date and prevalence estimates vary enormously across surveys, most probably reflecting methodologic differences between studies. At this stage, it is therefore difficult to provide a robust estimate for the prevalence of AD. Comparing rates of autism and of AD, it nevertheless seemed that the prevalence of AD was consistently lower than that for autism, and based on conservative calculations, a working rate of 2/10,000 can be taken as a starting point. This figure was arrived at indirectly and is bound to change in the future.

Table 2
Asperger syndrome (AS) in recent autism surveys: prevalence rates

	Autism		Asperger syndrome		Autism / AS
	Prevalence/10.000	N	Prevalence/10.000	N	Ratio
Sponheim and Skjeldal, 1998	4.9	32	.3	2	16.0
Taylor et al, 1999	8.7	427	1.4	71	6.0
Kadesjö et al, 1999	72.6	6	48.4	4	1.5
Powell et al, 2000	—	54	—	16	3.4
Baird et al, 2000	27.7	45	3.1	5	9.0
Chakrabarti and Fombonne, 2001	16.8	26	8.4	13	2.0
Overall		590		111	5.3

It is hoped that estimates will become more accurate as new epidemiologic research accumulates in the years ahead. To obtain more valid estimates, it will be important to focus on rates applying to slightly older age groups (ie, children aged 8–12 years) because children with AD are identified and diagnosed much later than children with typical autism and, as a consequence, estimates obtained in younger samples might underestimate the prevalence of Asperger disorder. Second, investigators should study large enough populations to obtain sufficient precision in their epidemiologic estimates. Third, case finding and case identification procedures need to be improved and instruments that target specifically the developmental and symptomatic features characteristic of AD should be incorporated in the design of future surveys. Obviously, general progresses in assessment and diagnostic procedures are first required to improve the quality of epidemiologic enquiries. Finally, the progressive broadening of the concept of autism spectrum disorder and a tendency by some investigators to overuse this concept call for a sharpening of the diagnostic definition, particularly vis-à-vis manifestations of the lesser variant of autism (broader phenotype) believed to index genetic susceptibility to autism spectrum disorders and of normal variability. To achieve this, the systematic inclusion of measures of impairment, independent of the measure of the disorder itself, is needed for clinical and epidemiologic studies.

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