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Brief Clinical Report

Very Superior Intelligence in a Child With Noonan Syndrome

Jo-Anne K. Finegan and Helen E. Hughes

Department of Psychology, The Hospital for Sick Children, Toronto, Canada (J.K.F.), and Institute of Medical Genetics, University Hospital of Wales, Cardiff, U.K. (H.E.H.)

Psychological assessment documents very superior intelligence in a child with Noonan syndrome.

Key words: intelligence, variability, Noonan syndrome

INTRODUCTION

Variability of expression is a characteristic of dominantly inherited conditions [Vogel and Motulsky, 1986]. Noonan syndrome, one syndrome that may show a dominant pattern of inheritance and variability of physical phenotype, has been well-described [Allanson, 1987; Allanson et al., 1985; Mendez and Opitz, 1985; Quazi et al., 1974]. Variability in intellectual phenotype also has been reported. Ample evidence is available that mental retardation is common [Collins and Turner, 1973; Duncan et al., 1981; Gorlin and Sedano, 1973; Jackson and Lefrak, 1969; Kaplan et al., 1968; Mendez and Opitz, 1985; Noonan and Ehmke, 1963; Wilroy et al., 1979]. Less prevalent are reports of average intelligence in these individuals [Allanson, 1987; Baird and DeJong, 1972; Bolton et al., 1974; Char et al., 1972; Money and Kalus, 1979; Noonan, 1968; Penchaszadeh et al., 1974]. Although high levels of intelligence have been inferred from level of academic achievement or occupational status [Berberich and Hall, 1976], only a single case report documents high intelli-

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Address reprint requests to Jo-Anne Finegan, Ph.D., Department of Psychology, The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, Canada, M5G 1X8.

Fig. 1. PC at 13 years, 3 months.

gence on the basis of a psychometric measure of ability [Money and Kalus, 1979]. In this report we describe an individual with Noonan syndrome whose IQ scores fell within the very superior range on a well-standardized measure of intellectual ability and thereby extend the range of intelligence observed in this syndrome.

CLINICAL REPORT

Reason for Referral

PC is an 11 6/12-year-old girl (Fig. 1) who is one of 3 children with Noonan syndrome in a family with an affected father. Her 2 affected brothers are functioning well academically. One brother [Allanson et al., 1985, Fig. 5] had an IQ score in the high average range. The other brother was not assessed psychometrically but is reported to have a very high level of academic achievement (i.e., in the enriched stream with a grade average in the 90's in his final year of high school). Her father also was not assessed but owns a successful business with 40–50 employees [Allanson, 1987, Fig. 5]. Psychological assessment of PC was requested to confirm a clinical impression of high level of intelligence.

General Observations

PC presented as an articulate and vocal young lady. Initially, she was anxious about testing and expressed worries that she would not do well, but once testing had begun, anxiety was not overtly evident. She approached tasks in an organized manner; she was persistent, and reflective and exhibited excellent attention and concentration.

Tests Administered

Bender Gestalt Test [Koppitz, 1968], Developmental Test of Visual-Motor Integration [Beery, 1982], Wechsler Intelligence Scale for Children - Revised [Wechsler, 1974], Wide Range Achievement Test - Revised (Level 1) [Jastak and Jastak, 1984]. These tests are widely-used for children in this age group and they are well-standardized.

Results

On the Wechsler scales, PC obtained the following IQ scores:

Scale	IQ Score	+/−Standard Error	Level	Centile
Verbal	122	119–125	Superior	93rd
Performance	138	134–142	Very superior	>99th
Full Scale	133	130–136	Very superior	>98th

The performance > verbal discrepancy of 16 points is reliable but is fairly common, in that a difference of this magnitude is obtained by 22% of children in the standardization sample [Kaufman, 1979].

Subtest analysis shows that PC has significantly elevated abilities in mental arithmetic and in social awareness and judgment (both at the 98th centile). Her fund of general factual knowledge was relatively weak (50th centile), but because this was the first subtest administered, it may reflect her anxiety level early in the testing session. All abilities measured by the performance subtests were equally well-developed (84th to 99th centile).

PC made no errors on the Bender Gestalt Test suggesting that visual-perceptual-motor systems were intact. Her age equivalency on the test of visual-motor integration was 12 7/12 years.

The findings of the screening test of academic achievement showed the following:

Sight reading/word recognition	47th centile
Spelling of dictated words	19th centile
Arithmetic	87th centile

Reading comprehension was not assessed. Given PC's level of intelligence, higher achievement in spelling and word recognition might have been expected. She showed a phonetic approximation of spelled words suggesting an appreciation of sound-symbol relationships. The presence of a learning disability cannot be inferred on the basis of the current screening tests and could be entertained only with evidence of academic underachievement, but PC currently shows average or above average abilities in all school subjects.

Conclusions

PC is a child with superior to very superior intellectual abilities, good organizational skills, concentration, and persistence. Relative strengths are noted in mental computation and social awareness and judgement.

Visual-motor-perceptual abilities are developed to above age level. Based on screening tests, level of achievement in spelling and word recognition is lower than PC's level of intelligence would predict.

DISCUSSION

Based on a well-standardized test of mental ability, this patient extends the range in level of intelligence in individuals with Noonan syndrome to include the possibility of very superior intelligence. Moreover, it demonstrates that visual-perceptual-motor

systems can be intact [cf. Money and Kahns, 1979]. We know of no other report on the pattern of strengths and weaknesses in academic achievement. Further investigation of spelling and reading abilities in high functioning individuals with Noonan syndrome may reveal whether the relative weakness in these language areas, as seen in our patient, is a common rather than chance finding. If it were a common finding, it would suggest leads to follow in attempting to unravel the possible neuropsychological manifestations of Noonan syndrome.

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