

# The Psycho-educational 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 was also problematic. Despite significant underachievement 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.

## **Objective of Paper**

The purpose of this article was to describe the intelligence and achievement characteristics, and the frequency and types of learning disabilities of boys with KS. This is done through a review of the existing research and the findings of a 20-year long study of boys with KS in Toronto, Canada.

## **Overview of Previous Research**

To summarise the findings on school achievement in boys with KS, the literature from 1970 to the present was examined. The one study of boys diagnosed before birth reported no evidence of language or school-related problems. It was felt that this could be due to a more supportive environment and higher socio-economic status in these families. A summary of the other studies on children indicated that boys with KS experienced significant school under-achievement. It appears that they are of normal intelligence, but may have difficulties with social relations, speech and language development, reading, spelling, and writing. Problems with maths seem to become more apparent as children reached the age of about 10. Some studies reported 60-80% of boys receiving special education.

The findings on boys with KS in late adolescence and early adulthood were presented at a conference in 1991. They indicated that, despite early problems, many boys with KS do succeed at school, and continue to the end of secondary education. However, levels of achievement are clearly below expectation, and there is an increased need for remedial help and special education. Although repeating a year is a common solution for under-achievement, a number of boys do complete secondary school, with a few even acquiring post-secondary education.

Although these results are more optimistic than those found previously, they do indicate an increased risk for learning problems and greater need for remedial services. In the light of

all these findings it was felt that a more detailed description was needed of the specific problems experienced in education by KS boys.

### **The Toronto Study**

The purpose of this section is to describe the 20-year long study conducted in Toronto on intelligence, school achievement and the incidence of specific learning difficulties in boys with KS.

Initially 41 boys with KS were identified from a large post-natal screening programme conducted between 1967 and 1971. Detailed study of their early development indicated delayed speech and motor development as well as hypo-activity (the opposite of hyper-activity). These results are published elsewhere. The Toronto study was conducted on 36 boys from the age of 6 onwards. Of the 36, 29 were followed until completion of the study in 1988, when the oldest child reached 20 years of age. The study used brothers and sisters as a comparison for the progress of the boys with KS.

### **Results of the Toronto Study**

The findings indicate that the boys with KS had a distinctly poorer verbal ability, whereas their non-verbal performance skills were relatively unaffected. Also, the boys with KS were more easily distractible, and they had more difficulty on tasks that require auditory memory, language comprehension and expressive abilities. The study also found that boys with KS scored less well than their siblings on a range of tests including spelling, writing, and maths. As they got older they fell increasingly further behind. Whereas other studies found only reading and language disabilities, this study found that the boys with KS also had problems with maths.

For the boys with KS, a generalised learning disability was most frequently found, whereas any problems amongst the controls were more specific. Also the likelihood of having a learning disability increased with age for the KS group, but not for the other children. Across age, KS was associated with a 3:1 increased risk of reading problems and a 5:2 increased risk of maths problems. In addition, boys with KS were much more likely to have failed one or more years, or to have received special education. These results seem to show that KS increases the risk of what appears to be primarily a language-based learning disability. This significantly affects the performance of these boys at school.

### **Why do these problems occur?**

It is believed that the difficulties experienced by KS boys are possibly a result of an underlying problem in working memory. From the results of previous studies, it seems that there is a basic problem in KS in recalling, retrieving, and applying information relating to language. This can lead to a delay in acquiring speech, poor sentence building, poor intonation, problems with articulation, and word finding. Previous studies have also shown slower processing speeds and shorter span for visual and auditory memory.

### **Implications of the findings for the classroom**

The present findings suggest a number of possible problems for boys with KS at school. They may be less able to take in as much information, or to do so as accurately as their peers. They may be less able to understand the meaning of information given to them, if they cannot properly process spoken information, they will be more easily confused than their peers in the classroom, and therefore less able to understand instructions or follow tasks. These problems would almost certainly increase with age, as conversation and instructions become increasingly complex.

Reading comprehension was also clearly a problem (though only looked at in the adolescents in this study). Given the difficulties experienced by boys with KS with

expressive language, it is not surprising that this would also affect various tasks at school. For example they would have difficulties in expressing their ideas in the written word. Problems recalling words, with object naming, and a lack of clear understanding of the words they read, would make reading comprehension more difficult. This could then affect the quality of the answers they produce (as a result of not understanding what they have read), and also their ability to tell the teachers what their specific needs are (which in turn may give rise to behaviour problems).

Difficulties observed in maths were not surprising, when the poorer recall and difficulties with comprehension are taken into account. Shorter memory span sizes and less adequate working memories would make it more difficult to memorise times tables or other maths facts. Carrying out step-wise tasks in maths could be made more difficult because of poorly organised mental frameworks, and difficulties in comprehension would make it harder to solve word problems. Further research is definitely recommended in order to determine the best remedial help.

The findings on distractibility were not felt to be a separate problem, but related to their difficulty in coping with verbal information, it is not surprising that they would not be able to attend if they could not follow the information that was being presented. Implications for the classroom teacher.

What are the implications of these findings, particularly in light of the fact that many of these children with this relatively common condition have never been diagnosed? Clearly they should be treated as any child who has a language-based learning difficulty, 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 more slowly, and be aware of the need to use shorter, simpler sentences, with specific meaning. Background noise should be reduced, and lots of repetition provided. With regard to reading, comprehension can be improved by giving help to organise story information and structuring the lesson so that small chunks are read, and checks are provided, in order to ensure that the passage is being understood. In maths, much repetition of maths facts will be necessary until they are achieved. Checks on procedures in sums should be in place to ensure that they are being carried out correctly. Extra training on word problems should also be given.

With newer and improved pre-natal screening techniques, it is expected that more of these children will be diagnosed before birth. With better counselling regarding the development of boys with KS, and better remedial teaching, more pregnancies may be continued in the future. If the stigma associated with this condition is reduced, more parents may want teachers to know about their son's condition so that specific help can be given early. As the disorder becomes more common in the classroom, it is important that it is dealt with appropriately. In our experience with these boys, those who received the most support and help from their families and school did better in the long term.

According to one research report, it is felt that a diagnosis of KS should be considered in all boys with language difficulties, learning problems, behavioural difficulties, and lack of co-ordination, and who are disproportionately tall (i.e. long legs and arms). Some boys may benefit from hormonal therapy in puberty. This can provide a more normal adolescent development, and can help avoid learning difficulties and behaviour problems. Clearly more research is needed to understand this condition.

*To order this reference from your librarian you will need the following Harvard Reference:  
Rovet J., Netley C., Keenan M., Bailey J., Stewart D, (1996), The Psychoeducational  
Profile of Boys with Klinefelter Syndrome, Journal of Learning Disabilities, Volume 29,  
Number 2, March 1996. pages 180-196*