

Turner's Syndrome: Further Demonstration of the Presence of Specific Cognitive Deficiencies*

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Previous neuropsychological studies of patients diagnosed clinically and cytogenetically as Turner's syndrome have indicated, on the basis of the Wechsler Intelligence Scales and the Benton Visual Retention Test, that these patients possess a specific cognitive defect in space-form perception (Shaffer, 1960; Money, 1963). The Wechsler test has also shown that a relative difficulty with numbers, a form of dyscalculia, is associated with the syndrome (Money, 1964). These deficiencies do not appear in every case, however, and in this sense they parallel the physical stigmata of the syndrome which, as described by Wilkins (1965), are widely variable.

According to the findings mentioned above, the Wechsler performance IQs in Turner's syndrome averaged approximately 20 points lower than the verbal IQs. Furthermore, application of Cohen's (1957; 1959) method of factor analysis of the subtest scores showed that the calculation factor (also defined as freedom from distractibility) and the factor of perceptual organization were significantly lower than the verbal comprehension factor.

In order to confirm these results, as well as to look for possible additional neurocognitive deficiencies, a group of Turner's syndrome patients has now been tested with the Science Research Associates (SRA) Primary Mental Abilities test (1958). This test, applicable for ages 11 to 17+, is specifically designed to measure separate factors of mental ability; namely, verbal-meaning, space, reasoning, number, and word-fluency. There are percentile scores for each factor and an IQ estimate can be obtained from the verbal-meaning and reasoning factor scores.

The SRA test was given individually to 16 patients

with an unequivocal diagnosis of Turner's syndrome, who met sample selection criteria designed to avoid intentional bias. All 16 had been originally referred from the Johns Hopkins Hospital Pediatric Endocrine Clinic by Dr. Lawson Wilkins and, more recently, by Dr. Robert M. Blizzard. The diagnosis of Turner's syndrome had been made clinically from the physical stigmata, including, among others, dwarfed stature, webbed neck, and pubertal failure, and was confirmed by buccal smear or chromosome count. Of the 16 patients in the sample, 14 were chromatin-negative or XO, and 2 were chromatin-positive XO/XX mosaics. The age range was from 11 to 24 years, the mean age being 15½ years. The mean Wechsler IQs for the sample were verbal 112, performance 87.4, and full scale 101, the respective ranges being 134 to 86, 110 to 65, and 119 to 80. The mean SRA estimated IQ and IQ range conform fairly closely to the Wechsler findings (see Table)

The Table also shows the SRA subtest results. The median and range scores (columns a and b) provide an indication of the group performance on each subtest. Columns c and d indicate the distribution of the percentile scores according to the number falling above the 75th and below the 25th percentiles, respectively. The group was also divided for each subtest into those whose scores fell at the 50th percentile or below, and those at the 51st percentile or above (column e). The observed frequency of scores in the two subgroups was compared with the expected (50/50) frequency and subjected to a χ^2 test. The results of this comparison, expressed as the probability that the observed distribution is within normal limits, are shown in column f.

The Primary Mental Abilities test results corroborate the earlier observations from the Wechsler test. The verbal-meaning scores approximate a normal distribution. The reasoning scores, though generally lower, are still within normal limits.

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TABLE
ANALYSIS OF SRA PRIMARY MENTAL ABILITIES TEST FINDINGS IN TURNER'S SYNDROME

Subtest	a Median Score (Percentile)	b Range of Scores (Percentile)	c Proportion Above 75th Percentile	d Proportion Below 25th Percentile	e Proportion Above 50th Percentile	f p Value for χ^2 †
Verbal-meaning	42	90-7	4/16	5/16	7/16	p > 0.80
Space	10.5	78-1	2/16	14/16	2/16	p < 0.01
Reasoning	28.5	82-6	1/16	8/16	4/16	p > 0.08
Number	26	84-1	1/16	7/16	3/16	0.01 < p < 0.05
Word-fluency	17	93-1	2/16	10/16	2/16	p < 0.01
IQ Estimate	40*	91-2†	3/16	6/16	6/16	p > 0.40

* Mean IQ 95.3
† IQ range 68-122.

‡ Comparing numbers of subjects above and below the 50th percentile.

The space scores, by contrast, are significantly below the normal level, thus corresponding with Cohen's perceptual-organization quotient on the Wechsler: 14 of the 16 girls rank below the 25th percentile, and 8 of these are at the 10th percentile or lower. The performance on the number subtest is also poor, though the difference from normal is only of borderline statistical significance.

An unexpected and heretofore unknown phenomenon is the low score on the word-fluency subtest. Although the scores on this subtest are in general not as low as on the space subtest, the score distribution is none the less significantly lower than normal.

The name word-fluency is possibly misleading for this subtest. It is perhaps more accurately described as an alliteration or controlled association test, because it requires the subject to write in five minutes as many words as he can that start with the letter s. As such, it requires mobilization of attention and memory for the entire duration of the test. It is possible that the factor being measured in this subtest is, like Cohen's factor of 'freedom from distractibility' in the use of numbers, a freedom from distractibility in controlled lexigraphic association and not generalized fluency in the use of language.

In summary, the SRA Primary Mental Abilities test results support the hypothesis that patients with Turner's syndrome have defective space-form perception and difficulty in numerical manipula-

tions. This SRA test also suggests the hypothesis of a deficiency in the factor measured under the name of word-fluency.

Summary

SRA Primary Mental Abilities test results confirmed earlier reports that many patients with the cytogenetic and clinical entity, Turner's syndrome, exhibited deficiencies in space-form perception and in dealing with numbers. In addition, the same SRA test indicated that a deficiency in speed and fluency of lexigraphic association was also present.

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