The Psychoeducational Profile of Boys with Klinefelter Syndrome

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Abstract

Klinefelter syndrome (KS) affects about 1 in 900 males due to an extra X chromosome. Although there are no obvious physical features associated with childhood KS, many boys demonstrate a cognitive deficit in verbal processing. The first section of this article integrates the extant literature on intelligence and achievement outcomes in boys with KS. The second section presents our findings from a 20-year study involving one of the largest unselected cohorts of boys with KS. We followed 36 boys with KS and 33 sibling controls from 6 until 20 years of age. Boys with KS are shown to demonstrate a verbal cognitive deficit and significant underachievement in reading and spelling, as well as in arithmetic. These problems, which are evident from early school years, increase with age such that by late adolescence, boys with KS are four to five grade levels behind. In addition, we also found that they were most likely to have a generalized type of learning disability, with very few boys indicating a pure reading or pure arithmetic problem. They also showed deficits in written language skills and acquisition of knowledge-based subject material were also problematic. Despite significant underachievment and frequent grade failure, many boys with KS had completed high school, and a few were also pursuing postsecondary educations. The discussion section examines how their language-based disability affects comprehension and learning, leading to underachievement.

Most individuals have two sex chromosomes: a pair of X chromosomes in the female, and an X and a Y chromosome in the male. However, a small proportion of the population has an abnormal complement of sex chromosomes. This includes persons with a missing X chromosome, as in Turner syndrome; an extra X chromosome, as in Klinefelter or trisomy X syndromes; and an extra Y chromosome. In addition, there are individuals with mosaic constitutions (a combination of normal and abnormal cell lines) or with variations of more than three sex chromosomes. Of all the sex-chromosome anomalies (SCA), Klinefelter syndrome (KS) is the most prevalent, with an estimated frequency of about 1 in 900 males.

An association between cognitive functioning and sex-chromosome abnormalities has long been recognized (Jones, 1988). In the fragile X syndrome, for example, mild mental retardation is often observed (Bregman, Dykens, Watson, Ort, & Leckman, 1987; Hagerman, Kemper, & Hudson, 1985); in conditions involving structural abnormalities or selective mutations of the X chromosome, as in Aarskog syndrome (Fryns & van den Berghe, 1989), mare severe retardation may occur. By contrast, conditions involving abnormal sex chromosome complements do not place the person at risk for mental retardation but, rather, for selective cognitive impairments. For example, spatial and math disabilities are frequently observed in Turner syndrome (Rovet, 1990, 1993), whereas poorer language skills and diminished reading and spelling capacity are observed in children with an extra X chromosome (Berch & Bender, 1990). An extra Y chromosome is typically associated with behavior problems but a lesser degree of cognitive dysfunction. Mosaicism reduces the risk of impairment, whereas a supernumerary complement beyond three increases the risk of mental retardation.

In Klinefelter syndrome, there are no obvious physical features, although affected boys tend to be taller than average and have smaller head circumferences and genitals (Robinson, Lubs, Bergsma, 1979; Stewart et al., 1982). Hormone abnormalities observed during early puberty may be associated with gynaecomastia (breast development) and sterility (Ratcliffe & Paul, 1986). Although there is no apparent facial dysmorphology, two recent studies have shown that craniofacial structure may be altered slightly (Ingerslev & Kreiborg, 1978; Kosowiu & Rzymski, 1975), but not to a degree that would be noticeable to most observers. No study has as yet identified whether there are physical markers of poorer school achievement.

because boys with KS are not physically distinguishable from other male children, behavioral information about them is often limited to selected cases that were referred to physicians and mental health specialists. The findings on these cases suggest significant academic underachievement and behavior problems. Recently, two alternate sources of information have provided

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reliable, and optimistic, findings: (a) studies begun in the late 1960s to estimate the incidence of SCAs in the general population and follow the children prospectively and (b) a recent follow-up study of children identified in utero, whose parents maintained the pregnancy (Robinson, Bender, &Linden, 1992). The findings from the latter study showed better outcome in the children ascertained prenatally. Because this may reflect the higher socioeconomic levels and greater motivation among parents seekingprenatal screening and choosing to continue these pregnancies (Robinson et al., 1992), and because newborn screening for SCA is no longer allowed in most jurisdictions, the few cohorts followed longitudinally are probably the most accurate source of information on these children.

Because Klinefelter syndrome is relatively prevalent, a child with this disorder will likely be encountered in the classroom, although usually unbeknownst to the teacher (and, often, the parents alike). The purpose of this article is to describe the achievement characteristics of boys with KS. This will be accomplished through an integrative review of the extant literature on psychoeducational outcome. We will also present our findings collected over a period of 20 years from a large cohort ascertained by neonatal screening. The objectives are to describe (a) the intellectual profiles of boys with KS, (b) their achievement characteristics, and (c) the prevalence and type(s) of learning disabilities.

Overview of Previous Research

To integrate the findings on school achievement in boys with KS, we culled all the findings in the literature from 1970 to the present. These are presented in Table 1. Note that the studies often combined groups with different SCAs, rendering the unique characteristics of the KS phenotype uninterpretable. Table 1 lists the results from 27 studies, with a study being defined as one or more publications on the sample from the same time period. It should be noted that these studies represent the findings from just 10 samples of boys in different stages of development. Three cohorts involve (a) case report findings (Nielsen et al., 1970), (b) a clinic survey (Funderburk & Ferjo, 1978), and (c) subjects with prenatal screening diagnoses (Robinson et al., 1992). The remaining seven cohorts are based on samples that were identified through neonatal screening and followed from early childhood to late adolescence. Controls, usually siblings, were available for most comparisons. Sample sizes for the total SCA group ranged from 6 in the Connecticut study (Leonard, 1991) to over 50 subjects in the Denver (Robinson, Puck, Pennington, Borelli, & Hudson, 1979), Toronto (Stewart et al., 1982), and Edinburgh (Ratcliffe, Tierney, et al., 1982) samples. Descriptions are often based on chromosomally heterogeneous groups.

Although the broad spectrum of tests varled among the different studies, it can be seen that there was considerable consistency as to choice of standardized intelligence test. This usually involved one or more tests from the Wechsler series. Achievement information was somewhat more variably obtained and involved parent or school reports or tests such as the Wide Range Achievement Test (WRAT or WRAT-R; e.g., Evans, de von Flindt, Greenberg, & Hammerton, 1991; Stewart, Bailey, Netley, & Park, 1991), the Peabody Individual Achievement Test (e.g., Pennington, Bender, Puck, Salbenblatt, Robinson, 1982), the Woodcock Johnson Psycho-Educational Battery (e.g., Robinson et al., 1986; Stewart, Bailey, Netley, Rovet, & Park, 1986), and the British Ability Scales (e.g., Ratcliffe, Jenkins, & Teague, 1990). In addition, the study by Graham, Bashir, Stark, Silbert, and Walzer (1988) also involved a concerted effort to describe in great detail the specific language disability of boys with KS.

The two studies involving clinic referred and clinic-screened cases reported significant school underachievement in a high percentage (82%) of children (Funderburk & Ferjo, 1978; Nielson et al., 1970). Also described were difficulties with social relations and speech and language development, although intelligence was reported as normal. The *one* study of boys with KS identified prenatally (Robinson et al., 1992) reported very good outcome, with no evidence of either language or school-related problems.

The findings on boys with KS identified via newborn chromosome screening indicated that they evidenced moderate to severe reading, spelling, and writing problems as early as age 7 or 8; however, their arithmetic skills were unaffected (Nielsen Sørensen, & Sorensen, 1982; Stewart et al., 1979; Walzer, Graham, Bashir & Silbert, 1982). By about age 10, boys with KS were performing about one grade below their expected grade level, but they showed no specific area of deficit (Evans, de von Flindt, Greenberg, Ramsay, & Hamerton, 1982). After age 10, difficulties in arithmetic, in addition to reading and spelling, were noted (Leonard, Sparrow & Schowalter, 1982; Leonard & Sparrow, 1986; Stewart et al., 1986; Stewart et al., 1982), with between 60% and 86% of boys with KS receiving special education (Robinson et al., 1986; Stewart et al., 1986; Stewart et al.,1982; Walzer et al., 1982). The majority also repeated at least one grade (Leonard & Sparrow, 1986). An increased incidence of learning diiabiities was additionally reported by Robinson, Bender, Linden, and Salbenblatt (1991) for all nonmosaic males with an extra X chromosome.

The findings on boys with KS in late adolescence and early adulthood were described in the proceedings of a recent conference (Evans, Hamerton, & Robinson, 1991). Robinson et al. (1991) reported that although most of the boys with KS in their study required help for learning difficulties, particu-

TABLE 1
Intelligence and Achievement Characteristics of Boys with Klinefetter Syndrome

Study	Country, city	N	Karyotype	Ascertainment	Age	Controls	Measures	Results	Conclusions
Nielsen et al. (1970)	Denmark, Aarhus	11	47,xXY 46,XY/47,XXY	CR	8.0–15.0 10.0–25.0	Yes	WISC WAIS School reports	Referred to psychiatric service due to problems at school; difficulty relating to other children. School problems in 82%.	Early diagnosis of SCA important so appropriate school placement, special teaching can be arranged.
Walzer et al. (1977)	USA. *Boston*	13	47,XXY	NS	5.0–7.0	Yes	Bayley WPPSI	All XXY past Grade 1 given reading evaluations; 3/5 had moderate–severe reading, spelling, writing problems and learning disabilities.	Developmental language deficits seen as expressive language problems *(temporal* sequencing, comprehension).
Funderburk & Ferjo (1978)	Sweden	11	47,XXY	CS	6.8–24.0	No	WISC WAIS	High frequency of speech and language problems but normal intelligence. School underachievement in 9 (82%).	Appropriate educational intervention may diminish some long-term intellectual and psychiatric problems in some XXY males.
Leonard et al. (1979)	USA. New Haven	11	47,xXY 47,xYY 47,XXX 45,x	NS	8.0–9.5	Yes	School records	Grades repeated by 55% of SCA children, 0% by controls. Delayed language development, deficits in vocab, articulation.	Mild learning disabilities decreased achievement not attributed solely to SCA.
Robinson et al. (1979)	USA, Denver	51	47,XXY 47,xYY 47,XXX	NS	5.0-l 1 .o	Yes	School	School problems in 50% XXY versus 3% controls.	Possible increased risk for learning problems based on karyotype.
Stewart et al. (1979)	Canada. Toronto	47	47,xXY 47,XYY 47,XXX 46,XX male X/XX	NS	0.0–0.8	Yes	McCarthy WISC-R WRAT Rutter Parent Vineland	XXY deficits in reading and spelling, not arithmetic.	XXY similar but less severe memory deficit in XXX.
Pennington et al. (1982)	USA, Denver	44	47,XXY 47,XXX 45,x Mosaics	NS	9.3-12.5	Yes	WPPSI PIAT School reports	School intervention in 69% pure SCA children, 26% controls. LDs in 60% SCA subjects. 26% in mosaics and controls. Specific reading LDs in 27% 47,XXY.	Depressed VIQ, reading and spelling delays in 47,XXY.
Ratcliffe, Bancroft, Axworthy, & McLaren (1962)	UK, Edinburgh	12	47,xXY	NS	16.0–18.0	Yes	WISC-R WAIS BSRI HSPQ	No difference between XXY and contmls until high school. Eight (6%) had learning, behavior problems compared to 2 (17%) controls. Five (63%) did not pass school at ordinary grade, 6 (6%) contmls passed at ordinary grade.	XXY boys at higher risk for problems with speech developments, school performance, social adjustment.

(table continues)

TABLE 1. Continued

Study	Country, city	*N*	Karyotype	Ascertain-ment	Age	Controls	Measures	Results	Conclusions
Ratcliffe, Tierney, et al. (1982)	UK. Edinburgh	67	47,XXY 47,XXX 47,XYY	NS	2.0-I 3.5	Yes	WISC-R Stanford-Binet Gesell Reynell	62% XXY required remedii education versus 45% XXY, 6.7% male controls. Reading difficulty predominant in XXY, lower VIQ than controls.	Higher incidence of language difficulties in 3 SCA groups. XXX girls have greater cognitive deficits than XXY, XYY boys.
Nielsen et al. (1982)	Denmark. Aarhus	25	47,XYY 47,XXX 45x	NS	7.9-I 1.9	Yes	WISC Bender Fiiure Drawing Lateral Dom Rod 8 Frame GFW And Dis	40% SCA group, 7% controls had poor school achievement. None with poor math skills.	Extra X or Y children need special stimulation. support and teaching efforts in learning to read and write.
Evans et al. (1962. 1966)	Canada, Winnipeg	13	47,XXY 47,XYY	NS	7.5–10.1	Yes	PIC Vineland WISC-R PPVT McCarthy WRAT	All XXYs one grade lower than expected for age but no specific area of de&it.	XXY and XXX at greater risk than XYY for teaming and school-related problems.
Stewart et al. (1982) Stewart et al. (1986)	Canada, Toronto	51-53	47,XXY 47,XXX 47,XYY Mosaics 46,XX Male	NS	9.0-I 2.0 12.5–16.5	Yes	WISC-R WRAT WJPB	XXY significantly lower reading, spell. ing, arithmetic shills than control. 59% XXY, 16% controls in special education.	Testostemne treatment did not produce any change In behavior *or* school performance in XXY. XXY subjects resemble other learning disabled children. Extra X children have limited short-term memory spans not accounted for by language impairment.
Waker et al. (1982, 1986)	USA. Boston	13	47,XXY	NS	5.0–7.0	Yes	WRAT Bayley WPPSI WISC-R	89%–92% required remedial help in reading, spelling. 77% XXY, 11% controls have early LD and in special ed. 66% XXY with LD had delays in speech and language developmem and significant VIQ–PIQ discrepancy.	IDeficits in verbal ability associated with decreased achievement in reading and spelling tasks. Deficits in auditory memory and pmcessrng implicatea in verbal and language impaimtents.
Leonard, Sparrow, a Schowalter, (1982); Leonard a Sparrow (1986)	USA, New Haven	11	47,XXY 47,XYY 47,XXX	NS	9.0-I 3.0 17.0-I 6.0	Yes	WISC-R VMI PPVT WRAT	All have some learning difficulties; 73% hive repeated a grade but 91% in regular classes. 64% had reading pmblems, 36% problems with math. Low-average, borderline intelligence levels.	Speech, language, and reading areas of most difficulty but no mental retardation due to SCA alone.
Robinson et al. (1986)	USA, Denver	48	46,XYY 47,xxx 45.x Mosaics	NS	9.9-I 9.0	Yes	VMI GFW Aud Disc Receptive Lang Boston Naming	XXY lower reading and written language, WJ scores than controls. LDs requiring special education in 86% XXY, 79% for reading.	XXY slow cognitive pmcessing, impaired language, verbal memory deficits. XXY deficits due to decreased processing speed. not lack of ability.

(table continues)

TABLE 1. Continued

Study	Country, city	N	Karyotype	Ascertainment	Age	Controls	Measures	Results	Conclusions
Ratcliffe, et al. (1990, 1991)	UK. Edinburgh	67	47,XXY 47,XYY 47,XXX XX male	NS	8.0-l 5.0	Yes	WISC-R Burt Reading Bristol Soc Adj BSQ British Abilii Scale	67% XXY received special help. less severe cognitive deficits than XXX girls.	XXY increases risk of LD.
Bender et al. (1987)	USA, Denver	46	47,XXY 47,xYY 47,XXX 45.x Mosaics	NS	10.5–15.5	Yes	Lang Test Battery, BOTMP, Family Dysfunctn, School Reports	language, school impairments more frequent in pure SCA karyotypes (21%–66%) than controls (0%–32%). More academic difficulties in pure SCA children from dysfunctional families.	Higher vulnerability in environmental stress in SCAs than controls.
Graham et al. (1988)	USA, Boston	14	47,XXY	NS	5.0-l 2.0	Yes	WISER, Token Test, TDA. ITPA, Syntactic Comp. DLA. PPVT, Boston Naming, Repetition, GFW Aud Disc., Sentence Memory, Gilmore. GM (speed and accur), Word Reading, Schonell Spelling	Lower verbal IQ and decreased achievement in reading and spelling due to pre-existing language disabilities. Problems in expressive language. audiiry processing, and memory abilities correlated with reduced achievement.	Specific deficits in expressive language implicates left hemisphere dysfunction.
Bender, Linden, & Robinson (1989)	USA, Denver	32	47,XXY 47,xxx	NS	8.0–18.0	Yes	WISC-R Vocab Thurstone WF PMA SR WJ SR	47,XXY deficient in rapid retneval of verbal information.	Additional time to learn complete work required by 47,XXY.
Robinson (1990)	USA, Denver	46	47,XXY 47,XYY 47,XXX 45,x Mosaics	NS	12.5–20.0	Yes	WISC Language Test BOTMP School Reports	LDs in 33 (87%) of nonmosaic SCA children. More LDs in males (90%) than females (75%) but LDs in nonmosaic females more global and severe. All three disorders (motor, lang, learning) in 53% SCAs, 0% in controls and mosaics.	SCA affects development of CNS as motor and cognitive abilities affected in nonmosaic SCA children.
Walzer et al. (1991)	USA. Boston	24	47,XXY 47,XYY	NS	9.0-l 2.0	Yes	WISC-R Direct Observn	Reading and spelling difficulties in 65% 47,XXY, 36% with LDs in full-time special ed. Reading LDs in 11% controls.	Severe reading and writing problems due to language deficits in both groups. XXY more impaired in expressive than receptive language.

(table continues)

TABLE 1. Continued

Study	Country, city	*N*	Karyotype	Ascertain- ment	Age	contmla	Measures	Results	Conclusions
Robinson et al. (1991)	USA, Denver	41	47,XXY 47,XYY 47,XXX 45,x Mosaics	NS	14.0–24.0	Yes	BOTMP Self-esteem BDI, BSRI PMA WF & SR WJ-SR, PPVT WAIS-R	86% XXY required help for learning difficulties, 79% for reading LDs. One XXY in university, 64% still in high school, one dropout	Deficits based on karyotype and related types of LDs, 47,XXY has generally depressed language skills, 45X deficits in spatial skills. and 47,XXX have globally reduced abilii.
Leonard (1991)	USA, New Haven	6	47,xXY 47,XXX 47,xw	NS	20.0–21.0	No	Question- naires, interviews	All had some learning problems with speech and language delays, all required remedial, had to work hard to succeed. Four now employed in meaning- ful jobs and one in technical school.	After high school. subjects felt learning problems much less significant. More satis- factory development in children diagnosed prenatally rather than through screening.
Stewart et al. (1991)	Canada, Toronto	38	47,XXY 47,XXX 46,XY/47,XXY 46,XX male	NS	18.0–21.0	Yes	WISC-R, WAIS-R, WRAT-R, Educ histories	XXY males below controls in reading, spelling, arithmetic. 61% XXY. 17% con- trols had special ed in past 5 years. 1 of3 mosaics mildly below average in intellec- tual ability and edu- cational performance.	Ability to cope suc- cessfully in school has potentially long-term implications for social adjustment in 47,XXY males.
Evans et al. (1991)	Canada, Winnipeg	10	47,XXY 47,XYY 47,XXX Mosaics	NS	15.8-18.9	No	WISC-R WAIS-R WRAT	44% SCA group had repeated a grade. Verbal defi- cits and verbal performance dis- crepancies frequemly observed.	Best functioning children from stable and supportive families.
Nielsen (1991); Nielsen & Wohlert (1991)	Denmark, Aarhus	25	47,XXY 47,XXX 47,XYY	NS	10.0-14.0 15.0–19.0	Yes	Parent interview, school reports	All SCA children in normal school; no mental retardation, 63% below-average school performance. Remedii teaching in both math (42%) and reading (35%). Children average or above-average at school if good child- hood conditions. Poor school perfor- mance and behavior problems if had poor childhood quality.	Prevention of deviation from normal in school performance, behavior, and adjustment if appropriate social. educational resources available and if parents willing to accept coun- seling and child's SCA.

(table continues)

TABLE 1. Continued

Study	Country, city	N	Karyotype	Ascertainment	Age	Controls	Measures	Results	Conclusions
Robinson et al. (1992)	USA, Denver	29	47,XXY 47,XYY 47,XXX Mosaics	PS NS	7.0–14.0	Yes	Questionnaires, school reports, WISC-R, physician report	No language deficits or school problems in prenatal XXY boys. Learning problems in 11 (79%) of prenatally diagnosed XXYs. Fewer language, learning deficits in prenatal XXY boys. all postnatal in special education.	Confirms impressions that prenatally diagnosed SCA cohort may develop differently than postnatally screened children. Effect may be due to more supportive environment and higher SES in prenatal families.

Note. CR = case report: NS = newborn screening; CS = clinic screening; PS = prenatal screening; BDI = Beck Depression Inventory; BOTMR = Bruinincks-Oseretsky Test of Motor Proficiency: BSO = British Screening Questionnaire; BSRI = Bem Sex Role Inventory; DLA = Detroit Learning Attitudes; GFW Aud Disc = Goldman-Fristoe-Woodcock Auditory Skills Test Battery: GM = Gates-McG initie Speed and Accuracy subtest; HBPO = High School Personality Questionnaire: ITPA = Illinois Test of Psycholinguistic Abilii PIAT = Peabody Individual Achievement Test: PIC = Personality Inventory for Children; PMA SR = Primary Mental Abilities Spatial Relations Test: PMA WF = Primary Mental Abilities Word Fluency; PPVT = Peabody Picture Vocabulary Test; TDA = Templin Dawley Articulation: Thurstone WF = Thurstone Word Fluency; VMI = Developmental Test of Visual Motor Integration; WAIS · Wechsler Adult Intelligence Scale: WISC = Wechsler Intelligence Scale for Children: WJPB = Woodcock-Johnson Psycho-Educational Battery; WJSR · Woodcock Johnson Spatial Relations; WPPSI = Wechsler Preschool and Primary Scale of Intelligence: WRAT = Wiie Range, Achievement Test: YDE = Yale Developmental Exam.

larly reading disabilities, 1 boy was in college, 64% were still in high school, and only 1 had dropped out of school early. Leonard (1991), who reported on 6 children with SCAs from the New Haven area, indicated that despite early learning, speech, and language problems and the need for remedii help, all of the boys had completed high school (but had to work hard to succeed), 4 of 6 were employed in meaningful jobs, and 1 was in technical school. Lower levels of achievement and a greater need for special education compared to sibling controls were observed for the children in the Toronto cohort (Stewart et al., 1991), frequent grade repetition was found in the Winnipeg cohort (Evans, de von Flindt, et al., 1991), and below-average school performance and increased need for remediation in math and reading were observed in the Denmark cohort (Nielsen & Wohlert, 1991).

These findings indicate that, despite early problems, many boys with KS do succeed at school to the point of graduating high school. However, levels of achievement are clearly below par, and there is an increased need for remediation and special education. Although grade retention is a frequent and common solution for underachievement, a number of boys do complete high school, with a few even acquiring postsecondary educations. Although these results are more optimistic than those based on children refererred to psychiatric services for school problems (Nielsen et al., 1970), they do indicate an increased risk for learning problems and greater need for remedial services. However, a more detailed and cohesive description of their specific psychoeducational characteristics is clearly needed. The next section presents our findings on intelligence and school achievement, which are additionally examined for changes over time and the incidence of specific learning disabilities.

The Toronto Cohort

Between 1967 and 1971, a cytogenetic survey was conducted in the Toronto area of the amniotic membranes from the placentae of over 72,000 consecutive newborns (Bell & Corey, 1974). The purpose of that study was to establish the incidence of X chromosome abnormalities in the general population of a circumscribed geographic region. This resulted in the identification of 78 children with an atypical sex chromosome complement, 41 of whom were boys with a 47,XXY karyotype, or KS. Upon identification of an SCA, the physician responsible for the child was notified and, at his or her discretion, parents were or were not informed. In the majority of cases in which parents were told of the finding, they agreed to partake in a study of their child's development. From the onset, children were studied by a team of developmental and endocrine specialists; after age 6, psychological investigations were included and continued until the study formally ended, in 1988, when the oldest children reached 20 years of age. Details of their early development, described by Haka-Ikse, Stewart, and Cripps (1978), indicated delayed speech and motor development as well as hypoactivity (see also Stewart et al, 1979). The present article deals only with the findings from age 6 on.

Method

Participants

Of the 41 boys with KS identified in the Bell and Corey (1974) project, 30 were selected for follow-up at birth while a further 5 were recruited later when a physical or behavioral problem became evident. The remaining 6 either were not notified by the physician or refused to participate. One boy was the brother of a 47,XXY child, who was identified during genetic investigations of the family. The final sample of 36 boys with KS spanned the full SES range, with the majority (86%) being Caucasian.

From the original 36 boys with KS, 29 (81%) were followed until completion of the study. There was no systematic bias in the subjects' completing versus not completing the study and no appparent reason for dropping out (Rovet, Netley, Bailey, Keenan, & Stewart, 1995). They did not differ in SES or early intellectual characteristics.

Controls consisted of 16 male and 17 female siblings of the entire group of children with an SCA (Rovet et al., 1995), only 24 of whom were siblings of boys with KS. Because siblings were tested less frequently than boys with KS, we chose to include both male and female siblings and siblings of other children with SCAs in order to increase statistical power. Most controls (75%) were older than the proband, given that they were recruited when the aneuploid child was born The median age difference between the boys with KS and the control group was approximately 3 years. Supplementary testing indicated that no sex differences existed among controls or differences between siblings of boys with KS and children with other SCAs.

Tests *and Procedure*

Details of the procedure and follow-up are described in various previous publications on this cohort (Netley, 1990; Netley & Rovet, 1984, 1987; Rovet & Netley, 1983; Stewart et al., 1991; Stewart et al., 1986; Stewart et al., 1982). Most children with an SCA were tested *on an* annual basis, when they were given various psychological tests. These usually included a measure of intelligence and achievement. During middle childhood, achievement testing was conducted somewhat more frequently than intelligence testing. Controls were seen less often than boys with KS; a few controls were seen only once.

The Wechlser Intelligence Scale for Children (Wechsler, 1949) or the Wechlser Intelligence Scale for Children-Revised (Wechsler, 1974) was given to boys below age 16, whereas boys above this age received the Wechsler Adult Intelligence Scale (Wechsler, 1955) or the Wechsler Adult Intelligence Scale-Revised (Wechsler, 1984). Because intelligence testing was sporadic over the 15-year period of the study, results were blocked into specific age intervals (6 to 8, 9 to 11, 12 to 14, 15 to 17, and 18 to 20 years) and one score, typically the oldest in an age interval, was recorded for each child. Determined for the Wechsler tests were the Full Scale IQ (FSIQ), Verbal IQ (VIQ), Performance IQ (PIQ), differences between VIQ and PIQ scores, and individual subtest scores.

Achievement was assessed with the Wide Range Achievement Test (WRAT; Jastak, Bijou, & Jastak, 1978) or, when it became available, the Wide Range Achievement Test-Revised (WRAT-R; Jastak & Wilkinson, 1984). These results were recorded for every age, with an age group including results obtained between -6 and +5 months of each year (i.e., the 6-year test range would include children between 6.5 and 7.5 years). Statistical comparisons were conducted between the KS group and the control group only when there were a minimum of 3 subjects per group at a specific age.

During early adolescence, boys in the KS group were also assessed with the Woodcock Johnson Psycho-Educational Test Battery (WJPTB; Woodcock & Johnson, 1977) and the Test of Written Language (Hammill & Larsen, 1983). These were not provided to controls.

Results

Intelligence

Table 2 shows the intelligence test results of the *boys* with KS and the controls at the five age intervals. For the KS group, mean VIQs ranged from 82 to 88 and for controls, from 93 to 103. *Mean* PIQs ranged from 93 to 101 for KS and from 99 to 111 for controls. The mean VIQ averaged across age intervals was 84.6 for KS and 104.2 for controls; their mean averaged PIQ values were 98.9 and 106.8, respectively. T tests indicated that the groups differed significantly in VIQ at all ages except 15 to 17 years; in contrast, they only differed in PIQ at 6 to 8 years. Although IQs were not observed to vary systematically with age, there does appear to be fall-off in FSIQ for the KS group (after age 15).

Analyses were also conducted to determine whether the groups differed in their difference scores between VIQ and PIQ scales. Boys with KS had consistently higher PIQ than VIQ scores, which differed significantly ($p < .001$) at every age interval. Between 86% (12 to 14 years) and 93% (6 to 11 years) of the boys had a profile involving a significantly (> 1 SD) lower VIQ value. Although controls also scored higher on PIQ than VIQ, the difference between scales was only significant ($p < .05$) at 6 to 8 years. Directly comparing the VIQ-PIQ difference scores of KS and non-KS subjects revealed that the groups differed significantly only at 9 to 11 years. When results were averaged across age intervals, KS had a mean VIQ-PIQ difference score of -14.2, compared to -5.2 for controls.

Regarding individual verbal subtests, boys with KS scored significantly below controls on the Information, Similarities, Arithmetic, Vocabulary, Comprehension, and Digit Span subtests at most, but not all, ages (see Table 2). They did not differ on any nonverbal subtests. Freedom from distractibility (FFD) factor scores were also computed from the mean of the Information, Arithmetic, Digit Span, and Coding subtests. As shown in Figure 1, boys with KS obtained lower FFD scores than controls at all ages, which were only significant ($p < .05$) at 9 to 11 years.

In summary, the present *findings* indicate that boys with KS demonstrate a distinct intellectual impairment in verbal ability, whereas their nonver-

TABLE 2
Intelligence Test Findings in Boys with Klinefelter Syndrome (KS) and Controls (C)

	Ages 6–8		Ages 9–11		Ages 12–14		Ages 15–17		Ages 18–20	
	KS	c	KS	C	KS	C	KS	C	KS	C
	(*n* = 28)	(*n* = 14)	(*n* = 29)	(*n* = 12)	(*n* = 21)	(*n* = 11)	(*n* = 26)	(*n* = 4)	(*n* = 22)	(*n* = 5)
Verbal IQ	87.9	103.3‴	82.1	103.3-	05.9	98.4’	82.7	93.0	83.4	99.0**
Performance IQ	100.6	110.5”	99.2	106.4	99.5	102.3	99.6	102.5	92.6	99.4
Full Scale IQ	93.5	109.5”	69.7	105.3*	91.4	99.0	96.6	96.5	86.6	106.3
Information	62	9.6	72	9.7*	7.2	9.1	6.9	9.0	5.7	92’
Similarities	7.3	112”	6.8	10.2**	6.6	9.6	6.9	8.6	6.5	10.4’
Arithmetic	62	9.7	6.1	10.4	7.6	8.8	7.7	9.0	6.7	9.5’
Vocabulary	8.8	11.9”	6.6	10.7**	7.2	9.6	7.0	8.5	6.1	8.8
Comprehension	8.3	10.6	7.4	11.5”	7.8	10.3’	8.7	9.3	8.1	9.4’
Digit span	7.9	92	7.1	93	8.6	9.1	7.4	10.5	7.3	11.4’
Picture Completion	10.8	10.1	11.2	10.8	8.7	9.1	10.8	10.5	9.1	9.2
Picture Arrangement	9.8	13.0	10.4	11.5	10.6	10.0	10.6	11.3	8.5	9.4
Block Design	102	11.9	9.9	10.7	10.0	9.6	9.7	10.3	9.3	10.2
Object Assembly	10.8	12.4	11.1	112	102	11.1	10.0	0.0	8.9	10.0
Coding	8.7	9.9	7.6	102	8.3	11.1	8.3	10.3	8.1	10.8

*$p < .05$. **$p < .01$. ***$p < .001$.

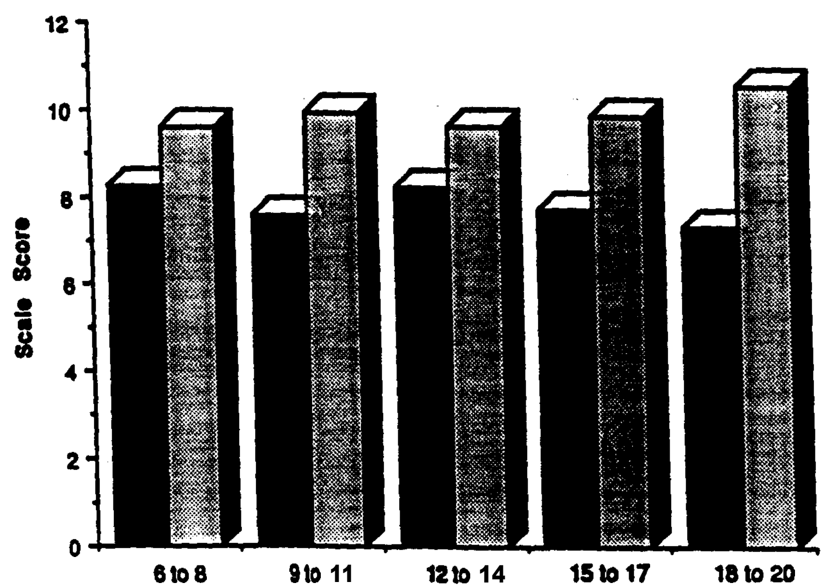

FIGURE 1. Mean freedom from distractibility saxes in KS end control groups. Boys with KS are shown with the solid bars, controls with the stippled bars.

bal performance skills are relatively unaffected. Marginally lower scores on the freedom from distractibility factor suggest that cognitive functioning may be mildly compromised by inadequate attention.

Achievement

Boys with KS were compared with controls on their WRAT or WRAT-R results. Statistical comparisons were carried out only at those ages with a minimum of 3 subjects per group. The percentile scores for Reading, Spelling, and Arithmetic subtests, and the respective grade-level equivalents, were recorded. The latter were converted to difference scores based on the child's expected grade level for his age (i.e., the grade he would have been in had he not failed). The decision to use expected rather than current grade level was based on the observation of the high frequency of grades failed in this population, which occurred at different times for different subjects. However, the results are very similar.

The mean percentile scores of boys with KS and controls are provided in Table 3. The KS group scored consistently lower in all domains and at all ages. Mann-Whitney *U* tests indicated that the groups differed significantly at nearly every age excect 6 years. For boys with KS, scores on the three subtests declined steadily with age. This was not observed for controls, except in their arithmetic performance during adolescence, which is not uncommon in Toronto teenagers (Lewington, 1993).

Table 4 presents the proportions of subjects in each group who evidenced mild (1 to 2 SD below the mean) or severe (≥ 2 SD) reading and arithmetic impairment. For reading, mild impairment ranged from 100% of the sample of boys with KS at age 20 to 13% at age 15; for controls, it ranged from 50% (13, 14, 17, and 18 years) to 0% (6, 7, and 10–18 years). For boys with KS, the mean (across ages) rates of mild and severe impairment were 39% and

17%, respectively; for boys without KS, they were 18% and 0%. The relative risk of combined mild and severe impairment was 3.1. Similar findings were observed for arithmetic: The average rate of mild impairment was 28% for boys with KS and 18% for controls and of severe impairment was 17% and 0%, respectively, for a relative risk of 2.5.

For all subjects with KS, grade-equivalent difference scores were computed between current level of performance (in grade equivalent units) and expected grade level. As shown in Figure 2, an increasing deviation from expected grade for age was found in both reading and arithmetic. By age 18, boys with KS were performing more than 5 grade levels below their expected placement (and 4.5 below their current grade level). The correlation with age was $-.92$ ($p < .001$) for reading and $-.97$ ($p < .001$) for arithmetic, signifying that they fell consistently and progressively behind expectations and then peers. When the grade-equivalent scores of boys with KS were compared with those of controls, results revealed that they differed significantly in reading and arithmetic at age 11 and in reading at age 12.

At a median age of 15 years (range = 12.8 to 17.0 years), 23 boys with KS were assessed via the WJPTB. This test provides individual scores for reading, math, knowledge, and written language. On the reading cluster, boys

TABLE 3
Mean Wide Range Achievement Test Percentile Scores for Klinefelter Syndrome (KS) and Control (C) Groups

			Reading			Spelling			Arithmetic		
Age	KS (n)	C (n)	KS	C	p^a	KS	C	p	KS	C	p
6	12	4	46.1	61.3	ns	40.5	55.6	ns	502	51.2	ns
6	24	6	29.5	65.5	.01	24.6	65.6	.01	25.0	56.0	.01
9	24	6	32.9	47.0	.05	26.4	48.2	.01	31.0	44.2	.01
10	20	3	25.1	71.0	.05	34.9	70.0	.05	32.2	67.0	.05
11	19	3	29.2	88.0	.05	25.2	72.5	.05	21.6	56.6	ns
12	13	6	20.4	45.8	.01	19.9	33.7	.01	11.5	27.0	.01
13	9	4	19.6	65.0	.01	19.3	61.3	.01	12.0	32.5	.01

'Data are analyzed by Mann Whitney Utests.

TABLE 4
Rate of Reading and Arithmetic Impairment in Klinefelter Syndrome (KS) and Control (C) Groups

			Reading				Arithmetic			
			KS		C		KS		C	
Age	KS (n)	C (n)	Mild	Severe	Mild	Severe	Mild	Severe	Mild	Severe
6	14	4	.29	.00	.00	.00	.15	.14	.00	.00
7	19	1	.21	.05	.00	.00	.11	.00	.00	.00
a	22	6	.32	.14	.29	.00	.27	.05	.29	.00
9	23	6	.48	.00	.13	.00	.31	.00	.00	.00
10	24	3	.25	.13	.00	.00	.34	.04	.00	.00
11	17	3	.41	.00	.00	.00	.59	.00	.00	.00
12	12	6	.33	.25	.17	.00	.42	.33	.30	.00
13	10	4	.30	.30	.50	.00	.70	.00	.50	.00
14	12	2	.34	.oa	.50	.00	.17	.33	.50	.00
15	15	1	.13	.07	.00	.00	.27	.13	1.00	.00
16	7	1	.15	.71	.00	.00	.29	.57	1.00	.00
17	20	0	.15	.30			.10	.35		
18	14	2	.oa	.36	.50	.00	.07	.43	.50	.00
19	9	0	.45	.33			.23	.33		
20	2	0	.00	1.00			.00	1.00		
Across age			.39	.17	.la	.00	.28	.17	.18	.00

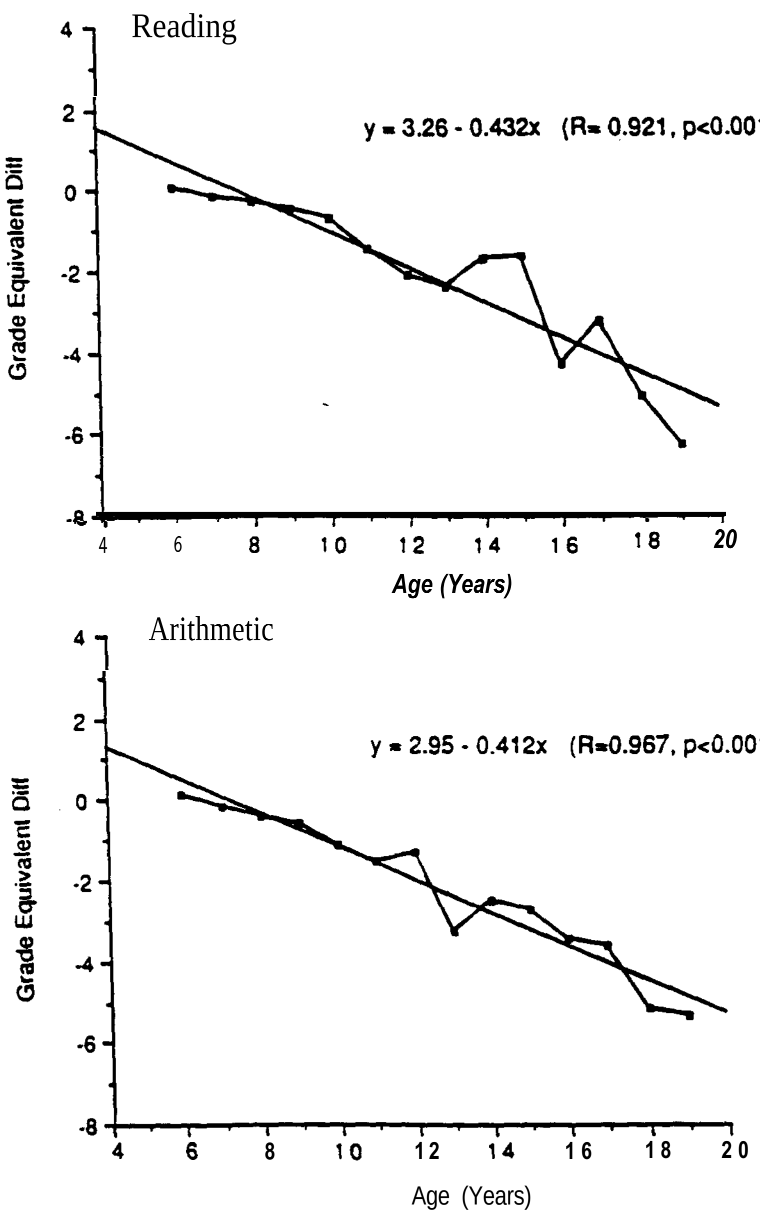

FIGURE 2. Mean reading and arithmetic grade-difference scores for boys with KS.

with KS obtained a mean percentile score of 25.4, with a range of 0 to 80. Fifty-seven percent scored in the below-average range (38% in the severely defective) in reading, whereas only 9% obtained above-average scores. Scores for decoding and comprehension components were equivalent. On the math cluster; they obtained a mean percentile score of 19.9, with no boys with KS obtaining an above-average score. In written language, their mean percentile score was 25.5, with 57% placing in the defective range and 15% scoring in the above-average to superior range. On the knowledge cluster, they obtained a mean percentile score of 20.1, with a range of 0 to 45 (with 42% of the group scoring in the defective range). Across clusters, results suggest that math and knowledge are affected to a greater degree than reading, while written language skills are the least affected. These results were significantly correlated (p c .001) with VIQ: reading: r(22) = .74; math: r(22)= .88; written language: r(22) = .64; knowledge: r(22) = .87, but not PIQ. Performance on the Vocabulary subtest was the strongest predictor of reading, written language, and knowledge achievement, whereas performance on the Arithmetic and Information subtests were the strongest predictors of math.

On the Test of Written Language, given to KS boys at an average age of 14.5 years (range = 12.8 to 16.51, they obtained a mean Written Language Quotient at the 20.5 *centile* (range = 1 to 861, with 56.5% of boys scoring below average; 39.1%, average; and 4.4% (1 child), above average Their subscale scores ranged from the 16th centile for Style to the 46th centile for Contextual Spelling, while Vocabulary, Thematic Maturity, Word Usage, and Handwriting were intermediary.

Learning Disabilities

The WRAT-R results were used to assign KS and control boys to different learning disability–subtype groups based on two methods involving different arbitrary cutoff criteria. Using the method of Rourke and his associates (Rourke, Fisk, & Strang, 19861, subjects were assigned to a reading disability (RD) group if they achieved below the 25th centile in reading and above the 34th in arithmetic; to an arithmetic disability (AD) group if they achieved below the 25th centile in arithmetic and above the 34th in reading; and to a generalized disability (GD) group if they achieved below the 25th centile in both reading and arithmetic. The second stringent criteria method (see Rovet, 1993) involved lower cutoff scores, whereby subjects were assigned to the RD group if they scored below the 10th centile in reading and above the 25th in arithmetic; to the AD group if they scored below the 10th in arithmetic and above the 25th in reading; and to the GD group if they scored below the 10th in both. These analyses were conducted for the 6 to 8, 9 to 11, and 12 to 14 age ranges only, as there were too few controls past this age to compare different subgroups.

Subjects' representations in the different learning disability-subtype groups are shown in Figure 3. With the Rourke criteria, boys with KS were assigned mostly to the GD category, with very few showing a pure reading or pure arithmetic disability in contrast to controls, who tended to be more equally distributed across categories. With the more stringent criteria, more children with KS had a significant learning disability; for KS, the most common classification was a GD, whereas for controls it varied across all categories. Also, the prevalence increased steadily with age, such that by early adolescence over 40% of boys with KS showed a severe learning disability, while the rate for no disability decreased dramatically (Rourke criteria: 23% at 6 to 8 years, 29% at 9 to 11 years, 8% at 12 to 14 years; stringent criteria: 23% at 6 to 8 years, 36% at 9 to 11 years, 13% at 12 to 14 years).

School Placement

Boys with KS were more likely than controls to have failed a grade (37% vs. 0%) or to have received special education (40% vs. 3%).

Discussion

This article uses the findings from two sources to describe the psychoeducational characteristics of boys with KS. The results of previous research were integrated to identify the cognitive deficit in these individuals, in order to provide information about their school achievement. Even though this is an exhaustive analysis of the findings on this population, we were limited because previous studies spanned wide age ranges, combined results across various SCA groups, and did not systematically test for achievement. The longitudinal approach used to analyze our own data represents an attempt to address these limitations. Yet, our study, too, is not without limitations. These include, first, our choice of control sample, which involved both male and female siblings and siblings from children with other SCAs. As controls were tested less frequently than probands, we were also limited in the number and type of statistical comparisons we were allowed. Furthermore, we were lacking data from controls on the more detailed tests of psychoeducational achievement and suffered an approximately 20% loss in sample size over the 15-year period. These limitations notwithstanding, we believe our data are unique and the most informative on this population ever to be published. Furthermore, we also present important information about these individuals' specific learning disabilities and the relative risks of different types of impairment.

The review of the literature on boys with KS, which is summarized in Table 1, shows that they do indeed have a chronic cognitive deficit in the area of verbal or language processing. Moreover, they achieve more poorly at. school and are at risk for dyslexia (Ratcliffe, Murray, & Teague, 1986; Robinson et al., 1986). Never-

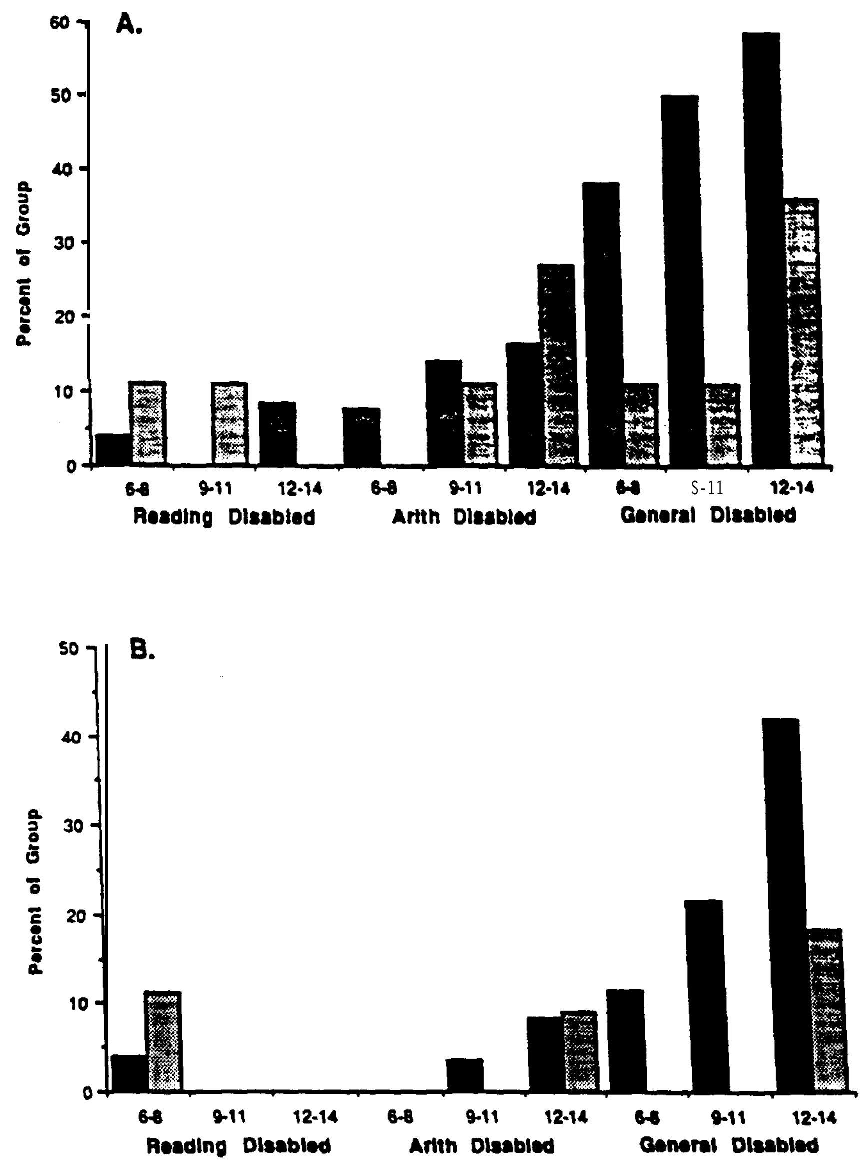

FIGURE 3. Proportion of subjects in KS and control groups assigned to different learning disability subtypes. A: Rourke criteria: B: stringent criteria. Boys with KS are shown with the solid bars, controls with the stippled bars.

theless, despite their poorer achievement, many did complete high school, and a few even attended postsecondary institutions. The need for special education is clearly indicated in this population.

Our findings showed that boys with KS were more likely than chromosomally normal subjects to have a specific cognitive impairment that persisted throughout childhood and adolescence. They demonstrated significantly depressed verbal ability but normal nonverbal ability and had more difficulty on tasks that made greater demands on auditory memory, language comprehension, and expressive abilities. This was observed in the majority of boys with KS, and they clearly differed from sibling controls in this respect. Although difficulties were also observed for tasks of attention, these effects were not as marked as those for verbal processing.

We also found that boys with KS showed significantly poorer achieve ment than sibling controls. They did more poorly on tests of word decoding, reading comprehension, spelling, written language skills, arithmetic and math problem solving, and the acquisition of conceptual knowledge in subject areas such as science and humanities. Their performance on standardized achievement tests was observed to fall progressively behind age and grade expectations, such that by age 18 to 20 years they were performing more than 5 grade levels behind expected grade levels for reading and arithmetic. Unlike previous studies indicating a selective learning disability in reading and language-related subject areas, we found that most KS boys were almost equally impaired in both reading and arithmetic Their most frequent type of learning disability was found to be a generalized learning disability, unlike controls, who were more equally represented among pure reading and pure arithmetic disability as well as combined reading and arithmetic subgroups. We also found that the likelihood of being learning disabled increased with age for the KS, but not the control, group. Across age, KS was associated with a 3.1 increased risk of reading impairment and a 2.5 increased risk of arithmetic impairment. In addition, boys with KS were much more likely to have failed one or more grades or to have received special educatiori. This is consistent with a report by Mandoki, Sumner, Hoffman, and Riconda (1991), in which 64% of boys with KS in the literature were reported to have severe academic problems, in contrast to 24% Of controls.

Detailed studies of the psychoeducational characteristics of our sample during early to midadolescence also showed that their written language skills and learning of knowledge-based subject areas were compromised. The degree of difficulty in these areas appeared to increase with age as the demands for logical and conceptual thinking increased, and were not specific to reading.

These results, therefore, signify that KS inueases the risk of what appears to be primarily a language-based learning disability. This disability significantly affects these students' performance at school. This difficulty, we believe, is a result of one or more basic cognitive impairments that may reflect an underlying problem in auditory temporal processing or working memory. Previous research has indicated that, from an early age, boys with KS show language problems reflecting delayed speech acquisition (Leonard, Schowalter, Landy, Ruddle, & Lubs, 1979; Robinson et al., 1979) and poor skills in sentence building intonation, and word naming. At a later age, they demonstrate poorer articulation and comprehension as well as deficits in verbal abstraction, syntax production, and word finding (Walzer et al., 1982). Graham et al. (1988) pointed out that these cognitive problems actually reflect a more basic underlying handicap in accessing, retrieving, and applying linguisitic information, which is central to their disability. Our previous studies comparing the performance of boys with KS and age- and IQ-matched controls from local schools on a number of experimental tasks of language and memory processing showed that those with KS were deficient in comprehending and using syntactic information to judge the veracity of sentences (Netley & Rovet, 1983) or facilitate recall (Stewart et al., 1982). They also differed on tasks of visual and auditory memory, demonstrating below-normal span sizes and slower processing speeds. In one study, in which subjects were required to repeat the final 3 digits from lists of varying lengths (12 to 50 digits) and varying speeds (1.75 to 8 digits/second), the KS group differed most from controls at normal speeds, whereas both groups deteriorated equally at fast speeds (Stewart et al., 1986). This suggested to us that boys with KS were less adept at processing auditory information in the normal range.

The implications of the present findings for the classroom suggest that auditory processing difficulties would affect the amount and accuracy of information processed by boys with KS. Also, because they are less able to deal with the syntactic aspects of language, they would be less likely to understand the meaning or intent of the information being conveyed. We found previously that when subjects were presented, at a slow rate, with word lists constituting (a) a meaningful sentence, (b) an anomolous sentence (syntax correct but meaning incongruous), or (c) a random list, they recalled all items as if they were random word lists (i.e., first and last items best) and not in sentence order, even if the lists constituted a meaningful sentence (Stewart et al., 1982). This signifies that if they cannot properly process orally presented verbal information, they will be more confused than nondisabled students in the classroom and therefore will be less able to understand instructions or follow tasks at hand. This should become more problematic as linguistic structures of conversation and instruction increase in complexity with age, in addition to the greater demands on integration, assimilation, and inferencing skills.

Reading comprehension, evaluated only during adolescence in our sample, was clearly problematic and indicated that boys with KS were severely restricted in their capacity to derive information from print. As with other children demonstrating comprehension deficits (Barnes & Dennis, 1992), this is most likely also due to their underlying cognitive impairment in mentally forming structures or templates for organizing the information they read. Lacking these frameworks, they would be severely restricted in recalling the information or in making inferences about it, as required on reading and other tests.

Given the difficulties in the expressive language domain experienced by boys with KS, it is not surprising that this would also adversely affect various productive tasks at school. For example, their written language skills were found to be clearly impaired, particularly in the stylistic component, which reflects their difficulty in expressing their ideas in print. Difficulties in word retrieval and object naming would place them at a disadvantage on tasks of reading comprehension, where the lack of a clear understanding of the words they read would make the meaning of the passage less comprehensible. This should also affect the quality of answers they produce (being wrong more often than others), and their ability to convey their specific needs to their teachers (which in turn may give rise to more behavior problems in the classroom).

Although we did not conduct detailed analyses of these students' arithmetic errors, the difficulties observed in this achievement domain are not suprising, given their poorer recall skills and difficulties with comprehension. For example, it is expected that their shorter span sizes and less adequate working memories would negatively affect table mastery and the memorization of other math facts. Also, one would expect that their procedural knowledge for carrying out step-wise componential skills of arithmetic processing would be less proficient, because of their poorly organized mental frameworks. Similarly, difficulties in comprehension would adversely affect their abiity to solve word problems. Additional studies to ascertain the specifics of their math processing difficulties using systematic and analytic approaches, as have been conducted on children with various forms of developmental dyscalculia (e.g., Rovet, Szekely, & Hockenberry, 1995; Sokol, 1993; Temple, 1989, 1991), are definitely recommended, as such information would be highly useful in determining proper remediation.

While our findings indicate that boys with KS performed below standard on the composite of subtests comprising the distractibility factor, we believe that problems with distraction do not represent a co-morbid condition but, rather, are a correlate of their basic cognitive difficulty in processing auditory information. For example, it is not surprising that they would fail to attend, if they could not adequately follow the information that was being presented. Furthermore, there is no evidence of increased hyperactivity—if anything, these boys are described as being less active than normal (Stewart et al., 1986).

What are the implications of the present findings for the classroom teacher or special educator, particularly in light of the fact that many of these children with this highly prevalent condition have never been diagnosed as having a genetic abnormality? Clearly, they should be treated as any child with a language-based learning disability and be given early speech and language therapy that stresses vocabulary building, trains sentence understanding, and teaches comprehension skills and word finding. In the classroom, teachers should speak slower and be cognizant of the need to use concrete, shorter sentences with simple syntax. Ambient noise should be reduced and considerable repetition provided. Strategies to improve memory and, if possible, training to expand memory span sixes should be provided. With regard to reading, comprehension can be improved by (a) presenting advance organizers to assist with the formation of mental templates to organize story information, and (b) structuring the lesson so that small chunks are read and continuous checks are provided in order to ensure that the passage is being followed. In math, drills for math facts should be given and continued until mastery is achieved; processing should be closely monitored to determine that component procedures are being carried out correctly. Extra training on word problems should also be given.

With newer and improved prenatal screening techniques, it is expected that more of these children will soon be identified in utero, and, with better counseling regarding the develop ment of boys with KS and improved techniques for remediating learning disabilities, more pregnancies may be continued in the future. If the stigma associated with this condition is reduced, more parents may want teachers to know a child's problem so that specific assistance can be implemented early. Therefore, as this disorder may become more coinmon in the classroom, it is important that it be dealt with appropriately. In our experience with these boys, those from more functional and supportive families and/or who received assistance and remediation at school fared better in the long term.

What if a teacher suspects that a child has Klinefelter syndrome? According to Mandoki et al. (1991), the diagnosis of KS should be considered in all boys with language difficulties, learning problems, behavioral difficulties, and lack of coordination, and who are disproportionately tall (i.e., long legs and arms). Some boys with KS may benefit from hormonal therapy in puberty as well as from other pharmacological treatments (Mandoki & Sumner, 1991); such therapy can provide a more normal adolescent development and serve as a prophylactic against learning disabilities and deviations in behavior (Mandoki et al., 1991). The issues of who should convey this information to parents and how they should convey it are beyond the scope of this article; however, procedures should be implemented for educating teachers about genetic abnormalities and how to deal with the various kinds they may encounter.

This article has centered on the male with an extra X chromosome. What about the female who also has an additional sex chromosome complement? The literature on this group is unfortunately quite diverse, and for the most part suggests greater impairment than in the extra X boy, including the greater likelihood of mild mental retardation (Pennington et al., 1980). As most studies of this subject are based' on relatively small sample sizes, the likelihood for bias and skew in the data is quite high. In our sample of 11 girls with a trisomy X karyotype, we observed that the pattern of physical characteristics (Stewart et al., 1982, 1986) and cognitive development (Rovet & Netley, 1983; Rovet, Netley, Bailey, Stewart, & Keenan, 1995; Stewart et al., 1986) were similar to those of the boys with extra X chromosomes. This included tall stature (within the normal range), and language, memory, and learning problems. However, as our sample of girls with trisomy X appeared to be more functional than those described in other studies in the literature (e.g., Bender, Linden & Robinson, 1989), there is clearly a need to pool data across studies and conduct detailed studies of those identified in utero. Because awareness of this phenotype may guide intervention and therapy goals by focusing on primary strengths and weaknesses, educators should also be cognizant of its existence and of information, albeit limited, currently available on this group. As with boys with langauge disabilities, the possiblity of genetic testing of girls fitting the description might also be considered.

In closing, we have described the psychoeducational characteristics of boys with a relatively common genetic disorder known as Klinefelter syndrome Although this disorder has few if any outward physical manifestations in childhood, it is associated with increased risk of language-based learning disabilities and has a significant impact on school learning and success. The implications of these findings for the general educator who may suspect a boy has this condition were discussed briefly. Clearly more research is needed, and the possibility of identification on the basis of behavioral characteristics may be considered, particularly in light of available chemical therapies (Mandoki & Sumner, 1991) and, presumably, behavioral therapies.

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