Problem Behavior of Individuals with Down Syndrome in a Nationwide Cohort Assessed in Late Adolescence

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Objective To assess problem behavior in adolescents with Down syndrome and examine the association with sex and severity of intellectual disability.

Study design Cross-sectional data of a Dutch nationwide cohort of Down syndrome children aged 16-19 years were collected using a written parental questionnaire. Problem behavior was measured using the Child Behavior Checklist and compared with normative data. The degree of intellectual disability was determined using the Dutch Social competence rating scale.

Results The response rate was 62.8% (322/513), and the mean age 18.3 years (SD ± 0.8). The total score for problem behavior was higher in adolescents with Down syndrome than in adolescents without Down syndrome (26.8 vs 16.5; $P < .001$). Overall, 51% of adolescents with Down syndrome had problem scores in the clinical or borderline range on 1 or more Child Behavior Checklist subscales; this is more than twice as high as adolescents without Down syndrome. Adolescents with Down syndrome had more internalizing problems than their counterparts without Down syndrome (14% and 9%, respectively, in the clinical range); the percentages for externalizing problems were almost equal (7% and 9%, respectively, in the clinical range). The highest problem scores in adolescents with Down syndrome were observed on the social problems and thought problems subscales (large to very large standardized differences). Male sex and/or more severe mental disabilities were associated with more behavioral problems.

Conclusions Serious problem behavior is more prevalent in adolescents with Down syndrome. This demonstrates the need for a focus on general behavior improvement and on the detection and treatment of specific psychopathology in individuals with Down syndrome. *(J Pediatr 2013;163:1396-1401)*.

Down syndrome (aka, trisomy 21) is the most prevalent cause of intellectual impairment. The prevalence of Down syndrome is estimated to be 12 per 10 000 live births in the US and 14.6 per 10 000 live births in The Netherlands (approximately 245 children with Down syndrome are live born annually).[1,2] Children with Down syndrome have delayed cognitive and motor development as well as specific medical problems such as congenital heart defects, gastrointestinal disorders, thyroid dysfunction, and visual impairment.[3,4] Moreover, it is known that children with Down syndrome are prone to psychopathology; prevalence estimates range from 18%-38%.[5-7] This risk is lower than in other forms of intellectual disability.[8]

Problem behavior in children varies with age, becoming particularly prevalent during adolescence because this period is characterized by hormonal, physical, psychological, and social changes.[9] Adolescents with Down syndrome also have to cope with puberty, sexual development, the start of emotional separation from parents, and development of social autonomy.[4,10] Some studies have confirmed that there are also changes in behavioral patterns during adolescence in children with Down syndrome: externalizing symptoms decrease and internalizing symptoms increase.[11,12]

The few studies of behavioral problems in adolescents with Down syndrome are limited, as are studies of the 'dual diagnosis' of intellectual disability and psychopathology, mainly because of the small sample sizes (<60) and broad age ranges (mostly 4-19 years).[6,7,13,14] We were unable to find any large-sample studies describing behavior in Down syndrome in the late teens. Furthermore, sex effects as a factor in behavioral problems have hardly been investigated in studies looking at Down syndrome, and sex differences can be expected because this is a familiar phenomenon in the general population.[11,15] This study examines problem behavior during late adolescence in a large nationwide cohort of individuals with Down syndrome and the association with sex and degree of intellectual disability.

Methods

Data were collected from a nationwide Dutch cohort of parents of children with Down syndrome assessed at the age of 16-19 years. This cohort included children

CBCL	Child Behavior Checklist
SRZ	Dutch social competence rating scale

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with Down syndrome born in 1992, 1993, and 1994. Of all children with Down syndrome born in this 3-year period, an estimated 595 adolescents (based on an 81% survival rate) were still living in The Netherlands.[2,16] The Dutch Down Syndrome Foundation (an organization for parents) had contact with 86% of these parents and sent them a written request. The only selection criterion for inviting parents to participate was the year of birth of the Down syndrome child. Parents could respond within 4 months after receiving the invitation. Reminders were sent after 4 and 8 weeks.

Parents completed a written questionnaire consisting of 2 validated tests and additional questions about background and level of functioning. Written informed consent was obtained from parents/next of kin of all participants. Permission of an ethical committee was not obtained, because it is not required in the Netherlands for this type of study or anonymous data collection.

The Dutch version of the Child Behavior Checklist (CBCL) for children aged 4-18 years was used to measure problem behavior.[17,18] Although the CBCL was developed for children with normal intelligence, there have been frequent reports stating that it is suitable for children with a developmental delay.[19,20] Normative data are available in the test manual for the age group of adolescents aged 12-18 years.[17] In addition, normative data about the mean scale scores for the CBCL were available for adolescents in the 15-18 age range in the general Dutch population as published by Bongers.[15] Both sets of normative data were based on parental reporting. Bongers' sample covers almost the same age range as our sample, and so these data were used to compare mean scale scores.

The CBCL contains 113 problem behavior items rated from 0 (not true) to 2 (very true or often true). A total problem score can be calculated using these items. The items of the CBCL can also be grouped into the following 8 subscales: withdrawn, somatic complaints, anxious/depressed, social problems, thought problems, attention problems, delinquent behavior, and aggressive behavior. A scale of internalizing problems is constructed by combining the subscales withdrawn, somatic complaints, and anxious/depressed. The externalizing problem scale is formed by combining the subscales delinquent behavior and aggressive behavior. Moreover, all scale scores can be grouped into scores in the normal, borderline, or clinical ranges for the scale.

The Dutch social competence rating scale (SRZ) was used to determine the degree of intellectual disability. This validated instrument specifically measures social independence in mentally disabled children aged 4-18 years and has already been established as a sensitive instrument for measuring changes in self-help skills in adults with Down syndrome.[21,22] The SRZ was selected for its ability to measure IQ scores in children with an intellectual disability, whereas some other intelligence tests—such as the Wechsler Intelligence Scale for Children[23]—are not designed to measure such low IQ scores. The SRZ contains 31 items that measure skills needed for independent functioning in daily life. On the basis of these items, the degree of intellectual disability (mild, moderate, severe, or profound) can be determined as a reflection of the following self-help skills: (1) profound intellectual disability (ie, hardly able to dress oneself, wash hands and face properly, use adequate toilet hygiene, likely able to eat independently [without the use of a knife], barely able to speak [IQ <20]); (2) severe intellectual disability (ie, able to undress, wash hands and face, use a knife and fork at dinner, clear up after dinner, speak using incomplete sentences with unclear pronunciation that can be understood only by close caregivers or familiar people [IQ 20-34]); (3) moderate intellectual disability (ie, dresses completely, washes hands and face properly, uses adequate toilet hygiene, uses a knife and fork at dinner including cutting meat [without a bone], is able to walk outside the home without supervision, and that speech can mostly be understood by others [IQ 35-49]); and (4) mild intellectual disability (ie, able to dress oneself completely, including footwear, maintain complete personal hygiene, set the table properly, walk several streets away from the home without supervision, use full or more compound sentences when speaking, and that speech and language can be understood by most others [IQ 50-69]).

Statistical Analyses

The general characteristics of the study population were determined and compared for boys and girls with Down syndrome, using *t* tests for continuous variables and χ^2 tests for categorical variables.

Mean raw CBCL scale scores for boys and girls with Down syndrome were compared with normative data from adolescents aged 15-18 in the general Dutch population as published by Bongers.[15] To evaluate the differences between mean values, *t* tests were used and the standardized differences were estimated by dividing the differences in mean scores between the subgroups by the pooled SD. Cohen's standardized differences (d) were used for the interpretation of relevant differences: $d < 0.2$ is considered to be a negligible difference, $0.2 \leq d < 0.5$ a small difference, $0.5 \leq d < 0.8$ a moderate difference, $0.8 \leq d < 1.3$ a large difference, and $d \geq 1.3$ a very large difference.[24]

Linear regression analysis was performed to assess the association between intellectual disability and the total CBCL problem score, adjusting for parental education and sex. In addition, to determine whether the effect of sex on the outcome variable was the same for all degrees of intellectual disability, the influence of interaction terms was assessed using linear regression analysis. For this purpose, cross products were computed between degree of intellectual disability and sex. These cross products were added as an extra step in the regression equation (which included all main effects).

In all analyses, the statistical tests were 2-tailed and statistical significance was defined as $P < .05$. The analyses were performed using Statistical Package for the Social Sciences, v 20.0 for Windows (SPSS Inc, Chicago, Illinois).