

INTELLECTUAL IMPAIRMENT IN A PATIENT WITH HEPATOLENTICULAR DEGENERATION (Wilson's Disease)

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Introduction

The present paper presents a description of the intellectual functioning of a patient with hepatolenticular degeneration (Wilson's Disease). This condition is rare and the pattern of intellectual impairment demonstrated in this case is of interest to students utilizing psychological test performance for the detection of organic brain impairment.

Wilson's Disease is a condition characterized by bilateral degeneration of the corpus striatum and cirrhosis of the liver, and is marked by tremor, spastic contractions, psychic disturbance, and increasing weakness and emaciation. The primary cause is the accumulation of copper in the body due to a deficient transport mechanism. Although in the brain, the greatest concentration of copper is found in the lenticular region, Wintrobe et al state that a substantial increase in the copper content of the cerebral cortex is commonly found (Wintrobe et al 1954).

The psychic disturbance has been described in many ways. Bickel, Neale and Hall describe reactions, including that of psychosis, with schizophrenic and paranoid ideation, olfactory and auditory hallucinations, mental decay and violence (Bickel et al, 1957); Uzman comments on the marked euphoria and general nervousness (Uzman, 1957); Heller found depression and apprehension over the deterioration of the body image (Heller, 1965); and Bearn states the patients may sometimes present symptoms suggesting schizophrenia, thus leading to an incorrect diagnosis (Bearn, 1960).

Only one article in the literature could be located which dealt specifically with the intellectual functioning of individuals with Wilson's Disease. Knehr and Bearn examined seven Ss on a battery designed to study disorganization of thinking in psychiatric patients and psychological impairment in patients with cerebral histopathologic changes or dysfunctions related to neurophysiological factors (Knehr and Bearn, 1956). They concluded that there was impairment in intellectual functioning in all cases. Losses in IQ from 12 to 34 points were estimated. The well habituated language function appeared to be fairly well preserved with the loss occurring in the efficiency with which new relationships are abstracted or perceived and the ease with which these can be used conceptually. No disturbance in perceptual accuracy was noted. Impairment was noted which was the result of changes in cortical functioning. Evidence suggested a loss in the capacity for conceptual thinking in all cases studied.

The present patient was administered the Wechsler Adult Intelligence Scale (WAIS) (Wechsler, 1958). The detection of organic brain dysfunction from the differential subtest performance on the Wechsler Scales has been a popular endeavour. Wechsler has stated, "Brain injury, irrespective of origin, if sufficiently extensive, results in a more or less typically organic syndrome" (Wechsler, 1958). The syndrome is characterized by a higher verbal than performance IQ and by special deficit or dysfunction in certain of the verbal as well as most of the performance subtests. Digit Span, Arithmetic and Similarities are more affected than Vocabulary, Information and Comprehension. In a review of the rather extensive literature on the

Wechsler subtest patterns associated with organic brain damage, Guertin, et al note that the subtest pattern produced is a function of the age and sex of the S and the extent, type, specificity, locale and chronicity of the dysfunction (Guertin et al 1966). As Yates points out, it is essential to regard the effects of brain damage from a hierarchical point of view and that brain damage will produce in any given individual a general deterioration in all aspects of functioning, differential effects depending on the location and the extent of the damage and that highly specific effects will occur as a result of certain highly specified areas of the brain being involved (Yates, 1966). The effects of brain damage will be manifested in different ways according to which hierarchical level is being tapped by any particular test.

Case History

The patient studied was a 23-year-old single female of Hawaiian—Chinese—Spanish ancestry, the second of seven children. She graduated from high school at 18 years of age in a college preparatory course and graduated 6th in a class of 111. She was elected, while in the 12th grade, to Junior Achievement, a National Honor Society. She entered nursing school after high school graduation and completed 2½ years of a 3-year course, leaving because of poor health. Her grades were average. She then entered business school training, completing 4 terms of a 5 term course, again leaving for health reasons. Her grades were reported to be average. Psychological tests given in September of 1957 revealed the following scores: Differential Aptitude Test, Verbal Reasoning 75th Percentile, Numerical 75th Percentile and a Californian Test of Mental Maturity IQ Score of 110. The diagnosis of Wilson's Disease was made in 1964. She apparently had been ill since age 9 with frequent absences from school. Previous diagnoses included: cirrhosis, hepatitis and anemia. One brother and one sister died at age 9, due to "liver trouble." It is now suspected that they also had Wilson's Disease. Treatment consisting of regulated diet and D-Penicillamine was begun after the diagnosis was made. She has been able to tolerate the medication with concomitant use of steroids. She was seen in September of 1966 at the request of the Department of Vocational Rehabilitation to assess her intellectual capacities for programme planning. At the time she was seen she was doing work in a sheltered workshop programme for the mentally retarded.

Results

She was pleasant, co-operative, neat and well groomed. She spoke rather slowly and enunciated very clearly. On one occasion a slight slur in her speech was detected. She evidenced no anxiety or apprehension in the test situation and even when she had considerable difficulty with test items, her affect remained rather cheerful and she gave no indication of being disturbed by her inability to perform adequately. Often brain damaged individuals show shock, apprehension, concern and fear about their inability to perform; this girl gave no indication of any traumatic response to her inabilities. She stated that her health was improved, that she was able to work more consistently and regularly and that she did not tire as she had in the past. She denies any difficulty with vision, clumsiness or motor impairment. She eats well, sleeps well, and has no difficulty with mood swings.¹

Her performance on the Wechsler Adult Intelligence Scale was as follows: Information 13; Comprehension 12; Arithmetic 10; Similarities 13; Digit Span 7;

¹Information received from her physician some five months following the testing indicates marked deterioration in the patient's functioning. She now demonstrates euphoria, poor judgement, personal untidiness, and has become irresponsible and a management problem in the home.

Vocabulary 13; *Verbal IQ* 108; Digit Symbol 8; Picture Completion 7; Block Design 8; Picture Arrangement 12; Object Assembly 11; *Performance IQ* 94; *Full-Scale IQ* 102. The Verbal IQ exceeds the performance IQ by 14 points. The one and two-tailed probabilities for such a difference are .075 and .150 respectively (Fisher, 1960).

The subtest pattern of this protocol has some agreement with Wechsler's typical organic test pattern but there are some exceptions. First, the higher Verbal than Performance IQ and lower scores on Digit Span, Digit Symbol and Block Design are all consistent with the organic syndrome. Picture Arrangement and Object Assembly, however, are at the same level as the average of the verbal subtests, excluding Digit Span. Similarities is the highest subtest score whereas this is a subtest which is thought to be sensitive to organic dysfunction. The discrepancy between Arithmetic and Digit Span is of interest. On Arithmetic, no credit was received for the last three items and one of these was not credited because of the patient taking longer than the required time. As compared with this good performance on Arithmetic, on Digit Span she was only able to repeat six digits forward and three digits backward. Even after a number of trials on four digits backwards she was not able to achieve success. Yet, on Arithmetic she was able to hold the instructions mentally and do the mental calculations. The difference here perhaps is one of structure and meaning in that in the Arithmetic tasks there is a context within which the information can be "held" whereas in Digit Span one is simply required to freely attend and give back the auditory stimuli without any meaningful context. Wechsler notes that a marked falling off in memory span is often one of the earlier indications of mental impairment. Information, Comprehension and Vocabulary were among her highest scores, which is typical of the brain damage syndrome. There was some slowing in her thought processes, some blockage noted and a groping for words for explanations in the Comprehension subtest but she was able to achieve success. The failures on Comprehension are on two of the proverbs items. Inability to interpret proverbs is often taken as a sign of inability to engage in abstract and conceptual thinking. On the 14 items on Comprehension she achieved full credit scores on 9 of the items. On Similarities, of the 13 items she achieved full credit scores on the first 9 items and level 1 responses (functional responses) on 2 of the last 4 items. She had much less difficulty with Similarities than with Comprehension and gave the abstract similarity rather quickly and easily. On the Vocabulary subtest she achieved full credit scores on 31 of the 40 items. Clinically it is of interest to note that the words she missed were regulate, remorse and calamity.

The mental impairment is seen most dramatically on the Digit Span, Picture Completion, Block Design and Digit Symbol subtests. On Digit Symbol she evidenced difficulty in learning new material and worked slowly and gave great effort to making the proper marks in the assigned squares. On Block Design, where one must make reproductions with coloured blocks from pictures of designs, she showed considerable difficulty and inconsistent performance. On the second item (consisting of 4 blocks) she had great difficulty and could not complete it successfully even after two demonstrations were given. She completed items 3 to 5 and missed item 6 because of excessive time. Item 8 was given full credit and she used the maximum time allotment on that item. She showed a good deal of confusion and difficulty in trying to reproduce the designs properly. She verbalized her confusion and her inability to perceptually discriminate and had difficulty in recognizing if her reproductions were accurate. Her affect here was one of bewilderment and confusion and some surprise at her inability. Object Assembly was done fairly well and she achieved all of the four assemblies correctly. On the last assembly she needed 164 of the 180 seconds allotted. She had a good deal of difficulty in recognizing the object on the last assembly and achieved the success through a trial and error approach. Her response on Picture Completion was of diagnostic importance and her perfor-

mance is very inconsistent here. She missed items 4, 6, 10, 11 and out of a total of 21 items only achieved success on nine. On Item 11 where there are insufficient stars on an American flag, after counting the stars she said that nothing was missing. In the latter items on Picture Completion she forgot the instructions (to tell what is missing) and started describing what she considered to be inaccuracies in the figures. It is of interest to note that on Item 17, which is a figure of a man with a missing finger, she stated that his head was too small for his body. On Item 18 where the correct answer is a missing shadow she described, at random, parts of the picture which should not be in the picture. Her lack of ability to remember the instructions and her beginning to describe inaccuracies is significant with respect to her organic condition. Her performance on Picture Arrangement, which requires an ability to comprehend and evaluate a total situation and place individual cards in a correct order so as to tell a story, was done with a good deal of accuracy. She apparently relied on the context again in this situation and came to successful solutions by using logic and by paying attention to the details present in the individual pictures. The comparison here between her ability to succeed on Picture Arrangement and her inability to succeed on Picture Completion is perhaps analogous to the comparison between her ability to succeed on Arithmetic and her inability to succeed on Digit Span in that in both instances where she had success she depended on the structure and meaning of the situation whereas on the subtests on which she failed there was a lack of inherent meaning and context within which she could operate.

At the time of examination this patient did not have marked involvement in the motor areas and she did not demonstrate tremor and dysarthria which occurs in the more advanced stage of the disease. She did demonstrate impaired intellectual performance and those areas which are deteriorated are those usually seen in organic brain conditions. Some subtests which usually are effected by brain dysfunctions were still performed satisfactorily in this patient. The reason here seems to be that there is a context within which she could function which has psychological meaning. In addition she did not show *gross* deficiencies in ability to engage in conceptual and abstract thinking. The functioning demonstrated in this case is similar to that found by Knehr and Bearn in that well habituated language functions and established learned material were intact whereas there was a loss in the efficiency with which new relationships were abstracted. Whereas Knehr and Bearn did not detect perceptual disturbances, such were found in this patient. In addition, it is quite noteworthy that in tasks which did not present a familiar structure within which she could utilize familiar cues, her performance was quite inferior. A rigidity and dependency on old habit patterns appear to aid her maximally in giving her present performance.

Summary

The intellectual performance of a patient with hepatolenticular degeneration was described. Some of the test performance demonstrated was that typically seen in individuals with organic brain dysfunction and some differences were noted. Some hypotheses as to the reasons for the differences were advanced.

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